The GALE ENCYCLOPEDIA *of* CHILDREN'S HEALTH

INFANCY THROUGH ADOLESCENCE

KRISTINE KRAPP AND JEFFREY WILSON, EDITORS

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PLEASE READ—IMPORTANT INFORMATION

The Gale Encyclopedia of Children's Health is a medical reference product designed to inform and educate readers about a wide variety of health issues related to children, ranging from prenatal to adolescence. Thomson Gale believes the product to be comprehensive, but not necessarily definitive. It is intended to supplement, not replace, consultation with a physician or other healthcare practitioner. While Thomson Gale has made substantial efforts to provide information that is accurate, comprehensive, and up-to-date, Thomson Gale makes no representations or warranties of any kind, including without limitation, warranties of merchantability or fitness for a particular purpose, nor does it guarantee the accuracy, comprehensiveness, or timeliness of the information contained in this product. Readers should be aware that the universe of medical knowledge is constantly growing and changing, and that differences of medical opinion exist among authorities. They are also advised to seek professional diagnosis and treatment for any medical condition, and to discuss information obtained from this book with their healthcare provider.

INTRODUCTION

The Gale Encyclopedia of Children's Health: Infancy Through Adolescence (GECH) is a one-stop source for medical information that covers common and rare diseases and medical conditions, immunizations and drugs, procedures, and developmental issues. It particularly addresses parents' concerns about their children's health from before birth through age 18. The book avoids medical jargon, making it easier for the layperson to use. *The Gale Encyclopedia of Children's Health* presents authoritative, balanced information and is more comprehensive than single-volume family medical guides.

SCOPE

Approximately 600 full-length articles are included in *The Gale Encyclopedia of Children's Health*. Articles follow a standardized format that provides information at a glance. Rubrics include:

Diseases/Disorders
- Definition
- Description
- Demographics
- Causes and symptoms
- Diagnosis
- Treatment
- Prognosis
- Prevention
- Parental concerns
- Resources
- Key terms

Procedures
- Definition
- Purpose
- Description
- Risks
- Normal results
- Parental concerns
- Resources
- Key terms

Immunizations/Drugs
- Definition
- Description
- General use
- Precautions
- Side effects
- Interactions
- Parental concerns
- Resources
- Key terms

Development
- Definition
- Description
- Common problems
- Parental concerns
- Resources
- Key terms

A preliminary list of diseases, conditions, procedures, drugs, and developmental issues was compiled from a wide variety of sources, including professional medical guides and textbooks, as well as consumer guides and encyclopedias. The advisory board, composed of seven doctors with specialties in pediatric medicine, evaluated the topics and made suggestions for inclusion. Final selection of topics to include was made by the medical advisors in conjunction with Thomson Gale editors.

INCLUSION CRITERIA

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ABOUT THE CONTRIBUTORS

The essays were compiled by experienced medical writers, including healthcare practitioners and educators, pharmacists, nurses, and other healthcare professionals. *GECH* medical advisors reviewed all of the completed essays to insure that they are appropriate, up-to-date, and medically accurate.

HOW TO USE THIS BOOK

The Gale Encyclopedia of Children's Health has been designed with ready reference in mind:

- Straight **alphabetical arrangement** allows users to locate information quickly.
- Bold faced terms function as *print hyperlinks* that point the reader to related entries in the encyclopedia.
- A list of **key terms** is provided where appropriate to define unfamiliar words or concepts used within the context of the essay. Additional terms may be found in the **glossary**.
- **Cross-references** placed throughout the encyclopedia direct readers to where information on subjects without their own entries can be found. Synonyms are also cross-referenced.
- A **Resources section** directs users to sources of further medical information.
- An appendix of updated **growth charts** from the U.S. Centers for Disease Control for children from birth through age 20 is included.
- An appendix of **common childhood medications** is arranged alphabetically and includes descriptions of each drug and important information about their uses.
- A comprehensive **general index** allows users to easily target detailed aspects of any topic, including Latin names.

GRAPHICS

The Gale Encyclopedia of Children's Health is enhanced with approximately 300 full-color images, including photos, tables, and customized line drawings.

ADVISORY BOARD

An advisory board made up of prominent individuals from the medical community provided invaluable assistance in the formulation of this encyclopedia. They defined the scope of coverage and reviewed individual entries for accuracy and accessibility. The editors would therefore like to express our appreciation to them.

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Abandonment

Definition

Abandonment is a legal term describing the failure of a non-custodial parent to provide support to his or her children according to the terms approved by a court of law. In common use, abandonment refers to the desertion of a child by a parent.

Description

Legal abandonment is an persistent issue that has received increasing attention since the 1970s. It refers to non-custodial parents who do not fulfill court-ordered financial responsibilities to their children, regardless of their involvement in their children's lives in other ways. Lack of such support is blamed for substantial poverty among **single-parent families**.

In 2002 it was estimated that up to 30 percent (19.8 million) of children in the United States, representing 11.9 million families, lived in single-parent households. While the number of single mothers has remained constant in recent years at 9.9 million, the number of single fathers has grown from 1.7 million in 1995 to 2 million in 2002, according to data from the U.S. Census Bureau. In 2002, some 19.8 million children lived with one parent. Of these, 16.5 million lived with their mother and 3.3 million with their father.

Fewer than half of single-parent children under the age of 18 received any financial support from the non-custodial parent. The income of more than a third of these households fell below the poverty level. The term "deadbeat dads" is often used in discussions about abandonment because most of the parents involved are fathers.

An increasing **divorce** rate and a rise in the number of infants born to unmarried mothers were in large part responsible for forcing the abandonment issue into public consciousness in the 1970s. Typically during the twentieth century, mothers involved in divorce or unwed births were routinely given physical custody of children, while fathers were granted visitation rights and ordered to pay a certain amount of money to help care for the children's needs. Many men ignored this financial responsibility, forcing some women to get jobs or to seek government support.

States have always taken on the main responsibility for ensuring the welfare of abandoned children. Federal involvement came as early as 1935, when the Social Security Act established the Aid to Dependent Children (ADC) program, primarily to assist widows. Over subsequent years, federal provisions strengthened the states' mandate. During the early 1970s, when the government's financial burden grew as more and more women turned to welfare, the U.S. Congress began to call for even stronger child-support enforcement provisions.

Enforcement laws vary from state to state. Garnishing wages, attacking bank accounts, and foreclosing on real estate are all used to force payment to affected children. All state enforcement systems are automated, allowing more efficient monitoring of payment and better tracking of violating parents. Some states have begun to deny drivers' and professional licenses to known delinquent parents. For example, in California, licenses for real estate salespersons, brokers, and appraisers can be revoked, suspended, or denied to applicants who are delinquent in child support payments. "Wanted" posters and other forms of advertising are more unconventional methods used occasionally to locate such parents.

Most states give priority to finding parents whose children, lacking parental support, are receiving government assistance. Some families with independent incomes turn to lawyers or private collection agencies to find offenders and bring them to court for nonpayment. In the late 1990s and early 2000s, hundreds of agencies specializing in child support collection, some of them unscrupulous, have been formed to meet the demand

forced by overburdened state agencies. They sometimes charge extremely high retainer or contingency fees, substantially reducing the size of the payment recovered by the **family**.

In the 1990s, the federal government adopted measures to further assist states in the support-collection effort. Military personnel files have become more available, and a program to confiscate federal tax refunds has contributed to keeping the issue in the spotlight. The 1992 Child Support Recovery Act allows courts to impose criminal penalties on parents who cross state lines to avoid child support payments.

Some support exists for consolidating child-support enforcement through the Internal Revenue Service (IRS) rather than the states. Proponents argue that only the IRS can efficiently confiscate deadbeat parents' income and return it to children. Opponents contend that the involvement of the federal bureaucracy would more likely add inefficiency to the enforcement process and only aggravate an already growing problem.

Abandonment can take on a broader form than just legal abandonment. The term is used to refer to the abandonment of a child by one or both parents, either through desertion, divorce, or death. Although death is not legally abandonment, many children experience feelings and **fear** of abandonment following the death of one or both parents.

Abandonment is about the loss of love and a loss of connectedness. To the abandoned adolescent, it involves feelings of betrayal, guilt, loneliness, and lack of **self-esteem**. Abandonment is a core fear in humans, and this fear is intensified in adolescents.

The abandonment of children is an extreme form of child neglect stemming from many causes. Some include family breakdown, irresponsible fatherhood or motherhood, premature motherhood, birth out of wedlock, or the death of one or both parents.

The problem is not new. In the nineteenth century, "ragamuffins" were a familiar part of London's urban scene, and parents in Paris abandoned their children at the rate of 20 percent of the live births in the city. In his 1987 book, *Children of the Sun* Morris West tells of the survival of street children in Naples in the 1950s. What is new, rather, is the growing scale of the problem. The United Nations estimates 60 million children and infants have been abandoned by their families and live on their own or in orphanages in the world. In the United States, more than 7,000 children are abandoned each year.

Infancy and toddlerhood

Children in this stage of development understand little, if anything, about abandonment. However, they are aware of the emotional climate of the family. For the remaining parent, it is important to cuddle and care for the infant or toddler warmly, frequently, and consistently. The parent-child relationship continues to be central to the child's sense of security and independence.

Preschool

Preschoolers tend to have a limited and mistaken perception of abandonment. They are highly self-centered with a strict sense of right and wrong. So when bad things happen to them, they usually blame themselves by assuming they did something wrong. Children this age often interpret the departure of a parent as a personal rejection. Youngsters are likely to deny the reality of the abandonment and wish intently for the parent to return. They can also regress to behaviors such as **thumb sucking**, bed wetting, temper **tantrums**, and clinging to a favorite blanket or toy. They also fear abandonment by the other parent. They generally become afraid of the dark and of being alone.

School age

By the time children reach the early school years, ages six to nine, they can no longer deny the reality of the abandonment. They are extremely aware of the pervasive **pain** and sadness. Boys, especially, mourn the loss of their fathers, and their anger is frequently directed at their mothers. Crying, daydreaming, and problems with friends and in school are common abandonment behaviors in children of this age.

In the age group of nine to 12, adolescents usually react to abandonment with anger. They may also resent the additional household duties expected of them. There is also a significant disruption in the child's ability to learn. **Anxiety**, restlessness, inability to concentrate, and intrusive thoughts about the abandonment take a toll and can lead to a drop in school performance and difficulties with classmates.

Feelings of sadness, loneliness, guilt, lack of self worth, and self-blame are common in nine to 12-year-olds. They also tend to have concerns about family life, worry about finances, and feel they are a drain on the remaining parent's resources.

In children ages 13 to 18, the feelings are usually the same as with the younger groups except more pronounced. They become concerned about their own futures. **Truancy** is high, school performance is low, and they have a distorted view of themselves. In this popula-

KEY TERMS

Contingencies—Naturally occurring or artificially designated reinforcers or punishers that follow a behavior.

Deadbeat dad—A father who has abandoned his child or children and does not pay child custody as required by a court.

Deadbeat parent—A mother or father who has abandoned his or her child or children and does not pay child custody as required by a court.

Non-custodial parent—A parent who does not have legal custody of a child.

Promiscuous—Having many indiscriminate or casual sexual relationships.

Ragamuffins—A term used in nineteenth-century London to describe neglected or abandoned children who lived on the streets.

Retainer—A fee paid in advance to secure legal services.

tion there is a high incidence of drug and alcohol abuse and **aggressive behavior**.

The teen may also withdraw from all relationships, including those with friends, family, and classmates, and become extremely dependent on the remaining parent. Teens may also react by becoming sexually promiscuous at an early age, sometimes to the point of **addiction**. Sometimes, however, the child makes valuable decisions about their own future and values.

Common problems

Problems to watch for include trouble sleeping, crying, aggression, deep anger and resentment, feelings of betrayal, difficulty concentrating, chronic fatigue, and problems with friends or at school.

Parental concerns

The remaining parent should be aware of the effects of the abandonment on the child and above all, reassure the child that the remaining parent will not abandon them.

When to call the doctor

Medical help may be needed if the abandoned child inflicts self-injury. Psychological counseling may also be needed to help the child understand and cope with the abandonment. This is especially true if any of the common reactions lasts for an unusual amount of time, intensifies over time, or if the child talks about or threatens **suicide**.

Resources

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ORGANIZATIONS

American Bar Association (ABA) Center on Children and the Law. 740 15th St., NW, Washington, DC 20005. Web site: <www.abanet.org/child/home.html>.

Children's Rights Council. 6200 Editors Park Drive, Suite 103, Hyattsville, MD 20782. Web site: <www.gocrc.com/>.

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Ken R. Wells

Abdominal wall defects

Definition

Abdominal wall defects are birth (congenital) defects that allow the stomach or intestines to protrude.

Description

Many unexpected events occur during the development of a fetus inside the womb. The stomach and intestines begin development outside the baby's abdomen and only later does the abdominal wall enclose them. Occasionally, either the umbilical opening is too large or it develops improperly, allowing the bowels or stomach to remain outside or squeeze through the abdominal wall. This results in one of two conditions, gastroschisis and omphalocele. Gastroschisis occurs when the abdominal wall does not close completely and the stomach and the small and large intestines appear outside the infant's body. In omphalocele, some of the internal organs protrude through the abdominal muscles in the area around the umbilical cord. Omphalocele may be minor, involving only a small portion of the intestines, or it may be severe with most of the abdominal organs, such as the intestines, liver, and spleen, outside the body.

Demographics

Abdominal wall defects, specifically gastroschisis and omphalocele, are rare and occur in only once in every 5000 births. Both boys and girls have these defects in equal numbers. While infants with gastroschisis rarely have defects other than those affecting the intestines, of children with omphalocele, 50 percent to 75 percent have associated congenital anomalies and 20 percent to 35 percent have chromosomal abnormalities.

Causes and symptoms

There are many causes for birth defects that still remain unclear. As of 2004, the causes of abdominal wall defects remained unknown. Any symptoms the mother may have had to indicate that the defects are present in the fetus are nondescript.

Diagnosis

At birth, the problem is obvious, because the base of the umbilical cord at the navel will bulge or, in worse cases, contain internal organs. Before birth, an ultrasound examination may detect the problem. It is always necessary in children with abdominal wall defects to look for other birth defects, because multiple anomalies are more likely to occur in these children.

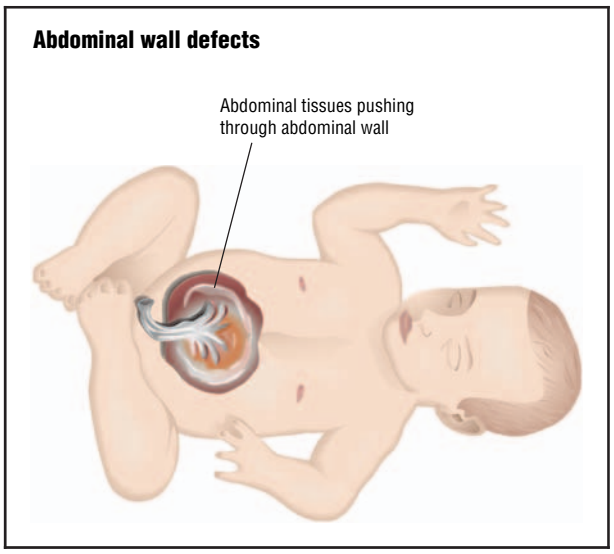

Illustration of an omphalocele, or umbilical hernia, in a newborn. *(Illustration by GGS Information Services.)*

Treatment

Abdominal wall defects are effectively treated with surgical repair. Unless there are accompanying anomalies, the surgical procedure is not overly complicated. However, if the defect is large, it may be difficult to fit all the organs into the small abdominal cavity.

Prognosis

If there are no other defects, the prognosis after surgical repair of this condition is relatively good. However, 10 percent of those with more severe or additional abnormalities die from it. The organs themselves are fully functional; the difficulty lies in fitting them inside the abdomen. The condition is, in actuality, a **hernia** requiring only replacement and strengthening of the passageway through which it occurred. However, after surgery, increased pressure in the stretched abdomen can compromise the function of the organs inside.

Prevention

Some, but by no means all, birth defects are preventable by early and attentive prenatal care, good **nutrition**, supplemental **vitamins**, and other elements of a healthy lifestyle, along with the diligent avoidance

KEY TERMS

Congenital—Present at birth.

Hernia—A rupture in the wall of a body cavity, through which an organ may protrude.

Umbilical—Refers to the opening in the abdominal wall where the blood vessels from the placenta enter.

of all unnecessary drugs and chemicals—especially tobacco.

Parental concerns

Most children with abdominal wall defects require immediate and intensive medical care. Some of these infants may have multiple surgeries, and serious complications such as feeding problems and infections may persist long term. Parents will need to work closely with a team of physicians during the treatment of their child. Children with abdominal wall defects may need additional services, especially those with omphalocele and associated chromosomal abnormalities and birth defects. These children require long-term treatment for both the physical and developmental difficulties they face. Parents may need support services in addition to the services provided by the healthcare team. They can contact the hospital's social work department to learn more about available resources.

Resources

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Abuse, child *see* **Child abuse**

Accessory digits *see* **Polydactyly and syndactyly**

Acetaminophen

Definition

Acetaminophen is a medicine used to relieve **pain** and reduce **fever**.

Description

Acetaminophen is used to relieve many kinds of minor aches and pains: headaches, muscle aches, backaches, toothaches, menstrual cramps, arthritis, and the aches and pains that often accompany colds.

Description

This drug is available without a prescription. Acetaminophen is sold under various brand names, including Tylenol, Panadol, Aspirin Free Anacin, and Bayer Select Maximum Strength **Headache** Pain Relief Formula. Many multi-symptom cold, flu, and sinus medicines also contain acetaminophen.

Studies have shown that acetaminophen relieves pain and reduces fever about as well as aspirin. But differences between these two common drugs exist. Acetaminophen is less likely than aspirin to irritate the stomach. However, unlike aspirin, acetaminophen does not reduce the redness, stiffness, or swelling that accompany arthritis.

Precautions

Most of the precautions for acetaminophen apply to adults rather than children but may apply to some teenagers.

The primary precaution in children's therapy is to watch the dosage carefully and follow the label instructions only. Acetaminophen for children comes in two strengths. Children's acetaminophen contains low concentrations of the drug, 160 milligrams in a teaspoonful

of solution. The infant drops contain a much higher concentration of acetaminophen, 100 milligrams in 20 drops, equal to 500 milligrams in a teaspoonful. The infant drops should never be given by the teaspoonful.

Parents should never give their child more than the recommended dosage of acetaminophen unless told to do so by a physician or dentist.

Patients should not use acetaminophen for more than 10 days to relieve pain (five days for children) or for more than three days to reduce fever, unless directed to do so by a physician. If symptoms do not go away or if they get worse, a physician should be contacted. Anyone who drinks three or more alcoholic beverages a day should check with a physician before using this drug and should never take more than the recommended dosage. A risk of liver damage exists from combining large amounts of alcohol and acetaminophen. People who already have kidney or liver disease or liver infections should also consult with a physician before using the drug. Women who are pregnant or breastfeeding should do the same.

Side effects

Acetaminophen causes few side effects. The most common one is lightheadedness. Some people may experience trembling and pain in the side or the lower back. Allergic reactions do occur in some people, but they are rare. Anyone who develops symptoms such as a rash, swelling, or difficulty breathing after taking acetaminophen should stop taking the drug and get immediate medical attention. Other rare side effects include yellow skin or eyes, unusual bleeding or bruising, weakness, fatigue, bloody or black stools, bloody or cloudy urine, and a sudden decrease in the amount of urine.

Overdoses of acetaminophen may cause **nausea**, **vomiting**, sweating, and exhaustion. Very large overdoses can cause liver damage. In case of an overdose, parents should get immediate medical attention for their child.

Interactions

Acetaminophen may interact with a variety of other medicines. When this happens, the effects of one or both of the drugs may change or the risk of side effects may be greater. Among the drugs that may interact with acetaminophen are the following:

- alcohol
- nonsteroidal anti-inflammatory drugs (NSAIDs) such as Motrin
- oral contraceptives
- the antiseizure drug phenytoin (Dilantin)
- the blood-thinning drug warfarin (Coumadin)
- the cholesterol-lowering drug cholestyramine (Questran)
- the antibiotic Isoniazid
- zidovudine (Retrovir, AZT)

Check with a physician or pharmacist before combining acetaminophen with any other prescription or nonprescription (over-the-counter) medicine.

Acetaminophen is generally safe when taken as directed. Acetaminophen is commonly mixed with other ingredients as part of combinations intended for colds, **influenza**, and other conditions. Parents should read the labels carefully in order to avoid giving an overdose of acetaminophen to their child. They need to be particularly cautious about liquid medicines that contain acetaminophen and alcohol.

Parental concerns

Acetaminophen is very safe when used properly. While most precautions are intended to reduce the risk of overdose, parents should not try to reduce the risk by giving a lower than normal dose. Children should not suffer pain if it can be safely treated.

See also Analgesics; Pain management.

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Acne

Definition

Acne is a skin disorder that leads to an outbreak of lesions called pimples or "zits." The most common form of the disease is called acne vulgaris—the rash that affects many adolescents. Acne vulgaris is triggered by the hormonal changes that occur in **puberty**.

Description

Acne is a condition in which pimples appear on the face, chest, and back. In teenagers, acne usually appears on the forehead, nose, and chin. It is caused by the overproduction of sebum. Sebum is an oily substance that forms in glands just under the surface of the skin called sebaceous glands. Sebum normally flows out hair follicles onto the skin to act as a natural skin moisturizer. The glands are connected to hair follicles that allow the sebum, or oil, to empty onto the skin through a pore.

If hair follicles become blocked by sebum, dead skin cells, and bacteria, acne is the result. The sebaceous gland units are most commonly found on the face, neck, and back.

During puberty, there are increased levels of the male hormone androgen. High levels of androgen cause excess sebum to form. Sometimes the sebum combines with dead, sticky skin cells and bacteria called *Propionibacterium acnes (P. acnes)* that normally live on the skin. The mixture of oil and cells allows the bacteria to grow in the plugged follicles. When this happens, a hard plug called a comedo can form. A comedo is an enlarged hair follicle. It can take the following forms:

- a blackhead, which is a comedo that reaches the skin's surface and looks black
- a whitehead, which is a comedo that is sealed by keratin, the fibrous protein produced by the skin cells and looks like a white bump.

In addition, pimples can form on the skin. Types of pimples include:

- papules, which are small, red bumps that may be tender to the touch
- pustules, which are pus-filled lesions that are often red at the base
- nodules, which are large, painful lesions deep in the skin
- cysts, which are painful pus-filled lesions deep in the skin that can cause scarring

Pimples form when the follicle is invaded by the *P. acnes* bacteria. The damaged follicle weakens and bursts open, releasing sebum, bacteria, skin cells, and white blood cells into surrounding tissues. Scarring happens when new skin cells are created to replace the damaged cells. The most severe type of acne includes both nodules and cysts.

Demographics

Acne affects as many as 17 million people in the United States, making it the most common skin disease. Acne usually begins at puberty and worsens during **adolescence**. Nearly 85 percent of people develop acne at some point between ages 12 to 25. As many as 20 million teens have the condition. Acne may appear as early as age 10, and even may be found in some newborns. Some people may continue to be affected by acne after age 30.

Causes and symptoms

The exact cause of acne is as of 2004 not known. There are several risk factors that make acne more likely to occur:

- Age. Adolescents are more likely to have acne.
- Disease. Certain hormonal disorders such as polycystic ovarian syndrome make acne more likely.

- Hormonal changes. Acne can flare up before **menstruation**. An increase in the male hormone androgen during puberty (seen in both males and females) causes the sebaceous glands to overproduce androgen. Boys have more severe acne than girls.
- Heredity. Some individual are genetically more susceptible to acne.
- Drugs. Steroids and performance enhancing drugs, **oral contraceptives**, **antibiotics**, **antidepressants**, and tranquillizers such as lithium are known to cause acne.
- Cosmetics. Oily cosmetics can plug up hair follicles.

Other factors can worsen acne or cause it to flare up:

- Environmental irritants. Air pollution and high humidity can worsen acne, as can exposure to greasy environments such as working in a fast food restaurant.
- Friction. Rubbing the skin vigorously or exposure to constant friction from backpacks or tight collars can worsen acne.
- Personal hygiene. Picking at pimples or scrubbing the skin too hard can result in worsened acne.

Factors that do not cause acne include:

- chocolate and greasy foods
- stress

When to call the doctor

A healthcare provider should be contacted under the following circumstances:

- Painful nodules and cysts are present.
- Over-the-counter medications have not been effective.
- Acne lesions are causing scarring.
- Acne is causing dark skin to have darker patches when lesions heal.
- Acne is causing embarrassment or self-consciousness.
- Acne is creating emotional upset.

Acne may be treated by the **family** doctor. More severe cases may be referred to a dermatologist (skin doctor) or an endocrinologist (doctor who treats hormonal/glandular disorders).

Diagnosis

Acne can be diagnosed by physical examination and a medical history of acne. The physician will take a medical history, including information about skin care, diet, medications, factors that can cause flare-ups, and prior treatment. Blood tests are not usually necessary unless a hormonal disorder is suspected.

Physical examination will include the face, neck, shoulders, back, and other affected areas. Using specialized lighting, the physician will examine the affected areas to see the following:

- what type and how many lesions are present
- how deep the lesions are
- whether they are inflamed
- whether scarring or skin discoloration is present

Treatment

Acne treatment consists of reducing the sebum production, removing dead skin cells, and killing bacteria with oral medication and drugs used on the skin (topical). The treatment depends on the severity of the condition.

Drugs

TOPICAL (SKIN) MEDICATION Treatment for mild noninflammatory acne consists of reducing the formation of new comedones with medications including topical tretinoin, benzoyl peroxide, adapalene, or salicylic acid. Tretinoin is especially effective because it increases turnover and replacement of skin cells. If lesions are inflamed, **topical antibiotics** may be added to the treatment regimen. Improvement is usually seen in two to four weeks.

Topical medications are available as cream, gel, lotion, or pad preparations of varying strengths. They include antibiotics (to kill bacteria) such as erythromycin, clindamycin (Cleocin-T), and meclocycline (Meclan); and comedolytics (agents that loosen hard plugs and open pores) such as the vitamin A acid tretinoin (Retin-A), salicylic acid, adapalene (Differin), resorcinol, and sulfur. Drugs that act as both comedolytics and antibiotics, such as benzoyl peroxide, azelaic acid (Azelex), or benzoyl peroxide plus erythromycin (Benzamycin), are also used. These drugs may be used for months to years to achieve disease control.

After the person washes with mild soap, the drugs are applied alone or in combination, once or twice a day over the entire affected area of skin. Possible side effects include mild redness, peeling, irritation, dryness, and an increased sensitivity to sunlight that requires use of a sunscreen.

ORAL DRUGS Oral antibiotics are taken daily for two to four months. The drugs used include tetracycline, erythromycin, minocycline (Minocin), doxycycline,

clindamycin (Cleocin), and trimethoprim-sulfamethoxazole (Bactrim, Septra). Possible side effects include allergic reactions, stomach upset, vaginal yeast infections, **dizziness**, and tooth discoloration.

The goal of treating moderate acne is to decrease inflammation and prevent new comedones from forming. One effective treatment is topical tretinoin, used along with a topical or oral antibiotic. A combination of topical benzoyl peroxide and erythromycin is also very effective. Improvement is normally seen within four to six weeks, but treatment is maintained for at least two to four months.

A drug reserved for the treatment of severe acne, oral isotretinoin (Accutane), reduces sebum production and cell stickiness. It is the treatment of choice for severe acne with cysts and nodules and is used with or without topical or oral antibiotics. Taken for four to five months, it provides long-term disease control in up to 60 percent of patients. If the acne reappears, another course of isotretinoin may be needed by about 20 percent of patients, while another 20 percent may do well with topical drugs or oral antibiotics. However there are significant side effects, including temporary worsening of the acne; dry skin; nosebleeds; vision disorders; and elevated liver enzymes, blood fats, and cholesterol. The drug also causes benign intracranial **hypertension** (pseudotumor cerebri) and mood changes. This drug must not be taken during pregnancy since it causes birth defects. Sexually active young women being treated with isotretinoin must use a reliable contractive, and they need to use **contraception** for up to one month after stopping use of the drug.

Anti-androgens, drugs that inhibit androgen production, are used to treat women who are unresponsive to other therapies. Certain types of oral contraceptives (for example, Ortho-Tri-Cyclen) and female sex hormones (estrogens) reduce hormone activity in the ovaries. Other drugs (for example, spironolactone and corticosteroids) reduce hormone activity in the adrenal glands. Improvement may take up to four months.

Oral corticosteroids, or anti-inflammatory drugs, are the treatment of choice for an extremely severe, but rare type of destructive inflammatory acne called acne fulminans, found mostly in adolescent males. Acne conglobata, a more common form of severe inflammation, includes numerous, deep, inflammatory nodules that heal with scarring. It is treated with oral isotretinoin and corticosteroids.

Other treatments

Several surgical or medical treatments are available to alleviate acne or the resulting scars:

- Comedone extraction. The comedo is removed from the pore with a special tool.
- Chemical peels. Glycolic acid is applied to peel off the top layer of skin to reduce scarring.
- Dermabrasion. The affected skin is frozen with a chemical spray and removed by brushing or planing.
- Punch grafting. Deep scars are excised and the area repaired with small skin grafts.
- Intralesional injection. Corticosteroids are injected directly into inflamed pimples.
- Collagen injection. Shallow scars are elevated by collagen (protein) injections.

Alternative treatment

Alternative treatments for acne focus on self care: proper cleansing to keep the skin oil-free; eating a well-balanced diet high in fiber, zinc, and raw foods; and avoiding alcohol, dairy products, tobacco, **caffeine**, sugar, processed foods, and foods high in iodine, such as salt.

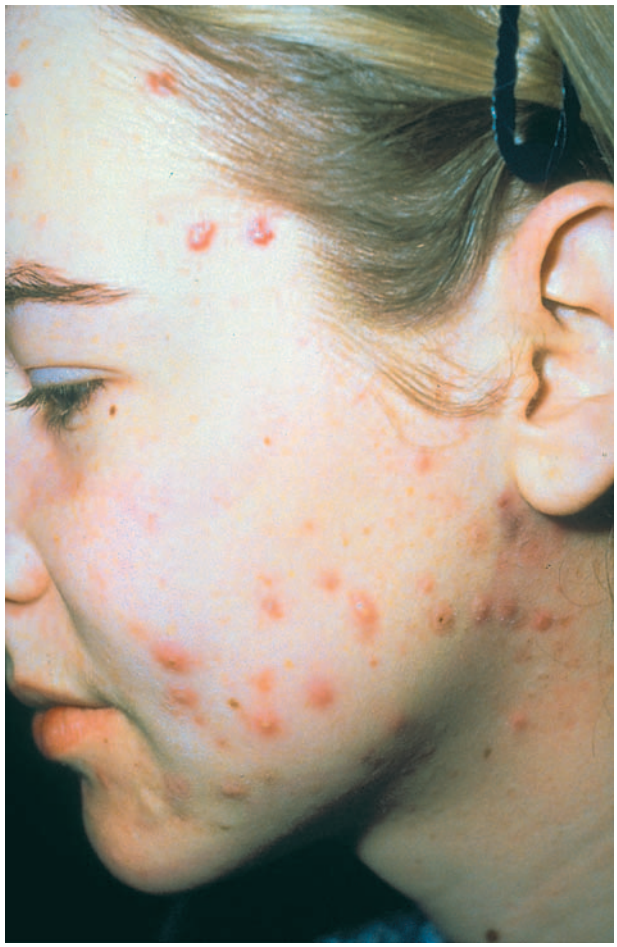
Supplementation with herbs such as burdock root (*Arctium lappa*), red clover (*Trifolium pratense*), and milk thistle (*Silybum marianum*), and with nutrients such as essential fatty acids, vitamin B complex, zinc, vitamin A, and chromium is also recommended. Chinese herbal remedies used for acne include cnidium seed (*Cnidium monnieri*) and honeysuckle flower (*Lonicera japonica*). Holistic physicians or nutritionists can recommend the proper amounts of these herbs.

Nutritional concerns

Acne is not caused or worsened by eating chocolate or oily foods.

Prognosis

Acne is not curable, although it can be controlled by proper treatment. Improvement can take two or more months. Long-term control is achieved in up to 60 percent of patients with severe acne who are treated with the drug isotretinoin. Acne tends to reappear when treatment stops, but spontaneously improves over time. Acne usually improves after adolescence, although some individuals continue to have lesions after age 30. Inflammatory acne may leave scars that require further treatment.



Teenage girl with acne. *(Photograph by Biophoto Associates. National Audubon Society Collection/Photo Researchers, Inc.)*

Prevention

There are no sure ways to prevent acne, but the following steps may be taken to minimize flare-ups:

- gentle washing of affected areas once or twice every day
- avoiding abrasive cleansers
- using noncomedogenic (does not clog pores) makeup and moisturizers
- shampooing often and wearing hair off the face
- eating a well-balanced diet, avoiding foods that trigger flare-ups
- unless told otherwise by the healthcare provider, giving dry pimples a limited amount of sun exposure
- not picking or squeezing blemishes

Parental concerns

Acne comes at a difficult time, during the adolescent years. While mild acne can be treated with over-the-counter medications, more severe acne needs medical attention. Experts advise against a wait-and-see attitude. Treatment options can help control acne and avoid scarring.

See also Antiacne drugs.

KEY TERMS

Androgens—Hormones (specifically testosterone) responsible for male sex characteristics.

Antiandrogen—A substance that blocks the action of androgens, the hormones responsible for male characteristics.

Antibiotics—Drugs that are designed to kill or inhibit the growth of the bacteria that cause infections.

Comedo—A hard plug composed of sebum and dead skin cells, also called a blackhead. The mildest type of acne.

Comedolytic drugs—Medications that break up comedones and open clogged pores.

Corticosteroids—A group of hormones produced naturally by the adrenal gland or manufactured synthetically. They are often used to treat inflammation. Examples include cortisone and prednisone.

Estrogen—Female hormone produced mainly by the ovaries and released by the follicles as they mature. Responsible for female sexual characteristics, estrogen stimulates and triggers a response from at least 300 tissues. After menopause, the production of the hormone gradually stops.

Isotretinoin—A powerful vitamin A derivative used in the treatment of acne.

Noncomedogenic—A substance that does not contribute to the formation of blackheads or pimples on the skin.

Sebaceous follicle—A structure found within the skin where a sebaceous gland opens into a hair follicle.

Sebum—An oily skin moisturizer produced by sebaceous glands.

Tretinoin—A drug, used in the treatment of acne, that works by increasing the turnover (death and replacement) of skin cells.

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Acromegaly and gigantism

Definition

Acromegaly is a disease in which an abnormality in the pituitary gland leads to an oversecretion of growth hormone. In adults, this condition results in an enlargement of bones; in children, the abnormality results in excessive height and is called gigantism.

Description

Acromegaly is a disorder in which the abnormal release of a particular chemical from the pituitary gland in the brain causes increased growth in bone and soft tissue, as well as a variety of other disturbances throughout the body. This chemical released from the pituitary gland is called growth hormone (GH). The body's ability to process and use nutrients like fats and sugars is also altered. In children whose bony growth plates have not closed, the chemical changes of acromegaly result in exceptional growth of long bones. This variant is called gigantism, with the additional bone growth causing unusual height. When the abnormality occurs after bone growth stops, i.e. in adults, the disorder is called acromegaly.

Demographics

Acromegaly is a relatively rare disorder, occurring in approximately 50 out of every 1 million people. Gigantism occurs even more rarely, with reported cases in the United States numbering only about 100 by 2004. Males and females are similarly affected. Onset of gigantism is usually at **puberty**, although some cases of toddlers and young children with gigantism have been reported.

Causes and symptoms

The pituitary is a small gland located at the base of the brain, which releases certain hormones that are important to the functioning of other organs or body systems. The pituitary hormones travel throughout the body and are involved in a large number of activities, including the regulation of growth and reproductive functions. The cause of acromegaly can be traced to the pituitary's production of GH.

Under normal conditions, the pituitary receives input from another brain structure, the hypothalamus, located at the base of the brain. This input from the hypothalamus regulates the pituitary's release of hormones. For example, the hypothalamus produces growth hormone-releasing hormone (GHRH), which directs the pituitary to release GH. Input from the hypothalamus should also direct the pituitary to stop releasing hormones.

In acromegaly, the pituitary continues to release GH and ignores signals from the hypothalamus. In the liver, GH causes production of a hormone called insulin-like growth factor 1 (IGF-1), which is responsible for growth throughout the body. When the pituitary refuses to stop producing GH, the levels of IGF-1 also reach abnormal peaks. Bones, soft tissue, and organs throughout the body begin to enlarge, and the body changes its ability to process and use nutrients like sugars and fats.

The most common cause of acromegaly and gigantism is the development of a noncancerous tumor within the pituitary, called a pituitary adenoma. In the case of pituitary adenomas, the tumor itself is the source of the abnormal release of GH. As these tumors grow, they may press on nearby structures within the brain, causing headaches and changes in vision. As the adenoma grows, it may disrupt other pituitary tissue, interfering with the release of other hormones. These disruptions may be responsible for changes in the menstrual cycle and abnormal production of breast milk in

women or delayed development of reproductive organs. In rare cases, acromegaly is caused by the abnormal production of GHRH, which leads to the increased production of GH. Certain tumors in the pancreas, lungs, adrenal glands, thyroid, and intestine can produce GHRH, which in turn triggers production of an abnormal quantity of GH.

In acromegaly, an individual's hands and feet begin to grow, becoming thick and doughy. The jaw line, nose, and forehead also grow, and facial features are described as coarse. The tongue grows larger, and because the jaw is larger, the teeth become more widely spaced. Due to swelling within the structures of the throat and sinuses, the voice becomes deeper and sounds hollower, and patients may develop loud snoring. Children and adolescents with gigantism show a characteristic lengthening and enlargement of bones, principally of the limbs. Some symptoms caused by various hormonal changes are as follows:

- heavy sweating
- oily skin
- increased coarse body hair
- improper processing of sugars in the diet (and sometimes actual diabetes)
- high blood pressure
- increased calcium in the urine (sometimes leading to kidney stones)
- increased risk of gallstones
- swelling of the thyroid gland

People with acromegaly have more skin tags, or outgrowths of tissue, than normal. This increase in skin tags is also associated with the development of growths, called polyps, within the large intestine that may eventually become cancerous. Patients with acromegaly often suffer from headaches and arthritis. The various swellings and enlargements throughout the body may press on nerves, causing sensations of local **tingling** or burning and sometimes result in muscle weakness.

When to call the doctor

Because early diagnosis and treatment of acromegaly and gigantism can often lead to the avoidance of more serious symptoms, a healthcare professional should be contacted if a child develops any of the early symptoms of the disease, such as a marked increase in height or height that is excessive for his or her age.

Diagnosis

Because acromegaly produces slow changes, diagnosis is often significantly delayed. In fact, the characteristic coarsening of the facial features is often not recognized by **family** members, friends, or long-time family physicians. Often, the diagnosis is suspected by a new physician who sees the patient for the first time and is struck by the patient's characteristic facial appearance. Comparing old photographs from a number of different periods often increases suspicion of the disease. By contrast, the effects of gigantism are typically dramatic, with remarkable changes over a short period of time.

Because the quantity of GH produced varies widely under normal conditions, demonstrating high levels of GH in the blood is not sufficient to merit a diagnosis of acromegaly. Instead, laboratory tests measuring an increase of IGF-1 (three to ten times above the normal level) are useful. These results, however, must be carefully interpreted because normal laboratory values for IGF-1 vary when the patient is pregnant, is pubescent, is elderly, or is severely malnourished. Normal patients will show a decrease in GH production when given a large dose of sugar (glucose). Patients with acromegaly will not show this decrease and will often show an increase in GH production. **Magnetic resonance imaging** (MRI) is useful for viewing the pituitary gland and for identifying and locating an adenoma. When no adenoma can be located, the search for a GHRH-producing tumor in another location begins.

Treatment

The first step in treatment of acromegaly is removal of all or part of the pituitary adenoma. Removal usually requires surgery, usually performed by entering the skull through the nose. While this surgery can cause rapid improvement of many acromegaly symptoms, most patients will also require additional treatment with medication. Bromocriptine (Parlodel) is a medication that can be taken by mouth, while octreotide (Sandostatin) must be injected every eight hours. Both of these medications are helpful in reducing GH production but must often be taken for life and produce their own unique side effects.

Alternative treatment

Some patients who cannot undergo surgery are treated with radiation therapy to the pituitary in an attempt to shrink the adenoma. Radiating the pituitary may take up to ten years, however, and may also injure or destroy other normal parts of the pituitary.

KEY TERMS

Adenoma—A type of noncancerous (benign) tumor that often involves the overgrowth of certain cells found in glands. These tumors can secrete hormones or cause changes in hormone production in nearby glands.

Gland—A collection of cells whose function is to release certain chemicals (hormones) that are important to the functioning of other, sometimes distantly located, organs or body systems.

Hormone—A chemical messenger secreted by a gland or organ and released into the bloodstream. It travels via the bloodstream to distant cells where it exerts an effect.

Hypothalamus—A part of the forebrain that controls heartbeat, body temperature, thirst, hunger, body temperature and pressure, blood sugar levels, and other functions.

Pituitary gland—The most important of the endocrine glands (glands that release hormones directly into the bloodstream), the pituitary is located at the base of the brain. Sometimes referred to as the "master gland," it regulates and controls the activities of other endocrine glands and many body processes including growth and reproductive function. Also called the hypophysis.

Nutritional concerns

Individuals with acromegaly or gigantism who have diabetes or diabetes-like symptoms should maintain a diet that helps normalize blood sugar levels.

Prognosis

Without treatment, patients with acromegaly are likely to die early because of the disease's effects on the heart, lungs, brain, or due to the development of **cancer** in the large intestine. With treatment, however, a patient with acromegaly may be able to live a normal lifespan.

Prevention

The initial onset of acromegaly or gigantism cannot as of 2004 be prevented. Once a pituitary adenoma has been removed, radiotherapy and/or medication may be recommended to prevent a recurrence of the tumor.

Parental concerns

In the great majority of children of tall stature, genetics and **nutrition** are the cause of the greater-than-average height, and linear growth ceases with the end of puberty. In individuals with gigantism who are not treated, linear growth can continue unchecked for several decades. It is important that a child with the symptoms of gigantism be assessed medically so that treatment can be implemented and abnormal linear height as well as potentially serious symptoms such as heart disease or colon cancer be minimized or avoided.

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Acting out

Definition

Acting out is defined as the release of out-of-control aggressive or sexual impulses in order to gain relief from tension or **anxiety**. Such impulses often result in antisocial or delinquent behaviors. The term is also sometimes used in regard to a psychotherapeutic release of repressed feelings, as occurs in psychodrama.

Description

Infants and toddlers

The earliest acting out behaviors are often referred to as temper **tantrums**. These behaviors are usually first observed in infants between the ages of 12 and 18 months of age. At that point, temper tantrums can be considered a normal part of growth and development. These early tantrums are simply an infant's attempt to communicate feelings of dissatisfaction or extreme disappointment. Observed behaviors in infants trying to express their anger or frustration usually include patently angry-sounding crying, kicking hands and feet, and possibly even trying to strike out. For toddlers, such violent outbursts of temper often include hitting, kicking, and biting others; and possibly self-injurious behaviors such as head-banging. The child's reaction to the supposed cause of the tantrum is often markedly disproportionate to the precipitating incident. An example would be the child who is told that he or she cannot have a lollipop and then proceeds to violently attack the mother, hitting and kicking her, while screaming as loudly as possible.

Acting out conduct can include any highly emotional, disruptive, and unacceptable outburst that appears to be the child's reaction to unmet needs or wishes. A primary reason for such emotional lack of control in a child, especially above the age of three or four, is having not learned how to cope with their own frustration. Such temper tantrums usually peak between the ages of two and three. Because under normal circumstances the child has learned the necessary lessons in how to deal with disappointment by the time he or she has reached the age of four, there is then a noticeable decrease in this sort of acting out behavior.

Preschool and school age children

When no medical or psychological determination is discovered for acting out behaviors in young children above the age of four, the assumption can be made that the temper tantrums are a learned behavior. Toddlers and **preschool** children very rapidly learn the effectiveness of such acting out. If parents or other caregivers acquiesce to the child's wishes each time to avoid the occurrence of a tantrum, it quickly becomes obvious to the child that this is the most successful means of getting his or her wishes fulfilled. Acting out quickly becomes a time-honored response to the word "No."

More critical negative behaviors including aggressive or abusive actions toward other children, animals, adults, or even themselves are usually a more serious and longer-lasting form of acting out. These are usually related to more momentous causes including mental illness or pathological conditions in the child's life, either in the home or in some other facet of the child's environment. Such negative childhood conduct is often seen in children who have been the victims of physical and/or sexual abuse or of severe neglect. Such acting out for this group of children is often referred to among mental health professionals as "a cry for help." Though certainly significant problems, acting out in the form of various rebellious behaviors that are not self-injurious or life-threatening is considered the less serious form of this "cry for help." These actions include disobeying parents and teachers, non-life-threatening alcohol or drug use, promiscuity, and exercising poor judgment in relationships and activities.

Common problems

High-profile crimes such as the Columbine High School shootings have made most people much more acutely aware of the potential danger involved in young people acting out in a highly antisocial manner. In 2000, the National Institute of Mental Health (NIMH) began to study child and adolescent violence in the United States. One of NIMH's initial findings indicates that though youth violence is indeed a serious problem, events such as Columbine are a rare occurrence. On average across the United States, every day six to seven young people are murdered by their peers. The overwhelming majority of these homicides occur within the confines of inner cities, and the average victim is a member of a minority group. These teenagers' deaths for the most part do not occur on school grounds. Moreover, many factors other than those that cause acting out may contribute to these crimes.

The NIMH found in its research that causes for serious acting out include the following:

- weak **bonding** with parents, caused by parents being physically or emotionally unavailable to the child
- impotent parenting as manifested by failing to watch over children, by being excessively strict, or by providing harsh and inconsistent discipline
- a home environment that exposes children to violence and supports and models aggressive and violent behaviors
- the impact of rejection by or competition with peers in early school years (In some cases this experience results in children who do not succeed socially or academically banding together to act out. This tendency to band together appeared to be true of the Columbine High School perpetrators.)
- gender (From approximately the age of four years, boys were found by the NIMH study to be more likely

than girls to engage in aggressive, acting out behaviors.)

- child psychopathology (The NIMH study suggests that children with behavioral difficulties are likely to have two or more psychological problems such as conduct or anxiety disorders or depression.)
- lower socioeconomic status (A correlation between low **family** income and antisocial acting out has been repeatedly noted.)
- heredity (Perhaps the most surprising factor of the NIMH study has been the possibility that genes may indeed influence behaviors. Exactly how genetics affects personality and mental illness is not clearly understood. How the environment interacts with this genetic component also remains a mystery. However, in 2004 this genetic ingredient in acting out behaviors was a topic of study for the NIMH.)

A 2001 Carleton University (Ottawa, Canada) study published in the *Journal of Research in Childhood Education* attempted to look at means of predicting which preschool children would be more apt to act out or experience academic or social difficulties in school. The study entitled "Solitary-Active **Play** Behavior: A Marker Variable for Maladjustment in the Preschool?" concluded that young children who play alone in a very active and boisterous manner were more likely to have adjustment problems in school.

Problems identified for this group of children include the following:

- exhibiting a lower attention span
- being more difficult to comfort
- being more timid and bashful
- showing more aggression and acting out
- doing poorer on academic testing for early skills
- showing a less positive response to being in school
- showing less ability to form positive relationships with other children

The Carleton study asserted two points: first, their research gave no evidence that these children had less learning ability than other children, but rather that they simply experienced problems with adjusting to school; second, it is normal for preschool age children to engage in solitary play. However it is the mode in which a child plays alone—in an overly boisterous, hyperactive manner—that seems to provide the marker for potential acting out problems later. The Canadian researchers undertook this study not to merely identify future acting out children, but to find those children who could benefit from specialized and comprehensive educational programs that address both academic and social needs.

Parental concerns

When to call the doctor

The parent whose child exhibits the negative behaviors called "acting out" probably wonders what is normal and what is not. Well-meaning friends and family may assure them that the child "will grow out of it" when the acting out behaviors are clearly not a normal part of growth and development. Some people consider acting out behaviors as simply part of the learning process for young children not requiring professional help. The results of appropriate parental intervention may suggest the real severity of the behavior. For example, the child who ceases having temper tantrums once he or she realizes that the tantrum will not get him or her the desired result has both identified the severity of the problem and resolved the identified problem for their parents.

Temper tantrums after the age of four or in children younger than four when very frequent and/or prolonged (that is, lasting longer than a half hour) should be evaluated by a healthcare professional as there may be a other medical or psychological causes. Any type of acting out behavior that can be termed unsafe, damaging to others, or self-injurious will probably need to be evaluated by the child's healthcare provider.

Among the forms of acting out behavior seen in children and teens that warrant professional attention are the following:

- pathological lying
- bullying others
- self-injury, such as cutting self or head-banging
- alcohol or drug abuse
- truancy
- **running away**
- participating in unsafe sexual activities
- getting into fights
- assault
- vandalism
- fire-setting
- stealing
- rape
- homicide

Coping with acting out in toddlers

For toddlers, most childcare professionals recommend that parents make it obvious that temper tantrums are not an appropriate way to handle disappointment. Giving a "time out," having the child go to his or her room or another quiet area for a set period of time or until able to interact in a socially acceptable manner, is an effective means of dealing with this form of acting out. Though small children often do not appear able to hear or comprehend reason, it is perfectly appropriate for parents to note that they understand the child's disappointment or frustration but that the child's negative behavior will not alter the situation. Some parents combine time outs with the message, "Deal with it." As time has gone on, the time outs become less necessary, and the direction "Deal with it" is enough for the child to regain self-control. The child learns that people do not always get what they want.

A research program under the auspices of NIMH has as of 2004 two decades of experience in the prevention of serious childhood acting out. The Nurse Home Visitation Program operates in Colorado, New York, and Tennessee. Nurses visit high-risk families beginning during a pregnancy and continuing through the child's second birthday. The selected families are considered at-risk because they have low income and/or a single parent. The goals of the visits are to improve the outcome in **childbirth**, promote the child's health and development, and aid in increasing the family's financial self-sufficiency. Follow-up of these children to the age of 15 show them to have fewer behavioral problems than comparable 15-year-olds without the service. Behavioral problems studied included use of drugs and alcohol, running away, sexual acting out, and arrests and convictions for crimes.

Coping with acting out in preschool and school age children

The Administration on Children, Youth, and Families (ACYF) has collaborated with NIMH in developing several **assessment** tools to identify children at risk for behavioral problems. Many of these tools are designed to pinpoint even preschool children at risk, and are provided within such programs as Head Start. It is now known that 70 to 80 percent of all children provided services for mental health problems in United States schools have these services delivered by the school system itself, by school guidance counselors and psychologists. Several recent initiatives by the NIMH have as their goal working with the child, classmates, parents, and teachers to reduce disruptive behavior. The Families and Schools Together (FAST) Track Program currently operates in North Carolina, Pennsylvania, Tennessee, and Washington. FAST Track has studied aggressive children from the age of six on for several years now, providing intervention as necessary. Follow-up studies have shown that those FAST Track Program children who received intervention required less **special educa-**

KEY TERMS

Antisocial—Actions described as impulsively aggressive, sometimes violent, that do not comply with established social and ethical codes.

Anxiety disorder—A mental disorder characterized by prolonged, excessive worry about circumstances in one's life. Anxiety disorders include agoraphobia and other phobias, obsessive-compulsive disorder, post-traumatic stress disorder, and panic disorder.

Attention deficit hyperactivity disorder (ADHD)—A condition in which a person (usually a child) has an unusually high activity level and a short attention span. People with the disorder may act impulsively and may have learning and behavioral problems.

Conduct disorder—A behavioral and emotional disorder of childhood and adolescence. Children with a conduct disorder act inappropriately, infringe on the rights of others, and violate societal norms.

Delinquent—A term applied to young people who behave in a manner in defiance of established social and ethical codes.

Depression—A mental condition in which a person feels extremely sad and loses interest in life. A person with depression may also have sleep problems and loss of appetite and may have trouble concentrating and carrying out everyday activities.

Psychodrama—A specific form of role play that focuses on acting out "scripts" of unresolved issues within the family, or helping family members adopt new approaches and understanding of one another.

Psychopathology—The study of mental disorders or illnesses, such as schizophrenia, personality disorder, or major depressive disorder.

Psychotherapy—Psychological counseling that seeks to determine the underlying causes of a patient's depression. The form of this counseling may be cognitive/behavioral, interpersonal, or psychodynamic.

tion services by grade 3 than children that did not participate in the program.

The Coping with Stress Course is a group educational program as of 2004 provided to adolescents in Maryland, Ohio, and Oregon. Its purpose is help young people develop strategies for coping positively with their negative thinking, tendencies toward depression, and acting out behaviors. Initial results from this course have shown successful outcomes for course participants. Among teens who did not take the course, symptoms of depression were reported twice as often. Adolescents taking the Coping with Stress Course showed a reduction in depressive symptoms and an increase in overall positive adjustment. However, it appears that time reduced the potency of this learning experience. Research over a longer period showed less difference between the two groups.

The NIMH study of acting out concluded that it is a multi-faceted problem involving the interactions between the child and his or her family, friends, classmates, school, and community. Children who have a warm, loving, and supportive childhood are far less likely to act out as a cry for help or to act out in a violent manner. Two types of teen **antisocial behavior** have been identified by NIMH: life course persistent and **adolescence** limited. Teens with life course persistent behaviors act out in violent ways from early childhood on, in a variety of situations. They are considered to have psychopathology, often including attention deficit hyperactivity disorder (ADHD). They usually continue with negative behaviors into adulthood. Those young people with adolescence limited behaviors typically act out in specific defined social situations and usually stop acting out behaviors before reaching adulthood.

See also Antisocial behavior.

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Adaptive behavior scales for infants and early childhood

Definition

Adaptive behavior scales are standardized tests used to describe and evaluate the behavior of infants, toddlers, and preschoolers, especially those at risk for communication delays and behavior impairments.

Purpose

Adaptive behaviors are learned. They involve the ability to adapt to and manage one's surroundings to effectively function and meet social or community expectations. Infants learn to walk, to talk, and to eat with a spoon. Older children learn to cross the street, to go to the store, and to follow a great variety of rules while interacting with people, such as when to say please and thank you. Good adaptive behavior promotes independence at home, at school, and in the community. Undesirable or socially unacceptable behaviors that interfere with the acquisition of desired skills and with the performance of everyday activities are classified as maladaptive behaviors, or more commonly, behavior problems. Maladaptive behavior interferes with child's achievement of independence because the child requires more supervision and assistance in order to learn how to behave appropriately.

Problems in developing adaptive skills can occur in children of any age. For example, difficulties can develop in mastering basic functional skills (such as talking, walking, or toileting), in learning academic skills and concepts, or in making social and vocational adjustments. Adaptive behavior scales are evaluation tools designed to help care providers improve their assessments of the abilities and needs of infants and children who have disabilities or are at risk for developmental delays.

Description

Many different adaptive behavior scales are used in the United States for **assessment** purposes. The most widely used are the Developmental Profile II (DPII), the Early Coping Inventory (ECI), the **Bayley Scales of Infant Development** (BSID), the Scales of Independent Behavior—Revised (SIB-R), the Vineland Adaptive Behavior Scales (VABS), and the Adaptive Behavior Scales (ABS).

Developmental Profile II (DPII)

The DP-II behavior scale is used to screen for developmental delays and compare a child's development to

that of other children who are in the same age group. This scale is a check-off list of 186 skills. A parent or therapist who knows the child well simply indicates whether the child has mastered the skill in question. The DPII, which can be administered from infancy to age nine, assesses development in the following areas:

- Physical development: Large and small muscle coordination, strength, stamina, flexibility, and sequential motor skills.
- Self-help development: Ability to cope independently with the environment, for example, to eat, dress, and take care of self and others.
- Social development: Interpersonal abilities, emotional needs, and how the child relates to friends, relatives, and other adults.
- Academic development: Intellectual abilities and skills required for academic achievement; IQ (intellectual quotient) score.
- Communication development: Expressive and receptive **communication skills**, including written, spoken, and body language.

Early Coping Inventory (ECI)

The ECI measures adaptive behavior. It is based on observation and is used to assess the coping-related behaviors that are used by infants and toddlers in everyday living. Analysis of a child's scores provides information about level of effectiveness, coping style, and specific coping strengths and weaknesses. The findings can then be used to plan educational and therapeutic interventions. The ECI can also be used to involve parents in its use as a means of increasing knowledge of the child. The ECI, which can be administered to infants aged four to 36 months or to children with disabilities, has 48 test items that are divided into three broad coping clusters:

- Sensorimotor organization: According to the famous developmental psychologist, Jean Piaget, infants learn, from birth to approximately age two, to coordinate all their sensory experiences (sights, sounds, etc.) with their motor behaviors. At this stage of development, children start to explore and understand the world around them by doing things like sucking, grasping, and **crawling**. This part of the ECI tests the child's level of sensorimotor skills: visual attention, reaction to touch, self-regulation of basic body functions, tolerance for various body positions, and activity level depending on various situations.
- Reactive behavior: This behavior includes a child's capacity to accept emotional warmth and support from other people and to react to the feelings and moods of others. The ECI can assess reactive behavior, including tolerance of frustration, ability to "bounce back" after stressful events, and capacity to adapt to changes in the environment.
- Self-initiated behavior: This part of the ECI tests the ability of a child to initiate action in order to communicate needs, to try new behaviors, to achieve a goal, as well as problem-solving abilities and level of persistence during activities.

Bayley Scales of Infant Development (BSID)

The BSID are used extensively to assess the development of infants from one to three years of age. The test is given on an individual basis and takes from 45 to 60 minutes to complete. It is administered by examiners who are experienced clinicians specifically trained in BSID test procedures. The examiner presents a series of test materials to the child and observes the child's responses and behaviors. The test also contains items designed to identify young children at risk for **developmental delay**. BSID evaluates three scales:

- Mental scale: This part of the evaluation assesses several types of abilities: sensory/perceptual acuities, discriminations, and response; memory-learning and problem-solving; vocalization and range of verbal communication; basis of abstract thinking; development of habits.
- Motor scale: This part of the BSID assesses the degree of body control, large muscle coordination, finer manipulatory skills of the hands and fingers, dynamic movement, postural imitation, and the ability to recognize objects by sense of touch (stereognosis).
- Behavior rating scale: This scale provides information that can be used to supplement information gained from the mental and motor scales. This 30-item scale rates the child's relevant behaviors and measures attention/arousal, orientation/engagement, emotional regulation, and motor quality.

Scales of Independent Behavior—Revised (SIB-R)

Children with developmental disabilities or who become handicapped through accident or illness often need special assistance at home and at school. The SIB-R assesses adaptive and maladaptive behavior to determine the type and amount of special assistance that children with disabilities may need. The SIB-R is widely used in **preschool** and **special education** programs for diagnosis, for intervention planning, and for assessing outcomes. The SIB-R evaluation can be completed by a teacher, psychologist, or social worker directly or with the help of special interview materials that involve par-

KEY TERMS

Adaptive behavior—The ability to do things on one's own without getting into trouble and to adapt to and manage one's surroundings.

Asperger syndrome—A developmental disorder of childhood characterized by autistic behavior but without the same difficulties acquiring language that children with autism have.

Autism—A developmental disability that appears early in life, in which normal brain development is disrupted and social and communication skills are retarded, sometimes severely.

Behavior—A stereotyped motor response to an internal or external stimulus.

Body language—Communication without words, also sometimes referred to as "non-verbal communication"; conscious or unconscious bodily movements and gestures that communicate to others a person's attitudes and feelings.

Fine motor skill—The abilities required to control the smaller muscles of the body for writing, playing an instrument, artistic expression and craft work. The muscles required to perform fine motor skills are generally found in the hands, feet and head.

Gross motor skills—The abilities required to control the large muscles of the body for walking, running, sitting, crawling, and other activities. The muscles required to perform gross motor skills are generally found in the arms, legs, back, abdomen and torso.

Maladaptive behavior—Undesirable and socially unacceptable behavior that interferes with the acquisition of desired skills or knowledge and with the performance of everyday activities.

Motor skills—Controlled movements of muscle groups. Fine motor skills involve tasks that require dexterity of small muscles, such as buttoning a shirt. Tasks such as walking or throwing a ball involve the use of gross motor skills.

Sensorimotor—Relating to the combination of sensory and motor coordination.

Sensory—Refers to network of nerves that transmit information from the senses to the brain.

Sequential motor skill—Ability to coordinate different motor skills in sequence, such as running followed by a jump.

Socialization—The process by which new members of a social group are integrated in the group.

Stereognosis—The ability to recognize objects by sense of touch.

ents. The SIB-R contents provide opportunity for team discussion, often eliciting information and opinions that parents might not otherwise bring up on their own.

The SIB-R adaptive behavior items include 14 subscales grouped into four clusters: motor skills, social interaction and communication skills, personal living skills, and community living skills. Each SIB-R adaptive behavior item is a statement of a task. (For example, "Child washes, rinses, and dries hair.") The examiner rates the child being assessed regarding each task, using a scale from zero to three:

- 0—never or rarely performs the task (even if asked)
- 1—does the task but not well or about 25 percent of the time (may need to be asked)
- 2—does the task fairly well or about 75 percent of the time (may need to be asked)
- 3—does the task very well always or almost always (without being asked)

The Vineland Adaptive Behavior Scales (VABS)

The VABS are designed to assess the personal and social self-sufficiency of individuals from birth to early adulthood. The scales are equally applicable to handicapped and non-handicapped children. The VABS assessment provides the information required for the diagnosis or evaluation of a wide range of disabilities, including **mental retardation**, developmental delays, functional skills impairment, and speech/language impairment. Vineland has also been proven to be an accurate resource for predicting **autism** and Asperger syndrome, among other diagnoses. The Vineland measures adaptive behavior in four domains:

- Communication: Vineland evaluates expressive and written communication skills, as well as the ability to listen.
- Daily living skills: These skills are assessed on a personal basis, in the **family** setting and in the wider community.

- Socialization: VABS evaluate interpersonal relationships, **play** and leisure time activities, and interpersonal coping skills.
- Motor skills: The test also evaluates both gross and fine motor skills.

The Adaptive Behavior Scales—School (ABS-S)

The ABS-S scale was developed to assess the personal independence of school-age children. Like other scales, it evaluates the personal and social skills used for everyday living. It is most frequently used to assess the current functioning of children being evaluated for evidence of mental retardation, for evaluating adaptive characteristics in autistic children, and for distinguishing behavior-disordered children who require special education assistance from those who can be educated in a regular school setting. The ABS-S is divided into two parts:

- ABS Part One: This part focuses on personal independence and is designed to evaluate coping skills considered important for developing personal independence and responsibility in daily living. The skills within Part One are grouped into nine behavior domains: independent functioning, physical development, economic activity, **language development**, numbers and time, prevocational/vocational activity, self-direction, responsibility, and socialization.
- ABS Part Two: This part evaluates social maladaptation. The behaviors assessed are assigned to seven domains that are measures of those adaptive behaviors that may lead to personality and behavior disorders: social behavior, conformity, trustworthiness, stereotyped and hyperactive behavior, self-abusive behavior, social engagement, and disturbing interpersonal behavior.

Precautions

Since behavior is socially defined, a child's performance must be considered within the context of the cultural environments and social expectations that affect his or her functioning.

Preparation

Before performing an adaptive behavior test, the examiner explains to the parents the purpose of the test. If the parents are required to provide the answers, they are reminded that accuracy is the best way to achieve a result that may help the child. If the test is given directly to the child, the examiner describes what will happen during the test procedure and the parents are asked not to talk to the child during the test to avoid skewing results.

Risks

There are no risks associated with adaptive behavior tests.

Parental concerns

Parental involvement in the developmental assessment of their children is very important. First, because parents are more familiar with their child's behavior, their assessment may indeed be more indicative of the child's developmental status than an assessment that is based on limited observation in an unfamiliar clinical setting. The involvement of parents in their child's development testing also improves their knowledge of child development issues and their subsequent participation in required intervention programs, if any.

See also Autism; Bayley Scales of Infant Development; Emotional development; Personality development; Personality disorders.

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American Academy of Pediatrics (AAP). 141 Northwest Point Boulevard, Elk Grove Village, IL 60007–1098. Web site: <www.aap.org>.

American Psychological Association (APA). 750 First Street, NE, Washington, DC 20002–4242. Web site: <www.apa.org>.

Child Development Institute (CDI). 3528 E Ridgeway Road, Orange, CA 92867. Web site: <www.childdevelopmentinfo.com>.

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ADD *see* **Attention deficit/Hyperactivity disorder**

Addiction

Definition

Addiction is a physical or mental dependence on a behavior or substance that a person feels powerless to stop.

Description

Addiction is one of the most costly public health problems in the United States. It is a progressive syndrome, which means that it increases in severity over time unless it is treated. The term has been partially replaced by the word "dependence" for substance abuse. Addiction has been extended, however, to include mood-altering behaviors or activities. Some researchers speak of two types of addictions: substance addictions (for example, **alcoholism**, drug abuse, and **smoking**); and process addictions (for example, gambling, spending, shopping, eating, and sexual activity). There was as of 2004 a growing recognition that many addicts are addicted to more than one substance or process. Substance abuse is characterized by frequent relapse or return to the abused substance. Substance abusers often make repeated attempts to quit before they are successful.

The National Survey on Drug Use and Health (NSDUH) is conducted annually by the Substance Abuse and Mental Health Services Administration (SAMHSA) of the U.S. Department of Health and Human Services. Among the findings of the 2003 study are the following:

- In 2003, an estimated 19.5 million Americans, or 8.2 percent of the population aged 12 or older, were current illicit drug users. Current illicit drug use means use of an illicit drug during the month prior to the survey interview. The numbers did not change from 2002.
- The rate of illicit drug use among youths aged 12–17 did not change significantly between 2002 (11.6%) and 2003 (11.2%), and there were no changes for any specific drug. The rate of current marijuana use among youths was 8.2 percent in 2002 and 7.9 percent in 2003. There was a significant decline in lifetime marijuana use among youths, from 20.6 percent in 2002 to 19.6 percent in 2003. There also were decreases in rates of past year use of LSD (1.3 to 0.6%), ecstasy (2.2 to 1.3%), and methamphetamine (0.9 to 0.7%).
- About 10.9 million persons aged 12–20 reported drinking alcohol in the month prior to the survey interview in 2003 (29.0 percent of this age group). Nearly 7.2 million (19.2%) were binge drinkers and 2.3 million (6.1%) were heavy drinkers. The 2003 rates were essentially the same as those from the 2002 survey.
- An estimated 70.8 million Americans reported current (past month) use of a tobacco product in 2003. This is 29.8 percent of the population aged 12 or older, similar to the rate in 2002 (30.4%). Young adults aged 18–25 reported the highest rate of past month cigarette use (40.2%), similar to the rate among young adults in 2002. An estimated 35.7 million Americans aged 12 or older in 2003 were classified as nicotine dependent in the past month because of their cigarette use (15% of the total population), about the same as for 2002.

Demographics

In 2003, the rate of substance dependence or abuse was 8.9 percent for youths aged 12–17 and 21 percent for persons aged 18–25. Among persons with substance dependence or abuse, illicit drugs accounted for 58.1

percent of youths and 37.2 percent of persons aged 18–25. In 2003, males were almost twice as likely to be classified with substance dependence or abuse as females (12.2% versus 6.2%). Among youths aged 12–17, however, the rate of substance dependence or abuse among females (9.1%) was similar to the rate among males (8.7%). The rate of substance dependence or abuse was highest among Native Americans and Alaska Natives (17.2%). The next highest rates were among Native Hawaiians and other Pacific Islanders (12.9%) and persons reporting mixed ethnicity (11.3%). Asian Americans had the lowest rate (6.3%). The rates among Hispanics (9.8%) and whites (9.2%) were higher than the rate among blacks (8.1%).

Rates of drug use showed substantial variation by age. For example, in 2003, some 3.8 percent of youths aged 12 to 13 reported current illicit drug use compared with 10.9 percent of youths aged 14 to 15 and 19.2 percent of youths aged 16 or 17. As in other years, illicit drug use in 2003 tended to increase with age among young persons, peaking among 18 to 20-year-olds (23.3%) and declining steadily after that point with increasing age. The prevalence of current alcohol use among adolescents in 2003 increased with increasing age, from 2.9 percent at age 12 to a peak of about 70 percent for persons 21 to 22 years old. The highest prevalence of both binge and heavy drinking was for young adults aged 18 to 25, with the peak rate of both measures occurring at age 21. The rate of binge drinking was 41.6 percent for young adults aged 18 to 25 and 47.8 percent at age 21. Heavy alcohol use was reported by 15.1 percent of persons aged 18 to 25 and 18.7 percent of persons aged 21. Among youths aged 12 to 17, an estimated 17.7 percent used alcohol in the month prior to the survey interview. Of all youths, 10.6 percent were binge drinkers, and 2.6 percent were heavy drinkers, similar to the 2002 numbers.

Rates of current illicit drug use varied significantly among the major racial-ethnic groups in 2003. The rate of illicit drug use was highest among Native Americans and Alaska Natives (12.1%), persons reporting two or more races (12%), and Native Hawaiians and other Pacific Islanders (11.1%). Rates were 8.7 percent for African Americans, 8.3 percent for Caucasians, and 8 percent for Hispanics. Asian Americans had the lowest rate of current illicit drug use at 3.8 percent. The rates were unchanged from 2002. Native Americans and Alaska Natives were more likely than any other racial-ethnic group to report the use of tobacco products in 2003. Among persons aged 12 or older, 41.8 percent of Native Americans and Alaska Natives reported using at least one tobacco product in the past month. The lowest current tobacco use rate among racial-ethnic groups in 2003 was observed for Asian Americans (13.8%), a decrease from the 2002 rate (18.6%).

Young adults aged 18 to 25 had the highest rate of current use of cigarettes (40.2%), similar to the rate in 2002. Past month cigarette use rates among youths in 2002 and 2003 were 13 percent and 12.2 percent, respectively, not a statistically significant change. However, there were significant declines in past year (from 20.3% to 19%) and lifetime (from 33.3% to 31%) cigarette use among youths aged 12 to 17 between 2002 and 2003. Among persons aged 12 or older, a higher proportion of males than females smoked cigarettes in the past month in 2003 (28.1% versus 23%). Among youths aged 12 to 17, however, girls (12.5%) were as likely as boys (11.9%) to smoke in the past month. There was no change in cigarette use among boys aged 12 to 17 between 2002 and 2003. However, among girls, cigarette use decreased from 13.6 percent in 2002 to 12.5 percent in 2003.

Causes and symptoms

Addiction to substances results from the interaction of several factors.

Drug chemistry

Some substances are more addictive than others, either because they produce a rapid and intense change in mood or because they produce painful withdrawal symptoms when stopped suddenly.

Genetics

Some people appear to be more vulnerable to addiction because their body chemistry increases their sensitivity to drugs. Some forms of **substance abuse and dependence** seem to run in families; a correlation that may be the result of a genetic predisposition, environmental influences, or a combination of the two.

Brain structure and function

Using drugs repeatedly over time changes brain structure and function in fundamental and long-lasting ways. Addiction comes about through an array of changes in the brain and the strengthening of new memory connections. Evidence suggests that those long-lasting brain changes are responsible for the distortions of cognitive and emotional functioning that characterize addicts, particularly the compulsion to use drugs. Although the causes of addiction remain the subject of ongoing debate and research, many experts as of 2004 considered addiction to be a brain disease, a condition caused by persistent changes in brain structure and function. However, having this brain disease does not absolve

the addict of responsibility for his or her behavior, but it does explain why many addicts cannot stop using drugs by sheer force of will alone.

Social learning

Social learning is considered the most important single factor in causing addiction. It includes patterns of use in the addict's **family** or subculture, **peer pressure**, and advertising or media influence.

Availability

Inexpensive or readily available tobacco, alcohol, or drugs produce marked increases in rates of addiction. Increases in state taxes on alcohol and tobacco products have not resulted in decreased use.

Personality

Before the 1980s, the so-called addictive personality was used to explain the development of addiction. The addictive personality was described as escapist, impulsive, dependent, devious, manipulative, and self-centered. Many doctors in the early 2000s believe that these character traits develop in addicts as a result of the addiction, rather than the traits being a cause of the addiction.

When to call the doctor

The earlier one seeks help for their teen's behavioral or drug problems, the better. How is a parent to know if their teen is experimenting with or moving more deeply into the drug culture? Above all, a parent must be a careful observer, particularly of the little details that make up a teen's life. Overall signs of dramatic change in appearance, friends, or physical health may signal trouble. If parents believe their child may be drinking or using drugs, they should seek help through a substance abuse recovery program, family physician, or mental health professional.

Diagnosis

In addition to noting a preoccupation with using and acquiring the abused substance, the diagnosis of addiction focuses on five criteria:

- loss of willpower
- harmful consequences
- unmanageable lifestyle
- increased tolerance or escalation of use
- withdrawal symptoms on quitting

Treatment

According to the American Psychiatric Association, there are three goals for the treatment of persons with substance use disorders: (1) the patient abstains from or reduces the use and effects of the substance; (2) the patient reduces the frequency and severity of relapses; and (3) the patient develops the psychological and emotional skills necessary to restore and maintain personal, occupational, and social functioning.

In general, before treatment can begin, many treatment centers require that the patient undergo detoxification. Detoxification is the process of weaning the patient from his or her regular substance use. Detoxification can be accomplished "cold turkey," by complete and immediate cessation of all substance use, or by slowly decreasing (tapering) the dose that a person is taking, to minimize the side effects of withdrawal. Some substances must be tapered because cold-turkey methods of detoxification are potentially life threatening. In some cases, medications may be used to combat the unpleasant and threatening physical and psychological symptoms of withdrawal. For example, methadone is used to help patients adjust to the tapering of heroin use.

The most frequently recommended social form of outpatient treatment is the 12-step program. Such programs are also frequently combined with psychotherapy. According to the American Psychological Association (APA), anyone, regardless of his or her religious beliefs or lack of religious beliefs, can benefit from participation in 12-step programs such as Alcoholics Anonymous (AA) or Narcotics Anonymous (NA). The number of visits to 12-step self-help groups exceeds the number of visits to all mental health professionals combined. There are 12-step groups for all major substance and process addictions.

Alternative treatment

Acupuncture and homeopathy have been used to treat withdrawal symptoms. Meditation, **yoga**, and reiki healing have been recommended for process addictions; however, the success of these programs has not been well documented through controlled studies.

Prognosis

The prognosis for recovery from any addiction depends on the substance or process, the individual's circumstances, and underlying personality structure. People who have multiple substance dependencies have the worst prognosis for recovery. It is not uncommon for someone in a treatment program to have a relapse, but the success rate increases with subsequent treatment programs.

Teenager smoking crystal methamphetamine, known to cause a very strong psychological addiction to the drug. *(© Houston Scott/Corbis Sygma.)*

Recovery from substance use is notoriously difficult, even with exceptional treatment resources. Although relapse rates are difficult to accurately obtain, the National Institute on Alcohol Abuse and Alcoholism cites evidence that 90 percent of alcohol dependent users experience at least one relapse within four years after treatment. Relapse rates for heroin and nicotine users are believed to be similar. Certain pharmacological treatments, however, have been shown to reduce relapse rates. Relapses are most likely to occur within the first 12 months of having discontinued substance use. Triggers for relapses can include any number of life stresses (problems in school or on the job, loss of a relationship, death of a loved one, financial stresses), in addition to seemingly mundane exposure to a place or an acquaintance associated with previous substance use.

Prevention

The most effective form of prevention appears to be a stable family that models responsible attitudes toward mood-altering substances and behaviors. Prevention education programs are also widely used to inform young people of the harmfulness of substance abuse.

KEY TERMS

Binge drinking—Consumption of five or more alcoholic drinks in a row on a single occasion.

Detoxification—The process of physically eliminating drugs and/or alcohol from the system of a substance-dependent individual.

Reiki—A form of energy therapy that originated in Japan. Reiki practitioners hold their hands on or slightly above specific points on the patient's body in order to convey universal life energy to that area for healing.

Parental concerns

Parents and guardians need to be aware of the power they have to influence the development of their kids throughout the teenage years. **Adolescence** brings a new and dramatic stage to family life. The changes that are required are not just the teen's to make; parents need to change their relationship with their teenager. It is best if parents are proactive about the challenges of this life stage, particularly those that pertain to the possibility of experimenting with and using alcohol and other drugs. Parents should not be afraid to talk directly to their kids about drug use, even if they have had problems with drugs or alcohol themselves. Parents should give clear, no-use messages about smoking, drugs, and alcohol. It is important for kids and teens to understand that the rules and expectations set by parents are based on parental love and concern for their well being. Parents should also be actively involved and demonstrate interest in their teen's friends and social activities. Spending quality time with teens and setting good examples are essential. Even if problems such as substance abuse already exist in the teen's life, parents and families can still have a positive influence on their teen's behavior.

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Adenoid hyperplasia

Definition

Adenoid hyperplasia is an enlargement of the lymph glands located above the back of the mouth.

Description

Located at the back of the mouth above and below the soft palate are two pairs of lymph glands. The tonsils below are clearly visible behind the back teeth; the adenoids lie just above them and are hidden from view by the palate. Together these four arsenals of immune defense guard the major entrance to the body from foreign invaders, the germs we breathe and eat. In contrast to the rest of the body's tissues, lymphoid tissue reaches its greatest size in mid-childhood and recedes thereafter. In this way children are best able to develop the immunities they need to survive in a world full of infectious diseases.

Beyond its normal growth pattern, lymphoid tissue grows excessively (hypertrophies) during an acute infection, as it suddenly increases its immune activity to fight off the invaders. Often it does not completely return to its former size. Each subsequent infection leaves behind a larger set of tonsils and adenoids. To make matters worse, the sponge-like structure of these hypertrophied glands can produce safe havens for germs where the body cannot reach and eliminate them. Before **antibiotics** and the reduction in infectious childhood diseases over the last few generations of the twentieth century, tonsils and adenoids caused even greater health problems.

Demographics

The true incidence of adenoid hyperplasia is difficult to assess. What is clear, however, is that tonsillectomy and adenoidectomy (T and A), the surgical treatment for the condition, is the most frequently performed major surgical procedure in the United States. Information current in 2004 on the exact number of these procedures performed was difficult to obtain because they are routinely performed in outpatient settings. Adenoid hypertrophy does not appear to affect any gender or racial group more than another.

Causes and symptoms

Most tonsil and adenoid hypertrophy is simply caused by the normal growth pattern for that type of tissue. Less often, the hypertrophy is due to repeated throat infections by cold viruses, **strep throat**, mononucleosis, and in the past, **diphtheria**. The acute infections are usually referred to as **tonsillitis**, the adenoids getting little recognition because they cannot be seen without special instruments. Symptoms include painful, bright red, often ulcerated tonsils, enlargement of lymph nodes (glands) beneath the jaw, **fever**, and general discomfort.

After the acute infection subsides, symptoms are generated simply by the size of the glands. Extremely large tonsils can impair breathing and swallowing, although that is quite rare. Large adenoids can impair nose breathing and require a child to breathe through the

mouth. Because they encircle the only connection between the middle ear and the eustachian tube, hypertrophied adenoids can also obstruct the tube and cause middle ear infections.

Diagnosis

A simple depression of the tongue allows an adequate view of the tonsils. Enlarged tonsils may have deep pockets (crypts) containing dead tissue (necrotic debris). Viewing adenoids requires a small mirror or fiberoptic scope. A child with recurring middle ear infections may well have large adenoids. A **throat culture** or mononucleosis test usually reveals the identity of the germ.

Treatment

It used to be standard practice to remove tonsils and/or adenoids after a few episodes of acute throat or ear infection. The surgery is called tonsillectomy and adenoidectomy (T and A). Medical opinions changed as it was realized that this tissue is beneficial to the development of immunity. For instance, children without tonsils and adenoids produce only half the immunity to oral **polio vaccine**. In addition, treatment of ear and throat infections with antibiotics and of recurring ear infections with surgical drainage through the eardrum (tympanostomy) has greatly reduced the incidence of surgical removal of these lymph glands.

Alternative treatment

There are many botanical/herbal remedies that can be used alone or in formulas to locally assist the tonsils and adenoids in their immune function at the opening of the oral cavity and to tone these glands. Keeping the eustachian tubes open is an important contribution to optimal function in the tonsils and adenoids. **Food allergies** are often the culprits for recurring ear infections, as well as tonsillitis and adenoiditis. Identification and removal of the allergic food(s) can greatly assist in alleviating the cause of the problem. Acute tonsillitis also benefits from warm saline gargles.

Prognosis

Hypertrophied adenoids are a normal part of growing up and should be respected for their important role in the development of immunity. Only when their size causes problems by obstructing breathing or middle ear drainage do they demand intervention.

KEY TERMS

Eustachian tube—A thin tube between the middle ear and the pharnyx. Its purpose is to equalize pressure on either side of the ear drum.

Hyperplastic—Refers to an increase in the size of an organ or tissue due to an increase in the number of cells.

Hypertrophy—An increase in the size of a tissue or organ brought about by the enlargement of its cells rather than cell multiplication.

Strep throat—An infection of the throat caused by *Streptococcus* bacteria. Symptoms include sore throat, chills, fever, and swollen lymph nodes in the neck.

Ulcerated—Characterized by the formation of an ulcer.

Prevention

Prevention can be directed toward prompt evaluation and appropriate treatment of sore throats to prevent overgrowth of adenoid tissue. Avoiding other children with acute respiratory illness also reduces the spread of these common illnesses.

Parental concerns

Adenoid hypertrophy is a relatively common childhood condition. If a child has repeated ear infections, a physician, usually an ear, nose, and throat (ENT) specialist, will recommend treatment options. To alleviate the discomfort experienced by the child and to prevent secondary complication such as delayed speech that can occur if the child's hearing is compromised because of the accompanying ear infections, frequently a surgery called an adenoidectomy is performed.

When to call the doctor

Following an adenoidectomy, parents should call the doctor if any of the following occurs:

- unexpected bright red bleeding
- fever over 101°F (38°C)
- **pain** that is not relieved by pain medications

See also Tonsillitis.

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Adenovirus infections

Definition

Adenoviruses are small infectious agents that cause upper respiratory tract infections, **conjunctivitis**, and other infections in humans.

Description

Adenoviruses were discovered in 1953. By 2004 about 49 different types had been identified, and about half of those were believed to cause human diseases. Adenovirus infections can occur throughout the year, unlike the seasonality associated with other respiratory viruses.

In children, adenoviruses most often cause acute upper respiratory infections with **fever** and runny nose. Adenovirus types 1, 2, 3, 5, and 6 are responsible for most of these infections. Occasionally more serious lower respiratory diseases, such as **pneumonia** or **bronchitis**, may occur. Adenoviruses can also cause acute **diarrhea** in young children, characterized by fever and watery stools. This condition is caused by adenovirus types 40 and 41 and can last as long as two weeks.

As much as 51 percent of all hemorrhagic **cystitis** (inflammation of the bladder and of the tubes that carry urine to the bladder from the kidneys) in American and Japanese children can be attributed to adenovirus infection. A child who has hemorrhagic cystitis has bloody urine for about three days, and invisible traces of blood can be found in the urine a few days longer. The child will feel the urge to urinate frequently but find it difficult to do so, for about the same length of time.

Other illnesses associated with adenovirus include:

- **encephalitis** (inflammation of the brain) and other infections of the central nervous system (CNS)
- **gastroenteritis** (inflammation of the stomach and intestines), which sometimes leads to enlarged lymph nodes in the intestines and rarely intussusception
- acute pharyngoconjunctival fever (inflammation of the lining of the eye [conjunctivitis] with fever)
- acute mesenteric **lymphadenitis** (inflammation of lymph glands in the abdomen)
- chronic interstitial fibrosis (abnormal growth of connective tissue between cells)
- intussusception (a type of intestinal obstruction)
- pneumonia that does not respond to antibiotic therapy
- **whooping cough** syndrome when *Bordetella pertussis* (the bacterium that causes classic whooping cough) is not found

Transmission

Specific adenovirus infections can be traced to particular sources and produce distinctive symptoms. In general, however, adenovirus infection is transmitted by the following:

- inhaling airborne viruses
- getting the virus in the eyes by swimming in contaminated water, using contaminated eye solutions or instruments, wiping the eyes with contaminated towels, or rubbing the eyes with contaminated fingers
- not washing the hands after using the bathroom and then touching the mouth or eyes

Infections often occur in situations in which individuals are in close contact with one another, such as the military, cruise ships, or college dormitories. Outbreaks among children are frequently reported at boarding schools and summer camps.

Most children have been infected by at least one adenovirus by the time they reach school age. Most adults have acquired immunity to multiple adenovirus types due to infections they had as children.

Demographics

Adenoviruses are responsible for 3 to 5 percent of acute respiratory infections in children. Most adenovirus infections occur between the ages of six months and five years. The incidence of adenovirus infection does not appear to differ among males and females or individuals of different race.

Causes and symptoms

In one mode of adenovirus infection (called lytic infection because it destroys large numbers of cells), adenoviruses kill healthy cells and replicate up to 1 million new viruses per cell killed, of which 1 to 5 percent are infectious. People with this kind of infection feel sick. In chronic or latent infection, a much smaller number of viruses are released, and healthy cells can multiply more rapidly than they are destroyed. People who have this kind of infection do not exhibit symptoms.

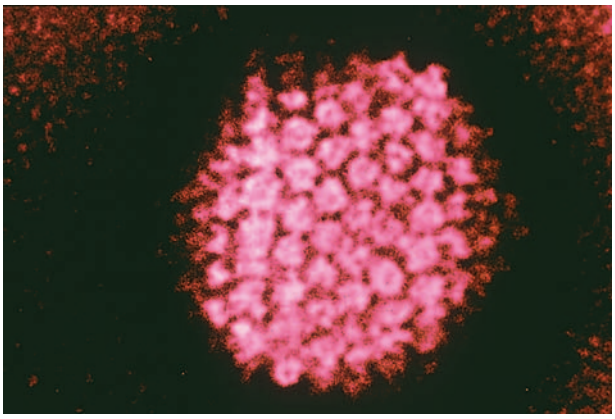
Children who have normal immune systems usually experience only minor symptoms when infected with adenovirus. The course of infection tends to be more serious in children who are immunocompromised, such as those undergoing **chemotherapy** or those who have a disease that disrupts normal immune response (e.g. human **immunodeficiency** syndrome [HIV]). In such children, the virus more often affects organs such as the lungs, liver, and kidneys, and the risk of fatality increases.

Symptoms common to respiratory illnesses caused by adenovirus infection include cough, fever, runny nose, **sore throat**, and watery eyes. In children with gastroenteritis caused by the adenovirus, symptoms may include diarrhea, fever, **nausea**, **vomiting**, and respiratory symptoms. Children with acute pharyngoconjunctival fever usually show signs of conjunctivitis, fever, sore throat, runny nose, and inflammation of the lymph glands in the neck (certical adenitis). More rarely, if the virus infects the lining of the brain and spinal cord (meninges) or the brain itself, **meningitis** or encephalitis may result; symptoms include fever, stiff or painful neck, irritability, changes in personality, or seizures.

When to call the doctor

Parents should contact a healthcare provider if the following applies to the infected child:

- The child is under three months of age.
- The child has symptoms that continue to worsen after one week.
- The child has difficulty breathing.
- The child shows symptoms of meningitis or encephalitis.
- The child has eye redness and swelling that becomes painful.
- The child shows signs of infection and is immunocompromised.



Magnification of an adenovirus. *(© Hans Gelderblom/Visuals Unlimited.)*

Diagnosis

Although symptoms may suggest the presence of adenovirus, distinguishing these infections from other viruses can be difficult. A definitive diagnosis is based on culture or detection of the virus in eye secretions, sputum, urine, or stool.

The extent of infection can be estimated from the results of blood tests that measure increases in the quantity of antibodies the immune system produces to fight it. Antibody levels begin to rise about a week after infection occurs and remain elevated for about a year.

Treatment

Treatment of adenovirus infections is usually supportive and aimed at relieving symptoms of the illness. Bed rest may be recommended along with medications to reduce fever and/or **pain**. (Aspirin should not be given to children because it is associated with Reye's syndrome.) Eye infections may benefit from topical corticosteroids to relieve symptoms and shorten the course of the disease. **Hospitalization** is usually required for severe pneumonia in infants and for keratoconjunctivitis (to prevent blindness). No effective **antiviral drugs** had been developed as of 2004.

Nutritional concerns

Because a child can become easily dehydrated if suffering from vomiting or diarrhea, it is important caregivers provide adequate fluid intake. Fluids such as water, breast milk or formula (if applicable), electrolyte replacement drinks, diluted juice, or clear broths should be encouraged. Drinks with **caffeine** should be avoided

KEY TERMS

Conjunctivitis—Inflammation of the conjunctiva, the mucous membrane covering the white part of the eye (sclera) and lining the inside of the eyelids also called pinkeye.

Reye's syndrome—A serious, life-threatening illness in children, usually developing after a bout of flu or chickenpox, and often associated with the use of aspirin. Symptoms include uncontrollable vomiting, often with lethargy, memory loss, disorientation, or delirium. Swelling of the brain may cause seizures, coma, and in severe cases, death.

Virus—A small infectious agent consisting of a core of genetic material (DNA or RNA) surrounded by a shell of protein. A virus needs a living cell to reproduce.

Whooping cough—An infectious disease of the respiratory tract caused by a bacterium, *Bordetella pertussis*. Also known as pertussis.

because of caffeine's diuretic effects (i.e. causes water to be lost through urine).

Prognosis

In otherwise healthy children, adenovirus infections are rarely fatal, and most patients recover fully. Immunocompromised children have a greater chance of serious side effects and death, with fatality rates as high as 50 to 69 percent (depending on the cause and extent of immunodeficiency).

Prevention

Practicing good personal hygiene and avoiding contact with people with infectious illnesses can reduce the risk of developing adenovirus infection. Proper hand washing can prevent the spread of the virus by oral-fecal transmission. Sterilization of instruments and solutions used in the eye can help prevent the spread of EKC, as can adequate chlorination of swimming pools.

A vaccine containing live adenovirus types 4 and 7 has been used to control disease in military recruits, but it is not recommended or available for civilian use. A recent resurgence of the adenovirus was found in a military population as soon as the **vaccination** program was halted. Vaccines prepared from purified subunits of adenovirus were as of 2004 under investigation.

Parental concerns

In the home setting, frequent hand washing should be encouraged, and children's **toys** and shared belongings should be frequently cleaned. Children who suffer from adenovirus infection should be kept home from school or daycare until they no longer show symptoms.

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ADHD *see* **Attention deficit/Hyperactivity disorder**

Adjustment disorders

Definition

Adjustment disorder is an umbrella term for several mental states characterized by noticeable behavioral and/or emotional symptoms. In order to be classified as an adjustment disorder, these symptoms must be shown to be a response to an identifiable stressor that has occurred within the past three months.

Description

The American Psychiatric Association (APA), in its *Diagnostic and Statistical Manual of Mental Disorders (DSM-IV)*, states that the behavioral and/or emotional signs observed must appear excessive for the stressor involved or have significant impact on the child's social and school functioning. The cause of the stress may be a single event affecting only the child, such as starting daycare or school, or an event that involves the entire **family**, such as a **divorce**. Multiple simultaneous stressors are also possible, such as starting daycare and having an abusive caretaker at the daycare or a divorce complicated by parental substance abuse. Chronic medical conditions of the child or parents, such as childhood leukemia or **cancer**, can also be a cause of stress.

Adjustment disorder, in some ways, is a hopeful diagnosis. Many mental health professionals consider it one of the less severe mental illnesses. It is normally a time-limited condition with manifestations arriving almost immediately after the appearance of the pressure-causing event and resolving within six months of the elimination of the stressor. However, the exception to this would be the duration of symptoms related to long-term stressors such as chronic illness or even the fall-out from divorce. Though these may appear within three months of the event, resolution may also take longer than six months.

Demographics

The diagnosis of adjustment disorder is a very common one for both children and teens, with a higher incidence among children than adults. Nearly one third (32%) of all adolescents are estimated to suffer from adjustment disorders during teenage years as opposed to a rate of occurrence of only 10 percent among adults. There is no identified difference between adjustment disorder rates between girls or boys. What provides the precipitating event and the symptoms manifested can vary, according to the culture in which a child lives. However, generally across all cultures, children and adolescents are more apt to experience **conduct disorder** symptoms manifested by **acting out** behaviors, while adults are more apt to experience depressive symptoms.

Causes and symptoms

Few descriptions of any mental illness specify its cause as precisely as the description of adjustment disorders does. An explicit incident or incidents causing stress for the child is always the precipitant. The cause of the stress seen in adjustment disorders can be events that for many children would be within the parameters of normal experience. These incidents are usually not the severe traumas associated with more serious stress-related illnesses such as post-traumatic stress disorder (PTSD). Though adjustment disorder precipitants are usually more "normal" events that can typically occur in the lives of most children, these events are still changes from everyday events. Especially for children, change is often the precursor of stress. For example, for a child who has always had daycare or **babysitters**, having caregivers other than his or her mother is a normal occurrence, so having a caregiver is not likely to be terribly stressful. However, a child who has never been separated from his or her mother may find going to daycare or kindergarten an extremely traumatic event.

Other examples of such childhood stressors include:

- divorce or separation of parents
- moving to a new place
- birth of a sibling
- natural disasters such as hurricanes or tornadoes
- illness of either the child or another loved one
- loss of a pet
- problems in school
- family conflict
- sexuality issues
- witnessing or being involved in an incidence of violence

Some psychological theorists and researchers consider adjustment disorders in adolescents less of an illness than a stage in establishing an identity. Adolescents may develop adjustment disorders as part of a defense mechanism meant to break their feelings of dependence on parents. This psychological maneuver may precipitate problems in families as adolescents begin seeking individuals outside the family as replacements for their parents. This behavior can be particularly destructive when these feelings of dependence are transferred to involvement with **gangs** or cults. However, it should be noted that the APA does classify adjustment disorder as a mental illness.

DSM-IV divides adjustment disorders into subgroups, based upon the symptoms manifested most prominently. These subgroups include:

- Adjustment disorder with depressed mood. This is characterized by feelings of sadness or hopelessness of varying degrees. However depression usually interferes with the child's ability to function, i.e. attending school or playing with friends. The sad feelings are sometimes accompanied by feelings of anger or frustration. It is important to note that though depressed mood adjustment disorder is less common among children, when it does occur, suicidal thoughts and even **suicide** attempts can be one of the symptoms. This symptom requires careful monitoring and the involvement of a mental health professional.
- Adjustment disorder with **anxiety**. This form typically includes agitation or nervous behavior and/or obsessive worrying. The child may feel or express **fear** of being separated from parents.
- Adjustment disorder with mixed anxiety and depressed mood. This condition combines the symptoms seen in both adjustment disorders with depression and with anxiety.
- Adjustment disorder with disturbance of conduct. Behavioral signs of this adjustment disorder include primarily actions that show a disregard for rules, laws, and the rights of others, such as picking fights, vandalism, **truancy**, and reckless driving for teens.
- Adjustment disorder with mixed disturbance of emotions and conduct. This condition combines depression and anxiety symptoms with those of disturbance of conduct.
- Unspecified adjustment disorders. This phrase is the catch-all term to describe any adjustment disorder not showing a predominance of any one set of the above-listed symptoms.

When to call the doctor

In order to even establish a diagnosis of adjustment disorder, a mental health professional needs to meet and evaluate the child or teen. As this illness can be debilitating, making it quite difficult for the child to function, that evaluation should take place as soon as possible after symptoms are observed. As noted above, suicidal ideation can be a potential facet of depressed mood adjustment disorders, and untreated adjustment disorder with depressed mood can lead to more serious mental illness, including major depression. These two facts give additional impetus to quickly involving a psychiatrist or psychologist.

Diagnosis

One of the primary measurements used in diagnosing adjustment disorder is the occurrence of the stress-causing event within the past three months. The only usual life-stressor not considered a possible cause for adjustment disorder is bereavement. Adjustment disorders are also differentiated from other reactions to stress such as PTSD by both symptoms and the relative severity of the causative event. Adjustment disorders can be caused by almost any stressor and manifest a wide variety of symptoms, while PTSD is normally associated with severe stress-causing life events and has a more specific set of symptoms.

The child being evaluated for an adjustment disorder needs to meet the following criteria in order to confirm the diagnosis:

- has had a psychological evaluation
- has experienced a psychological stressor within the past three months
- shows symptoms that appear disproportionate to the stressful event
- does not appear to be suffering from any other underlying mental or physical illness

Treatment

The most important goal in the treatment of adjustment disorder is relieving the symptoms a child or teen experiences so that they can return to the same level of functioning they possessed prior to the onset of illness. Treatment depends upon the age and overall health of the child as well as the severity of the symptoms. Medication is only ordered on an extremely limited basis or not ordered at all because psychotropic medications have been shown to have little efficacy in treating adjustment disorders. Age-appropriate cognitive-behavioral individual psychotherapy, focusing on problem solving, communication, impulse control, and stress and anger-management is a usual component of treatment. **Family therapy** to improve communication between the child or teen and parents and siblings is often helpful, as is group therapy with peers (other children also suffering from adjustment disorder).

Prognosis

Early detection and treatment of adjustment disorders in children has been shown to appreciably reduce the severity of symptoms and improve their quality of life. Most recoveries from adjustment disorder uncomplicated by other mental illness are both rapid and complete returns to the child's former level of functioning.

KEY TERMS

Anxiety—Worry or tension in response to real or imagined stress, danger, or dreaded situations. Physical reactions, such as fast pulse, sweating, trembling, fatigue, and weakness, may accompany anxiety.

Cognitive-behavioral therapy—A type of psychotherapy in which people learn to recognize and change negative and self-defeating patterns of thinking and behavior.

Defense mechanisms—Indirect strategies used to reduce anxiety rather than directly facing the issues causing the anxiety.

Depression—A mental condition in which a person feels extremely sad and loses interest in life. A person with depression may also have sleep problems and loss of appetite and may have trouble concentrating and carrying out everyday activities.

Post-traumatic stress disorder (PTSD)—A disorder that occurs among survivors of extremely stressful or traumatic events, such as a natural disaster, an airplane crash, rape, or military combat. Symptoms include anxiety, insomnia, flashbacks, and nightmares. Patients with PTSD are unnecessarily vigilant; they may experience survivor guilt, and they sometimes cannot concentrate or experience joy.

Psychological evaluation—Examination of a patient by a psychologist through interviews, observation of behavior, and psychological testing with the goal of determining personality adjustment, identifying problems, and helping to diagnose and plan treatment for a mental disorder.

Psychotherapy—Psychological counseling that seeks to determine the underlying causes of a patient's depression. The form of this counseling may be cognitive/behavioral, interpersonal, or psychodynamic.

Psychotropic drug—Any medication that has an effect on the mind, brain, behavior, perceptions, or emotions. Psychotropic medications are used to treat mental illnesses because they affect a patients moods and perceptions.

Stressor—A stimulus, or event, that provokes a stress response in an organism. Stressors can be categorized as acute or chronic, and as external or internal to the organism.

Prevention

The National Institute for Mental Health (NIMH) notes that there is no way to predict who will develop an adjustment disorder given the appearance of certain life-situation stressors. Since there is also no known way to prevent the occurrence of these stressors, prevention seems impossible. However, it is known that understanding and support from family and friends can help.

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Adolescence

Definition

Sometimes referred to as teenage years, youth, or **puberty**, adolescence is the transitional period between childhood and maturity, occurring roughly between the ages of 10 and 20.

Description

The word adolescence is Latin in origin, derived from the verb *adolescere,* which means "to grow into adulthood." Adolescence is a time of moving from the immaturity of childhood into the maturity of adulthood. There is no single event or boundary line that denotes the end of childhood or the beginning of adolescence. Rather, experts think of the passage from childhood into and through adolescence as composed of a set of transitions that unfold gradually and that touch upon many aspects of the individual's behavior, development, and relationships. These transitions are biological, cognitive, social, and emotional.

Puberty

The biological transition of adolescence, or puberty, is perhaps the most observable sign that adolescence has begun. Technically, puberty refers to the period during which an individual becomes capable of sexual reproduction. More broadly speaking, however, puberty is used as a collective term to refer to all the physical changes that occur in the growing girl or boy as the individual passes from childhood into adulthood.

The timing of physical maturation varies widely. In the United States, menarche (onset of **menstruation**) typically occurs around age 12, although some youngsters start puberty when they are only eight or nine, others when they are well into their teens. The duration of puberty also varies greatly: 18 months to six years in girls and two to five years in boys.

The physical changes of puberty are triggered by hormones, chemical substances in the body that act on specific organs and tissues. In boys a major change incurred during puberty is the increased production of testosterone, a male sex hormone, while girls experience increased production of the female hormone estrogen. In both sexes, a rise in growth hormone produces the adolescent growth spurt, the pronounced increase in height and weight that marks the first half of puberty.

Perhaps the most dramatic changes of puberty involve sexuality. Internally, through the development of primary sexual characteristics, adolescents become capable of sexual reproduction. Externally, as secondary sexual characteristics appear, girls and boys begin to look like mature women and men. In boys primary and secondary sexual characteristics usually emerge in a predictable order, with rapid growth of the testes and scrotum, accompanied by the appearance of pubic hair. About a year later, when the growth spurt begins, the penis also grows larger, and pubic hair becomes coarser, thicker, and darker. Later still comes the growth of facial and body hair, and a gradual lowering of the voice. Around mid-adolescence internal changes begin making a boy capable of producing and ejaculating sperm.

In girls, sexual characteristics develop in a less regular sequence. Usually, the first sign of puberty is a slight elevation of the breasts, but sometimes this is preceded by the appearance of pubic hair. Pubic hair changes from sparse and downy to denser and coarser. Concurrent with these changes is further **breast development**. In teenage girls, internal sexual changes include maturation of the uterus, vagina, and other parts of the reproductive system. Menarche, the first menstrual period, happens relatively late in puberty. Regular ovulation and the ability to carry a baby to full term usually follow menarche by several years.

Cognitive transition

A second element of the passage through adolescence is a cognitive transition. Compared to children, adolescents think in ways that are more advanced, more efficient, and generally more complex. This is evident in five distinct areas of cognition.

First, during adolescence individuals become better able than children to think about what is possible, instead of limiting their thought to what is real. Whereas children's thinking is oriented to the here and now (i.e., to things and events that they can observe directly), adolescents are able to consider what they observe against a backdrop of what is possible—they can think hypothetically.

Second, during the passage into adolescence, individuals become better able to think about abstract ideas. For example, adolescents find it easier than children to comprehend the sorts of higher-order, abstract logic inherent in puns, proverbs, metaphors, and analogies. The adolescent's greater facility with abstract thinking also permits the application of advanced reasoning and logical processes to social and ideological matters. This is clearly seen in the adolescent's increased facility and interest in thinking about interpersonal relationships, politics, philosophy, religion, and morality—topics that involve such abstract concepts as friendship, faith, democracy, fairness, and honesty.

Third, during adolescence individuals begin thinking more often about the process of thinking itself, or metacognition. As a result, adolescents may display increased introspection and self-consciousness. Although improvements in metacognitive abilities provide important intellectual advantages, one potentially negative byproduct of these advances is the tendency for

adolescents to develop a sort of egocentrism, or intense preoccupation with the self. Acute adolescent egocentrism sometimes leads teenagers to believe that others are constantly watching and evaluating them. Psychologists refer to this as the imaginary audience.

A fourth change in cognition is that thinking tends to become multidimensional, rather than limited to a single issue. Whereas children tend to think about things one aspect at a time, adolescents describe themselves and others in more differentiated and complicated terms and find it easier to look at problems from multiple perspectives. Being able to understand that people's personalities are not one-sided, or that social situations can have different interpretations, depending on one's point of view, permits the adolescent to have far more sophisticated and complicated relationships with other people.

Finally, adolescents are more likely than children to see things as relative, rather than absolute. They are more likely to question others' assertions and less likely to accept "facts" as absolute truths. This increase in relativism can be particularly exasperating to parents, who may feel that their adolescent children question everything just for the sake of argument.

Emotional transition

Adolescence is also a period of emotional transition, marked by changes in the way individuals view themselves and in their capacity to function independently. As adolescents mature intellectually and undergo cognitive changes, they come to perceive themselves in more sophisticated and differentiated ways. Compared with children, who tend to describe themselves in relatively simple, concrete terms, adolescents are more likely to employ complex, abstract, and psychological self-characterizations. As individuals' self-conceptions become more abstract and as they become more able to see themselves in psychological terms, they become more interested in understanding their own personalities and why they behave the way they do.

For most adolescents, establishing a sense of autonomy, or independence, is as important a part of the emotional transition out of childhood as is establishing a sense of identity. During adolescence, there is a movement away from the dependency typical of childhood toward the autonomy typical of adulthood. For example, older adolescents do not generally rush to their parents whenever they are upset, worried, or in need of assistance. They do not see their parents as all-knowing or all-powerful, and often have a great deal of emotional energy wrapped up in relationships outside the **family**. In addition, older adolescents are able to see and interact with their parents as people, not just as their parents. Many parents find, for example, that they can confide in their adolescent children, something that was not possible when their children were younger, or that their adolescent children can easily sympathize with them when they have had a hard day at work.

Being independent, however, means more than merely feeling independent. It also means being able to make decisions and to select a sensible course of action. This is an especially important capability in contemporary society, where many adolescents are forced to become independent decision makers at an early age. In general, researchers find that decision-making abilities improve over the course of the adolescent years, with gains continuing well into the later years of high school.

Many parents wonder about the susceptibility of adolescents to **peer pressure**. In general, studies that contrast parent and peer influences indicate that in some situations, peers' opinions are more influential, while in others, parents' are more influential. Specifically, adolescents are more likely to conform to their peers' opinions when it comes to short-term, day-to-day, and social matters—styles of dress, tastes in music, and choices among leisure activities. This is particularly true during junior high school and the early years of high school. When it comes to long-term questions concerning educational or occupational plans, however, or values, religious beliefs, and ethical issues, teenagers are influenced in a major way by their parents.

Susceptibility to the influence of parents and peers changes during adolescence. In general, during childhood, boys and girls are highly oriented toward their parents and less so toward their peers; peer pressure during the early elementary school years is not especially strong. As they approach adolescence, however, children become somewhat less oriented toward their parents and more oriented toward their peers, and peer pressure begins to escalate. During early adolescence, conformity to parents continues to decline and conformity to peers and peer pressure continues to rise. It is not until middle adolescence that genuine behavioral independence emerges, when conformity to parents as well as peers declines.

Social transition

Accompanying the biological, cognitive, and emotional transitions of adolescence are important changes in the adolescent's social relationships. Developmentalists have spent considerable time charting the changes that take place with friends and with family members as the individual moves through the adolescent years.

One of the most noteworthy aspects of the social transition into adolescence is the increase in the amount of time individuals spend with their peers. Although rela-

tions with age-mates exist well before adolescence, during the teenage years they change in significance and structure. For example, there is a sharp increase during adolescence in the sheer amount of time individuals spend with their peers and in the relative time they spend in the company of peers versus adults. In the United States, well over half of the typical adolescent's waking hours are spent with peers, as opposed to only 15 percent with adults, including parents. Second, during adolescence, peer groups function much more often without adult supervision than they do during childhood, and more often involve friends of the opposite sex.

Finally, whereas children's peer relationships are limited mainly to pairs of friends and relatively small groups—three or four children at a time, for example—adolescence marks the emergence of larger groups of peers, or crowds. Crowds are large collectives of similarly stereotyped individuals who may or may not spend much time together. In contemporary American high schools, typical crowds are "jocks," "brains," "nerds," "populars," "druggies," and so on. In contrast to cliques, crowds are not settings for adolescents' intimate interactions or friendships, but instead serve to locate the adolescent (to himself and to others) within the social structure of the school. As well, the crowds themselves tend to form a sort of social hierarchy or map of the school, and different crowds are seen as having different degrees of status or importance.

The importance of peers during early adolescence coincides with changes in individuals' needs for intimacy. As children begin to share secrets with their friends, loyalty and commitment develop. During adolescence, the search for intimacy intensifies, and self-disclosure between best friends becomes an important pastime. Teenagers, especially girls, spend a good deal of time discussing their innermost thoughts and feelings, trying to understand one another. The discovery that they tend to think and feel the same as someone else becomes another important basis of friendship.

One of the most important social transitions that takes place in adolescence concerns the emergence of sexual and romantic relationships. In contemporary society, most young people begin dating sometime during early adolescence. Dating during adolescence can mean a variety of different things, from group activities that bring males and females together (without much actual contact between the sexes); to group dates, in which a group of boys and girls go out jointly (and spend part of the time as couples and part of the time in large groups); to casual dating as couples; and to serious involvement with a steady boyfriend or girlfriend. More adolescents have experience in mixed-sex group activities like parties or dances than dating, and more have experience in dating than in having a serious boyfriend or girlfriend.

Most adolescents' first experience with sex falls into the category of "autoerotic behavior," sexual behavior that is experienced alone. The most common autoerotic activities reported by adolescents are erotic fantasies and **masturbation**. By the time most adolescents are in high school, they have had some experience with sexual behaviors in the context of a relationship. The Youth Risk Behavior Surveillance System (YRBSS), a self-reported survey of a national representative sample of high school students in grades nine to 12, indicated that in 2003, 46.7 percent of the students reported having had sex. By grade level, the rates were 32.8 percent for ninth grade, 44.1 percent for tenth grade, 53.2 percent for eleventh grade, and 61.6 percent for twelfth grade.

Common problems

Generally speaking, most young people are able to negotiate the biological, cognitive, emotional, and social transitions of adolescence successfully. Some adolescents, however, are at risk of developing certain problems, such as:

- eating disorders such as **anorexia nervosa**, bulimia, or obesity
- drug or alcohol use
- depression or suicidal ideation
- violent behavior
- anxiety, stress, or **sleep** disorders
- unsafe sexual activities

Parental concerns

Many parents dread the onset of adolescence, fearing that their child will become hostile and rebellious and begin to reject his or family. Although it is incorrect to characterize adolescence as a time when the family ceases to be important, or as a time of inherent and inevitable family conflict, adolescence is a period of significant change and reorganization in family relationships. Family relationships change most around the time of puberty, with increasing conflict and decreasing closeness occurring in many parent-adolescent relationships. Changes in the ways adolescents view family rules and regulations may contribute to increased disagreement between them and their parents. Family conflict during this stage is more likely to take the form of bickering over day-to-day issues than outright fighting. Similarly, the diminished closeness is more likely to be manifested in increased privacy on the part of the adolescent and diminished physical affection between teenagers and parents, rather than any serious

KEY TERMS

Anorexia nervosa—An eating disorder marked by an unrealistic fear of weight gain, self-starvation, and distortion of body image. It most commonly occurs in adolescent females.

Bulimia nervosa—An eating disorder characterized by binge eating and inappropriate compensatory behavior, such as vomiting, misusing laxatives, or excessive exercise.

Hormone—A chemical messenger secreted by a gland or organ and released into the bloodstream. It travels via the bloodstream to distant cells where it exerts an effect.

Menarche—The first menstrual cycle in a girl's life.

Metacognition—Awareness of the process of cognition.

loss of love or respect between parents and children. Research suggests that this distancing is temporary, and that family relationships may become less conflicted and more intimate during late adolescence.

When to call the doctor

Although changes—biologically, cognitively, emotionally, and socially—are to be expected during adolescence, certain inappropriate behaviors, drastic changes in personality or physical appearance, or abnormal sexual development may warrant a phone call to a physician or counselor. These include:

- extreme changes in weight (loss or gain) or excessive dieting
- sleep disturbances
- social withdrawal or loss of interest in activities
- sudden personality changes
- signs of alcohol or drug use
- talk or threats of suicide
- violent or aggressive behavior
- atypical (early or late) onset of puberty; in girls, failure to menstruate by the age of 16

See also Puberty.

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Society for Research on Adolescence, 3131 S. State St., Suite 302, Ann Arbor, MI 48108-1623. Web site: <www.s-r-a.org>.

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Adoption

Definition

Adoption is the practice in which an adult assumes the role of parent for a child who is not the adult's biological offspring. The process usually involves some legal paperwork.

Description

The ancient practice of adoption was a way of ensuring male heirs to childless couples in order to preserve **family** lines and religious traditions. In the 1850s the Children's Aid Society of New York City began to move dependent children out of city institutions. Between 1854 and 1904 orphan trains carried an estimated 100,000 children to families on farms in the Midwest; these children were to provide farm work in exchange for care.

Modern U.S. adoption laws are designed with the best interests of the child in mind, not the best interests of the adult who intends to adopt. Throughout most of the twentieth century, adoptions were conducted in secret, and records were often sealed to protect those involved from the social stigma of birth out of wedlock. After World War I, the advent of commercial formula facilitated raising babies without their being fed by breast. Adults were trained in parenting, and childless couples became interested in adopting. Because of the rapidly increasing interest in infant adoptions, many state laws demanded investigations of prospective adoptive parents and court approval before the adoption could be completed.

In the early 2000s, state laws on adoption vary. Adoptions can be conducted privately between individuals, between independent agencies and individuals, and between public agencies (such as a state's child protective services) and individuals. Adoptees may be infants or older children, they may be adopted singly or as sibling groups, and they may come from the local area or from other states or countries. Adoptive parents may be married couples, single men or women, or nontraditional couples. Adoptive parents may be childless or have other children.

Demographics

In the 1990s, roughly 120,000 children were adopted annually in the United States. This number remained proportionate to the U.S. population throughout that decade and into the early 2000s. During this period, nearly 10,000 children were adopted from abroad.

Types of adoptions

PUBLIC ADOPTIONS In 2000 and 2001, about 127,000 children were adopted annually in the United states. Since 1987, the number of adoptions annually has remained relatively constant, ranging from 118,000 to 127,000. Adoptions through publicly funded child welfare agencies accounts for about 40 percent of all adoptions. More than 50,000 public agency adoptions in each year (2000 and 2001) accounted for 40 percent of adoptions, up from 18 percent in 1992 for 36 states that reported public agency adoptions in that year.

PRIVATE ADOPTIONS In a private adoption, children are placed in non-relative homes through a nonprofit agency licensed by the state in which it operates. In an independent or non-agency adoption, children are placed in non-relative homes directly by the birthparents or through the services of a licensed or unlicensed facilitator, certified medical doctor, member of the clergy, or attorney.

About 40 percent of the 127,000 adoptions in 2000 and 2001 were primarily private agency, kinship, or tribal adoptions. There were 58,420 adoptions (46%) private adoptions reported in 2000–2001. With the available data, it is not possible to separate figures within this group for types of adoptions. However, in 1992, for example, stepparent adoptions (a form of kinship adoption) alone accounted for 42 percent of all adoptions.

Informal adoptions occur when a relative or stepparent assumes permanent parental responsibilities without court involvement. However, legally recognized adoptions need a court or other government agency to award permanent custody of a child to adoptive parents.

The U.S. Department of Health and Human Services, Administration for Children and Families Interim Estimates for 2000 as of August 2002 reports 30,939 foster parent adoptions and 10,612 relative adoptions through the foster parent system. (Relatives who were also foster parents were counted as relatives.)

The U.S. Census is the principal source of data on adopted children and their families on a national level. The report for 2000 presents information on 2.1 million adopted children and 4.4 million stepchildren of householders, as estimated from the census sample, which collected from approximately one out of every six households. Together, these children represented approximately 8 percent of the 84 million sons and daughters of householders. In 2000 there were more than twice as many stepchildren as adopted children in U.S. households, with stepchildren representing 5 percent of children in the household. While these data are non-specific, it is safe to say that a significant number of the stepchildren were neither kinship nor stepparent adoptions. Since almost all adoptions by related applicants are independent, it is likely that most independent adoptions were by relatives.

TRANSRACIAL In transracial adoptions, children are placed with an adoptive family of another race. These adoptions may be through public and private agencies or be independent, but most transracial adoptions take place through the public child welfare system. The civil rights movement of the 1960s led to an increase in transracial adoptions involving black children and white parents. This practice peaked in 1971, and one year later the National Association of Black Social Workers issued a statement opposing transracial adoption. The association argued that white families were unable to foster the

growth of psychological and cultural identity in black children.

An estimated 15 percent of the 36,000 adoptions of foster children in 1998 were transracial or transcultural adoptions. Many Americans continue to be troubled by these adoptions. The National Association of Black Social Workers called them a form of cultural genocide. That point aside, there are in fact not enough African American adults willing to adopt to fill the need of African American children in need of adoption.

INTRANATIONAL AND INTERNATIONAL In response to a shortage of healthy, Caucasian infants, prospective adoptive white parents started adopting children from Japan and Europe. In 2003, approximately 21,616 children were adopted through international adoption. International adoptions accounted for more than 15 percent of all U.S. adoptions, an increase from 5 percent between 1992 and 2001. This practice showed a dramatic increase between the mid-1990s and the early 2000s.

Between 1999 and 2004, international adoptions grew in popularity in the United States as more families recognized the global humanitarian need to provide homes for waiting children. Besides this pressing need, international adoptions have proven to be safe and successful, so they provide an attractive option for people who have been trying without success to adopt within the United States.

Though U.S. citizens adopted children from 106 different countries in 2001, nearly three-fourth of all children came from only five countries: China (25%), Russia (22%), South Korea (10%), Guatemala (8%), and Ukraine (6%). The Chinese government's population control policy, which penalizes families who have more than one child, and the greater value placed on male heirs in Chinese culture have led many families to abandon female Chinese infants. These babies constituted a bountiful source of adoption candidates for American families. In 2003, U.S. interest in adopting from Kazakhstan also grew as many U.S. families reported a fast, smooth adoption experience there. Americans adopt children from Peru, Colombia, El Salvador, Mexico, and the Philippines. Some adoptions come from Vietnam. Adoption from India, however, is difficult for non-Indian parents. In 2002, Cambodia and Romania stopped international adoptions.

SINGLE PARENT According to the United States Department of Health and Human Services, 33 percent of adoptions from **foster care** are by single parents. Most of these single parents are women. Single women are more likely to adopt an older child than an infant. Single men adopted some children, and unmarried couples adopted some children in the same period. As one-parent households increase in number and become more acceptable, adoptions in these households also become more common. More than one half of African-American children, nearly one third of Hispanic children, and one fifth of Caucasian children live with a single parent because of **divorce** and unmarried mothers. This prevalence gives adoption agencies a more open-minded approach toward single parent adoptions. Also, the issue of personal finances and single income families has become less important since adoption subsidies are available nationwide.

Treatment of adoption information

Through most of the nineteenth century and into the twentieth century, adoptions were often informal and unofficial. Agencies, counselors, doctors, and private attorneys were generally not involved. If a young woman was pregnant out-of-wedlock, the baby's adoption might be arranged by the mother's parents with the help of the head of her extended family. Some family member or close friend took in the child. The child might refer to the adoptive parents as aunt and uncle, but people in the immediate social circle might know the child's biological parent.

In the early twentieth century, as governmental and independent agencies became involved with adoption, information about the individuals involved tended to be restricted. Decisions about who could adopt which baby were often made solely by agency personnel. In closed adoptions, mothers gave up parental rights immediately after birth. They did not see or hold their babies.

In the later twentieth century and in the early 2000s, information about adoptions is open to the participants. The birth mother may room in with the baby in the hospital. The birth mother and adoptive parents may have a contract before delivery and a formal or informal agreement about shared responsibility for the baby. The birth parent may have visitation rights after adoption takes place. This arrangement often occurs between a teenage birth parent and grandparents who become the legal parents through adoption. Open adoptions may also take place between surrogate and adopting parents.

Fraud by adoption agencies

Adoption fraud may involve the misrepresentation and fraudulent concealment of a child's pre-adoption history. Some state laws require full disclosure in good faith of information pertaining to the child's health. This information helps adopting parents anticipate any special needs the child may have. Full disclosure by

Parents with their adopted children. *(Photograph by John Hart. AP/Wide World Photos.)*

the adoption agency facilitates the child's receiving appropriate intervention and treatment as needed.

Parental concerns

Adoptions are expensive. Most of the financial expenses are attorney or court fees, and the cost of preparing the home for a new child. Expense results from parents' lost wages for time off to meet with social agencies or to have their homes inspected. Adoptions are emotionally taxing as well. The adoptive parents deal with uncertainty, and if there are other children in the household already, the parents deal with those children's responses and feelings as everyone involved prepares for the possibility of a new family member. Time must be spent with a social worker whose task it is to evaluate the home.

In some cases, the adoptive child is placed in the adoptive home before the legal termination of parental rights has freed the child for adoption. In these cases, child protective services are fairly certain the courts will decide in favor of the adoptive placement, but this tentative situation imposes a potentially uncomfortable arrangement on the adoptive family and their household.

Adoption is challenging for the adopting parents, for other children if they have them, and for the adopted child. Soon after the new child arrives, the adoptive parents should schedule a medical exam. Adopted children from other countries may be at greater risk for certain illnesses or conditions related to possible substandard care they received before their arrival in their new home. Medical evaluation may identify special needs the adoptive parents can then address.

Adopted children should be told early that they are adopted. Knowing from early childhood of the adoption is better for children than learning about it later. Three-year-old children can understand the story of their adoption.

Adolescents may have questions about identity that are connected to their not knowing their biological parents. It is common for them to spend time tracing records and trying to find their birth parents. This activity does not

KEY TERMS

Abandonment—Legally, the refusal to provide adequate financial support for one's dependent child; the failure to maintain a parental relationship with one's dependent child.

Adoptee—A person who has been adopted.

Adoption subsidy—A short-term or long-term financial payment, either in the form of cash or services, to help an adoptive family provide for the on-going care of an adopted child. A subsidy can be medical insurance for the child, counseling services for the family, respite care for the adoptive parents; or a monthly cash allowance to help cover other extraordinary expenses and services associated with the adoption.

Birth parents—The biological parents of a child.

Custody—The care, control, and maintenance of a child, which in abuse and neglect cases can be awarded by the court to an agency or in divorce to parents. Foster parents do not have legal custody of the children who are in their care.

Disclosure—Release of information.

Relinquishment—Giving up parental rights to a child, so someone else can adopt the child.

Severance of parental rights—The end of parental rights; the involuntarily removal of parental rights of a parent that has abandoned a child; has without just cause failed to support a child; has neglected or abused a child or has stood by and allowed others to neglect or abuse a child; or who because of extended incarceration in prison, is unavailable to parent or nurture the child. Once the parental rights of both parents of a child are removed the child will become available for adoption by another family.

Trans-racial adoptions—Adoption in which a family of one race adopts a child of another race.

necessarily constitute a rejection of the adopting parents. Children seek out their birth parents because they need information about themselves in order to shape a sense of who they are and where they belong in the world.

Sometimes the adopted child will feel loss, **abandonment**, and resentment toward the birth parent and the adopting parents. For a period, the adoptive family may not be able to compensate the child who faces the loss of the birth family.

Parenting the adopted child

Adopting parents who intuitively understand the sense of loss and **separation anxiety** experienced by an adopted child and communicate with their child about the adoption can develop closeness. Even tiny infants have a bond with their mother before birth. A child knows his mother and instinctively wants to be with her. Even babies may experience loss of the natural mother and a sense of confusion regarding the stranger who assumes the role of mother. Parental separation from the child can also be traumatic. The adoptive parents need to be attuned to the child's emotional responses to loss.

In the absence of genetic markers (facial features, gestures, body language, basic personality, interests, and talents) both adoptive parents and the adopted child must learn how to communicate. The adopted child may have trouble fitting into the adoptive family when genetic traits are not mirrored or reflected.

There are many ways for adoptive parents to help an older child deal with sorrow, anger, **anxiety**, and low **self-esteem** caused by separation from the biological parents:

- Celebrate birthdays a week or so before the birthday, if the birthday is really the date of separation from the natural parent.
- Take extra time to prepare the child for changes in routine, a new school, and family life.
- Listen more and talk less to the adopted child.
- Respond to painful feelings with support, rather than by discounting them in any way.
- Respect and value the differences between the child and other members of the family.
- Encourage the child's talents and interests, even if they are different from the adoptive family.

Parenting an adopted child is parenting plus. But with intuition, information, understanding, and empathy, it can be a rewarding experience.

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Adrenoleukodystrophy *see* **Peroxisomal disorders**

AFP test *see* **Alpha-fetoprotein test**

Aggressive behavior

Definition

Aggressive behavior is reactionary and impulsive behavior that often results in breaking household rules or the law; aggressive behavior is violent and unpredictable.

Description

Aggression can a problem for children with both normal development and those with psychosocial disturbances. Aggression constitutes intended harm to another individual, even if the attempt to harm fails (such as a bullet fired from a gun that misses its human target). There is no single theory about the causes of aggressive behavior in humans. Some believe aggression is innate or instinctive. Social theorists suggest the breakdown in commonly shared values, changes in traditional **family** patterns of child-rearing, and social isolation lead to increasing aggression in children, adolescents, and adults. Aggression in children correlates with family unemployment, strife, criminality, and psychiatric disorders.

Differences exist between levels of aggression in boys and girls in the same families. Boys are almost always more aggressive than girls. Larger children are more aggressive than smaller ones. Active and intrusive children are also more aggressive than passive or reserved ones.

Aggressive behavior may be intentional or unintentional. Many hyperactive, clumsy children are accidentally aggressive, but their intentions are compassionate. Careful medical evaluation and diagnostic assessments distinguish between intentional behaviors and the unintentional behaviors of emotionally disturbed children.

Children in all age groups learn that aggressive behavior is a powerful way to communicate their wishes or deal with their likes and dislikes.

Infancy

Infants are aggressive when they are hungry, uncomfortable, fearful, angry, or in **pain**. Parents can tell what babies need by the loudness and pitch of crying and the flailing of arms and legs. Crying is an infant's defense, the way to communicate feelings and needs.

Toddlers

Children between two and four years of age show aggressive outbursts such as temper **tantrums** and hurting others or damaging **toys** and furniture because they are frustrated. Usually the aggression in this age group is expressed toward parents as a way to get their compliance with the child's wishes. Verbal aggression increases as vocabulary increases.

Preschool

Children between four and five years of age can be aggressive toward their siblings and peers. Because of greater social interaction, children need to learn the differences between real and imaginary insults, as well as the difference between standing up for their rights and attacking in anger.

School-age and adolescence

Aggressive boys between three to six years of age are likely to carry their behavior style into **adolescence**. In extreme cases, they may show aggression by purse snatching, muggings, or robbery, or in less overt ways by persistent **truancy**, **lying**, and vandalism. Girls younger

than six years of age who have aggressive styles toward their peers do not tend to continue being aggressive when they are older, and their earlier aggression does not correlate with adult competitiveness.

Common problems

Frustration is a response to conditions that keep children from achieving goals important to **self-esteem**. Frustration and aggression are closely associated. If children learn that being aggressive when frustrated is tolerated or gives them special treatment, the behavior is reinforced and may be repeated. Aggression may be a way for children to face obstacles or solve problems. It is important not to attribute malice to children who are responding to **anxiety**, feelings of incompetence, or a sense of low self-esteem.

Through the media, including film, the U.S. culture reinforces violence and aggressive behavior in children. Police brutality, crime-based television programs, and governmental reliance on military aggression to solve political and economic differences all create a climate in which violence is presented to children as a legitimate solution to problems.

Violent behavior in children and adolescents

CULTURAL VIOLENCE Violence includes a wide range of behaviors: explosive temper tantrums, physical aggression, fighting, and threats or attempts to hurt others (including homicidal thoughts). Violent behaviors also include the use of weapons, cruelty toward animals, setting fires, and other intentional forms of destruction of property.

PREDISPOSITION TO VIOLENCE Some children are supersensitive, easily offended, and quick to anger. Many children are tense and unusually active, even as infants. They are often more difficult to soothe and settle as babies. Beginning in the **preschool** years, they are violent toward other children, adults, and even animals. They often lash out suddenly, sometimes for no obvious reason. When they hurt someone in their anger, they tend not to be sorry and may tend not to take responsibility for their actions. Instead, they blame others for their own actions. Parent should give this behavior serious attention and take measures to correct it.

Children may go through a brief period of aggressive behavior if they are worried, tired, or stressed. If the behavior continues for more than a few weeks, parents should talk to the pediatrician. If it becomes a daily pattern for more than three to six months, it could be a serious problem.

Factors that increase risk of violent behavior

Parents and teachers should be careful not to play down aggressive behaviors in children. In fact, certain factors put some children at risk for developing violent behaviors as adults. These factors include the following:

- being the victim of physical and sexual abuse
- exposure to violence in the home and community
- exposure to violence in media (TV, movies)
- use of drugs and alcohol
- presence of firearms in home
- combination of stressful family socioeconomic factors (poverty, severe deprivation, marital breakup, single parenting, unemployment, loss of support from extended family)
- brain injury

Parents can teach children nonviolence by controlling their own tempers. If parents express anger in quiet, assertive ways, children may follow their parent's example. Children need to understand when they have done something wrong so they can learn to take responsibility for their actions and learn ways to make amends. Responsible parenting does not to tolerate violence or use it in any way.

Violence prevention strategies

Efforts should be directed at dramatically decreasing the exposure of children and adolescents to violence in the home, community, and through the media. Clearly, violence leads to violence. Parents can use the following strategies to reduce or prevent violent behavior:

- prevent **child abuse** in the home
- provide sex education and parenting programs for adolescents
- provide early intervention programs for violent youngsters
- monitor children's TV programs, videos, and movies

The most important step that parents can take with aggressive children is to set firm, consistent limits and be sure that everyone caring for the children acts in accord with the parents' rules and expectations.

Parents should know the importance of helping children find ways to deal with anger without resorting to violence. Children can learn to say no to their peers, and they can learn how to settle differences with words instead of physical aggression. When children control their violent impulses, they should be praised.

KEY TERMS

Anxiety—Worry or tension in response to real or imagined stress, danger, or dreaded situations. Physical reactions, such as fast pulse, sweating, trembling, fatigue, and weakness, may accompany anxiety.

Consequences—Events that occur immediately after the target behavior.

Misbehavior—Behavior outside the norms of acceptance within the group.

Time-out—A discipline strategy that entails briefly isolating a disruptive child in order to interrupt and avoid reinforcement of negative behavior.

Parental concerns

All children have feelings of anger and aggression. Children need to learn positive ways to express these feelings and to negotiate for what they want while maintaining respect for others. Parents can help their children develop judgment, self discipline, and the other tools children need to express feelings in more acceptable ways and to live with others in a safe way.

Understanding the aggressive child

When children lose their sense of connection to others, they may feel tense, frightened, or isolated. These are the times when they may unintentionally lash out at other children, even children to whom they are close. Parents should be careful not to let children think aggression is acceptable.

When children are overcome with feelings of isolation or despair, they may run for the nearest safe person and begin to cry. They immediately release the terrible feelings, trusting that they are safe from danger and criticism. Effective parents listen and allow the child to vent without becoming alarmed.

Disciplining aggressive behavior

Parents can control the aggressive child in various ways. They should intervene quickly but calmly to interrupt the aggression and prevent the their child from hurting another child. Younger children may need a time-out to calm down and before rejoining a group. Simple rules about appropriate behavior are easier for a child to understand than lengthy explanations. Parents can affirm feelings while stressing that all feelings cannot be acted upon.

Parents can reach older children with eye contact, a stern voice, and physical contact. Older children can be told that they need to learn a better way to handle conflicts. Parents can suggest that, for instance, the child ask an adult to intervene before lashing out at a classmate. Any disciplinary measures should be explained as a simple consequence to the child's aggression.

When parents arrive after conflict occurs, it may be useful to listen to the child's explanation. Having a parent listen can encourage the child to develop trust in the parent.

Parents should not expect the aggressive child to be reasonable when he or she is upset. The child may need time to calm down. Sometimes the child may feel trapped and may need adult support. Parents should encourage the aggressive child to come to them when they are upset, hopefully before violence occurs.

Resources

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AIDS *see* **HIV infection and AIDS**

Albinism

Definition

Albinism is an inherited condition that is present at birth. It is characterized by a lack of melanin, the

pigment that normally gives color to the skin, hair, and eyes. Many types of albinism exist, all of which involve lack of pigment in varying degrees. The condition, which is found in all races, may be accompanied by eye problems and may ultimately lead to skin **cancer**.

Description

The most common type of albinism is oculocutaneous albinism, which affects the eyes, hair, and skin. In its most severe form, hair and skin remain completely white throughout life. People with a less severe form are born with white hair and skin that turn slightly darker as they age. Everyone with oculocutaneous albinism experiences abnormal flickering eye movements (**nystagmus**) and sensitivity to bright light. There may be other eye problems as well, including poor vision and crossed or ''lazy'' eyes (**strabismus**).

The second most common type of the condition is known as ocular albinism, in which only the eyes lack color; skin and hair are normal. Some types of ocular albinism cause more problems, especially eye problems, than others.

Albinism is also referred to as hypopigmentation.

Demographics

Albinism is a rare disorder found in fewer than five people per 100,000 in the United States and Europe. Although albinism can affect all races, other parts of the world have a much higher rate; for example, albinism is found in about 20 out of every 100,000 people in southern Nigeria. The parents of most children with albinism have normal hair and eye color for their ethnic background and do not have a **family** history of albinism.

Causes and symptoms

Albinism is an inherited problem caused by an alteration in one or more of the genes that are responsible for directing the eyes and skin to produce or distribute melanin, which is a photoprotective pigment that absorbs ultraviolet (UV) light coming from the sun so that the skin is not damaged. Sun exposure normally produces a tan, which is an increase in melanin pigment in the skin. Many people with albinism do not have melanin pigment in their skin, do not tan with exposure to the sun, and as a result develop **sunburn**. Over time, people with albinism may develop skin cancers if they do not adequately protect their skin from sun exposure.

Melanin is also important in the eyes and brain, but it is not known what role melanin plays in those areas. Parts of the retina do not develop correctly if melanin pigment is not present during development. Also nerve connections between the retina and brain are altered if melanin is not present in the retina during development.

Albinism is a autosomal recessive disease, which means that a person must have two copies of the defective gene to exhibit symptoms of the disease. The child therefore inherits one defective gene responsible for making melanin from each parents. Because the task of making melanin is complex, there are many different types of albinism, involving a number of different genes.

It is also possible to inherit one normal gene and one albinism gene. In this case, the one normal gene provides enough information to make some pigment, and the child has normal skin and eye color. The child has one gene for albinism. About one in 70 people are albinism carriers, with one defective gene but no symptoms; they have a 50 percent chance of passing the albinism gene to their child. However, if both parents are carriers with one defective gene each, they have a one in four chance of passing on both copies of the defective gene to the child, who will have albinism. There is also a type of ocular albinism that is carried on the X chromosome and occurs almost exclusively in males because they have only one X chromosome and, therefore, no other gene for the trait to override the defective one.

People with albinism may experience a variety of eye problems, including one or more of the following:

- They may be very far-sighted or near-sighted and may have other defects in the curvature of the lens of the eye (astigmatism) that cause images to appear unfocused.
- They may have a constant, involuntary movement of the eyeball called nystagmus.
- They may have problems in coordinating the eyes in fixing and tracking objects (strabismus), which may lead to an appearance of having ''crossed eyes'' at times.
- They may have reduced depth perception due to altered nerve connections from the retina to the brain.
- Their eyes may be very sensitive to light (photophobia) because their irises allow stray light to enter their eyes. It is a common misconception that people with albinism should not go outside on sunny days, but wearing sunglasses can make it possible to go outside quite comfortably.

One of the myths about albinism is that it causes people to have pink or red eyes. In fact, people with

African child with albinism surrounded by normally pigmented friends. *(© Silvia Morara/Corbis.)*

albinism can have irises varying from light gray or blue to brown. (The iris is the colored portion of the eye that controls the size of the pupil, the opening that lets light into the eye.) If people with albinism seem to have reddish eyes, it is because light is being reflected from the back of the eye (retina) in much the same way as happens when people are photographed with an electronic flash. In addition, albinism does not cause blindness.

In addition to characteristically light skin and eye problems, people with a rare form of albinism called Hermansky-Pudlak syndrome (HPS) also have a greater tendency to have bleeding disorders, inflammation of the large bowel (colitis), lung (pulmonary) disease, and kidney (renal) problems.

When to call the doctor

The doctor should be called when a person with albinism exhibits symptoms such as photophobia that cause discomfort. Also the doctor should be consulted if there are any skin changes that might be an early sign of skin cancer.

The parent of a child with albinism should also call the doctor if the child bruises easily or has unusual bleeding, such as repeated nosebleeds or bloody **diarrhea**. The child may have the rare Hermansky-Pudlak syndrome, which requires additional medical care.

Diagnosis

It is not always easy to diagnose the exact type of albinism a person has. The specific type is sometimes determined by developing a thorough family history and by examining the patient and several close relatives. In the early 2000s, a blood test has been developed that can identify carriers of the gene for some types of albinism; a similar test during **amniocentesis** can diagnose some types of albinism in an unborn child. A chorionic villus sampling test during the fifth week of pregnancy may also reveal some types of albinism.

There are also two tests available that can identify two types of the condition. The hairbulb pigmentation test is used to identify carriers by incubating a piece of the person's hair in a solution of tyrosine, a substance in

KEY TERMS

Amino acid—An organic compound composed of both an amino group and an acidic carboxyl group. Amino acids are the basic building blocks of proteins. There are 20 types of amino acids (eight are "essential amino acids" which the body cannot make and must therefore be obtained from food).

Astigmatism—An eye condition in which the cornea doesn't focus light properly on the retina, resulting in a blurred image.

Carrier—A person who possesses a gene for an abnormal trait without showing signs of the disorder. The person may pass the abnormal gene on to offspring. Also refers to a person who has a particular disease agent present within his/her body, and can pass this agent on to others, but who displays no symptoms of infection.

DNA—Deoxyribonucleic acid; the genetic material in cells that holds the inherited instructions for growth, development, and cellular functioning.

DOPA—The common name for a natural chemical (3,4-dihydroxyphenylalanine) made by the body during the process of making melanin.

Enzyme—A protein that catalyzes a biochemical reaction without changing its own structure or function.

Gene—A building block of inheritance, which contains the instructions for the production of a particular protein, and is made up of a molecular sequence found on a section of DNA. Each gene is found on a precise location on a chromosome.

Hairbulb—The root of a strand of hair from which growth and coloration of the hair develops.

Hermansky-Pudlak syndrome—A rare type of albinism, most common in the Puerto Rican community, which can cause pigment changes, lung disease, intestinal disorders, and blood disorders.

Melanin—A pigment that creates hair, skin, and eye color. Melanin also protects the body by absorbing ultraviolet light.

Nystagmus—An involuntary, rhythmic movement of the eyes.

Strabismus—A disorder in which the eyes do not point in the same direction. Also called squint.

Tyrosinase—An enzyme in a pigment cell which helps change tyrosine to dopa during the process of making melanin.

Tyrosine—An amino acid synthesized by the body from the essential amino acid phenylalanine. It is used by the body to make melanin and several hormones, including epinephrine and thyroxin.

food which the body uses to make melanin. If the hair turns dark, it means the hair is making melanin (a positive test); light hair means there is no melanin. This test is the source of the names of two types of albinism: "ty-pos" and "ty-neg."

The tyrosinase test is more precise than the hairbulb pigmentation test. It measures the rate at which hair converts the amino acid tyrosine into another chemical (3,4-dihydroxyphenylalanine, or DOPA), which is then made into pigment. The hair converts tyrosine with the help of an enzyme called tyrosinase. In some types of albinism, a genetic defect in tyrosinase means that the amino acid tyrosine cannot be converted by tyrosinase into melanin.

Treatment

There is no treatment that can replace the lack of melanin that causes the symptoms of albinism. In addition, doctors can only treat, but not cure, the eye problems that often accompany the lack of skin color. Glasses or corrective lenses and low vision aids, for example, magnifiers, monoculars (handheld telescopes used with only one eye), or bioptics (glasses with small telescopic lenses mounted in the standard lenses) can improve vision but usually cannot correct vision to 20/20. The lenses can be tinted to ease **pain** from too much sunlight. For reading, children with albinism may or may not need materials with large print text, depending on the severity of their vision problems. There is no cure for involuntary eye movements (nystagmus), and treatments for focusing problems (surgery or **contact lenses**) are not effective in all cases.

Crossed eyes (strabismus) can be treated during infancy by using eye patches, surgery, or medicine injections. Treatment may improve the appearance of the eye, but it can do nothing to cure the underlying condition.

Patients with albinism should avoid excessive exposure to the sun, especially between 10 a.m. and 2 p.m., and should wear long sleeves and pants if possible. If exposure cannot be avoided, they should use a UVA-UVB sunblock with an SPF rating at least 30.

Most children with albinism function satisfactorily in a mainstream classroom as long as the school provides classroom assistance for their vision needs. The child's eye doctor, the classroom teacher, and the school's vision resource teacher should work as a team with the parents to determine what classroom modifications and vision aids will best help the child. The local school district or the state agency for the blind should be contacted during the child's infancy or during **preschool** years to determine what assistive services might be available for the child. Early intervention allows parents and teachers to develop an educational plan for the child.

Prognosis

In the United States, people with this condition can expect to have a normal lifespan. However, one of the greatest health hazards for people with albinism is excessive exposure to sun without protection, which may lead to skin cancer. Wearing opaque clothes and sunscreen with at least an SPF rating of 30, people with albinism can safely work and **play** outdoors even during the summer.

Prevention

Genetic counseling should be considered for individuals with a family history of albinism.

Parental concerns

Children with albinism may experience complex social problems because of their unusual appearance, especially when a member of a normally dark-skinned ethnic group has albinism. The eyes of children with albinism may move rapidly and not focus together, and the children may have to squint, tilt their heads, and hold reading materials close in order to see. These behaviors may result in the child being treated badly by peers. Modifications made for the children in the classroom and in physical education classes to compensate for low vision may make them feel even more isolated. Parents often feel that teasing and name-calling, insensitivity, and ignorance are the greatest challenges that they face with regards to their child's albinism. The support and love of these families of children with albinism are essential to helping the children understand and accept themselves. Support groups for the children and for their families, as well as counseling, may be useful for developing means of coping with the social effects of albinism.

Resources

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ORGANIZATIONS

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American Foundation for the Blind. 11 Penn Plaza, Suite 300 New York, NY 10001. Web site: <www.afb.org/>.

Hermansky-Pudlak Syndrome Network Inc. One South Road, Oyster Bay, NY 11771–1905. Web site: <www.hpsnetwork.org/HTML/frame.html>.

National Organization for Albinism and Hypopigmentation (NOAH). PO Box 959, East Hampstead, NH 03826–0959. Web site: <www.albinism.org>.

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Alcoholism

Definition

Alcoholism, or alcohol dependence, is described in the *Diagnostic and Statistical Manual of Mental Disorders (DSM-IV)* as "A maladaptive pattern of alcohol use, leading to clinically significant impairment or distress."

That maladaptive pattern is manifested, according to the *DSM-IV*, by the following behaviors occurring any time within one 12-month period:

- tolerance for alcohol
- withdrawal from alcohol

- alcohol taken in larger amounts and over a longer period of time than was intended
- persistent desire or unsuccessful efforts to cut down or control alcohol use
- much time spent in activities necessary to obtaining alcohol
- various important activities, for example, in socializing or at work, are given up or reduced because of alcohol use
- alcohol use continued regardless of the pattern of physical or psychological problems that it causes or worsens

Alcohol abuse has the same definition but is manifested by one (or more) of the following behaviors occurring within the same 12-month period:

- repeated alcohol use leading to failure to fulfill major role obligations at work, school, or home
- repeated alcohol use in situations in which it is physically hazardous
- repeated alcohol-related legal problems
- persistent alcohol use despite its causing social and interpersonal problems or exacerbating them

This definition and the criteria established by *DSM-IV* apply to both adults and children. The American Academy of Pediatrics (AAP) states that the pattern of use and abuse of alcohol in children and adolescents is not observably different from what is manifested in young people using marijuana or cocaine.

Description

The AAP divides the progression into childhood alcoholism into three stages:

- Stage 1: Experimentation with alcohol. Normally there is no change noted in physical status or behavior, and the drinking usually occurs only on weekends during social occasions with peers, making it the least detectable of the three. **Peer pressure** to use alcohol "just for fun."
- Stage 2: Actively trying to obtain alcohol. Alcohol use happens during the week to provide relief when stress is felt or to "feel good" when depressed.
- Stage 3: Preoccupation with alcohol. The child or adolescent has nearly lost the capacity for controlling alcohol use. Setting limits results in withdrawal symptoms, including depression, moodiness, or irritability. Severe withdrawal can result in serious medical problems, including delirium tremens.

Demographics

Prevalence and severity of the problem

Based on findings mostly coming from the 1990s, the National Council on Alcohol and Drug Dependence (NCAAD) cites the following in its fact sheet *Youth, Alcohol and Other Drugs*:

- Approximately 10.4 million Americans ages 12 to 20 have at least one drink per month. One fifth of these (2.1 million) are heavy drinkers who have five or more drinks on a minimum of five different occasions. More than half of these (6.8 million) are considered binge drinkers who have five or more drinks on a single occasion.
- Eighty percent of all high school seniors have tried alcohol.
- Eight percent of eighth graders, 24 percent of tenth graders, and 32 percent of high school seniors have been intoxicated from alcohol in the past month.
- Children who begin **smoking** tobacco before the age of 13 are significantly more at risk for alcohol problems.
- Among high school seniors, alcohol use is more prevalent among Caucasian and Hispanic students than among African American students.
- Junior, middle, and senior high school students consume 35 percent of all wine coolers sold in the United States as well as 1.1 billion cans of beer.
- A Southern Illinois University study showed that students with overall grades of D or F drank on average three times as much alcohol as students with overall grades of A.
- A United States Department of Justice survey showed that nearly one third of children below the age of 18 incarcerated in juvenile institutions are under the influence of alcohol at the time of their arrest.
- More than half (56%) of children and teens in grades five through 12 report that alcohol advertising encourages them to drink.
- Thirty percent of children in grades four through six state that they have received pressure from peers to drink beer.
- Two thirds of teenagers who drink report that they are able to make their own alcohol purchases.
- The total cost of alcohol use by young people, including automobile crashes, violent crime, alcohol **poisoning**, **burns**, drowning, **suicide** attempts, and **fetal alcohol syndrome** is more than 58 billion dollars each year.
- Eighty percent of teenagers do not know that a 12-ounce can of beer has the same amount of alcohol as a shot of whiskey or a five-ounce glass of wine.

Causes and symptoms

In their article "Early Identification and Intervention for Adolescent Alcohol Use," Mark Werner and Hoover Adjer Jr., both fellows at the American Academy of Pediatrics (AAP), state that attitudes regarding alcohol use are developed quite early in life, usually by the age of eight. Parental attitudes regarding alcohol and behaviors related to alcohol use have a major impact on how children and young adults view drinking alcohol. Not every child or teen who experiments with alcohol becomes an alcoholic, but NCADD studies have shown that children who drink before the age of 15 are four times more likely to become alcoholic than those who begin drinking after the age of 21. Some evidence supports a genetic component to this disease. Parents who are themselves alcoholic or problem drinkers are more likely to have children who develop alcohol dependence. Statistically, one in five children who have an alcoholic parent becomes an alcoholic, too.

Physical symptoms seen in adult alcoholics, such as gastritis, pancreatitis, hepatitis, or even cirrhosis, usually are absent in childhood alcoholics. Such physical damage normally takes longer to develop and is more typical of long-term adult alcoholics. More often in potential childhood alcoholics, behavioral symptoms provide the most significant clues.

These behavioral warning signs, according to the AAP, typically include the following:

- decline in school functioning, decreased attendance, poorer grades, and/or general deterioration in social functioning in school
- increased isolation outside school; rejection of usual long-term friendships in favor of new or different friends
- frequent arguments or less communication with **family** members; being more secretive
- marked changes in grooming and clothing styles
- noticeable increase in unexplained injuries and fights
- running away from home
- depressive symptoms such as weight loss, **sleep** problems, lethargy, feelings of hopelessness, mood swings, suicidal feelings, or suicide attempts
- evidence of the presence of risk-taking behaviors such as either driving while under the influence of alcohol or driving with others who are intoxicated, engaging in violent behaviors such as fights, or participating in unsafe sex

When to call the doctor

It is worth noting that these behavioral warning flags can appear in non-alcoholic children or teens and also are usually not observed before the second or third stage of childhood alcoholism. Parents observing some or all of these warning signs need professional help to both clarify diagnosis and plan treatment. Individual and family denial is considered a large portion of any alcohol problem. Parents need objectivity and open and honest communication with their children in order to deal effectively with childhood alcoholism and to know when to seek help.

Diagnosis

As noted, behavioral symptoms help to determine the diagnosis, but not usually until the second and third stage of the disease. There are assessments available that can provide both earlier identification and intervention for childhood alcoholism.

Diagnostic assessments for alcoholism, according to the APA, include:

- CAGE, a mnemonic that points to four key questions by highlighting key words: "Cut down," "Annoyed," "Guilty," and "Early" (see below)
- Alcohol Use Disorders Inventory Test (AUDIT)
- Personal Experience Screening Questionnaire (PESQ)
- Problem Oriented Screening Instrument for Teenagers (POSIT)

CAGE is an assessment guide containing the following four questions:

- C: Have you ever felt the need to *cut down* on your drinking?
- A: Do you get *annoyed* at criticism by others about your drinking?
- G: Have you ever felt *guilty* about your drinking or something you have done while drinking?
- E: Have you ever felt the need for a drink *early* in the morning?

Treatment

Once assessment has led to a diagnosed problem with alcohol, its severity determines the treatment needed. In "Early Identification and Intervention for Adolescent Alcohol Use," Werner and Adjer divide problem teen drinkers into three groups:

- The first category includes those teens who are using alcohol occasionally but still doing well emotionally and developmentally and who are not drinking and driving. The treatment objectives for this

group are to encourage abstinence and re-enforce **safety** by fostering the continuation of not driving while drinking and not driving with others who are drinking.

- The second category includes those teens who are more at-risk because while they are maintaining stability in physical, developmental, and emotional status, they are also drinking and driving. Professionals dealing with members of this group may not be able to maintain confidentiality, and people in this group may benefit from an introduction to organizations such as Students Against Drunk Driving (SADD).
- The third category includes those showing serious signs of impairment, including inability to follow through on obligations at school or on a job, alcohol-related encounters with police or the justice system, and mental health problems such as **anxiety**, depression, or oppositional-defiant behavior. These children may experience frequent acute intoxication or withdrawal symptoms, medical complications, or an inability to stop or reduce their alcohol intake. Werner and Adjer suggest that professionals dealing with members of this group probably need to set aside confidentiality in order to involve parents in the treatment process. Treatment may include detoxification in an in-patient facility and/or rehabilitation in a youth-centered substance abuse program.

Treatment options

The following key issues should be considered in determining which treatment option is appropriate:

- severity of the problem and evidence to suggest other mental health problems (e.g. depression, suicide attempts)
- staff credentials of those treating the child or teen, and what forms of therapy (e.g., family, group, medications) are to be used
- nature of family involvement
- how education is to be continued during treatment
- if an in-patient program is necessary, what length it should be
- what aftercare is to be provided following discharge
- what portion of treatment is to be covered by health insurance and what needs to be paid out of pocket

Alcoholics Anonymous

Since its inception in the 1930s, Alcoholics Anonymous (AA) has been an important non-medical means of treating alcoholism with millions of members worldwide, many of whom are teenagers. It is a spiritual but non-religious program that fosters abstinence from alcohol based upon a belief that the person suffering from alcoholism is "powerless" over their **addiction**. AA suggests that people can stay free of alcohol by using an attitude that focuses on "one day at a time" and that consciously seeks spiritual support from "a power greater than themselves." AA is generally a part of most in-patient treatment and rehabilitation programs.

Prognosis

Prevention provides the best possible prognosis for alcohol abuse and dependence. The National Council on Alcoholism and Drug Dependence estimates that parents who talk with their children regularly about the danger from drugs (including alcohol) have children who are 42 percent less likely to use these substances. Once alcoholism is present, abstinence is the only known completely successful treatment. Children suffering from alcohol dependence continue for the rest of their lives to be at risk for problems with alcohol if they again drink. The prognosis is excellent for young alcoholics who remain alcohol-free and who do not substitute other drugs for alcohol, sometimes called "chewing their booze" in AA.

Prevention

Alcohol use and abuse has been a feature of Western culture for centuries, a facet of American life since Europeans arrived in North America, literally arriving with the pilgrims on the Mayflower. It is typically part of U.S. celebrations and even some American-observed religious rites. Because alcohol overuse and abuse has been so much a part of Western experience, there is clearly a tremendous need–among children and adults—for better education about both alcohol consumption and alcoholism. The statistics indicate that parents, teachers, and healthcare professionals need to begin educating children as early as possible regarding the risks involved in alcohol use. Parents who provide the example of limiting their own alcohol and other drug use can help their children inestimably. It should never be inferred that difficult situations can be better coped with by having a drink or that getting drunk is either helpful or amusing. Moreover, parents and other adults need to set the example by not driving a car or operating machinery while they are drinking or under the influence of alcohol.

The APA suggests the following as the most effective ways that parents can aid their children in resisting drinking:

- Provide children with self-confidence by building **self-esteem** and not engaging in constant criticism. Good self-esteem is the best defense against peer pressure to drink.
- Listen to children. Parents who listen attentively and provide support during difficult times give their children invaluable aid in coping with pressures.
- Get to know the children's friends.
- Provide supervision and discourage teens from attending parties where alcohol is served or parents are absent, and band together with other parents to arrange alcohol-free social events for children.
- Be available and encourage children and teens to call home for a ride rather than drive with someone who has been drinking; assure children there will be no recrimination, as SADD recommends.
- Teach therapeutic coping mechanisms by modeling how to handle stress, **pain**, or tension in healthy ways, by exercising, using **yoga** and meditation, and talking about feelings.
- Understand the tremendous importance of child and adolescent issues, including alcohol and other drug use and acceptance by peers; be ready and able to discuss these subjects with children.
- Encourage and participate in enjoyable, worthwhile activities with children; be reassuring that there is time enough for both work and fun.
- Be willing to learn about alcohol abuse; attend, along with their children, programs offered by schools, churches, and other groups providing information about the prevention of alcohol abuse.
- Maintain healthy lines of communication with children; remember the saying, "You are only as sick as the secrets you keep."

Nutritional concerns

Most childhood alcoholics do not reach the serious state of **malnutrition** that chronic adult alcoholics can reach. However, severe cases of alcohol abuse and dependence may result in a child or teen not eating normally, resulting in weight loss and vitamin deficiencies (B-vitamins particularly). Resumption of normal eating habits and possible addition of vitamin supplements can help in regaining normal **nutrition**.

Parental concerns

Parents of alcoholic children often encounter persistent and highly traumatic worries regarding serious physical, emotional, social, and legal problems for the affected child as well as the terrifying possibility of that child's death or serious injury. Clearly, these concerns can take a huge toll in a family. The denial mentioned earlier is often a complicating factor. The notion of "Not my son or daughter!" can actually hinder treatment and recovery for a child. Parents are also often embarrassed by their child's alcohol abuse and may believe that it is somehow their fault. It is not uncommon for parents to feel isolated and to feel as if they are the only ones with this problem. High school programs such as the earlier-described SADD can address some of the fear regarding drunk driving. Parental support groups such as *Tough Love* programs and twelve-step groups such as Al Anon can help parents to better understand the problem they facing and can help make them aware that they not alone and that they have options.

Al Anon is a twelve-step program (that is, a program based on the twelve steps employed by Alcoholics Anonymous) that provides support and spiritual recovery for the families and loved ones of alcoholics. This program was begun in the early 1940s by the wives of some of AA's earliest members and founders, including Lois Wilson, wife of AA founder Bill Wilson. Twelve-step programs have spun off over the years to meet the needs of specific populations, including adult children of alcoholics and teens who are alcoholics.

Because parents of alcoholic children often believe they are responsible for their child's drinking, the *Three C's* that Al Anon offers its participants may be particularly helpful. These *Three C's* state:

- "I didn't cause anyone else's alcoholism."
- "I can't control anyone else's alcoholism."
- "I can't cure anyone else's alcoholism."

Tough Love, a program begun by Phyllis and David York, co-authors of a book by the same title, is designed to help families and especially parents cope with a variety of problems, including alcoholism and drug abuse, that often affect children and ultimately the whole family. *Tough Love*'s approach is different from that of Al Anon. The ten beliefs listed below form the basis for this program and show clearly the difference in philosophy:

- Parents are people too.
- Parents' material and emotional resources are limited.
- Parents and kids are not equal.
- Blaming keeps people helpless.

KEY TERMS

Alcohol Use Disorders Inventory Test (AUDIT)—A test for alcohol use developed by the World Health Organization (WHO). Its ten questions address three specific areas of drinking over a 12-month period: the amount and frequency of drinking, dependence upon alcohol, and problems that have been encountered due to drinking alcohol.

Binge drinking—Consumption of five or more alcoholic drinks in a row on a single occasion.

CAGE—A four-question assessment for the presence of alcoholism in both adults and children.

Delirium tremens—A complication that may accompany alcohol withdrawal. The symptoms include body shaking (tremulousness), insomnia, agitation, confusion, hearing voices or seeing images that are not really there (hallucinations), seizures, rapid heart beat, profuse sweating, high blood pressure, and fever.

Detoxification—The process of physically eliminating drugs and/or alcohol from the system of a substance-dependent individual.

Personal Experience Screening Questionnaire (PESQ)—A questionnaire for alcoholism.

Problem Oriented Screening Instrument for Teenagers (POSIT)—A questionnaire used specifically for teenagers to assess alcohol and drug use.

Students against Drunk Driving (SADD)—An organization that offers a "Contract for Life" that asks teens to discuss substance use with parents, to call home for a ride if safe transportation is needed, and to wear a seat belt. Parents in turn promise to arrange for that safe transportation home "regardless of the time or circumstances," without discussion of the incident until both teens and parents are calm.

Tolerance—A condition in which an addict needs higher doses of a substance to achieve the same effect previously achieved with a lower dose.

Twelve-step programs—Several programs to assist in breaking addictions, offering either support to addicted people or to friends and loved ones of addicted people. These programs are spiritual but not religious and are based on the twelve steps that are the basis of Alcoholics Anonymous (AA). Programs include AA, Narcotics Anonymous (NA), Al-Anon, Adult Children of Alcoholics (ACOA), Alateen, and Co-Dependence Anonymous (CODA).

Withdrawal—The characteristic withdrawal syndrome for alcohol includes feelings of irritability or anxiety, elevated blood pressure and pulse, tremors, and clammy skin.

- Kids' behavior affects parents. Parents' behavior affects kids.
- Taking a stand precipitates a crisis.
- From a controlled crisis comes the possibility of positive change.
- Families need to give and get support in their own community in order to change.
- The essence of family life is cooperation, not togetherness.

Al Anon and *Tough Love* are offered in a variety of formats to the families of alcoholic children through treatment centers, churches, and other community services. It is clear that there are dramatic differences between these two philosophies. But perhaps the best way for parents to decide which approach makes sense to them is to take the advice Al Anon offers all newcomers: "Take what you like and leave the rest."

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Allergic purpura

Definition

Allergic purpura (AP), a form of vasculitis (inflammation of the blood vessels), is a disease characterized by inflammation of the small arterial vessels (capillaries) in the skin, kidneys, and intestinal tract. Symptoms include a purple spotted skin rash, abdominal **pain**, gastrointestinal upsets, and joint inflammation, swelling, and pain. Although the exact cause of the disease is unknown, it often develops following a recent viral or bacterial infection of the respiratory tract and is an abnormal reaction of the immune system to the infection.

Description

AP may occur suddenly, or it may develop slowly over a period of weeks. The characteristic rash is always present in the disease. The rash begins as areas of redness and as small **hives**, which may develop anywhere on the body, but especially on the legs and buttocks, and may itch. The rash is caused by inflamed capillaries rupturing, allowing small amounts of blood to accumulate in the surrounding tissues. Through time the rash changes color from red to a bruised, purple color. Each rash spot will last about five days, and the rash can reoccur several times. The skin rash is the most obvious symptom of AP but is not the most serious, for the joints, gastrointestinal tract, and kidneys may also be affected.

Joint inflammation (arthritis), especially of the knees and ankles, occurs in two-thirds of the children affected by AP. The joints become swollen, tender, and painful with movement; the pain may be debilitating. However, the arthritis usually clears up with no permanent damage.

Gastrointestinal symptoms are a result of inflammation and bleeding of the capillaries in the gastrointestinal tract, including the mouth, esophagus, stomach, and intestines. Most children with AP experience severe abdominal pain, **vomiting** (possibly with blood), and bloody stools.

The most serious complication of AP is kidney inflammation (nephritis), which occurs in almost half of older affected children. Symptoms include blood and protein in the urine. Most children whose kidneys are affected recover fully, but about 10 percent later develop more serious chronic kidney disease. In 80 percent of those with kidney involvement, the kidney disease develops within the first four weeks of illness.

Children younger than three years who contract AP have a shorter, milder course of the disease with fewer recurrences. Older children are more likely to have more serious symptoms.

AP is also called Henoch-Schonlein purpura, named after the two German physicians who first recognized and described it in the 1880s. AP is also referred to as anaphylactoid purpura or vascular purpura.

Demographics

AP is the most common acute vasculitis affecting children. In the United States, the prevalence of AP is approximately 14 to 15 cases per 100,000 population.

Approximately 75 percent of cases occur in children between the ages of two and 11, with peak prevalence in children aged five years. AP is rare in infants and younger children. Boys are affected more often than girls (the female-to-male ratio is 1.5-2:1). Most cases occur in late fall and winter. Adults can also develop the disease.

Causes and symptoms

Causes

AP is caused by reactions of antibodies binding with foreign proteins, called antigens. In some cases, the antigen-antibody complexes become too large to remain suspended in the bloodstream. When this occurs, they precipitate out and become lodged in the capillaries, which can cause the capillary to burst, resulting in a local hemorrhage.

The source of the antigens that cause AP is unknown. Antigens may be introduced by bacterial or viral infection, because more than 75 percent of children with AP report having had an infection of the throat, upper respiratory tract, or gastrointestinal system several weeks before the onset of AP. AP may also be caused by allergens, which are otherwise harmless substances that stimulate an immune reaction. Drug allergens that may cause AP include penicillin, ampicillin, erythromycin, and quinine. Vaccines possibly linked to AP include those for typhoid, **measles**, cholera, and yellow fever. Food allergens, cold exposure, and insect **bites** have also been associated with AP.

Symptoms

The onset of AP may be preceded by a **headache**, fever, and loss of appetite. Most children first develop an itchy skin rash. The rash is red, either flat or raised, and may be small and freckle-like. The rash may also be larger, resembling a bruise. **Rashes** become purple and then rust-colored over the course of a day, and fade after several weeks. Rashes are most common on the buttocks, abdomen, and lower extremities. Rashes higher on the body may also occur, especially in younger children.

Joint pain and swelling is common, especially in the knees and ankles. Abdominal pain occurs in almost all children with AP, along with blood in the stools. About half of all affected children show blood in the urine, low urine volume, or other signs of kidney involvement. Kidney failure may occur due to widespread obstruction of the capillaries in the filtering structures called glomeruli. Kidney failure develops in about 2–5 percent of all affected children and in 15 percent of those with elevated blood or protein in the urine.

Less common symptoms include prolonged headache, fever, and pain and swelling of the scrotum, scalp, eyelids, lips, ears, backs of the hands and feet, and perineum. Involvement of other organ systems may lead to heart attack (myocardial infarction), inflammation of the pancreas (pancreatitis), intestinal obstruction, bowel perforation, or acute intussusception (a twisting inversion of the lining of the bowel).

Diagnosis

Diagnosis of AP is based on the symptoms and their development, a careful medical history, and blood and urine tests. **X rays** or computed tomography (CT) scans may be performed to assess complications in the bowel or other internal organs. In some cases a renal biopsy may be useful to determine the extent of kidney involvement.

When to call the doctor

A doctor should be consulted if a child exhibits symptoms of AP. After a child has had an episode of AP, the doctor should be called if the child experiences sudden increases in abdominal pain, which may indicate a bowel infarction or perforation, or if the child exhibits decreased urine output, indicating kidney disease.

Treatment

Most cases of AP resolve completely without treatment. Nonetheless, a hospital stay with supportive treatment is usually required because of the possibility of serious complications. Non-aspirin pain relievers may be given for joint pain. Corticosteroids (such as prednisone) are sometimes used to alleviate gastrointestinal tract inflammation but have not been shown to be effective for associated kidney problems. Kidney involvement requires monitoring and correction of blood fluids and electrolytes. Salt intake should be restricted. A child with AP should be monitored until abnormal urinary findings subside.

Children with severe kidney complications may require a kidney biopsy so that tissue can be analyzed. Even after all other symptoms subside, elevated levels of blood or protein in the urine may persist for months and require regular long-term monitoring. **Hypertension** or kidney failure may develop months or even years after the acute phase of the disease. Kidney failure requires dialysis or transplantation.

Surgery may be necessary to correct acute intussusception of the bowel.

KEY TERMS

Capillaries—The tiniest blood vessels with the smallest diameter. These vessels receive blood from the arterioles and deliver blood to the venules. In the lungs, capillaries are located next to the alveoli so that they can pick up oxygen from inhaled air.

Glomerulus—Plural, glomeruli; a network of capillaries located in the nephron of the kidney where wastes are filtered from the blood.

Prognosis

AP may be mild, lasting only two or three days. However, for those children with moderate to severe symptoms, AP may last for four to six weeks, with relapses in about half of all children within six weeks, especially if the child contracts another respiratory infection or is exposed to the allergic agent. Relapses can occur up to seven years after the initial disease. Full recovery occurs in most cases without kidney involvement. However, one fourth of children who have kidney symptoms still have detectable problems years later. There is a higher likelihood of permanent renal damage with a higher number of recurrences.

Prevention

If the initiating trigger for a case of AP is identified in an affected child, everything possible should be done to ensure that the child is not exposed to that substance again. If the cause is thought to be a bacterial infection, such as **strep throat**, prophylactic antibiotic treatment is sometimes given once the infection has been treated to prevent recurrence.

Parental concerns

Parents should be vigilant regarding recurrence of symptoms after their child has had AP. Parents should also realize that although severe kidney involvement is rare, if it does occur, it may require aggressive treatment and long-term care. If the child does have long-term kidney problems, the stress of the illness can often be mitigated by parents joining a support group in which members share common experiences and problems.

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Judith Sims
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Allergic rhinitis

Definition

Allergic **rhinitis**, more commonly referred to as hay fever, is an inflammation of the nasal passages caused by allergic reaction to airborne substances.

Description

Allergic rhinitis (AR) is the most common allergic condition and one of the most common of all minor afflictions. AR affects up to 20 percent of children and 15 to 30 percent of adolescents in the United States. **Antihistamines** and other drugs used to treat allergic rhinitis make up a significant fraction of both prescription and over-the-counter drug sales each year.

There are two types of allergic rhinitis: seasonal and perennial. Seasonal AR occurs in the spring, summer, and early fall, when airborne plant pollens are at their highest levels. In fact, the term hay fever is really a misnomer, since allergy to grass pollen is only one cause of symptoms for most children. Perennial AR occurs all year and is usually caused by airborne pollutants in the home and other places. A child can be affected by one or both types. Symptoms of seasonal AR are worst after being outdoors, while symptoms of perennial AR are worst after spending time indoors.

Both types of **allergies** can develop at any age, although onset in childhood through early adulthood is most common. Although allergy to a particular substance is not inherited, increased allergic sensitivity may be genetic (inherited). While allergies can improve on their own over time, they can also become worse over time.

Demographics

AR affects up to 20 percent of children and 15 to 30 percent of adolescents. Boys are twice as likely to get allergic rhinitis as girls. Half of children develop the

condition before age 10, and half after that time. Some regions of the country are more likely to have the pollens that cause AR, so those areas will have more children with the condition. Other risk factors include having a mother with **asthma** or having asthma oneself, having others in the **family** with AR, being the oldest in the family, having a family dog, being breast fed for more than a month and having a higher socioeconomic level.

Causes and symptoms

Causes

Allergic rhinitis is a type of immune reaction. Normally, the immune system responds to foreign microorganisms, or particles like pollen or dust, by producing specific proteins, called antibodies. Antibodies are capable of binding to identifying molecules (antigens) on the foreign particle. This reaction between antibody and antigen sets off a series of reactions designed to protect the body from infection. Sometimes this same series of reactions is triggered by harmless, everyday substances. This is the condition known as allergy, and the offending substance is called an allergen.

Like all allergic reactions, AR involves a special set of cells in the immune system known as mast cells. Mast cells, found in the lining of the nasal passages and eyelids, display a special type of antibody called immunoglobulin type E (IgE) on their surfaces. Inside, mast cells store reactive chemicals in small packets called granules. When the antibodies encounter allergens, they trigger release of the granules, which spill out their chemicals onto neighboring cells, including blood vessels and nerve cells. One of these chemicals, histamine, binds to the surfaces of these other cells, through special proteins called histamine receptors.

Interaction of histamine with receptors on blood vessels causes neighboring cells to become leaky, leading to the fluid collection, swelling, and increased redness characteristic of a runny nose and red, irritated eyes. Histamine also stimulates **pain** receptors, causing the itchy, scratchy nose, eyes, and throat common in allergic rhinitis.

The number of possible airborne allergens is enormous. Seasonal AR is most commonly caused by grass and tree pollens, since their pollen is produced in large amounts and is dispersed by the wind. Showy flowers like roses or lilacs that attract insects produce a sticky pollen that is less likely to become airborne. Different plants release their pollen at different times of the year, so seasonal AR sufferers may be most affected in spring, summer, or fall, depending on which plants provoke a response. The amount of pollen in the air is reflected in the pollen count, often broadcast on the daily news during allergy season. Pollen counts tend to be lower after a good rain that washes the pollen out of the air and higher on warm, dry, windy days.

Virtually any type of tree or grass may cause AR. A few types of weeds that tend to cause the most trouble include the following:

- ragweed
- sagebrush
- lamb's-quarters
- plantain
- pigweed
- dock/sorrel
- tumbleweed

Perennial AR is often triggered by house dust, a complicated mixture of airborne particles, many of which are potent allergens. House dust contains some or all of the following:

- House mite body parts. All houses contain large numbers of microscopic insects called house mites. These harmless insects feed on fibers, fur, and skin shed by the house's larger occupants. Their tiny body parts easily become airborne.
- Animal dander. Animals constantly shed fur, skin flakes, and dried saliva. Carried in the air, or transferred from pet to owner by direct contact, dander can cause allergy in many sensitive people.
- Mold spores. Molds live in damp spots throughout the house, including basements, bathrooms, air ducts, air conditioners, refrigerator drains, damp windowsills, mattresses, and stuffed furniture. Mildew and other molds release airborne spores that circulate throughout the house.

Other potential causes of perennial allergic rhinitis include the following:

- cigarette smoke
- perfume
- cosmetics
- cleansers
- copier chemicals
- industrial chemicals
- construction material gases

Symptoms

Inflammation of the nose, or rhinitis, is the major symptom of AR. Inflammation causes **itching**, sneezing,

runny nose, redness, and tenderness. Sinus swelling can constrict a child's eustachian tube that connects the inner ear to the throat, causing a congested feeling and "ear popping." The drip of mucus from the sinuses down the back of the throat, combined with increased sensitivity, can also lead to throat irritation and redness. AR usually also causes redness, itching, and watery eyes. Fatigue and **headache** are also common.

When to call the doctor

AR that is not successfully treated by over-the-counter medication will benefit from an evaluation and treatment by a healthcare professional.

Diagnosis

Diagnosing seasonal AR is usually easy and can often be done without a medical specialist. When a child's symptoms appear in spring or summer and disappear with the onset of cold weather, seasonal AR is almost certainly the culprit. Other causes of rhinitis, including infection, can usually be ruled out by a physical examination and a nasal smear, in which a sample of mucus is taken on a swab for examination.

Along with a runny nose and reddened eyes, other symptoms may include dark circles under the eyes caused by nasal congestion, the "alleric salute" in which a child rubs a hand along the side of the nose, mouth breathing, sleepiness during the day, and learning problems caused by inability to concentrate during school.

Allergy tests including skin testing and provocation testing can help identify the precise culprit, but may not be done unless a single source is suspected and subsequent avoidance is possible. Skin testing involves placing a small amount of liquid containing a specific allergen on the skin and then either poking, scratching, or injecting it into the skin surface to observe whether redness and swelling occurs. Provocation testing involves challenging an individual with either a small amount of an inhalable or ingestible allergen to see if a response is elicited.

Perennial AR can also usually be diagnosed by careful questioning about the timing of exposure and the onset of symptoms. Specific allergens can be identified through allergy skin testing.

Treatment

Avoidance of the allergens is the best treatment, but this is often not possible. When it is not possible to avoid one or more allergens, there are two major forms of medical treatment: drugs and immunotherapy. Always read the package label for directions or consult your doctor or pharmacist before treating children with over-the-counter medications. Children are not small adults, but have different physiology. They are more susceptible than adults to the effects of certain medicines and may have unexpected reactions.

Drugs

ANTIHISTAMINES Antihistamines block the histamine receptors on nasal tissue, decreasing the effect of histamine release by mast cells. They may be used after symptoms appear, though they may be even more effective when used preventively, before symptoms appear. A wide variety of antihistamines are available.

Older (first generation) antihistamines often produce drowsiness as a major side effect. Such antihistamines include the following:

- Diphenhydramine (Benadryl and generics). May be used for children age 2 and up, depending on the type of delivery (capsule, liquid).
- Chlorpheniramine (Chlor-trimeton and generics). May be used for children age 6 and up.
- Brompheniramine (Dimetane and generics). May be used for children age 2 and up.
- Clemastine (Tavist and generics). May be used for children age 12 and up.

Newer antihistamines (second generation) that do not cause drowsiness are available by prescription or over-the-counter include the following:

- Loratidine (Claritin). May be used for children age 2 and up.
- Cetirizine (Zyrtec). May be used for children age 2 and up.
- Fexofenadine (Allegra). May be used for children age 6 and up.
- Azelastin HCl (Astelin). May be used for children age 5 and up.

DECONGESTANTS **Decongestants** constrict blood vessels to counteract the effects of histamine. Nasal sprays are available that can be applied directly to the nasal lining and oral systemic preparations are available. Decongestants are stimulants and may cause increased heart rate and blood pressure, headaches, and agitation. Use of topical decongestants for longer than several days can cause loss of effectiveness and rebound congestion, in which nasal passages become more severely swollen than before treatment.

TOPICAL CORTICOSTEROIDS Topical corticosteroids reduce mucous membrane inflammation and are

available by prescription. Allergies tend to become worse as the season progresses because the immune system becomes sensitized to particular antigens and can produce a faster, stronger response. Topical corticosteroids are especially effective at reducing this seasonal sensitization because they work more slowly and last longer than most other medication types. As a result, they are best started before allergy season begins. Side effects are usually mild, but may include headaches, nosebleeds, and unpleasant taste sensations.

However, a larger skin surface area to body weight ratio may make children more susceptible to adrenal gland problems such as growth retardation and delayed weight gain. Topical corticosteroids administration to children should be limited to the least amount possible to achieve therapeutic effect.

MAST CELL STABILIZERS Cromolyn sodium prevents the release of mast cell granules, thereby preventing release of histamine and the other chemicals contained in them. It acts as a preventive treatment if it is begun several weeks before the onset of the allergy season. It can be used for perennial AR as well. Cromolyn sodium is so low in side effects that it is recommended for children as young as two years of age.

Immunotherapy

Immunotherapy, also known as desensitization or **allergy shots**, alters the balance of antibody types in the body, thereby reducing the ability of IgE to cause allergic reactions. Immunotherapy is preceded by allergy testing to determine the precise allergens responsible. Injections involve very small but gradually increasing amounts of allergen, over several weeks or months, with periodic boosters. Full benefits may take up to several years to achieve and are not seen at all in about one in five patients. Individuals receiving all shots will be monitored closely following each shot because of the small risk of **anaphylaxis**, a condition that can result in difficulty breathing and a sharp drop in blood pressure. Allergy shots can be given to children as young as five years.

Alternative treatment

Alternative treatments for AR often focus on modulation of the body's immune response, and frequently center around diet and lifestyle adjustments. Chinese herbal medicine can help rebalance a person's system, as can both acute and constitutional homeopathic treatment. Vitamin C in substantial amounts can help stabilize the mucous membrane response. For symptom relief, western herbal remedies including eyebright (*Euphrasia officinalis*) and nettle (*Urtica dioica*) may be helpful. Bee pollen may also be effective in alleviating or eliminating AR symptoms.

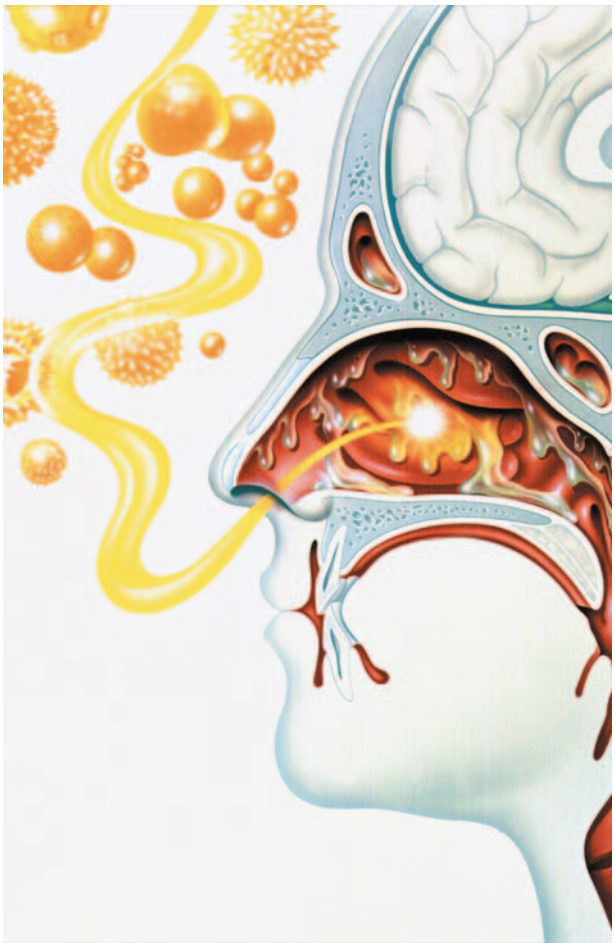

Illustration depicting excessive mucus production in the nose after inhalation of airborne pollen. *(Photograph by John Bavosi. Photo Researchers, Inc.)*

Prognosis

Most children with AR can achieve adequate relief with a combination of preventive strategies and treatment. While allergies may improve over time, they may also get worse or expand to include new allergens. Early treatment can help prevent an increased sensitization to other allergens.

Prevention

Reducing exposure to pollen may improve symptoms of seasonal AR. Strategies include the following:

- staying indoors with windows closed during the morning hours, when pollen levels are highest
- keeping car windows up
- avoiding uncut fields

KEY TERMS

Allergen—A foreign substance that provokes an immune reaction or allergic response in some sensitive people but not in most others.

Anaphylaxis—Also called anaphylactic shock; a severe allergic reaction characterized by airway constriction, tissue swelling, and lowered blood pressure.

Antibody—A special protein made by the body's immune system as a defense against foreign material (bacteria, viruses, etc.) that enters the body. It is uniquely designed to attack and neutralize the specific antigen that triggered the immune response.

Antigen—A substance (usually a protein) identified as foreign by the body's immune system, triggering the release of antibodies as part of the body's immune response.

Granules—Small packets of reactive chemicals stored within cells.

Histamine—A substance released by immune system cells in response to the presence of an allergen. It stimulates widening of blood vessels and increased porousness of blood vessel walls so that fluid and protein leak out from the blood into the surrounding tissue, causing localised inflammation of the tissue.

Mast cells—A type of immune system cell that is found in the lining of the nasal passages and eyelids. It displays a type of antibody called immunoglobulin type E (IgE) on its cell surface and participates in the allergic response by releasing histamine from intracellular granules.

- learning which trees are producing pollen in which seasons, and avoiding forests at the height of pollen season
- washing clothes and hair after being outside
- cleaning air conditioner filters in the home regularly
- using electrostatic filters for central air conditioning

Moving to a region with lower pollen levels is rarely effective, since new allergies often develop in children.

Preventing perennial AR requires identification of the responsible allergens.

Mold spores:

- keeping the house dry through ventilation and use of dehumidifiers
- using a disinfectant such as dilute bleach to clean surfaces such as bathroom floors and walls
- having heating/air conditioning ducts cleaned and disinfected
- cleaning and disinfecting air conditioners and coolers
- throwing out moldy or mildewed books, shoes, pillows, or furniture

House dust:

- vacuuming frequently, and changing the bag regularly (Use a bag with small pores to catch extra-fine particles.)
- cleaning floors and walls with a damp mop
- installing electrostatic filters in heating and cooling ducts, and changing all filters regularly

Animal dander:

- avoiding contact if possible
- washing hands after contact
- vacuuming frequently
- keeping pets out of the child's bedroom, and off furniture, rugs, and other dander-catching surfaces
- having pets bathed and groomed frequently

Parental concerns

AR can lead to daytime sleepiness in school and affect school performance.

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Christine Kuehn Kelly

Allergies

Definition

Allergies are abnormal reactions of the immune system that occur in response to otherwise harmless substances.

Description

Allergies are among the most common of medical disorders. It is estimated that 60 million Americans, or more than one in every five people, suffer from some form of allergy, with similar proportions throughout much of the rest of the world. Allergy is the single largest reason for school absence and is a major source of lost productivity in the workplace.

Allergies are a type of immune reaction. Normally, the immune system responds to foreign microorganisms, or particles, like pollen or dust, by producing specific proteins, called antibodies, that are capable of binding to identifying molecules, or antigens, on the foreign particle. This reaction between antibody and antigen sets off a series of reactions designed to protect the body from infection. When this same series of reactions is triggered by harmless, everyday substances, it is called an allergy. The substance that causes the allergy is called an allergen.

All allergic reactions involve a special set of cells in the immune system known as mast cells. Mast cells, found in the lining of the nasal passages and eyelids, display a special type of antibody, called immunoglobulin type E (IgE), on their surface. Inside, mast cells store reactive chemicals in small packets, called granules. When the antibodies encounter allergens, they trigger release of the granules, which spill out their chemicals onto neighboring cells, including blood vessels and nerve cells. One of these chemicals, histamine, binds to the surfaces of these other cells, through special proteins called histamine receptors. Interaction of histamine with receptors on blood vessels causes neighboring cells to become leaky, leading to the fluid collection in the body's tissues, swelling, and increased redness characteristic of a runny nose and red, irritated eyes. Histamine also stimulates **pain** receptors, causing the itchy nose, eyes, and throat common in **allergic rhinitis**. In the gastrointestinal tract, these reactions lead to swelling and irritation of the intestinal lining, which causes the cramping and **diarrhea** typical of food allergy. Allergens that enter the circulation may cause **hives**, angioedema, **anaphylaxis**, or **atopic dermatitis**. Allergens on the skin usually cause a delayed hypersensitivity reaction. This type of allergic response may develop over several days following contact with the allergen, and symptoms may persist for a week or more.

Demographics

According to the National Institute of Allergy and Infectious Diseases, annually, more than 50 million Americans suffer from allergic diseases, with approximately 36 million suffering from allergic **rhinitis**. Up to 6 percent of the general population suffers from an allergy to latex, and children with **spina bifida** who have had multiple surgical procedures are at higher risk for allergic reactions to latex. Atopic **dermatitis** is one of the most common skin conditions and occurs commonly in infants and children. Prevalence in the United States is about 10 percent. **Food allergies** occur in 8 percent of children aged six years and younger. Peanut or other nut allergies affect about 3 million Americans and produce the most severe reactions. Acute allergic hives affect from 10 percent to 20 percent of Americans at some time during their lifetime, and half of those affected have symptoms for more than six months. Allergies to stinging insects occur in about 3.5 percent of Americans. According to the American Academy of Allergy, **Asthma**, and Immunology, if one parent has an allergic disease, a child has a 48 percent risk of developing allergies. If both parents have allergies, risk increases to 70 percent.

Causes and symptoms

Allergens enter the body through four main routes: the airways, the skin, the gastrointestinal tract, and the circulatory system.

Airborne allergens cause the sneezing, runny nose, and itchy, bloodshot eyes of allergic rhinitis (hay fever). Airborne allergens can also affect the lining of the lungs, causing asthma, or the conjunctiva of the eyes, causing allergic **conjunctivitis**. The most common airborne allergens are the following:

- plant pollens
- animal fur and dander
- body parts and excrement from dust mites (microscopic creatures found in all houses)

- excrement from cockroaches
- house dust
- mold spores
- cigarette smoke
- solvents
- cleaners

Allergens in food can cause **itching** and swelling of the lips and throat, cramps, and diarrhea. When absorbed into the bloodstream, they may cause hives (urticaria) or more severe reactions involving recurrent, non-inflammatory swelling of the skin, mucous membranes, organs, and brain (angioedema). Some food allergens may cause anaphylaxis, a potentially life-threatening condition marked by tissue swelling, airway constriction, and drop in blood pressure. Common food allergens include the following:

- nuts, especially peanuts, walnuts, and brazil nuts
- fish, mollusks, and shellfish
- eggs
- wheat
- milk
- food additives and preservatives

In contact with the skin, allergens can cause reddening, itching, and blistering, called **contact dermatitis**. Skin reactions can also occur from allergens introduced through the airways or gastrointestinal tract. This type of reaction is known as atopic dermatitis. Dermatitis may arise from an allergic response (such as from **poison ivy**) or exposure to an irritant causing nonimmune damage to skin cells (such as soap, cold, and chemical agents). Injection of allergens, from insect **bites and stings** or drug administration, can introduce allergens directly into the circulation, where they may cause system-wide responses (including anaphylaxis), as well as the local ones of swelling and irritation at the injection site.

Common causes of contact dermatitis include the following:

- poison ivy, oak, and sumac
- nickel or nickel alloys
- latex

Insects and other arthropods whose **bites** or **stings** typically cause allergy include the following:

- bees, wasps, and hornets
- mosquitoes
- fleas

The following types of drugs commonly cause allergic reactions:

- penicillin or other **antibiotics**
- flu vaccines
- tetanus toxoid vaccine
- gamma globulin

Children and adolescents with allergies are not equally sensitive to all allergens. Some may have severe allergic rhinitis but no food allergies, for instance, or be extremely sensitive to nuts but not to any other food. Allergies may get worse over time. For example, childhood ragweed allergy may progress to year-round dust and pollen allergy. On the other hand, a child may outgrow allergic sensitivity. Infant or childhood atopic dermatitis disappears in almost all people. More commonly, what seems to be loss of sensitivity is instead a reduced exposure to allergens or an increased tolerance for the same level of symptoms.

Symptoms depend on the specific type of allergic reaction. Allergic rhinitis is characterized by an itchy, runny nose, sneezing, and often a scratchy or irritated throat due to postnasal drip. Inflammation of the thin membrane covering the eye (allergic conjunctivitis) causes redness, irritation, and increased tearing in the eyes. Asthma causes wheezing, coughing, and shortness of breath. Symptoms of food allergies depend on the tissues most sensitive to the allergen and whether the allergen spread systemically by the circulatory system. Gastrointestinal symptoms may include swelling and tingling in the lips, tongue, palate or throat; **nausea**; cramping; diarrhea; and gas. Contact dermatitis is marked by red, itchy, weepy skin blisters, and an eczema that is slow to heal. It sometimes has a characteristic pattern from the object containing the allergen, such as a glove allergy with clear demarcation on the hands, wrist, and arms where the gloves are worn, or on the earlobes by wearing earrings.

Whole-body or systemic reactions may occur from any type of allergen but are more common following ingestion or injection of an allergen. Skin reactions include the raised, red, and itchy patches called hives that characteristically blanch with pressure and resolve within 24 hours. A deeper and more extensive skin reaction, involving more extensive fluid collection and pain, is called angioedema. This response usually occurs on the extremities, fingers, toes, and parts of the head, neck, and face. Anaphylaxis is marked by airway constriction, blood pressure drop, widespread tissue swelling, heart rhythm abnormalities, and in some cases, loss of consciousness. Other symptoms may include **dizziness**, weakness, seizures, coughing, flushing, or cramping. The symptoms may begin within five minutes after

exposure to the allergen up to one hour or more later. Commonly, this is associated with allergies to medications, foods, and insect venoms. In some individuals, anaphylaxis can occur with **exercise**, plasma exchange, hemodialysis, reaction to insulin, radiocontrast media used in certain types of medical tests, and on rare occasions during the administration of local anesthetics.

When to call the doctor

Parents should consult a physician when a child has repeated and prolonged symptoms. Allergic rhinitis may be mistaken for a cold or other upper respiratory infection. Usually, a fever indicates an infection. Food allergies and allergies to insect stings or medications can be especially dangerous, causing anaphylactic reactions that require emergency treatment.

Diagnosis

Allergies can often be diagnosed by a careful medical history, matching the onset of symptoms to the exposure to possible allergens. Allergy is suspected if the symptoms presented are characteristic of an allergic reaction, and this occurs repeatedly upon exposure to the suspected allergen. **Allergy tests** can also be conducted to determine allergens.

Skin tests

Skin tests are performed by administering a tiny dose of the suspected allergen by pricking, scratching, puncturing, or injecting the skin. The allergen is applied to the skin as an aqueous extract, usually on the back, forearms, or top of the thighs. Once in the skin, the allergen may produce a classic immune wheal and flare response (a skin lesion with a raised, white, compressible area surrounded by a red flare). The tests usually begin with prick tests or patch tests that expose the skin to small amounts of allergen to observe the response. A positive reaction occurs on the skin even if the allergen is at levels normally encountered in food or in the airways. Reactions are usually evaluated approximately 15 minutes after exposure. Intradermal skin tests involve injection of the allergen into the dermis of the skin. These tests are more sensitive and are used for allergies associated with risk of death, such as allergies to antibiotics. Skin testing may be painful for children.

Provocation tests

These tests involve the administration of allergen to elicit an immune response. Provocation tests, most commonly done with airborne allergens, present the allergen directly through the route normally involved. Delayed allergic contact dermatitis diagnosis involves similar methods by application of a skin patch with allergen to induce an allergic skin reaction. Food allergen provocation tests require abstinence from the suspect allergen for two weeks or more, followed by ingestion of a measured amount of the test substance administered as an opaque capsule along with a placebo control. Provocation tests are not used if anaphylaxis is a concern given the patient's medical history.

Common childhood allergies

Type of allergy	Common triggers
Food allergies	Eggs, dairy products, peanuts, soy, wheat
Allergic rhinitis and asthma	Pollens, molds, dust mites, animal dander, cigarette smoke
Atopic dermatitis (eczema)	Food allergy (see above), irritating laundry or body soaps, scratchy fabrics, rubbing of fabric on skin, overheating
Other allergies	Insect stings, medications, latex (for children who are often exposed in a medical setting), poison ivy, oak, and sumac

(Table by GGS Information Services.)

Treatment

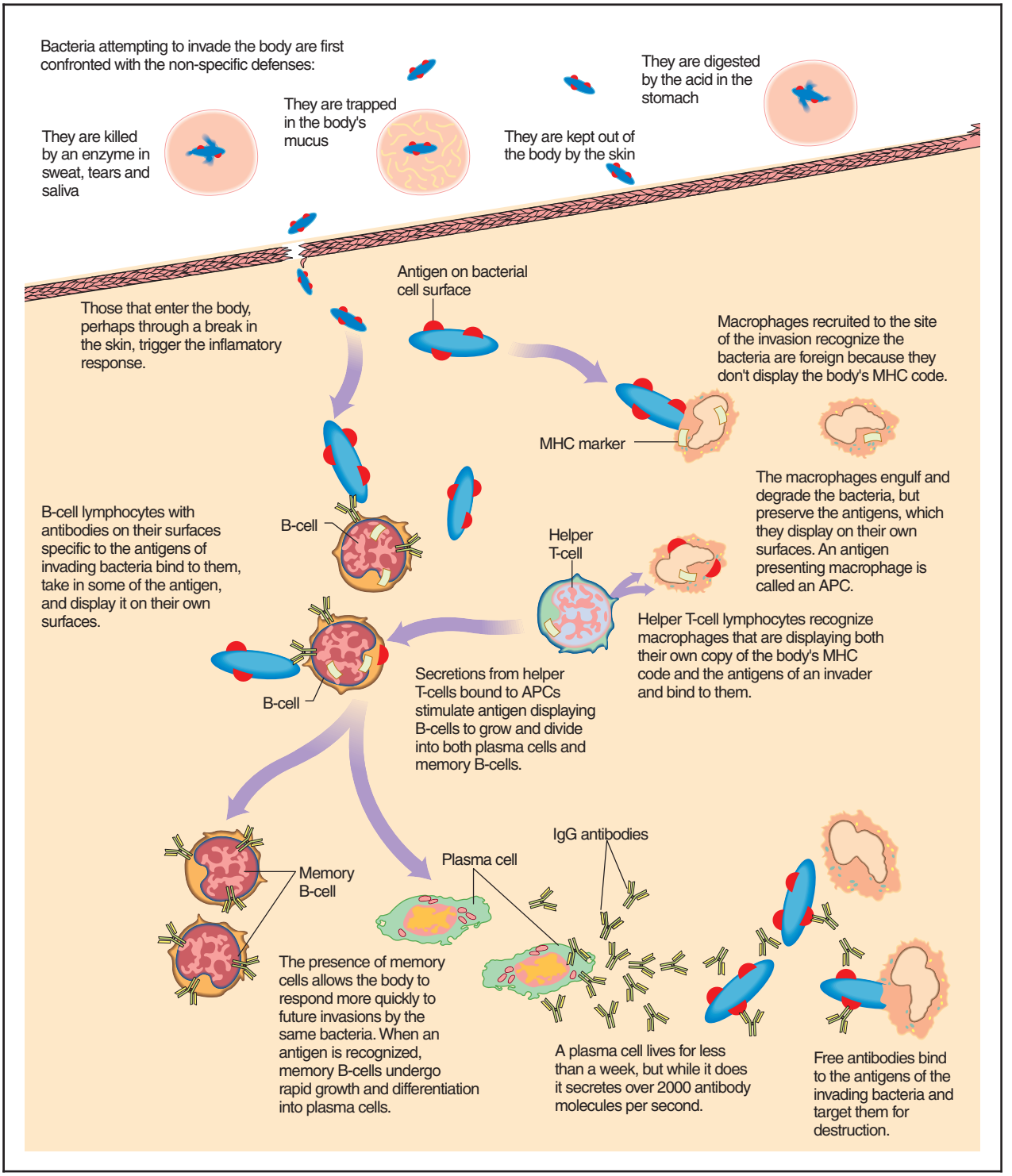
Avoiding allergens is the first line of defense to reduce the possibility of an allergic attack. However, complete environmental control is often difficult to accomplish; hence, therapeutic interventions are usually necessary. A large number of prescription and over-the-counter drugs are available for treatment of immediate hypersensitivity reactions. Most of these products work by decreasing the ability of histamine to provoke symptoms. Other drugs counteract the effects of histamine by stimulating other systems or reducing immune responses in general.

Antihistamines

Antihistamines are drugs used to treat the symptoms of allergic rhinitis by blocking the action of histamine, a chemical released by the immune system in allergic reactions. Antihistamines are available as prescription and over-the-counter tablets, topical gels or creams, nasal sprays, and eye drops.

Commonly used antihistamines include the following:

- diphenhydramine (Benadryl)
- loratadine (Claritin)
- cetirizine (Zyrtec)



Flow chart depicting a response to an allergen introduced into the body, causing an allergic response. *(Illustration by Hans & Cassidy.)*

- fexofenadine (Allegra)
- clemastine fumarate (Tavist)
- chlorpheniramine (Chlor Trimeton)
- brompheniramine (Dimetapp)

Decongestants

Decongestants dry up nasal passage tissues and reduce swollen nasal membranes so as to relieve congestion. Decongestants are available as nasal sprays or drops, oral tablets, or syrups. Decongestants are stimulants and may cause increased heart rate and blood pressure, headaches, and agitation. Use of nasal spray decongestants for longer than four to five days can cause loss of effectiveness and rebound congestion, in which nasal passages become more severely swollen than before treatment. Saline nasal sprays, which do not contain decongestants, may be used for longer periods of time to help congestion and nasal passage irritation.

Commonly used decongestants include the following:

- oxymetazoline (Afrin)
- pseudoephedrine (Sudafed)
- phenylephrine (Neo Synephrine)

Corticosteroids

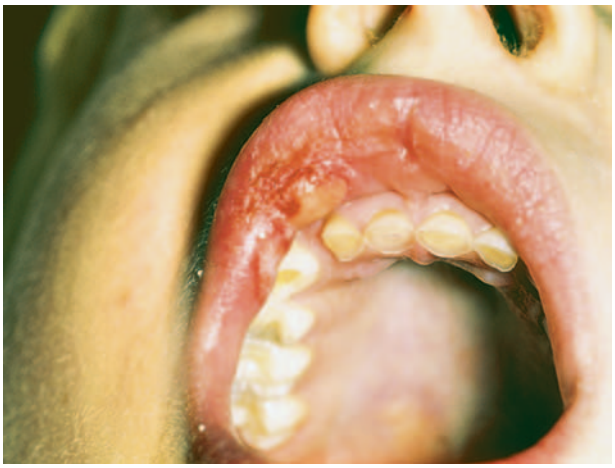
Corticosteroids reduce mucous membrane inflammation and are available by prescription and taken as a series of oral tablets. Corticosteroids are also available as nasal sprays. Allergies tend to become worse as the season progresses because the immune system becomes sensitized to particular antigens and can produce a faster, stronger response. Corticosteroids are especially effective at reducing this seasonal sensitization because they work more slowly and last longer than most other medication types. Side effects may include headaches, nosebleeds, and unpleasant taste sensations. Long-term use of oral corticosteroids may cause more serious side effects, such as weight gain, cataracts, weakening bones, high blood pressure, elevated blood sugar, and easy bruising.

Mast cell stabilizers

Cromolyn sodium prevents the release of mast cell granules, thereby preventing release of histamine and the other chemicals contained in them. Cromolyn sodium is available in nasal sprays or via an inhaler. It is most frequently prescribed when allergic rhinitis is accompanied by asthma.

Immunotherapy

Immunotherapy, also known as desensitization therapy or **allergy shots**, alters the balance of antibody types in the body, thereby reducing the ability of IgE to cause allergic reactions. Immunotherapy is preceded by allergy testing to determine the precise allergens responsible. Injections involve very small but gradually increasing amounts of allergen, over several weeks or months, with periodic boosters. Full benefits may take up to several years to achieve and are not seen at all in about one in five patients. Individuals receiving all shots are monitored closely following each shot because of the small risk of anaphylaxis, a condition that can result in difficulty breathing and a sharp drop in blood pressure.



Close-up view of a boy with an allergic reaction on his lip as a result of contact with a latex glove. *(© Dr. P. Marazzi/Photo Researchers, Inc.)*

Treatment of contact dermatitis

An individual suffering from contact dermatitis should initially take steps to avoid possible sources of exposure to the offending agent. Calamine lotion applied to affected skin can reduce irritation somewhat, as can cold-water compresses. Topical antihistamine and corticosteroid sprays, gels, and creams are available to reduce itching. Side effects of topical agents may include overdrying of the skin. In the case of acute contact dermatitis, short-term oral corticosteroid therapy may be appropriate. Moderately strong corticosteroids can also be applied as a wrap for 24 hours. Healthcare workers are especially at risk for hand eruptions due to latex glove use.

Treatment of anaphylaxis

The emergency condition of anaphylaxis is treated with injection of adrenaline, also known as epinephrine. Children and adolescents who are prone to anaphylaxis because of food or insect allergies often carry an Epi-pen containing adrenaline in a hypodermic needle. Other medications may be given to aid the action of the Epi-pen. Prompt injection can prevent a more serious reaction from developing. Particular care should be taken to

assess the affected child's airway status, and he or she should be placed in a recumbent pose and vital signs determined. Emergency treatment may be required for severe reactions.

Nutritional concerns

For children and adolescents with food allergies, all foods must be monitored to make sure that the allergen is not an ingredient or was not used during preparation. In individuals with severe food allergies to peanuts, peanut oil used to fry foods, or even the fumes produced during cooking with peanut oil have been known to cause anaphylactic shock.

Parents whose children have allergies to foods, like milk and gluten, which are common ingredients in many other foods, can purchase gluten-free foods and lactose-free foods in most grocery stores. Cookbooks dealing with allergies to these foods are also available.

Prognosis

Allergies can improve over time, although they often worsen. While anaphylaxis and severe asthma are life threatening, other allergic reactions are not. Learning to recognize and avoid allergy-provoking situations allows most children and adolescents with allergies to lead normal lives.

Prevention

Avoiding allergens is the best means of limiting allergic reactions. For food allergies, there is no effective treatment except avoidance. By determining the allergens that are causing reactions most people can learn to avoid allergic reactions from food, drugs, and contact allergens such as poison ivy or latex. Airborne allergens are more difficult to avoid. Preventive measures for airborne allergens include the following:

- staying indoors with windows closed during the morning hours, when pollen levels are highest
- keeping car windows up while driving
- using a surgical face mask when outside
- avoiding uncut fields
- learning which trees are producing pollen in which seasons and avoiding forests at the height of pollen season
- washing clothes and hair after being outside
- regularly cleaning air conditioner filters in the home
- using electrostatic filters for central air conditioning

For mold spores, the following steps will help:

- keeping the house dry through ventilation and use of dehumidifiers
- using a disinfectant such as diluted bleach to clean surfaces such as bathroom floors and walls
- having air ducts cleaned and disinfected
- cleaning and disinfecting air conditioners and coolers
- throwing out moldy or mildewed books, shoes, pillows, or furniture

For house dust, the following steps will help:

- vacuuming frequently and changing the bag regularly; using a bag with small pores to catch extra-fine particles
- cleaning floors and walls with a damp mop
- installing electrostatic filters in heating and cooling ducts and changing all filters regularly

For animal dander, the following steps will help:

- avoiding contact if possible
- washing hands after contact
- vacuuming frequently
- keeping pets out of the bedroom and off furniture, rugs, and other dander-catching surfaces
- bathing and grooming pets frequently

Parents may find it helpful to keep an allergy journal for their child to track occurrence of allergic responses. For seasonal allergic rhinitis, they may use a calendar to note when symptoms begin and end. Documenting the level of seasonal allergens at the time can help determine when seasonal allergies tend to occur and what allergens affect the child. Local weather reports on television and on Web sites provide detailed allergen maps of pollen and mold/mildew spores. Antihistamines can then be taken as a preventive measure before symptoms begin each season. For children with allergies to foods, keeping a journal of foods eaten can help identify specific food allergens.

Parental concerns

For children who resist taking pills, many antihistamines are available as flavored chewable tablets, tablets that easily dissolve on the tongue, or flavored syrups. Because many over-the-counter allergy medicines contain multiple drugs, parents should be sure to read the prescribing and dosage information for any antihistamine their children are taking to ensure safe use.

Parents of children and adolescents with severe food and insect sting allergies that might result in sudden ana-

KEY TERMS

Allergen—A foreign substance that provokes an immune reaction or allergic response in some sensitive people but not in most others.

Allergic rhinitis—Swelling and inflammation of the nasal membranes caused by sensitivity to airborne matter like pollen or cat hair.

Anaphylaxis—Also called anaphylactic shock; a severe allergic reaction characterized by airway constriction, tissue swelling, and lowered blood pressure.

Angioedema—Patches of circumscribed swelling involving the skin and its subcutaneous layers, the mucous membranes, and sometimes the organs frequently caused by an allergic reaction to drugs or food. Also called angioneurotic edema, giant urticaria, Quincke's disease, or Quincke's edema.

Antibody—A special protein made by the body's immune system as a defense against foreign material (bacteria, viruses, etc.) that enters the body. It is uniquely designed to attack and neutralize the specific antigen that triggered the immune response.

Antigen—A substance (usually a protein) identified as foreign by the body's immune system, triggering the release of antibodies as part of the body's immune response.

Asthma—A disease in which the air passages of the lungs become inflamed and narrowed, causing wheezing, coughing, and shortness of breath.

Atopic dermatitis—An intensely itchy inflammation often found on the face, in the bend of the elbow, and behind the knees of people prone to allergies. In infants and young children, this condition is called infantile eczema.

Conjunctivitis—Inflammation of the conjunctiva, the mucous membrane covering the white part of the eye (sclera) and lining the inside of the eyelids also called pinkeye.

Contact dermatitis—Skin inflammation as a result of contact with a foreign substance.

Granules—Small packets of reactive chemicals stored within cells.

Histamine—A substance released by immune system cells in response to the presence of an allergen. It stimulates widening of blood vessels and increased porousness of blood vessel walls so that fluid and protein leak out from the blood into the surrounding tissue, causing localised inflammation of the tissue.

Immune hypersensitivity reaction—An allergic reaction that is mediated by mast cells and occurs within minutes of allergen contact.

Mast cells—A type of immune system cell that is found in the lining of the nasal passages and eyelids. It displays a type of antibody called immunoglobulin type E (IgE) on its cell surface and participates in the allergic response by releasing histamine from intracellular granules.

phylactic reactions should make sure that their children and any other **family** members and caregivers fully understand the severity of the allergic response and the need for immediate administration of epinephrine. Parents should consider having children with these severe allergies wear a medical alert bracelet.

Children with severe food allergies to whole food groups, such as milk or wheat, may require dietary management by a dietitian or nutritionist to ensure they receive the proper nutrients and a well-balanced diet. Breastfeeding mothers of highly allergic infants may need to eliminate suspected food allergens from their diets, because food proteins ingested by a mother can be transferred to the infant via breast milk. Special formulas are available for infants sensitive to breast milk, cow's milk, and soy milk.

Resources

BOOKS

Borrel, Marie. *60 Tips: Allergies*. London: Hachette Illustrated, 2004.

Cross, Linda. *How to Manage Your Child's Life-Threatening Food Allergies*. Lake Forest, CA: Plumtree Press, 2004.

Dadamo, Peter J. *Eat Right for Your Type Allergies*. East Rutherford, NJ: Penguin Group, 2005.

Ford, Jean. *Breathe Easy: A Teen's Guide to Allergies and Asthma*. Broomall, PA: Mason Crest Publishers, 2005.

Taylor, R., et al. *Allergy Relief and Prevention*, 3rd. ed. Vancouver: Hartley and Marks, 2000.

PERIODICALS

Ansel, K. "Working with the Food-Allergic Child. *Today's Dietitian*. 4 (November 2002): 36–39.

ORGANIZATIONS

Allergy and Asthma Network. 3554 Chain Bridge Road, Suite 200. Web site: <www.aanma.org>.

American Academy of Allergy, Asthma, and Immunology. 611 East Wells St, Milwaukee, WI 53202. Web site: <www.aaaai.org>.

Asthma and Allergy Foundation of America. 1125 15th Street NW, Suite 502, Washington, DC 20005. Web site: <www.aafa.org/>.

WEB SITES

"All About Allergies." *Nemours Foundation*. Available online at <www.kidshealth.org/parent/medical/allergies/allergy.html> (accessed October 26, 2004).

Jennifer E. Sisk, MA

Allergy elimination diet *see* **Elimination diet**

Allergy medication *see* **Antihistamines**

Allergy shots

Definition

Allergy shots, also called immunotherapy, are a form of treatment that reduces a person's allergic reaction to a particular allergen. Allergy shots can reduce symptoms of **allergic rhinitis** (hay fever) and allergic **asthma**. Allergy shots are less effective against molds and are not a useful method for treating **food allergies**.

Description

Allergy shorts are a series of injections with a solution containing the allergens that cause an allergic reaction. Treatment begins with a weak solution given once or twice a week. The strength of the solution gradually increases with each dose. The incremental increases of the allergen cause the child's immune system to become less sensitive to the substance by producing a "blocking" antibody. Once the strongest dose is reached, the child's sensitivity has decreased, and the injections are given monthly to control allergic symptoms.

General use

The term allergy refers to a person's immunologic sensitivity to any substance that causes an allergic reaction. **Allergies** can become obvious in the first few months of life in the form of nasal congestion, **cough**, rash, or **diarrhea** following food intake. Allergies affect all races and occur in all parts of the world.

Depending on the severity and nature of the allergies, allergen avoidance and allergy medications alone may not effectively manage symptoms in children. Allergens that doctors most commonly use in immunotherapy treatments for allergic **rhinitis**, allergic **conjunctivitis**, and allergic asthma include extracts of inhalant allergens from tree, grass, and weed pollens; mold spores; and dust mites. The doctor selects the treatment based on the patient's particular patterns of allergic response.

Allergy shots are not recommended for food allergies. However, if these allergies are left untreated, infants and children may be more likely to develop chronic allergies, asthma, and respiratory infections later in life. Furthermore, knowing and managing the child's sensitivities to food help in isolating the antigens that respond to immunotherapy. Parents can follow a few simple steps to reduce the child's risk to allergies:

- Pay attention to symptoms that persist, like eczema, earaches, or runny nose. See the pediatrician for treatment.
- Review the **family** history. If allergies run in the family, the child is likely to have them too.
- Minimize exposure to new foods in the first year. Avoid cow's milk until after the first birthday, eggs until the second birthday, and peanut butter or fish until age three. Introduce new foods in small servings.
- Eliminate from the child's diet foods suspected of being an allergy trigger and see if the symptom diminishes. Gradually reintroduce the food to see if the symptom returns. If a reaction recurs, avoid the food in the future.
- A simple blood test can help determine if the infant has allergies to certain foods and other substances. As children often outgrow allergies, they should have a second blood test to see if the allergies persist.

When to use immunotherapy

Parents considering allergy shots for their child should be referred to a board certified allergist. An allergist will follow specific steps to determine if allergy shots are necessary. The allergist will consider the child's age and general health status in deciding to start allergy treatments. The allergist will also inquire as to the child's environment (indoor and outdoor) and related symptoms to decide if testing is necessary. Allergy testing provides convincing evidence of specific antibodies to which the child is reacting. A pediatric allergist can diagnose the specific cause of the allergic reaction and

provide the correct allergen extracts. He or she may also consider the following factors when deciding whether a child would benefit from allergy shots:

- The child's particular allergen may be difficult or impossible to avoid because of exposure to environment.
- Expensive medications producing side effects that adversely affect the child's health and quality of life are necessary to manage allergy symptoms. If the child's deterioration in health and the cost of allergy drugs outweigh their benefits, allergy shots may be appropriate.
- A parental commitment to see the child through the therapy is necessary for immunotherapy to work. The treatments demand a significant investment of parental time and support.

Allergy shots are also effective in treating allergic asthma. Allergy shots can help relieve the allergic reactions that trigger asthma episodes and decrease the need for asthma medications.

Skin testing

The allergist may decide to conduct skin testing on a child to determine the specific allergen that is causing a reaction in the child. In a skin test, a small dose of suspect antigens is injected under the skin. The physician looks at the injected area 20 minutes later; if it is red, with a raised area (wheal) in the center, the reaction is positive.

Skin testing should only be performed under the supervisions of a board certified allergist. The child should be followed closely during a skin test, because occasionally skin testing causes a severe reaction. This condition is unusual, since the amount of allergen used is small; however, it can happen if the child is highly sensitive to the allergen.

Preparing the shot

Once the testing is finished, the allergist prepares an allergen abstract (serum) specially for the child. If the child is sensitive to multiple allergens, the physician may mix similar abstracts in one vial. Preparing the vials in combination extracts ensures that the child receives only one shot for each group of extracts, thus reducing the number of injections needed for effective therapy.

INJECTIONS The first intramuscular injection (shot) is important because based on this experience the parent and child build an attitude toward future injections. If the child is old enough to understand, the nurse or physician will explain why the child must receive the injection. The procedure should be explained in simple terms, and should proceed quickly and as gently as possible. The child should be allowed to express his **fear** and resentment of needles.

Although the healthcare professional will record the allergy shot in the medical record, the parent may want to keep an updated record of the treatment for quick reference during emergencies and when the child is traveling.

INJECTION SITES The following injections sites are recommended for children:

- Infants: Outer front thigh. The parent should place the child in a secure position to prevent movement of the extremity. The parent should hold and cuddle the infant following the injection.
- Toddlers and school-age children: Buttocks, upper outer side. The buttocks do not develop until the child begins to walk, so this site is used only when the child has been walking for one year or more.
- Older children: Upper portion of the buttocks. This site provides a dense muscle mass in older children, which eliminates the possibility of injuring the nervous and vascular organs. The disadvantage of this site is that it is visible to the child who may become apprehensive when the injection is given. Older children can also receive shots in the upper arm and in the upper outer part of the thigh.

After the injection, the parent and nurse should take time to praise the child for his cooperation. Infants and small children should be cuddled and given affection for a few minutes so they do not associate the experience only with the **pain**. A small child may be given a toy to divert attention. Older children may be allowed to select the site for the injection. In addition, nasal sprays are in the early 2000s being developed as a painless alternative to injection delivery.

Precautions

Strict adherence is essential to an effective immunotherapy program for children. Parents should maintain as exactly as possible the injection schedule that the allergist prescribes. They should also report immediately to their healthcare provider any adverse reactions to the treatment.

Children who receive allergy shots may develop serum sickness or other problems in reaction to the treatment. Serum sickness, an allergic reaction to serum contained in the allergy shot, may occur with the first injection or as a delayed reaction. Children may have a

KEY TERMS

Allergen—A foreign substance that provokes an immune reaction or allergic response in some sensitive people but not in most others.

Allergic conjunctivitis—Inflammation of the membrane lining the eyelid and covering the eyeball; congestion of the conjunctiva, with mucus secretion.

Allergic reaction—An immune system reaction to a substance in the environment; symptoms include rash, inflammation, sneezing, itchy watery eyes, and runny nose.

Anaphylaxis—Also called anaphylactic shock; a severe allergic reaction characterized by airway constriction, tissue swelling, and lowered blood pressure.

Antibody—A special protein made by the body's immune system as a defense against foreign material (bacteria, viruses, etc.) that enters the body. It is uniquely designed to attack and neutralize the specific antigen that triggered the immune response.

Antigen—A substance (usually a protein) identified as foreign by the body's immune system, triggering the release of antibodies as part of the body's immune response.

Asthma—A disease in which the air passages of the lungs become inflamed and narrowed, causing wheezing, coughing, and shortness of breath.

Desensitization—A treatment for phobias which involves exposing the phobic person to the feared situation. It is often used in conjunction with relaxation techniques. Also used to describe a technique of pain reduction in which the painful area is stimulated with whatever is causing the pain.

Leukotriene antagonist—An agent or class of drugs which exerts an action opposite to that of another.

Pediatric allergist—A board certified physician specializing in the diagnosis and treatment of allergic conditions in children.

Rhinitis—Inflammation and swelling of the mucous membranes that line the nasal passages.

Urticaria—An itchy rash usually associated with an allergic reaction. Also known as hives.

moderate fever, local redness and **itching**, or pain at the site of injection and a generalized skin eruption, most commonly urticaria (**hives**) associated with severe itching. The child may be uncomfortable but not seriously ill. A more severe and less common variety of serum sickness includes several of the following symptoms: malaise, protein in the urine, joint pains, swelling of mucous membranes with hoarseness and cough, vertigo, **nausea**, and **vomiting**. A rare and still more severe variety of serum illness produces extreme weakness approaching collapse; the child's temperature may be subnormal and the pulse weak. The rarest and most severe reaction, called **anaphylaxis**, produces immediate shock and can be fatal.

The symptoms of serum disease, particularly urticaria, may occur when a child acquires sensitivity to several drugs. Penicillin is the most frequent and important offender, and in these circumstances the symptoms are often delayed until days or even weeks after the penicillin therapy begins. Any of the symptoms of serum sickness may be mimicked. Immediate severe reactions to penicillin are almost unknown in children but can occur in adults.

There is an increased risk of a reaction with a variation of allergy shots which "rushes" the first phase of the treatment. In this treatment, steadily increasing doses of allergen extract are given every few hours instead of every few days or weeks. Rush immunotherapy should only be performed in a hospital under close supervision. Also, children who take medication that contain beta blockers for unstable heart conditions should not be given allergy shots unless the allergist thinks the benefits of starting immunotherapy outweigh the risks associated with suspending cardiac inhibitors.

Parental concerns

The goal shared by both physicians and parents in treating childhood asthma and allergies is to minimize medication side effects while maximizing the chance for children to lead normal lives. Parents can take the following steps to increase their child's comfort:

- Remove carpet, launder bed linens in hot water, and keep windows closed at night and in the early morning hours to minimize exposure to outdoor allergens.
- Avoid exposure to perfume and tobacco, or other forms of smoke.

- Monitor weather and seasonal changes in an effort to minimize exposure to pollen.
- Schedule outdoor playtime or **exercise** at non-peak pollen periods, such as afternoons or early evening.
- Have the child wear a mask when helping in the garden or cutting the lawn, vacuuming, or dusting.
- Involve the allergist in decisions regarding child's lifestyle.

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Aliene Linwood, RN, DPA

Allergy tests

Definition

Allergy tests evaluate levels of allergic sensitivity to commonly encountered allergens, which may be foods, pollen, chemicals, or other substances in the environment.

Purpose

When recurring symptoms in children indicate sensitivity to certain substances in the environment, allergy testing can be used to identify the particular allergens that my be triggering reactions. Using special testing techniques, the child will be exposed to small amounts of specific allergens to determine which of these might provoke a reaction or a "positive" result. Laboratory tests may also be conducted to identify allergens that react with allergy-related substances in the child's blood serum. Positive results from any allergy test may be used to narrow candidates for the allergen responsible for the reaction. Identification of the allergen may help parents avoid exposing their child to the substance and thereby reduce allergic reactions. In addition, allergy testing may be done on children with **asthma**, eczema, or skin **rashes** to determine if an allergy is causing the condition or making another condition worse. Allergy tests may also be done before allergen desensitization treatments to ensure the safety of additional exposure.

Description

Allergy is the reaction of the immune system to substances foreign to the body. It is normal for the immune system to respond to foreign microorganisms and particles, like pollen or dust, by producing antibodies against those substances. Antibodies are specific proteins the immune system manufactures to bind to corresponding molecules (antigens) on the cell surfaces of foreign organisms in an attempt to render them harmless. This antigen/antibody reaction is the body's way of protecting itself from invasion by harmful substances and the allergic responses or possible illness that may follow. In some sensitive individuals, excessive antibody production can be triggered by seemingly harmless, everyday substances in the environment. This reactive condition is commonly known as allergy, and the offending substance is called an allergen. Allergic disease arises in the sensitive child from either acute or chronic exposure to certain allergens by inhaling, ingesting, or touching them. Allergic reactions may be dose dependent; that is, longer exposure or exposure to larger amounts of the offending allergen may cause a greater response of the immune system and result in a stronger reaction. Common inhaled allergens include pollen, dust, cat dander, and insect parts from tiny house mites. Common food allergens, all protein-based, include nuts, shellfish, and milk. Allergic reactions can also be triggered by insect **bites**, molds and fungi, certain prescription drugs, plants such as **poison ivy** and **poison oak**, and irritating or toxic substances released into the air.

Allergic reactions involve a special set of cells in the immune system known as mast cells. Mast cells serve as guards in the tissues where the body meets the outside world: the skin; the mucous membranes of the eyes (conjunctiva), nose and throat (nasal and oral mucosa); and the linings of the respiratory and digestive (gastrointestinal) systems. Mast cells produce a special class of antibody, immunoglobulin E (IgE), that coats cell surfaces. Inside the mast cells are reactive chemicals in small

packets or granules. When the antibodies encounter allergens, mast cells release granules, which spill out their chemicals onto the cells of nearby tissues, including blood vessels and nerves. Histamine is the most notable of these chemicals, binding quickly to histamine receptors on cell surfaces. Interaction of histamine with receptors on blood vessel, nerve, and tissue cells causes inflammation and the accumulation of intracellular fluid released by the cells. The characteristic swelling and redness that accompanies **allergies** are the result, seen especially in an irritated nose and throat, a runny nose, and red, irritated eyes. Histamine also stimulates **pain** receptors, causing the itchy, scratchy nose, eyes, and throat common in **allergic rhinitis**.

Allergy tests may be performed on the skin or using blood serum in a test tube. During skin tests, potential allergens are placed on the skin and the reaction is observed. In radio-allergosorbent allergy testing (RAST), blood serum is combined with a specific concentration of potential allergens in a test tube, and the mixture is tested for antibody/antigen reactions. Provocation testing involves direct exposure to a likely allergen, either through inhalation or ingestion. It is sometimes performed to determine if symptoms develop on exposure to allergens identified in skin or RAST tests.

The range of allergens used for allergy testing is chosen to reflect possible sources in the environment and may include the following:

- pollen from a variety of trees, common grasses, and weeds
- mold and fungus spores
- house dust
- house mites
- animal skin cells (dander) and saliva
- food extracts
- antibiotics
- insect venoms

Skin testing is the most common type of allergy test. There are two forms of skin tests: percutaneous and intradermal. In percutaneous or prick testing, a drop of each allergen to be tested is placed on the skin, usually on the forearm or the back. A typical battery of tests may involve two dozen allergen drops, including a drop of saline solution that should never provoke a reaction (negative control) and a drop of histamine that should always provoke a reaction (positive control). A small needle is inserted through the drop to prick the skin below. A new needle is used for each prick. The sites are examined over the next 20 minutes for evidence of swelling and redness, indicating a positive reaction. In some instances, a tracing of the set of reactions may be made by placing paper over the tested area. Scratch testing, in which the skin is scratched instead of punctured, is used less often, but the principle is the same.

Intradermal testing involves directly injecting allergen solutions into the skin. Separate injections are made for each allergen tested. Observations are made over the next 20 minutes. As in percutaneous tests, a reddened, swollen spot develops at the injection site for each substance to which the child is sensitive. Skin reactivity can be seen for allergens whether they usually affect the skin. In other words, airborne and food allergens that are inhaled or ingested are capable of causing skin reactions when contact is made with mast cells.

Radio-allergosorbent testing (RAST) is a laboratory test performed on those who may be too sensitive to risk exposure to allergens through skin testing or when medications or skin conditions make testing unreliable. RAST testing involves obtaining a blood sample, usually venous blood from a vein in the arm. The sample will be centrifuged in the laboratory to separate the antibody-containing serum from the blood cells. The serum is then exposed to allergens bound to a solid-phase medium. If antibodies against a particular allergen are present, those antibodies will bind to the solid medium and remain attached after being rinsed. The antigen/antibody complex can be detected in the laboratory by adding specific immunoglobulins that are linked with a radioactive dye. The test is read by locating radioactive spots on the solid-phase medium, and a positive result is reported in each test in which reactive allergens are found.

Testing for **food allergies** is usually done through diet by a process of elimination, that is, by removing the suspect food from the diet for two weeks and then eating a single portion of the suspect food, followed by careful monitoring for symptoms. A slightly different method is to eat a simple, bland, prescribed diet for a period of two weeks, removing all possible food allergens. Suspect foods are then added to the diet one at a time and the individual is observed for reactions.

Provocation testing is done in some cases to confirm associations between exposure to certain allergens and the subsequent development of symptoms when skin testing or RAST tests have indicated possible sensitivity. In provocation challenges, the skin, nasal and oral mucosa, and lining of the lungs and gastrointestinal tract are exposed to suspected allergens. A purified preparation of the allergen is inhaled or ingested in increasing concentrations to determine if it will provoke symptoms. Oral food challenges with foods are more tedious than inhalation testing, since full passage through the

digestive system may take a day or more. The test involves gradual ingestion of increasing amounts of the suspect food, usually at timed intervals. The test is discontinued with a particular food when either gastrointestinal symptoms occur or it becomes clear that the food is tolerated. In bronchial provocation challenges, the individual inhales increasingly concentrated solutions of a particular allergen prepared in a nebulizer. Each inhalation is followed by measuring the exhalation capacity with a measuring tool called a spirometer. Only one allergen is tested per day. Because provocation tests may actually provoke an allergic reaction in sensitized individuals, treatment medications such as **antihistamines** are typically available during and following the tests, for administration as needed.

Precautions

While allergy tests are quite safe for most people, the testing involves additional exposure to allergens. The possibility of causing an exaggerated allergic response, a dangerous condition known as **anaphylaxis**, does exist. Anaphylaxis can result in difficulty breathing and a sharp drop in blood pressure. Individuals who have had prior anaphylactic episodes should inform the testing clinician. Skin tests should never include a substance to which the individual has had severe allergic reactions or that has previously caused anaphylaxis.

Provocation tests may provoke an allergic reaction by exposing the individual to reactive allergens. Treatment medications such as injectable antihistamines should, therefore, be available during and following the tests, to be administered if needed.

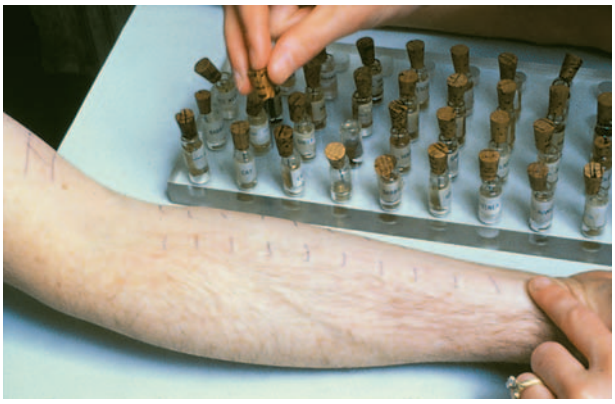
Preparation

Skin testing is preceded by a brief examination of the skin. The patient should refrain from using antiallergy drugs for at least 48 hours before testing. Prior to inhalation testing, children with asthma who can tolerate it may be asked to stop asthma medications. Testing for food allergies usually requires the child to avoid all suspect food for at least two weeks before testing.

The RAST test will usually require that a venous blood sample be drawn to obtain sufficient serum for the test. Parents can explain the procedure briefly to the child ahead of time to help reduce fears and encourage cooperation.

Aftercare

Skin testing does not usually require any aftercare. A generalized redness and swelling may occur in the test area, but it will usually resolve within a day or two.



A scratch test is used to identify reactions to allergens. *(Custom Medical Stock Photo Inc.)*

Inhalation tests may cause delayed asthma attacks, even if the antigen administered in the test initially produces no response. Severe initial reactions may justify close professional observation for at least 12 hours after testing.

If a blood sample has been drawn for RAST testing, a bandage may be applied to the venipuncture site to help keep it clean and to stop slight bleeding that may occur. Unusual bleeding or bruising of the site should be reported to the pediatrician.

Children should be observed closely for signs of allergic reactions after allergy testing using skin tests, inhalation tests, or provocation tests.

Risks

Intradermal testing may inadvertently result in the injection of the allergen into the circulation, with an increased risk of adverse reactions. Inhalation tests may provoke an asthma attack. Exposure to new or unsuspected allergens in any test carries the risk of anaphylaxis. Because patients are monitored following allergy testing, an anaphylactic reaction is usually recognized and treated promptly to reverse the condition. Occasionally, a delayed anaphylactic response can occur that requires immediate care. It is critical that physicians provide education about how to recognize anaphylaxis and tell patients what to do if it occurs at home.

Normal results

Lack of redness or swelling on a skin test indicates no allergic response. In an inhalation test, the exhalation capacity should remain unchanged. In a food challenge, no symptoms should occur.

KEY TERMS

Allergen—A foreign substance that provokes an immune reaction or allergic response in some sensitive people but not in most others.

Anaphylaxis—Also called anaphylactic shock; a severe allergic reaction characterized by airway constriction, tissue swelling, and lowered blood pressure.

Antibody—A special protein made by the body's immune system as a defense against foreign material (bacteria, viruses, etc.) that enters the body. It is uniquely designed to attack and neutralize the specific antigen that triggered the immune response.

Antigen—A substance (usually a protein) identified as foreign by the body's immune system, triggering the release of antibodies as part of the body's immune response.

Antihistamine—A drug used to treat allergic conditions that blocks the effects of histamine, a substance in the body that causes itching, vascular changes, and mucus secretion when released by cells.

Histamine—A substance released by immune system cells in response to the presence of an allergen. It stimulates widening of blood vessels and increased porousness of blood vessel walls so that fluid and protein leak out from the blood into the surrounding tissue, causing localised inflammation of the tissue.

Immunoglobulin G (IgG)—Immunoglobulin type gamma, the most common type found in the blood and tissue fluids.

Intradermal—An injection into a deep layer of skin.

Mast cells—A type of immune system cell that is found in the lining of the nasal passages and eyelids. It displays a type of antibody called immunoglobulin type E (IgE) on its cell surface and participates in the allergic response by releasing histamine from intracellular granules.

Abnormal results

Presence of redness or swelling, especially over 5 mm (0.25 inch) in diameter, indicates an allergic response. This reaction does not mean the substance actually causes the child's symptoms, however, since he or she may have no regular exposure to the allergen. In fact, the actual allergen may not have been included in the test array.

Following allergen inhalation, reduction in exhalation capacity of more than 20 percent, and for at least 10 to 20 minutes, indicates a positive reaction to the allergen and the sensitivity of the individual being tested.

Gastrointestinal symptoms within 24 hours following the ingestion of a suspected food allergen indicates a positive response and sensitivity to that food allergen.

Parental concerns

Parents who are already confronted with their child's allergies may be reluctant to have the child undergo testing. Physicians and medical personnel can assure parents that careful observation is involved in testing procedures and that allergens to which the child may have had severe reactions will be avoided. Appropriate medications will be available to treat the child immediately if a reaction is provoked by testing. It is important for parents to tell the child how the tests will be done so that the child is not anxious or apprehensive, which in some cases may increase the likelihood of false positive reactions.

See also Asthma; Allergic rhinitis; Drug allergies/sensitivities.

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Richard Robinson

Allowance and money management

Definition

An allowance is money earned or given to a child at regular intervals to teach the child how to manage money.

Purpose

Parents differ in their opinions about giving allowances to their children. Some parents believe that they should provide for the material needs of their children, and there is no reason a child should have to manage money until they are old enough to understand the working world and mature enough to make responsible purchases. Other parents feel that giving their children allowances is a good way to teach them about money and financial responsibility. If parents decide to give their children allowances, there are several ways to do it. How much they receive, how often they receive it, what they may spend it on, and whether the children must earn their allowance by completing chores at home are all questions for parents to consider.

Description

School-age children are starting to develop the cognitive skills necessary to understand basic monetary ideas, such as identifying coins, counting change, and matching small amounts of money to items they want to buy. Apart from introducing children to basic economics, money lessons have other benefits. Money illustrates parental values and teaches children about the relative worth of things, time, and effort.

When to start an allowance

Children younger than five are not mature enough to understand money management. They usually resist saving money and tend to spend money right way. Older children are more likely to take responsibility for their money. As they learn math skills, children are more able to calculate expenses. They can begin to figure how much they need to save for a item or how much they will have left over after buying something.

By the time children are five or six years old, they may be responsible enough to handle small amounts of money. Before starting an allowance, parents may discuss budgets with children and what children want to buy. They may shop with their children in stores on online and discuss prices.

The amount of allowance is a personal choice for parents. Young children may be given one or two dollars, while teenagers may be given ten dollars or more. Some families will give close-in-age siblings the same amount of allowance, but general practice is to give older children more money than younger children.

Young children may be given enough money to buy small items such as trading cards, hair clips, or ice-cream bars. The next time parents go shopping, the children can bring their own money if they think they might want to buy something. If they have already spent their allowance, then they have to wait for the next allowance before buying something else.

Building financial skills

Parents may want to reinforce lessons the children learn in school by making a chart that shows basic money equivalents. They may choose to post it on the refrigerator or in the child's room. Playing store and putting price tags on things around the house teach children relative worth of items.

Like adults, children may have trouble saving money. If a child wants to buy an item that costs more than his or her allowance, parents may choose to be flexible. They can allot the child an extra allowance or help the child figure out how long it would take to save the amount from future allowances. Parents may offer to provide matching funds, contributing a dollar for every dollar the child saves.

Some parents devise a category system to help their children manage their allowances. The first category is short-term expenses, money the child may spend right away on whatever he or she wants. The second category is savings, money put in a special jar, where its gradual accumulation is visible. This money is used for items the child wants that cost more than the weekly allowance. The third category is charity, money for church donation or a local cause, for example, or for gifts. The parents may decide how a younger child's allowance should be divided among the three categories, or the budgeting may be left up to an older child.

Allowances and chores

Many people believe that child's allowance should not be tied to household chores. Children should help out around the house because they are part of the **family**, not because they are paid. Allotting children chores that are proper to their abilities teaches responsibility and makes them feel the worth of their contribution to the family. The sense of belonging and empowerment gained by being an inherent part of a family team is important for

children. Children learn to contribute something valuable and realize that others depend on them to do their part. This relationship raises **self-esteem** and allows children to see themselves as active and valuable participants in others' lives.

Guidelines for spending the allowance

If parents expect children to pay for their needs, such as school clothes, **sports** gear, music lessons, or a comic book collection, or services the parents would otherwise pay for, the allowance has to be large. These items should be in the budget developed with the children's help.

Children need to learn that they can increase the money they have by saving it or working for it. Parents may create a list of jobs children can do above and beyond their regular chores, listing the amount of money the parents are willing to pay for these jobs.

Parental concerns

Some parents stop giving allowances to their teenagers at a certain age and encourage them to get a part-time job. Although teenagers may earn money from jobs outside the home, they may still need parental guidance to develop correct spending habits. They need to know how to save money and give to charity as well.

Parents can be authoritative without being dictatorial in teaching their children how to manage money. They can invite suggestions from the teenager about what he or she should buy and how much money to spend on it. Teenagers may be asked to buy gas for the family car when they drive it to work or to social events. They may be asked to use their earnings to pay their telephone service if they own a cell phone and to buy gifts with their own money for their friends and for the family.

Some families put teenagers in charge of all their own expenses, so they learn to budget. But some parents also maintain that teenagers should not take large sums of money from their account to make important purchases such as a car or motorcycle without parental permission. Money can become a difficult issue between parents and children of any age, and learning to be flexible can help each member of the family become more financially responsible.

Money management

Children's financial education extends throughout childhood. It occurs as they listen to parents talk about money; in their parents' wage earning patterns; and in their increasing experience with earning and spending.

Children can absorb a lot of knowledge from their parents about money management. Parent can **play** money games with them, using mail-order catalogs and price tags in stores, to teach values. When children are old enough, parents can discuss banking and investments. Rather than let children keep their financial gifts in a piggy bank in the house, parents can help children open a passbook savings account or a mutual fund and let them file the receipts for these accounts. Parents can also discuss with their children the impact of advertisement on personal spending habits and introduce their children to consumer advocacy publications such as *Consumers Report*.

KEY TERMS

Budget—A summary of income (allowance) and expenses (spending) for a given period of time.

Charity—Giving money or providing help to the poor and needy. To make a donation of money to a religious organization.

Chores—A small or minor job; a routine duty of a household or farm.

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Alopecia

Definition

Alopecia is the partial or complete loss of hair—especially on the scalp—either in patches (alopecia areata), on the entire head (alopecia totalis), or over the entire body (alopecia universalis).

Description

A basic understanding of hair biology and normal hair development is essential in distinguishing normal versus abnormal hair loss in children and adolescents.

Hair consists of the shaft and the root, which is anchored into a follicle beneath the epidermis. Hair is formed by rapid divisions of cells at the base of the follicle. Except for a few growing cells at the base of the root, hair, which is composed of keratin and other proteins, is dead tissue.

An individual hair follicle has a long growth phase, producing steadily growing hair for two to six years. About 80 percent to 90 percent of hair follicles are involved in this active growing period called the anagen phase. Next is a brief transitional phase (of about three weeks' duration)—the catagen phase—during which the hair follicle degenerates. About 5 percent of follicles are involved in the catagen phase.

Then a dormant period known as the telogen phase occurs. About 10 percent to 15 percent of hairs are involved in this phase, which lasts for approximately three months. Following the telogen phase, the growth phase begins again, and the growth cycle repeats.

Each person has about 100,000 hairs on their scalp. Although it is normal to lose between 25 and 100 hairs per day, any disruption of the hair growth cycle may cause abnormal hair loss.

Demographics

It is estimated that alopecia affects several million children in the United States and that hair loss is responsible for about 3 percent of all pediatric office visits.

Alopecia areata affects both sexes and all ages but is most common in children five to 12 years old. About one per 1000 children has alopecia areata. Approximately 5 percent of children with alopecia areata go on to develop alopecia totalis, and some of these children may develop alopecia universalis.

Tinea capitis (**ringworm**) affects an estimated 10 percent to 20 percent of susceptible children, and although the demographics are sketchy, telogen effluvium is the most common type of alopecia in both children and adults.

Causes and symptoms

Although in children and adolescents, hair loss may be caused by a wide variety of factors, most children experience hair loss as a result of one of four major causes:

- fungal infections
- alopecia areata
- trauma to the hair shaft
- telogen effluvium

A fungal infection called tinea capitis, which is similar to athlete's foot, is a common cause of hair loss, particularly among toddlers and early school-aged children. Tinea capitis, which affects the hair root, is a highly contagious condition and is often transmitted when a child uses the comb, brush, hat, or bed linen of an infected child. Tinea capitis seldom occurs after **puberty**.

Children with this condition usually have patchy hair loss with some broken hairs visible just above the surface of the scalp. The patches of hair loss are usually round or oval but are sometimes irregular in shape. When broken off at the surface, the hairs resemble small black dots on the scalp. Occasionally gray flakes or scales are present.

Alopecia areata, or localized baldness, is the sudden appearance of sharply defined circular or oval patches of hair loss, most often on the scalp. These patches are smooth and without inflammation, scaling, or broken hairs and may appear overnight or over the course of a few days. This condition may affect scalp hair, the eyebrows, eyelashes, genital area, and occasionally the underarms. The hair loss is not accompanied by other visible evidence of scalp disease, and the condition is not contagious.

In alopecia areata, immune system cells (white blood cells) attack the rapidly growing cells in the hair follicles that produce hair. The affected hair follicles decrease in size and hair production slows drastically. Because the stem cells that continually supply the follicles with new cells do not appear to be affected, the follicle retains the potential to regrow hair.

Although it is uncertain why the hair follicles undergo these changes, it is thought that a combination of genes may predispose some children and adults to the disease. In those who are genetically predisposed, some type of trigger—perhaps a virus or something in the child's environment—brings on the attack against the hair follicles.

Trauma to the hair shaft is another common cause of hair loss in children. Often the trauma is caused by traction resulting from, for example, tight braids, ponytails, or by friction (hats, hair bands, or rubbing against a bed). Trauma may also be caused by chemicals or **burns**.

Another important cause of hair trauma is called trichotillomania—a habit similar to thumb-sucking or nail-biting—of twirling or pulling out the hair. **Trichotillomania** is generally considered to be a nervous habit and may include the pulling of eyebrows and eyelashes.

The hair loss associated with trichotillomania is patchy and is characterized by broken hairs of varying length. Within the patches, hair loss is not complete. If the hair trauma is not severe or chronic enough to cause scarring, the child's hair usually regrows when the trauma ceases.

Telogen effluvium, another common cause of hair loss, affects both children and adults. This condition is responsible for more hair loss than any other cause except adult male-pattern baldness. In telogen effluvium, there is a physiologic basis to the hair loss; something happens to interrupt the hair's normal growth cycle and to drive many or all of the hairs into the telogen phase. Between six and 16 weeks later, partial or complete baldness occurs.

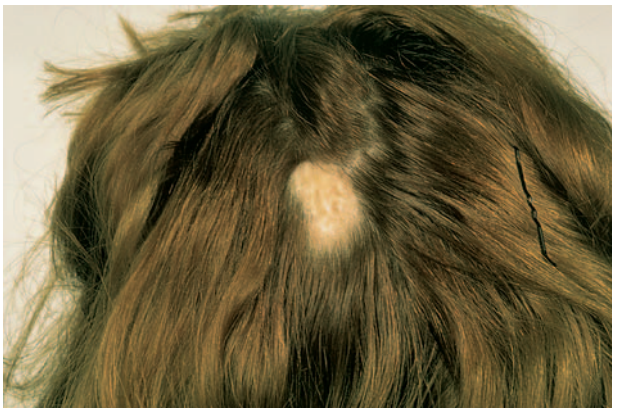
Many factors can cause telogen effluvium, including the following:

- high fever
- medications, including chemotherapy
- crash diets
- excessive vitamin A
- emotional stress
- surgery
- severe injury

In the telogen phase, a child's hair undergoes growth spurts and pauses. During the rest phase between spurts, the bulb at the end of the hair root decreases in volume and the hair loosens. Although exaggerated during **adolescence**, particularly in girls (due the influence of female hormones), even preadolescents may experience excessive hair loss on a daily basis. The scalp hair, however, appears normal in this condition.

When to call the doctor

It is important to consult a dermatologist or pediatrician if a child sheds hair in large amounts (more than 100 hairs per day for longer than four weeks) after combing, brushing, or shampooing or if the hair becomes significantly thinner. Also, if a child's scalp show signs of infection (redness, swelling, tenderness, warmth), consulting a physician is advised.



Bald spot on the scalp from the effects of alopecia. *(© Medis-can/Visuals Unlimited.)*

If children are observed pulling out their hair, eyelashes, or eyebrows, parents should consult a physician in order to determine the underlying cause of the habit.

Diagnosis

Because hair loss is caused by a variety of conditions, a physician diagnoses the cause of the child's hair loss based on medical history, **family** history of hair loss, medications (including **vitamins**), nutritional status, hair-care habits, and a physical examination.

If the physician suspects a fungal infection of the scalp, a hair sample may be tested by microscopic examination in the laboratory. Microscopic examination of a hair plucked at the periphery of the hair loss area often reveals a characteristic disruption of the integrity of the hair shaft. The infection may be confirmed by culturing the scalp for fungal organisms.

Blood tests or a scalp biopsy may be required if a medical condition—such as lupus erythematosus, thyroid dysfunction, iron deficiency, or hormonal imbalance—is suspected.

Treatment

Treatment varies with the cause of the hair loss. In some cases, early treatment is important in restoring the hair. Often congenital and hereditary hair loss and hair shaft abnormalities, however, have no effective treatment.

For fungal infections such as tinea capitis, treatment usually requires a systemic approach with an oral antifungal prescription medication such as griseofulvin (Fulvicin). This medication, which must be taken for four to eight weeks, is very effective in curing the infection and

restoring the hair. Early treatment is important in preventing possible permanent hair loss.

Topical creams or antifungal shampoos containing 2 percent ketoconazole are often used two to three times per week for eight weeks. Although shampoos and topical antifungal creams may decrease scaling, the infection usually returns because these products do not penetrate the hair follicle deeply enough to eradicate the infection.

A wide variety of treatments are available for alopecia areata. There has been some success with use of medications that suppress the immune system, including dinitrochlorobenzene (DNCB) and diphenylcyclopropenone (DPCP). The side effects of these drugs, however, may outweigh the benefits for a disease that most often resolves on its own.

In addition, topical creams or lotions such as minoxidil, cortisone (also injected into the scalp), or anthralin are sometimes used. Because such treatment triggers hair growth in bald patches but does not eradicate the disease, however, new bald patches can occur in other parts of the scalp even if new growth occurs.

Hair loss resulting from telogen effluvium or drug side effects usually requires no treatment. Hair loss from poor **nutrition** or medical illness usually stops with the adoption of a healthy diet and treatment of the underlying medical condition. Once the stressful event is over, complete hair growth usually occurs within six months.

Alternative treatment

There is some evidence to suggest that aromatherapy is a safe and effective treatment for alopecia areata. Aromatherapy involves rubbing scented essential oils into the skin to treat localized and systemic disease.

Massaging the essential oils of rosemary, lavender, sage, thyme, and cedar into the scalp is believed to increase circulation and reduce stress. About three to six drops of essential oil are added to 1 tablespoon of jojoba or grape seed oil and massaged into the scalp.

In addition to aromatherapy, stress reduction techniques such as **yoga**, meditation, or creative visualization may increase blood flow to the scalp and stimulate hair growth.

Prognosis

The prognosis for children with alopecia varies with the cause of hair loss. Certain types of alopecia respond more readily to treatment. For example, hair loss in telogen effluvium usually occurs over several weeks to months, then stops. Hair then grows back over the next several months.

KEY TERMS

Bulb—The hair bulb is the expanded portion on the lower end of the hair root.

Epidermis—The outermost layer of the human skin.

Follicle—A pouch-like depression.

Keratin—A tough, nonwater-soluble protein found in the nails, hair, and the outermost layer of skin. Human hair is made up largely of keratin.

Shaft—The portion of the hair that extends from the follicle and goes beyond the surface of the epidermis.

Overall, the outlook for children with alopecia areata is good. Alopecia areata usually resolves with time, although alopecia totalis is less likely to remit. With appropriate treatment, from 60 percent to 95 percent of children regrow all of their hair within one year.

Early treatment and the proper antifungal medications can cure tinea capitis, although patience is required because the condition may take several months to resolve.

Prevention

Although it may not be possible to prevent all types of alopecia—such as alopecia areata or hair loss associated with medical conditions—certain forms of hair loss may be prevented. Highly contagious fungal infections such as tinea capitis, for example, may be prevented by keeping hair clean and by teaching children not to share hats, combs, or hair brushes. In addition, adolescent girls should be cautioned not to share makeup.

It is important to teach children and adolescents to handle their hair with care, especially when shampooing, drying, brushing, combing, braiding, and using chemical processes. Hair is more fragile when it is wet, so vigorous towel drying and rough combing and brushing should be avoided. Wide-toothed combs and brushes with smooth tips are recommended.

Nutritional concerns

Children may experience hair loss or excessive thinning as a result of certain nutritional deficiencies. To prevent such hair loss, it is essential to include B-6, biotin, and folic acid—either in the diet or in supplement form.

It has been found that certain **minerals**, including magnesium, sulfur, silica, and zinc are also important for

maintaining healthy hair. Beta-carotene, which is converted to vitamin A in the body, is also essential to healthy skin, hair, and nails. Beta-carotene is found in green and yellow vegetables and fruits.

Because hair is composed of protein, a diet that is too low in protein may cause hair thinning or a disruption of the growth cycle. Thus eating a protein-rich diet often results in improved hair growth. In addition to lean meat, good food sources of protein include fish, eggs, dairy products, and beans.

Parental concerns

Because society has placed so much emphasis on appearance, hair loss, particularly if it is severe, may be emotionally devastating to children and adolescents. Hair loss can lead to embarrassment, low **self-esteem**, and depression. Thus it is important for parents to consult a physician as soon as possible to minimize not only the physical but also the emotional impact of hair loss on their child.

See also Dermatitis; Malnutrition; Trichotillomania.

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Genevieve Slomski, Ph.D.

Alpha-fetoprotein test

Definition

The alpha-fetoprotein (AFP) test is a blood test that is performed during pregnancy to screen the fetus for certain conditions; it is also used to screen for certain diseases in infants and children. The screening test measures the level of AFP in the mother's blood and indicates the probability that the fetus has one of several serious birth defects. The level of AFP can also be determined by analyzing a sample of amniotic fluid. This screening test cannot diagnose a specific condition; it only indicates the increase of risk for several birth defects. In infants and children, the AFP test is used to detect liver disease, certain cancerous tumors, and to monitor the progress of **cancer** treatment.

Purpose

Alpha-fetoprotein is a substance produced by the liver of a fetus, by tumors of the liver, by testes and ovaries, and by certain other diseases of the liver. The exact function of this protein was as of 2004 unknown. After birth, the infant's liver stops producing AFP; an adult liver contains only trace amounts. During pregnancy, the fetus excretes AFP in urine, and some of the protein crosses the fetal membranes to enter the mother's blood. The level of AFP can then be determined by analyzing a sample of the mother's blood.

By analyzing the amount of AFP found in a blood or amniotic fluid sample, doctors can determine the probability that the fetus is at risk for certain birth defects. It is very important that the doctor know precisely how old the fetus is when the test is performed, because the AFP level changes over the length of the pregnancy. AFP screening is used as an indicator of risk and then an appropriate line of testing (like **amniocentesis** or ultrasound) follows, based on the results.

Abnormally high AFP may indicate that the fetus has an increased risk of a neural tube defect, the most common and severe type of disorder associated with increased AFP. These types of defects include spinal column defects (**spina bifida**) and anencephaly (a severe and usually fatal brain abnormality). If the tube that becomes the brain and spinal cord does not close correctly during fetal development, AFP may leak through this abnormal opening and enter the amniotic fluid. This leakage creates abnormally high levels of AFP in amniotic fluid and in maternal blood.

Other fetal conditions that can raise AFP levels above normal include: cysts at the end of the spine,

blockage in the esophagus or intestines, some liver diseases, defects in the abdominal wall, kidney or urinary tract defects or disease, and brittle bone disease.

Levels may also be high if there is too little fluid in the amniotic sac around the fetus, more than one developing fetus, or a pregnancy that is farther along than estimated. For unknown reasons, abnormally low AFP may indicate that the fetus has an increased risk of **Down syndrome**. Down syndrome is a condition that includes **mental retardation** and a distinctive physical appearance linked to an abnormality of chromosome 21 (called trisomy 21). If the maternal screening test indicates an abnormally low AFP, amniocentesis is used to diagnosis the problem. Abnormally low levels of AFP can also occur when the fetus has died or when the mother is overweight.

AFP is often part of a triple-check blood test that analyzes three substances as risk indicators of possible birth defects: AFP, estriol, and human chorionic gonadotropin (HCG). When all three substances are measured in the mother's blood, the accuracy of the test results increases. Although AFP in human blood gradually disappears after birth, it never disappears entirely. It may reappear in liver disease, or tumors of the liver, ovaries, or testicles. The AFP test is used to screen people at high risk for these conditions. After a cancerous tumor is removed, an AFP test can monitor the progress of treatment. Continued high AFP levels suggest the cancer is growing.

Description

The AFP maternal screening test is usually performed at week 16 of pregnancy. In both pregnant mothers (whose fetus is being screened) and in children, blood is drawn from a vein, usually on the inside of the elbow. For a fetus, AFP can also be measured in the sample of amniotic fluid taken at the time of amniocentesis. Test results are usually available after about one week.

Precautions

It is very important that the doctor know precisely how old the fetus is when the test is performed, because the AFP level considered normal changes over the length of the pregnancy. Errors in determining the age of the fetus lead to errors when interpreting the test results. Since an AFP test is only a screening tool, more specific tests must follow to make an accurate diagnosis. An abnormal test result does not necessarily mean that the fetus has a birth defect. The test has a high rate of abnormal results (either high or low) in order to prevent missing a fetus that has a serious condition.

KEY TERMS

Amniotic fluid—The liquid in the amniotic sac that cushions the fetus and regulates temperature in the placental environment. Amniotic fluid also contains fetal cells.

Fetus—In humans, the developing organism from the end of the eighth week to the moment of birth. Until the end of the eighth week the developing organism is called an embryo.

Preparation

There is no specific physical preparation for the AFP test.

Aftercare

Other than making sure the bleeding stops from the needle puncture site and watching for any signs of infection at the needle site, there is no specific aftercare involved with this blood test.

Risks

The risks associated with drawing blood are minimal but may include bleeding from the puncture site, feeling faint or lightheaded after the blood is drawn, or blood accumulating under the puncture site (hematoma).

Normal results

Alpha-fetoprotein is measured in nanograms per milliliter (ng/mL) and is expressed as a probability. The probability 1:100, for example, translates into the chance that the fetus has a one in 100 chance, for example, of having the defect. An AFP level less than or equal to 50 ng/mL is considered normal.

Abnormal results

The doctor inform the mother of the fetus about the specific increased risk as compared to the normal risk of a standard case. If the risk of Down syndrome is greater than the standard risk for women who are 35 years old or older (1:270), then amniocentesis is recommended. Again, the test has a high rate of showing an abnormal AFP level in order to prevent missing

a fetus that has Down syndrome. This screening test only predicts risk; appropriate diagnostic testing follows an abnormal screening result. In neonatal liver disease testing, an AFP level greater than 40 ng/mL is considered abnormal. An AFP level greater than 20 ng/mL may be associated with tumors of the ovary or testes.

Parental concerns

A parent might be concerned about drawing blood from a child, but the **pain** from the needle puncture only lasts a moment.

When to call a doctor

If there is excess bleeding from the needle puncture site, or if hours to days later, the puncture site looks infected (red and swollen), then a doctor should be contacted.

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National Cancer Institute. Building 31, Room 10A31, 31 Center Drive, MSC 2580, Bethesda, MD 20892–2580. Web site: <www.nci.nih.gov>.

Mark A. Best

Alternative school

Definition

An alternative school is an educational setting designed to accommodate educational, behavioral, and/or medical needs of children and adolescents that cannot be adequately addressed in a traditional school environment.

Purpose

Alternative schools have been established since about the 1970s to meet the needs of children and adolescents who cannot learn effectively in a traditional school environment (i.e., conventional public or parochial schools) due to learning disabilities, certain medical conditions, psychological and behavioral issues, or advanced skills. In general, alternative schools have more comprehensive educational and developmental objectives than conventional schools. They often have curriculum elements that focus on improving student **self-esteem**, fostering growth of individuality, and enhancing social skills. Alternative schools are more flexible in their organization and administration, which allows for more variety in educational programs.

Once available primarily for disruptive students and those at risk for dropping out of a traditional school environment, alternative schools have expanded significantly in function as educators, parents, and wider communities recognize that many children cannot learn effectively in a traditional school environment. For children and adolescents with psychological and behavioral issues, such as **personality disorders**, substance use and abuse, depression, and violence, alternative schools can provide a safer therapeutic environment and more individualized attention than traditional schools. For children and adolescents with learning disabilities and certain medical conditions, such as attention deficit hyperactivity disorder (ADHD), **dyslexia**, and Asperger's syndrome, alternative schools can provide integrated education and clinical services in one place to facilitate learning.

Description

Alternative school structure and curriculum varies depending on the educational goals and desired student population. Alternative schools may be available and accessible locally or may require additional daily travel or boarding by the student. Usually, local alternatives to public schools do not require tuition, while private schools do require parents to pay tuition for student attendance.

A number of different types of alternative schools exist, including the following:

- local alternatives to public schools, for example, charter schools, magnet schools, at-risk programs
- special-needs day schools
- independent private schools
- therapeutic wilderness programs
- emotional growth boarding schools

For parents who desire a local alternative to traditional public and parochial schools, several charter and magnet schools may be available, especially in urban areas. Charter schools are independent, publicly funded schools run by teachers, parents, or foundations that are often formed to meet local community needs as an alternative to public schools. Charter schools may have a special focus, such as technical skills or music. As of 2004, virtual charter schools have been formed that offer all courses via the Internet or other distance learning methods for students who need to remain at home or whose parents wish them to remain at home. Magnet schools are public schools that offer specialized programs designed to attract students wishing to enhance particular skills. Magnet schools were originally formed in the 1960s and 1970s to promote voluntary racial desegregation in urban school districts. Magnet schools often advertise themselves as ''centers of excellence'' in a certain area, such as performing arts, science, or mathematics. Both charter and magnet schools generally have smaller classes and enhanced extracurricular offerings.

For children and adolescents identified as ''at-risk'' by the public school district, alternative programs may be available. Usually, at-risk alternative programs are offered at a special location within the public school district or at a location that is accessible to and serves multiple public schools (e.g., a county-wide program). At-risk students usually have undergone school psychological and behavioral evaluation that identifies them as requiring specialized attention not available in the traditional school environment. Suitable programs can include emotionally disturbed, oppositional, and disruptive students and offer smaller classes, specially trained staff, and closer supervision. Some programs may be dedicated to serving a particular group of at-risk students, such as pregnant teens and teen mothers. Researchers have estimated that more than 280,000 at-risk students in the United States are in alternative programs offered by school districts or private boarding schools (see below).

Special-needs day schools focus on **special education** programs to meet the needs of children and adolescents with learning disabilities and learning challenges. Students with severe ADHD, moderate-to-severe physical or behavioral obstacles, and other specialized educational needs receive customized instruction with individualized lesson plans, special counseling, adaptive physical education, speech therapy, and other supportive services to ensure that they can learn despite educational barriers caused by a medical condition or learning disability.

Independent private schools are privately funded schools controlled by an individual or non-government organization. Private schools may be day schools or boarding schools. Private schools require that parents pay tuition and usually have a competitive admissions process requiring students to complete an application and interview. Private schools usually emphasize academic and/or athletic achievement, and student acceptance is based on academic and athletic potential, as well as enthusiasm for being active in school community life. Private schools have smaller classes, a more structured learning environment, a variety of **extracurricular activities**, and individualized opportunities for developing student **creativity** and intellect.

Therapeutic wilderness programs involve group and individual therapy in an outdoor adventure setting. Depending on the program, academics may or may not be included. Usually, therapeutic wilderness programs do not run for a full school year and thus are not alternative schools per se; however, these programs generally run for a full summer or school semester (six to eight weeks) and may, therefore, be considered alternative education. Therapeutic wilderness programs use the outdoors to rapidly influence adolescents with at-risk behaviors through physical and emotional challenges that help them understand unhealthy behaviors and gain a more positive sense of self and responsibility. Group therapy employed in a wilderness setting helps adolescents learn how to successfully interact with peers. Therapeutic wilderness programs are appropriate for adolescents who have exhibited extreme defiance; who have a history of **running away**, substance abuse, sexual promiscuity, poor school performance (failing), and violence; and have not responded to other treatment programs. Therapeutic wilderness programs often serve as a transition to long-term therapeutic placement in a residential treatment center or emotional growth boarding school, depending on the needs of the adolescent.

Emotional growth boarding schools integrate therapeutic programs with academics to provide for students whose emotional, psychological, and behavioral issues prevent them from learning effectively in a traditional school environment. Therapeutic components of these schools include daily and weekly group and individual therapy, highly structured learning and living environments, experiential learning, and individualized academic programming. Because the root of many emotional and behavioral problems is low self-esteem and a negative perception of self, emotional growth programs focus on helping students permanently change negative self-perceptions, discovering and healing emotional trauma, and identifying and changing negative behaviors. Emotional growth boarding schools usually offer rolling admission; that is, students are accepted year-round and academics are available year-round. This type of operation helps parents whose children need emergency placement. Candidates for emotional growth

boarding schools are enrolled from therapeutic wilderness programs or undergo psychological and educational testing to determine their academic and therapeutic needs. Poor academic performance, a symptom of many emotional problems, is expected, and trained staff, counselors, and teachers provide support to improve student performance. While emotional growth boarding schools use different therapeutic models, depending on the school, most programs do use incentive-based learning and therapy, wilderness therapy, and intensive counseling to improve student decision-making, interpersonal skills, academic performance, and emotional coping skills. These schools also use **sports**, the arts, and interaction with animals as part of therapy.

Precautions

Parents considering alternative schools should thoroughly investigate the school's credentials, staff training, available curriculum, student support services, and student population to make sure that the needs of their child will be met.

There are a number of wilderness programs available for different types of students, and not all have a therapeutic component. In addition, some wilderness programs employ "boot camp" methods that may be unsafe for children and adolescents. A therapeutic wilderness program should have trained and/or certified wilderness counselors and medical support services, as well as provide training in wilderness skills for participants.

Preparation

Making the decision to place a child in an alternative school can be difficult and involves a number of factors. For independent private schools and schools that focus on a specific skill or talent, interviews and applications may be necessary, and advanced students and students with special talents have to complete an often-rigorous application process. Parents and students should be prepared to visit all schools under consideration and participate in interviews with school staff.

For children with special medical needs, clinical care may need to be coordinated with current physicians and clinical staff at the new alternative school. Parents and students should be prepared to undergo additional medical and educational testing to determine the student's needs for individualized lesson plans.

Schools that accept at-risk children and adolescents require psychological and educational testing, as well as references or recommendations from a professional (usually a psychologist, psychiatrist, or therapist). In some situations where the child or adolescent is a danger to himself/herself and/or others, emergency transport services to the therapeutic school are available; specially trained individuals escort the student from their home to the school, even via air travel, to ensure the child's **safety**. Parents of at-risk children and adolescents should be prepared emotionally to handle such situations and also to participate in regular **family therapy** sessions during the alternative program.

Public schools are obligated to provide access to a free and safe education for students, and if their curriculum and support services cannot handle the needs of a particular student, the public school may also be obligated to financially support the student in an alternative school that can better address the student's needs. To prepare for obtaining such financial support, parents of children whose needs are not being met in the public school should request an official evaluation by a school psychologist and the formulation of an individualized education plan (IEP), which should detail how the public school will meet the child's needs. Having an independent psychologist or psychiatrist complete testing as well can provide a second opinion. If the IEP does not address the child's problems, parents can request that the school find and pay for an alternative school program. An educational consultant and attorney specializing in educational issues can help guide parents through this process.

Aftercare

Students graduating or transferring from alternative schools may continue to require special support, such as counseling, group therapy, or medical care. Support and encouragement from **family** members is important.

Parental concerns

Choosing an alternative school is often difficult, particularly for parents of at-risk children and adolescents. Parents who feel that their local school district is not adequately addressing the educational needs of their child should consider an alternative school. Reasons for choosing an alternative school vary, depending on the child, who may:

- be unusually gifted or motivated
- have a special talent or interest, such as music or science, that cannot be further developed in the present school
- be an underachiever or failing and require more individualized attention

KEY TERMS

Asperger syndrome—A developmental disorder of childhood characterized by autistic behavior but without the same difficulties acquiring language that children with autism have.

Attention deficit hyperactivity disorder (ADHD)—A condition in which a person (usually a child) has an unusually high activity level and a short attention span. People with the disorder may act impulsively and may have learning and behavioral problems.

Dyslexia—A type of reading disorder often characterized by reversal of letters or words.

Individualized educational plan (IEP)—A detailed description of the educational goals, assessment methods, behavioral management plan, and educational performance of a student requiring special education services.

- have special needs due to a learning disability or medical condition
- be exhibiting behaviors such as substance abuse, inappropriate sexual activity, **acting out**, and oppositional defiance
- have engaged in petty criminal behaviors and is becoming more self-destructive
- have been diagnosed with emotional and/or psychological problems that require a more structured therapeutic environment

An educational consultant can help parents choose an alternative school. Educational consultants usually have visited any school they recommend and will consider the student's psychological evaluations and other test results to determine the alternative school that will best meet their needs. An attorney specializing in educational issues can help parents obtain financial support for alternative therapeutic programs from the public school.

At-risk children and adolescents involved in an emotional growth school require significant involvement and support from family members, since many psychological and behavioral issues are rooted in family dynamics and history (e.g., bitter **divorce**). Hence, parents may need to take family medical leave from their work or make significant changes in their family lifestyle to support therapy for their child. Joining a parent support group can help, and most emotional growth schools have parent networks. Alternative schools for at-risk children and adolescents may seem too structured and too rigorous with regard to emotional therapy for some parents. However, outcomes research for these types of schools has shown a high success rate; more than 85 percent of students completing such programs have improved family and peer relationships, attend a college or find a job, and remain free from substance use.

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Jennifer E., Sisk, M.A.

Ambiguous genitalia *see* **Intersex states**

Amblyopia

Definition

Amblyopia refers to diminished vision in either one or both eyes, for which no cause can be discovered upon examination of the eye. Amblyopia is the medical term used when the vision in one of the eyes is reduced because the eye and the brain are not working together properly. The eye itself looks normal, but it is not being used normally because the brain is favoring the other eye. This condition is also sometimes called lazy eye.

Description

Lazy eye is a common non-medical term used to describe amblyopia because the eye with poorer vision does not seem to be doing its job of seeing. Amblyopia is the most common cause of impaired vision in childhood. It affects approximately two or three out of every 100 children. Vision is a combination of the clarity of the images of the eyes (visual acuity) and the processing of those images by the brain. If the images produced by the two eyes are substantially different, the brain may not be able to fuse the images. Instead of seeing two different images or double vision (diplopia), the brain suppresses the blurrier image. This suppression can lead to amblyopia. During the first few years of life, preferring one eye over the other may lead to poor visual development in the blurrier eye. Unless it is treated successfully in early childhood, amblyopia usually persists into adulthood and is the most frequent cause of monocular (one eye) visual impairment among children.

Demographics

The prevalence of amblyopia is difficult to assess, with estimates ranging from 1.0 to 3.5 percent in healthy children to 4.0 to 5.3 percent in children with other vision problems. It is seen in similar numbers in both sexes and in all races.

Causes and symptoms

Amblyopia may be caused by any condition that adversely affects normal visual development or use of the eyes. All babies are born with poor eyesight. As babies grow, however, their eyesight usually progresses. Good eyesight needs a clear, focused image that is the same in both eyes. If the image is not clear in one eye, or if the image is not the same in both eyes, the vision pathways will not develop as they should. In fact, the pathways may actually worsen. Anything that blurs the vision or causes the eyes to be crossed during childhood may cause amblyopia. Some of the major causes of amblyopia are as follows:

- **Strabismus**. A misalignment of the eyes is the most common cause of functional amblyopia. The two eyes are looking in two different directions at the same time. The eyes may turn in, out, up, or down. Strabismus may be diagnosed at birth, or it may develop later in childhood. The brain is sent two different images and this creates confusion. Images from the misaligned or "crossed" eye are turned off to avoid double vision.
- Anisometropia. A difference of refractive states exists between the two eyes (in other words, a difference in prescription between the two eyes). For example, one eye may be more nearsighted than the other eye, or one eye may be farsighted and the other eye nearsighted. Because the brain cannot fuse the two images, the brain suppresses the blurrier image, causing the eye to become amblyopic.
- Cataract. Clouding of the lens of the eye causes the image to be blurrier than the other eye. The brain prefers the clearer image, and the eye with the cataract may become amblyopic.
- Ptosis. If light cannot enter the eye because of the drooping lid, the eye is essentially going unused, which can lead to amblyopia. However, ptosis is rarely related to the development of amblyopia, unless the droopy eyelid completely obscures the pupil.

Barring the presence of strabismus or ptosis, children may or may not show signs of amblyopia. Children may position their heads at an angle while trying to favor the eye with normal vision. They may have difficulty seeing or reaching for things when approached from the side of the amblyopic eye. Parents should see if one side of approach is preferred by the child or infant. If an infant's good eye is covered, the child may cry.

When to call the doctor

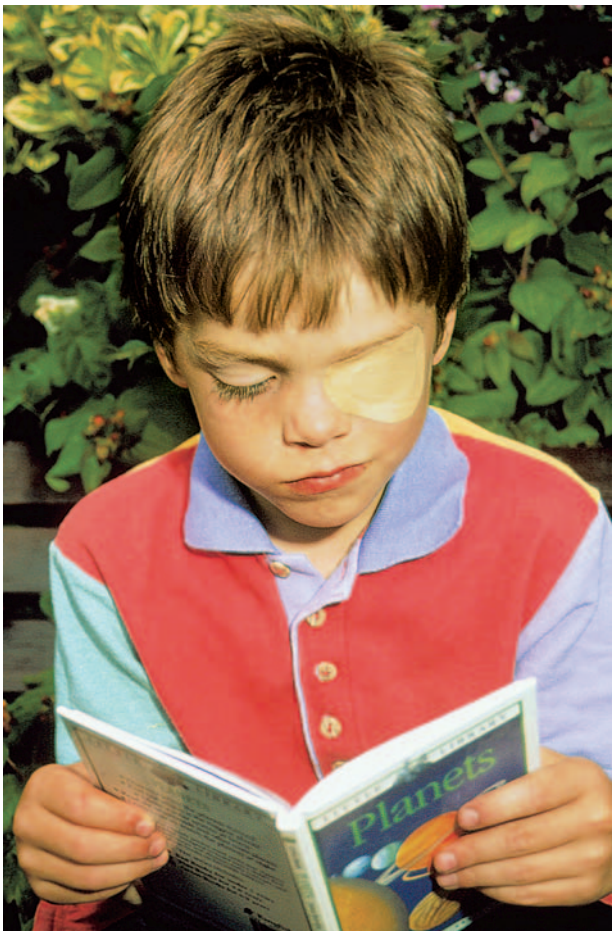
Parents should call the doctor if their child demonstrates any signs associated with amblyopia, including the appearance of crossed eyes, lazy eye, a drooping eyelid, difficulty seeing, or if the child seems to favor one side of approach over the other. However, since children do not always show symptoms of amblyopia, it is important to get their eyes examined at or before the age of three and no later than age five, while the disorder is more easily treated.

Diagnosis

It is not easy to recognize amblyopia. A child may not be aware of having one strong eye and one weak eye. Unless the child has a misaligned eye or other obvious abnormality, there is often no way for parents to tell that something is wrong. Because children with outwardly normal eyes may have amblyopia, it is important to have regular vision screenings performed for all children. While there is some disagreement regarding the age children should have their first vision examination, their eyes can, in actuality, be examined at any age, even on the first day of life.

Some people recommend that children have their vision checked by their pediatrician, **family** physician, ophthalmologist, or optometrist at or before six months of age. Others recommend testing by at least the child's fourth birthday. There may be a critical period in the development of vision, and amblyopia may not be treatable after age eight or nine. The earlier amblyopia is found, the better chance there is for a positive outcome. Most physicians test vision as part of a child's medical examination. If there is any sign of an eye problem, they may refer a child to an eye specialist.

There are objective methods, such as retinoscopy, by which to measure the refractive status of the eyes. This form of examination can help diagnose anisometropia. In retinoscopy, a hand-held instrument is used to shine a light in the child's (or infant's) eyes. While the doctor uses hand-held lenses, he can obtain a rough prescription. Visual acuity can be determined using a variety of methods. Many different eye charts are available (e.g. tumbling E, pictures, or letters). In amblyopia, single letters are easier to recognize than when a whole line is shown. This is referred to as the "crowding effect" and helps in diagnosing amblyopia. Neutral density filters may also be held over the eye to aid in the diagnosis. Sometimes visual fields to determine defects in the area of vision will be performed. Color vision testing may also be done. Again, it must be emphasized that amblyopia is a diagnosis of exclusion. Various medical problems can also cause a decrease in vision. An examination of the eyes and visual system is very important when there is an unexplained decrease in vision.



Boy wearing eye patch used to treat amblyopia. The patch is worn over the stronger eye to build the weaker one's strength. *(© Mark Clarke/Photo Researchers, Inc.)*

Treatment

Amblyopia treatment is most effective when done early in the child's life, usually before age seven. It is important that any anisometropia and refractive problems be treated initially, because sometimes amblyopia can be resolved with glasses alone.

The next step is to make the child use the eye with the reduced vision (weaker eye). As of 2004, there are two ways to do this:

- Patching. An opaque, adhesive patch is worn over the stronger eye for weeks to months. This therapy forces the child to use the eye with amblyopia. Patching stimulates vision in the weaker eye and aids the section of

KEY TERMS

Anisometropia—An eye condition in which there is an inequality of vision between the two eyes. There may be unequal amounts of nearsightedness, farsightedness, or astigmatism, so that one eye will be in focus while the other will not.

Cataract—A condition in which the lens of the eye turns cloudy and interferes with vision.

Occulsion therapy—A type of treatment for amblyopia in which the good eye is patched for a period of time, thus forcing the use of the weaker eye.

Visual acuity—Sharpness or clearness of vision.

the brain that manages vision to develop more completely. Patching may be part-time or full-time. Studies in the early 2000s have shown that less time patching the eye may be as effective as more. In the case of moderate amblyopia, two hours of daily patching for four months gave the same benefit as six hours of daily patching for the same period of time. Compliance with the patching regimen was also improved with the shorter daily patching time. The treatment plan should be discussed with the doctor to determine how long the patch should be worn. When the child is wearing the patch, prescribed eye exercises may force the amblyopic eye to focus and work. This is called vision therapy or vision training. Even after the child's vision has been restored in the weak eye, part-time patching may be required over a period of years to maintain the improvement.

- Atropine. This therapy is generally reserved for children who will not wear a patch or where compliance may be an issue. A drop of a drug called atropine is placed in the stronger eye once a day to temporarily blur the vision so that the child will prefer to use the eye with amblyopia. Treatment with atropine also stimulates vision in the weaker eye and helps the part of the brain that manages vision to develop more fully.

Prognosis

The younger the child, the better the chance for improvement with occlusion and vision therapy. Success in the treatment of amblyopia also depends on the amblyopia's severity, its specific type, and the child's compliance with treatment. It is important to diagnose and treat amblyopia early because significant vision loss can occur if it is left untreated. The best outcomes result from early diagnosis and treatment.

Prevention

Early recognition and treatment of amblyopia in children can help to prevent permanent visual deficits. All children should have a complete eye examination at least once between age three and five to avoid the risk of allowing unsuspected amblyopia to go beyond the age where it can be treated successfully.

Nutritional concerns

There are some rarer forms of amblyopia caused by various nutritional deficiencies. In these cases, the doctor recommends the proper diet and perhaps supplementation in order to resolve the problem.

Parental concerns

It is vital that parents bring their child for an eye exam sometime between the ages of three and five to prevent amblyopia from becoming untreatable.

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Amenorrhea

Definition

Amenorrhea is the medical term for the absence of **menstruation**. There are two types of amenorrhea, primary and secondary. Primary amenorrhea refers to delayed menarche (the first menstrual period) and is defined as any one of three conditions:

- the absence of menarche by age 16 in a girl with otherwise normal pubertal development (development of breasts and/or pubic hair)
- the absence of menarche by age 14 combined with delayed pubertal development
- the absence of menarche two years after **puberty** is otherwise completed

Secondary amenorrhea is defined as the absence of menstruation after menarche has taken place. Although it is not uncommon for a girl's menstrual periods to be irregular during early **adolescence**, most girls' periods usually become regular within 18 months after the first one. After that time, it is considered abnormal for an adolescent to miss three consecutive periods.

Description

Normal menstrual periods are the result of proper functioning and synchronization of the hypothalamus, pituitary gland, and ovaries. The hypothalamus is the part of the brain that controls body temperature, cellular metabolism, and such basic functions as appetite for food, the sleep/wake cycle, and reproduction. The hypothalamus also secretes hormones that regulate the pituitary gland. The pituitary gland in turn produces hormones that stimulate the ovaries to secrete two hormones known as estradiol and progesterone. These ovarian hormones encourage the growth of the endometrium, which is the tissue that lines the uterus. If pregnancy does not occur, the endometrium breaks down and the uterus sheds the extra tissue during the next menstrual period.

Amenorrhea can result from an interruption at any of several points in the normal cycle:

- The hypothalamus and pituitary may fail to produce enough hormone to stimulate the ovaries to produce their hormones.
- The ovaries may fail to produce enough estradiol to stimulate the growth of the endometrium.
- There may be structural abnormalities in the uterus, cervix, or vagina that prevent the shed tissue from leaving the body.

Demographics

Secondary amenorrhea is more common in females in North America than primary amenorrhea. One study estimates that about 5 percent of menstruating women have an episode of secondary amenorrhea each year.

The average age for the onset of the menses in girls in the United States and Canada is 12.77 years. There is no evidence as of the early 2000s that the incidence of either primary or secondary amenorrhea is related to race or ethnic background.

Causes and symptoms

Causes

There are a number of possible causes of amenorrhea:

- Pregnancy: An adolescent with amenorrhea most likely does not have a serious underlying medical problem. All teenagers with amenorrhea should seek medical care, and an adolescent who has had sexual intercourse even once and then missed a period should assume she is pregnant until a reliable pregnancy test proves otherwise. It should be noted that spotting or even bleeding is not unusual during early pregnancy. In addition, it is possible for a girl to conceive before she has had even one period.
- Disorders of the hypothalamus or the pituitary gland: These problems may be associated with brain tumors.
- Ovarian disorders: These disorders may include premature ovarian failure or may be the side effects of **chemotherapy** or radiation therapy for **cancer**. Premature ovarian failure accounts for about 10 percent of cases of secondary amenorrhea.
- Hyperandrogenism: The overproduction of male hormones (androgens) by the girl's body can interrupt menstruation. Male hormones are produced in small quantities

by all women, but some individuals produce excessive amounts, leading to such conditions as polycystic ovarian syndrome (PCOS), hirsutism (excessive growth of body hair), or abnormalities of the external genitalia. PCOS in adolescents is often triggered by **obesity**.

- Genetic disorders: Some genetic disorders that affect the X chromosome, such as Turner's syndrome, prevent normal sexual maturation in girls.
- Psychiatric disorders: Depression, **obsessive-compulsive disorder**, eating disorders, and **schizophrenia** can all cause disturbances of the menstrual cycle.
- Abuse of alcohol or other drugs: Excessive alcohol intake can lead to **malnutrition**, while cocaine and opioids (narcotics) can affect the menstrual cycle directly.
- **Immunodeficiency** disorders or conditions.
- Emotional stress: This disturbance can interfere with the brain's hormonal signals to the ovaries. It is not uncommon for a girl's period to be delayed when she is having problems with school, work, or relationships. A change in environment (the first year of college or taking a new job, for example) can also cause a young woman's period to be late.
- Female athlete triad: Female athletes at the high school or college level are at increased risk for a triad of disorders: excessive dieting or disordered eating, amenorrhea, and loss of bone **minerals** leading to osteoporosis. The triad was first formally named in 1993 but had been known to doctors for decades before. Girls who are involved in **sports** that emphasize weight control or a slender body build (gymnastics, track and field, cheerleading) are at greater risk than those who **play** field hockey, basketball, softball, or other sports that emphasize strength.

Symptoms

Amenorrhea may be associated with the symptoms of other disorders; for example, girls with an eating disorder will often have eroded tooth enamel, tiny pinpoint hemorrhages around the eyes, an abnormal heart rhythm, low blood pressure, and other signs of frequent **vomiting**. Girls whose amenorrhea is part of the female athlete triad may have a record of bone **fractures** or other evidence of bone mineral loss. Hot flashes and night sweats may indicate premature ovarian failure. Headaches or visual disturbances may suggest a brain tumor.

When to call the doctor

Girls who have not had a menstrual period by age 16 or who have not shown any signs of **breast development** or other indications of puberty by age 14 should be examined for causes of primary **dysmenorrhea**. Girls who have begun to menstruate and have missed three periods should be evaluated for secondary amenorrhea. If they are sexually active, they should have a pregnancy test after missing even one period.

Diagnosis

History and physical examination

The first part of diagnosing amenorrhea is a careful history, including a record of medications and any surgical procedures involving the abdomen or genitals. The doctor will ask detailed questions about stress, dieting, sexual activity, and athletic participation, as well as questions about chronic diseases or disorders of the central nervous system. **Family** history should be taken into consideration in any adolescent with primary amenorrhea, as mothers who started to menstruate late will often have daughters who also menstruate late.

In the case of female athletes, the doctor may need to establish a relationship of trust with the patient before asking about such matters as diet, practice and workout schedules, and the use of such drugs as steroids or ephedrine. The presence of stress fractures in young women should be investigated. In some cases, the doctor may give the patient the Eating Disorder Inventory (EDI) or a similar screening questionnaire to help determine whether the patient is at risk for developing anorexia or bulimia.

The doctor will then perform a physical examination to evaluate the patient's weight in proportion to her height as well as her general nutritional status; to check for breast development, pubic hair, and other signs of normal female sexual development; to make sure the heart rhythm, blood pressure, and other vital signs are normal; and to palpate (feel) the thyroid gland for evidence of swelling. The physical examination may include a pelvic examination to check for abnormalities in the structure of the vagina or cervix.

Laboratory tests

To rule out specific causes of amenorrhea, the doctor may order a pregnancy test in sexually active young women as well as blood tests to check the level of thyroid hormone. Based on the initial test results, the doctor may want to perform additional tests to determine the level of other hormones that play a role in reproduction. A special type of blood test called a karyotype may be done to analyze the girl's chromosomes if the doctor suspects Turner's syndrome or another genetic disorder.

One way to determine whether a teenager's ovaries and uterus are functioning is a progesterone challenge test. In this test, an amenorrheic teenager is given a dose of progesterone either orally or as an injection. If her ovaries are producing estrogen and her uterus is responding normally, she should have a menstrual period within a few days of the progesterone dose. This challenge indicates that the ovaries and uterus are functioning normally, and the cause of the amenorrhea is probably in the brain.

Imaging studies

In some cases the doctor may order an ultrasound study of the pelvic region to check for anatomical abnormalities or **x rays** or a bone scan to check for bone fractures. In some cases the doctor may order an MRI to rule out tumors affecting the hypothalamus or pituitary gland.

Psychiatric interview

Teenagers whose amenorrhea may be related to depression, family stress, eating disorders, or other mental health issues may be referred to a psychiatrist for further evaluation.

Treatment

The most frequent risk associated with amenorrhea is osteoporosis (thinning of the bone) caused by low estrogen levels. Because osteoporosis can begin as early as adolescence, hormone replacement therapy is sometimes recommended for teenagers with chronic amenorrhea.

Amenorrhea associated with hormonal, genetic, psychiatric, or immunodeficiency disorders may require a variety of different medications and other treatments administered by specialists. Tumors of the hypothalamus and the pituitary gland or abnormalities of the reproductive organs usually require surgery.

Alternative treatment

As with conventional medical treatments, alternative treatments are based on the cause of the condition. If a hormonal imbalance is revealed by laboratory testing, hormone replacements that are more natural for the body (including tri-estrogen and natural progesterone) are recommended. Glandular therapy can assist in bringing about a balance in the glands involved in the reproductive cycle, including the hypothalmus, pituitary, thyroid, ovarian, and adrenal glands.

Since homeopathy and acupuncture work on deep energetic levels to rebalance the body, these two forms of therapy may be helpful in treating amenorrhea. Western and Chinese herbal medicines also can be very effective. Herbs used to treat amenorrhea include dong quai (*Angelica sinensis*), black cohosh (*Cimicifuga racemosa*), and chaste tree (*Vitex agnus-castus*). Herbal preparations used to bring on the menstrual period are known as emmenagogues. For some adolescents, meditation, guided imagery, and visualization can play a key role in the treatment of amenorrhea by relieving emotional stress.

Nutritional concerns

Diet and adequate **nutrition**, including adequate protein, essential fatty acids, whole grains, and fresh fruits and vegetables are important for every female past puberty, especially if deficiencies are present or if she regularly exercises very strenuously. Girls who are abusing alcohol or other drugs should be evaluated for possible malnutrition as part of treatment for substance abuse.

Female athletes at the high school or college level should consult a nutritionist to make sure that they are eating a well-balanced diet that is adequate to maintain a healthy weight for their height. Girls participating in dance or in sports that emphasize weight control or a slender body type (gymnastics, track and field, swimming, and cheerleading) are at higher risk of developing eating disorders than those that are involved in such sports as softball, weight lifting, or basketball. In some cases the athlete may be given calcium or vitamin D supplements to lower the risk of osteoporosis.

Prognosis

The prognosis of either primary or secondary amenorrhea depends on the underlying cause.

Prevention

Amenorrhea related to pregnancy, the female athletic triad, drug or alcohol abuse, or eating disorders is preventable insofar as these are lifestyle choices. Primary or secondary amenorrhea associated with genetic mutations or other systemic diseases or disorders is not preventable.

Parental concerns

Amenorrhea is a fairly dramatic symptom of menstrual dysfunction that often causes parents to consult a doctor about a girl's health. Parental concerns about

KEY TERMS

Anorexia nervosa—An eating disorder marked by an unrealistic fear of weight gain, self-starvation, and distortion of body image. It most commonly occurs in adolescent females.

Emmenagogue—A type of medication that brings on or increases a woman's menstrual flow.

Endometrium—The mucosal layer lining the inner cavity of the uterus. The endometrium's structure changes with age and with the menstrual cycle.

Female athlete triad—A combination of disorders frequently found in female athletes that includes disordered eating, osteoporosis, and oligo- or amenorrhea. The triad was first officially named in 1993.

Hyperandrogenism—The excessive secretion of androgens.

Menarche—The first menstrual cycle in a girl's life.

Osteoporosis—Literally meaning "porous bones," this condition occurs when bones lose an excessive amount of their protein and mineral content, particularly calcium. Over time, bone mass and strength are reduced leading to increased risk of fractures.

Turner syndrome—A chromosome abnormality characterized by short stature and ovarian failure caused by an absent X chromosome. It occurs only in females.

amenorrhea, however, should be directed to the underlying cause. Amenorrhea related to emotional stress, dieting, or excessive **exercise** usually goes away when the stress is relieved or when the girl makes appropriate lifestyle adjustments. On the other hand, amenorrhea associated with glandular disturbances, tumors, genetic or anatomical abnormalities, diabetes, or other systemic disorders is part of a larger and more worrisome picture. Parents should discuss their concerns about the long-term effects of amenorrhea on the girl's health, whether she will be able to have children in adult life, and how they can help her manage her condition with the doctors, nutritionists, and other healthcare professionals who are treating her.

See also Anorexia nervosa; Bulimia nervosa; Menstruation; Oligomenorrhea; Sports.

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American Academy of Child and Adolescent Psychiatry. 3615 Wisconsin Avenue, NW, Washington, DC 20016–3007. Web site: <www.aacap.org>.

American College of Obstetricians and Gynecologists (ACOG). 409 12th Street, SW, PO Box 96920, Washington, DC 20090–6920. Web site: <www.acog.org>.

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Gail Slap, MD

Amniocentesis

Definition

Amniocentesis is a procedure used to diagnose fetal defects in the early second trimester of pregnancy. A sample of the amniotic fluid, which surrounds a fetus in the womb, is collected through a pregnant woman's abdomen using a needle and syringe. Tests performed on fetal cells found in the sample can reveal the presence of many types of genetic disorders, thus allowing doctors and prospective parents to make important decisions about early treatment and intervention.

Purpose

Since the mid-1970s, amniocentesis has been used routinely to test for **Down syndrome**, by far the most common, nonhereditary, genetic birth defect, afflicting about one in every 1,000 babies. By 1997, approximately 800 different diagnostic tests were available, most of them for hereditary genetic disorders such as **Tay-Sachs disease**, **sickle cell anemia**, **hemophilia**, **muscular dystrophy**, and **cystic fibrosis**.

Amniocentesis, often called amnio, is recommended for women who will be older than 35 on their due-date. It is also recommended for women who have already borne children with birth defects, or when either of the parents has a **family** history of a birth defect for which a diagnostic test is available. Another reason for the procedure is to confirm indications of Down syndrome and certain other defects which may have shown up previously during routine maternal blood screening.

The risk of bearing a child with a nonhereditary genetic defect such as Down syndrome is directly related to a woman's age—the older the woman, the greater the risk. Thirty-five is the recommended age to begin amnio testing because that is the age at which the risk of carrying a fetus with such a defect roughly equals the risk of miscarriage caused by the procedure–about one in 200. At age 25, the risk of giving birth to a child with this type of defect is about one in 1,400; by age 45 it increases to about one in 20. Nearly half of all pregnant women over 35 in the United States undergo amniocentesis and many younger women also decide to have the procedure. Notably, some 75% of all Down syndrome infants born in the United States each year are to women younger than 35.

One of the most common reasons for performing amniocentesis is an abnormal alpha-fetoprotein (AFP) test. Alpha-fetoprotein is a protein produced by the fetus and present in the mother's blood. A simple blood screening, usually conducted around the 15th week of pregnancy, can determine the AFP levels in the mother's blood. Levels that are too high or too low may signal possible fetal defects. Because this test has a high false-positive rate, another test such as amnio is recommended whenever the AFP levels fall outside the normal range.

Amniocentesis is generally performed during the 16th week of pregnancy, with results usually available within three weeks. It is possible to perform an amnio as early as the 11th week, but this is not usually recommended because there appears to be an increased risk of miscarriage when done at this time. The advantage of early amnio and speedy results lies in the extra time for decision making if a problem is detected. Potential treatment of the fetus can begin earlier. Important, also, is the fact that elective abortions are safer and less controversial the earlier they are performed.

Precautions

As an invasive surgical procedure, amnio poses a real, although small, risk to the health of a fetus. Parents must weigh the potential value of the knowledge gained, or indeed the reassurance that all is well, against the small risk of damaging what is in all probability a normal fetus. The serious emotional and ethical dilemmas that adverse test results can bring must also be considered. The decision to undergo amnio is always a matter of personal choice.

Description

The word amniocentesis literally means "puncture of the amnion," the thin-walled sac of fluid in which a developing fetus is suspended during pregnancy. During the sampling procedure, the obstetrician inserts a very fine needle through the woman's abdomen into the uterus and amniotic sac and withdraws approximately one ounce of amniotic fluid for testing. The relatively painless procedure is performed on an outpatient basis, sometimes using local anesthesia.

The physician uses ultrasound images to guide needle placement and collect the sample, thereby minimizing the risk of fetal injury and the need for repeated nee-

dle insertions. Once the sample is collected, the woman can return home after a brief observation period. She may be instructed to rest for the first 24 hours and to avoid heavy lifting for two days.

The sample of amniotic fluid is sent to a laboratory where fetal cells contained in the fluid are isolated and grown in order to provide enough genetic material for testing. This takes about seven to 14 days. The material is then extracted and treated so that visual examination for defects can be made. For some disorders, like Tay-Sachs, the simple presence of a telltale chemical compound in the amniotic fluid is enough to confirm a diagnosis. Depending on the specific tests ordered, and the skill of the lab conducting them, all the results are available between one and four weeks after the sample is taken.

Cost of the procedure depends on the doctor, the lab, and the tests ordered. Most insurers provide coverage for women over 35, as a follow-up to positive maternal blood screening results, and when genetic disorders run in the family.

An alternative to amnio, now in general use, is chorionic villus sampling, or CVS, which can be performed as early as the eighth week of pregnancy. While this allows for the possibility of a first trimester abortion, if warranted, CVS is apparently also riskier and is more expensive. The most promising area of new research in prenatal testing involves expanding the scope and accuracy of maternal blood screening as this poses no risk to the fetus.

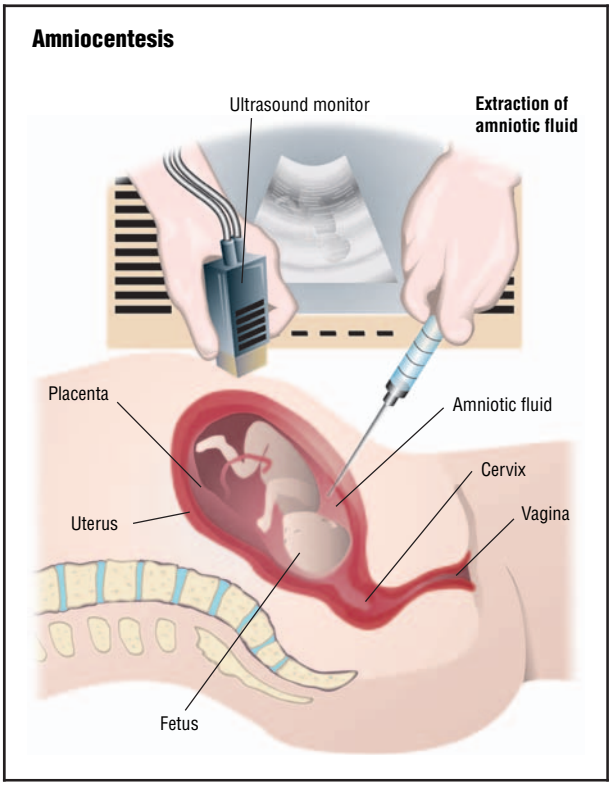
Preparation

It is important for a woman to fully understand the procedure and to feel confident in the obstetrician performing it. Evidence suggests that a physician's experience with the procedure reduces the chance of mishap. Almost all obstetricians are experienced in performing amniocentesis. The patient should feel free to ask questions and seek emotional support before, during and after the amnio is performed.

Aftercare

Necessary aftercare falls into two categories, physical and emotional.

PHYSICAL AFTERCARE During and immediately following the sampling procedure, a woman may experience **dizziness**, **nausea**, a rapid heartbeat, and cramping. Once past these immediate hurdles, the physician will send the woman home with instructions to rest and to report any complications requiring immediate treatment, including:



To perform amniocentesis, a physician uses an ultrasound monitor to visualize the fetus while inserting a syring to extract amnniotic fluid for analysis. *(Illustration by GGS Information Services.)*

- Vaginal bleeding. The appearance of blood could signal a problem.
- Premature labor. Unusual abdominal **pain** and/or cramping may indicate the onset of premature labor. Mild cramping for the first day or two following the procedure is normal.
- Signs of infection. Leaking of amniotic fluid or unusual vaginal discharge, and **fever** could signal the onset of infection.

EMOTIONAL AFTERCARE Once the procedure has been safely completed, the anxiety of waiting for the test results can prove to be the worst part of the process. A woman should seek and receive emotional support from family and friends, as well as from her obstetrician and family doctor. Professional counseling may also prove necessary, particularly if a fetal defect is discovered.

Risks

Most of the risks and short-term side effects associated with amniocentesis relate to the sampling procedure and have been discussed above. A successful amnio

KEY TERMS

Alpha fetoprotein (AFP)—A substance produced by a fetus' liver that can be found in the amniotic fluid and in the mother's blood. Abnormally high levels of this substance suggests there may be defects in the fetal neural tube, a structure that will include the brain and spinal cord when completely developed. AFP may also be found at elevated levels in the blood of adults with liver, testicular, and ovarian cancer.

Anencephaly—A genetic defect resulting in the partial to complete absence of the brain and malformation of the brainstem.

Chorionic villus sampling—A procedure used for prenatal diagnosis at 10–12 weeks gestation. Under ultrasound guidance a needle is inserted either through the mother's vagina or abdominal wall and a sample of the chorionic membrane. These cells are then tested for chromosome abnormalities or other genetic diseases.

Chromosome—A microscopic thread-like structure found within each cell of the human body and consisting of a complex of proteins and DNA. Humans have 46 chromosomes arranged into 23 pairs. Chromosomes contain the genetic information necessary to direct the development and functioning of all cells and systems in the body. They pass on hereditary traits from parents to child (like eye color) and determine whether the child will be male or female.

Down syndrome—A chromosomal disorder caused by an extra copy or a rearrangement of chromosome 21. Children with Down syndrome have varying degrees of mental retardation and may have heart defects.

Genetic—Refers to genes, the basic units of biological heredity, which are contained on the chromosomes.

Hereditary—Something which is inherited, that is passed down from parents to offspring. In biology and medicine, the word pertains to inherited genetic characteristics.

Maternal blood screening—Screening that is normally done early in pregnancy to test for a variety of conditions. Abnormal amounts of certain proteins in a pregnant woman's blood raise the probability of fetal defects. Amniocentesis is recommended if such a probability occurs.

Tay-Sachs disease—An inherited disease caused by a missing enzyme that is prevalent among the Ashkenazi Jewish population of the United States. Infants with the disease are unable to process a certain type of fat which accumulates in nerve and brain cells, causing mental and physical retardation, and, finally, death.

Ultrasonography—A medical test in which sound waves are directed against internal structures in the body. As sound waves bounce off the internal structure, they create an image on a video screen. Ultrasonography is often used to diagnose fetal abnormalities, gallstones, heart defects, and tumors. Also called ultrasound imaging.

sampling results in no long-term side effects. Risks include:

- Maternal/fetal hemorrhaging. While spotting in pregnancy is fairly common, bleeding following amnio should always be investigated.
- Infection. Infection, although rare, can occur after amniocentesis. An unchecked infection can lead to severe complications.
- Fetal injury. A very slight risk of injury to the fetus resulting from contact with the amnio needle does exist.
- Miscarriage. The rate of miscarriage occurring during standard, second trimester amnio appears to be approximately 0.5%. This compares to a miscarriage rate of 1% for CVS. Many fetuses with severe genetic defects miscarry naturally during the first trimester.
- The trauma of difficult family-planning decisions. The threat posed to parental and family mental health from the trauma accompanying an abnormal test result can not be underestimated.

Normal results

Negative results from an amnio analysis indicate that everything about the fetus appears normal and the pregnancy can continue without undue concern. A negative result for Down syndrome means that it is 99% certain that the disease does not exist.

An overall "normal" result does not, however, guarantee that the pregnancy will come to term, or that the fetus does not suffer from some other defect. Laboratory tests are not 100% accurate at detecting tar-

geted conditions, nor can every possible fetal condition be tested for.

Abnormal results

Positive results on an amnio analysis indicate the presence of the fetal defect being tested for, with an accuracy approaching 100%. Prospective parents are then faced with emotionally and ethically difficult choices regarding treatment options, the prospect of dealing with a severely affected newborn, and the option of elective abortion. At this point, the parents need expert medical advice and counseling.

Parental concerns

There is a risk of miscarrage with this procedure.

When to call a doctor

If there is excess bleeding, a doctor should be contacted.

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Mark A. Best

Amoxicillin *see* **Penicillins**

Amphetamines *see* **Stimulant drugs**

Amputation *see* **Traumatic amputations**

Anabolic steroids

Definition

Anabolic steroids are compounds, derived from testosterone, which promote tissue growth and repair. Because they have been used improperly by body builders and other athletes, they are controlled substances under United States federal law.

Description

As of 2004, there are four anabolic steroids available:

- nandrolone
- oxandrolone
- oxymetholone
- stanzolol

Although these products have different labeled uses, they are very similar in action and side effects and may be used interchangeably, subject to differences in route of administration and duration of action.

General use

Anabolic steroids are used for the following conditions:

- catabolic states such as chronic infections, extensive surgery, **burns**, or severe trauma
- anemia associated with renal insufficiency, **sickle cell anemia**, aplastic anemia, and bone marrow failure
- angioedema
- growth failure, including the short stature associated with Turner's syndrome

Precautions

Anabolic steroids are not recommended for young children because the drugs may cause an early end to the growth of long bones, which results in short stature. Anabolic steroids should be used with great care in girls, because the drugs have masculinizing properties. The drugs should be reserved for situations in which the benefits outweigh the risk. Anabolic steroids have been associated with liver **cancer**, and they have psychological effects, such as contributing to rage attacks.

Bottle of the injectable anabolic steroid, Durabolin. *(Custom Medical Stock Photo, Inc.)*

KEY TERMS

Anabolic—Refers to metabolic processes characterized by the conversion of simple substances into more complex compounds.

Catabolism—A process of metabolism that breaks down complex substances into simple ones.

Testosterone—Male hormone produced by the testes and (in small amounts) in the ovaries. Testosterone is responsible for some masculine secondary sex characteristics such as growth of body hair and deepening voice. It also is sometimes given as part of hormone replacement therapy to women whose ovaries have been removed.

Turner syndrome—A chromosome abnormality characterized by short stature and ovarian failure caused by an absent X chromosome. It occurs only in females.

Side effects

Anabolic steroids cause masculinization of females, including hair growth or loss, enlargement of the clitoris, and deepening of the voice. These effects are not reversible, even when the drug is promptly discontinued. In males past the age of **puberty**, side effects include increased urinary frequency, breast tenderness and enlargement, and frequent erections. In both males and females, anabolic steroids cause swelling of the feet, liver problems, and stomach upset.

This list of side effects is incomplete; many additional effects have been reported. Specialized drug references maintain for a complete list for each individual drug, including analysis of psychological effects of these drugs that contribute to rage attacks.

Interactions

Anabolic steroids have an anticoagulant effect. They should be used with care in combination with other drugs that have the same effect, including warfarin, **nonsteroidal anti-inflammatory drugs** (NSAIDs), and aspirin. Simultaneous use of anabolic steroids and corticosteroids will increase the risk of foot and ankle swelling. The combination is very likely to cause **acne**. Anabolic steroids may also lower blood glucose levels. They should be used with extreme care in patients taking insulin or other antidiabetic drugs. Other drug interactions have also been reported.

Parental concerns

Because of the nature of the adverse effects of anabolic steroids, their use should be restricted to cases where the benefits clearly outweigh the risks. Regular blood testing for blood counts and liver function is essential. Parents should observe the patient carefully for signs of liver damage, including **headache**, unpleasant breath odor, and black tarry stools.

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Samuel Uretsky, PharmD

Analgesics

Definition

Analgesics are medicines that relieve **pain**.

Description

Analgesics are those drugs whose primary purpose is pain relief. The primary classes of analgesics are the narcotics, including additional agents that are chemically based on the morphine molecule but have minimal abuse potential; **nonsteroidal anti-inflammatory drugs** (NSAIDs) including the salicylates; and **acetaminophen**. Other drugs, notably the tricyclic **antidepressants** and anti-epileptic agents, such as gabapentin, have been used to relieve pain, particularly neurologic pain, but are not routinely classified as analgesics. Analgesics provide symptomatic relief but have no effect on causation, although clearly the NSAIDs, by virtue of their dual activities as pain relievers and anti-inflammatories, may be beneficial in both regards.

Description

Pain has been classified as "productive" and "non-productive." While this distinction has no physiologic meaning, it may serve as a guide to treatment. Productive pain has been described as a warning of injury and so may be both an indication of need for treatment and a guide to diagnosis. Non-productive pain by definition serves no purpose either as a warning or diagnostic tool.

Although pain syndromes may be dissimilar, the common factor is a sensory pathway from the affected organ to the brain. Analgesics work at the level of the nerves, either by blocking the signal from the peripheral nervous system or by distorting the interpretation by the central nervous system. Selection of an appropriate analgesic is based on consideration of the risk-benefit factors of each class of drugs, based on type of pain, severity of pain, and risk of adverse effects. Traditionally, pain has been divided into two classes, acute and chronic, although severity and projected patient survival are other factors that must be considered in drug selection.

Acute pain

Acute pain is self limiting in duration and includes post-operative pain, pain of injury, and **childbirth**. Because pain of these types is expected to be short term, the long-term side effects of analgesic therapy may routinely be ignored. Thus, these patients may safely be treated with narcotic analgesics without concern for their addictive potential, or NSAIDs with only limited concern for their ulcerogenic (ulcer-causing) risks. Drugs and doses should be adjusted based on observation of healing rate, switching patients from high to low doses and from narcotic analgesics to non-narcotics when circumstances permit.

An important consideration of **pain management** in severe pain is that patients should not be subject to the return of pain. Analgesics should be dosed adequately to assure that the pain is at least tolerable and frequently enough to avoid the anxiety that accompanies the anticipated return of pain. Generally analgesics should not be dosed on an as-needed basis but should be administered often enough to assure constant blood levels of analgesic. This applies to both the narcotic and non-narcotic analgesics.

Chronic pain

Chronic pain, pain lasting over three months and severe enough to impair function, is more difficult to treat, since the anticipated side effects of the analgesics are more difficult to manage. In the case of narcotic analgesics this means the **addiction** potential, as well as respiratory depression and **constipation**. For the NSAIDs, the risk of gastric ulcers may be dose limiting. While some classes of drugs, such as the narcotic agonist/antagonist drugs bupronophine, nalbuphine, and pentazocine, and the selective COX-2 inhibitors celecoxib and rofecoxib represent advances in reduction of adverse effects, they are still not fully suitable for long-term management of severe pain. Generally, chronic pain management requires a combination of drug therapy, life-style modification, and other treatment means.

Narcotic analgesics

The narcotic analgesics, also termed opioids, are all derived from opium. The class includes morphine, codeine, and a number of semi-synthetics including meperidine (Demerol), propoxyphen (Darvon), and others. The narcotic analgesics vary in potency, but all are effective in treatment of visceral pain when used in

adequate doses. Adverse effects are dose related. Because these drugs are all addictive, they are controlled under federal and state laws. A variety of dosage forms are available, including oral solids, liquids, intravenous and intrathecal injections, and transcutaneous patches.

NSAIDs are effective analgesics even at doses too low to have any anti-inflammatory effects. There are a number of chemical classes, but all have similar therapeutic effects and side effects. Most are appropriate only for oral administration; however, ketorolac (Toradol) is appropriate for injection and may be used in moderate to severe pain for short periods.

Three new NSAIDs, celecoxib, rofecoxib, and valdecoxib may reduce the risk of gastric ulcers in long-term use for adults and have been widely advertised. As of 2004 these drugs had not been properly tested in children, and even in adults, their advantages were not well established. These drugs should not be given to infants and are not well documented for use in older children.

Acetaminophen is a non-narcotic analgesic with no anti-inflammatory properties. It is appropriate for mild to moderate pain. Although the drug is well tolerated in normal doses, it may have significant toxicity at high doses. Because acetaminophen is largely free of side effects at therapeutic doses, it has been considered the first choice for mild pain, including that of osteoarthritis.

General use

Appropriate dosage varies by drug and should consider the type of pain, as well as other risks associated with patient age and condition. For example, narcotic analgesics should usually be avoided in patients with a history of substance abuse but may be fully appropriate in patients with **cancer** pain. Similarly, because narcotics are more rapidly metabolized in patients who have used these drugs for a long period, higher than normal doses may be needed to provide adequate pain management.

Precautions

Narcotic analgesics may be contraindicated in patients with poor respiratory function. NSAIDS should be used with care in patients with insufficient kidney function or **coagulation disorders**. NSAIDs are contraindicated in patients who are allergic to aspirin.

Side effects

Parents of children taking analgesics should review adverse effects of each drug individually. Drugs within a class may vary in their frequency and severity of adverse effects.

The primary adverse effects of the narcotic analgesics are addiction, constipation, and poor respiratory function. Because narcotic analgesics stimulate the production of enzymes that cause the metabolism of these drugs, patients on narcotics for a prolonged period may require increasing doses. This physical tolerance is not the same thing as addiction and is not a reason for withholding medication from patients in severe pain.

NSAIDs may cause kidney problems. Gastrointestinal discomfort is common, although in some cases, these drugs may cause ulcers without the prior warning of gastrointestinal distress. NSAIDs may cause blood to clot less readily, although not to the same extent as if seen with aspirin.

Interactions

Parents should study information on interactions for specific drugs their children are taking.

Analgesics will interact with other drugs that have similar side effects. Nonsteroidal anti-inflammatory drugs should be used with care with other drugs that may cause stomach upset, such as aspirin. Narcotic analgesics should be used with care when taken in combination with drugs that inhibit respirations, such as the benzodiazepines.

Parental concerns

Regarding acetaminophen, parents should never confuse baby formulations, which are high concentration, with children's formulas. The infant formulas are meant to be given by the drop, never by the teaspoonful. Children's liquids are for teaspoonful dosing. Parents must read labels carefully and use the appropriate measure.

Aspirin should never be given to children under the age of 16 who have **chickenpox** or **influenza**, because children who have received aspirin for these conditions seem to have a higher than expected frequency of developing **Reye's syndrome**. High dose aspirin may be given to children for treatment of rheumatism, but this should only be done under medical supervision.

Regarding narcotics, although addiction is a concern when narcotic analgesics are used, this concern is not a problem when the medications are given appropriately. When a child is in severe pain, these pain relievers should not be withheld.

KEY TERMS

Acute pain—Pain in response to injury or another stimulus that resolves when the injury heals or the stimulus is removed.

Anodyne—A medicinal herb or other drug that relieves or soothes pain.

Chronic pain—Pain that lasts over a prolonged period and threatens to disrupt daily life.

Inflammation—Pain, redness, swelling, and heat that develop in response to tissue irritation or injury. It usually is caused by the immune system's response to the body's contact with a foreign substance, such as an allergen or pathogen.

Juvenile arthritis—A chronic inflammatory disease characterized predominantly by arthritis with onset before the sixteenth birthday.

Osteoarthritis—A noninflammatory type of arthritis, usually occurring in older people, characterized by degeneration of cartilage, enlargement of the margins of the bones, and changes in the membranes in the joints. Also called degenerative arthritis.

See also Acetaminophen; Nonsteroidal anti-inflammatory drugs; Pain management.

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Anaphylaxis

Definition

Anaphylaxis is a severe, sudden, and potentially fatal allergic reaction to a foreign substance or antigen that affects multiple systems of the body.

Description

Anaphylaxis is a severe, whole-body allergic reaction. After initial exposure to a substance such as wasp sting toxin, the allergic child's immune system becomes sensitized to that allergen. On a subsequent exposure to the specific allergen, an allergic reaction, which can involve a number of different areas of the body, occurs. Anaphylaxis is thought to result from antigen-antibody interactions on the surface of mast cells, connective tissue cells that are believed to contain a number of regulatory, or mediator, chemicals. Specifically, an immunoglobulin antibody protein, IgE, is produced in response to the presence of the allergen. IgE binds to the mast cells, causing them to suddenly release a number of chemicals, including histamine, heparin, serotonin, and bradykinin. Once released, these chemicals produce the bodily reactions that characterize anaphylaxis: constriction of the airways, causing wheezing and difficulty in breathing; and gastrointestinal symptoms, such as abdominal **pain**, cramps, **vomiting**, and **diarrhea**. Shock can occur when the released histamine causes the blood vessels to dilate, which lowers blood pressure; histamine also causes fluids to leak from the bloodstream

EpiPen Jr., a syringe containing a child's dosage of adrenaline, is used for the emergency treatment of anaphylactic shock. *(© Mark Thomas/Photo Researchers, Inc.)*

into the tissues, lowering the blood volume. Pulmonary edema can result from fluids leaking into the alveoli (air sacs) of the lung.

Substances that can trigger an anaphylactic reaction include:

- insect **stings** from hornets, wasps, yellow jackets, honey bees, or fire ants
- medications, including penicillin, cephalosporin, anesthetics, streptokinase, and others
- foods (ingesting even tiny amounts or simply being near the offending food), including peanuts, tree nuts (such as walnuts or almonds), fish, shellfish, eggs, milk, soy, and wheat
- vaccines, including **allergy shots** and egg- and gelatin-based vaccines
- hormones, including insulin and possibly progesterone
- rubber latex products
- animal and human proteins, including seminal fluid and horse serum (which is used as snake anti-venom)

Anaphylactoid (meaning "anaphylactic-like") reactions are similar to those of true anaphylaxis but do not require an IgE immune reaction. These are usually caused by direct stimulation of the mast cells. The same chemicals as with anaphylaxis are released, with the same effects, so the symptoms are treated the same way. However, an anaphylactoid reaction can occur on initial exposure to an allergen as well as on subsequent exposures, since no sensitization is required.

There is also a rare kind of food allergy, called exercise-induced allergy, that is caused by eating a specific food and then exercising. It can produce **itching**, lightheadedness, **hives**, and anaphylaxis. The offending food does not cause a reaction without **exercise**, and, alternately, exercise does not cause a reaction without ingesting the food beforehand.

Demographics

Although likely an underestimate, about 10,000 cases of anaphylaxis occur per year in North America, with about 750 fatalities a year. The exact prevalence of anaphylaxis is unknown, because milder reactions may be attributed to **asthma** attacks or sudden cases of hives, and more serious or fatal episodes might be reported as heart attacks, as the initial symptoms of hives, asthma, and swollen throat can fade quickly.

Causes and symptoms

The symptoms of anaphylaxis may occur within seconds of exposure, or be delayed 15 to 30 minutes and sometimes even an hour or more later, if the allergen is aspirin or other similar drugs. The sooner the symptoms occur after exposure, the more severe the anaphylactic reaction is likely to be.

The first symptoms of an anaphylactic reaction are associated with the skin: flushing (warmth and redness), itching (often in the groin or armpits), and hives. These symptoms are often accompanied by **anxiety**; a rapid, irregular pulse; and a sense of impending doom. Then the throat and tongue swell, the voice becomes hoarse, and swallowing and breathing become labored. Symptoms of **rhinitis** or asthma may also occur, causing a runny nose, sneezing, wheezing, and abnormal high-pitched breathing sounds, further worsening the breathing problems. Gastrointestinal effects may also develop, including vomiting, diarrhea, and stomach cramps. The child may be confused and have slurred speech. In about 25 percent of the cases, the chemicals flooding the blood stream will cause a generalized opening of capillaries (tiny blood vessels), resulting in a drop in blood pressure, lightheadedness, and even a loss of consciousness, which are typical symptoms of anaphylactic shock. The child may exhibit blueness of the skin (cyanosis), lips, or nail beds.

After the original symptoms occur, there are three possible outcomes:

- The symptoms may be mild and fade spontaneously or be quickly ended by administering emergency medication. The anaphylactic episode is over for that particular exposure.
- After initial improvement, the symptoms may reoccur after four to 12 hours (a late phase recurrent reaction)

and require additional treatment and monitoring. Late phase reactions occur in about 10 percent of cases.

- The reaction may be persistent and severe, requiring extensive medical treatment and **hospitalization**. This condition occurs in about 20 percent of cases.

When to call the doctor

The child should be given immediate emergency care, if possible, and then taken to the emergency room or the local emergency number (e.g., 911) should be called if symptoms of anaphylaxis develop.

Diagnosis

A child having an anaphylactic reaction will exhibit typical symptoms of anaphylaxis, such as hives and swelling of the eyes or face, blue skin from lack of oxygen, or pale skin from shock. The airway may be blocked, and the child may be wheezing as well as confused and weak. The pulse will be rapid and the blood pressure may be low. Anaphylaxis is an emergency condition that requires immediate professional medical attention.

Once a child has had an anaphylactic reaction, an allergist should be consulted to identify the specific allergen that caused the reaction. The allergist will take a detailed medical history and use blood or skin tests to identify the allergen. The allergist will ask about activities that the child participated in before the event, food and medications the child may have ingested, and whether the child had contact with any rubber products.

Treatment

Because of the severity of these reactions, treatment must begin immediately. The most common emergency treatment involves injection of epinephrine (adrenaline) to stop the release of histamines and relax the muscles of the respiratory tract. The injection is given in the outer thigh and can be administered through light fabric such as trousers, skirts, or stockings. Heavier clothing may have to be removed prior to the injection. After the injection, emergency services or 911 should be called immediately. A child with known severe allergic reactions should be carrying an allergy kit with epinephrine; if not, treatment will have to be delayed until emergency personnel can provide the required medication. For reactions to insect stings or allergy shots, a tourniquet should be placed between the puncture site and the heart; the tourniquet should be released every 10 minutes. If the child is conscious, he or she should lie down and elevate the feet. If trained, the parents or others present should administer CPR if the child stops breathing or does not have a pulse. After 10 to 15 minutes, if symptoms are still significant, another dose of epinephrine can be injected. Even after the reaction subsides, the child should still be taken to the emergency room immediately and monitored for three to four hours, since symptoms can redevelop. Other treatments may be given by medical personnel, including oxygen, intravenous fluids, breathing medications, and possibly more epinephrine. The epinephrine may make the child feel shaky and have a rapid, pounding pulse, but these are normal side effects and are only dangerous to those with heart problems. Steroids and **antihistamines** may also be given but are usually not as helpful initially as epinephrine. However, they may be useful in preventing a recurrent delayed reaction.

If the child is being treated with beta blocker medications commonly used to treat high blood pressure, angina, thyroid disorders, migraines, or glaucoma, it may be difficult to reverse an anaphylactic reaction.

Prognosis

Anaphylaxis is a severe disorder that has a poor prognosis without prompt treatment. Symptoms are usually resolved with appropriate therapy; therefore, immediate emergency care is essential.

Prevention

For children with known reactions to **antibiotics**, foods, insect stings, specific foods, or any of the allergens that can induce an anaphylactic reaction, avoidance of the symptom-inducing agent is the best form of prevention.

Specific avoidance measures that are recommended include:

Drugs/medications:

- Parents should advise healthcare personnel of the childs **allergies**.
- Parents should ask the doctor whether prescribed medications could contain the drug(s) to which the child is allergic.
- The child should take all medications by mouth, if possible, since the risk of anaphylaxis is greater with injections.
- Any child should stay in a doctors office or near medical care for a period of time after receiving injections of an antibiotic or vaccine.

Insect stings:

- The child should avoid areas where insects breed and live.
- The child should not wear bright clothing, perfume, hair spray, or lotions that might attract insects.
- If possible the child should wear long sleeves, long trousers, and shoes when out of doors.

Food:

- The child must be instructed to never again eat that kind of food that causes an anaphylactic reaction.
- Parents should carefully read all ingredient labels of foods that the child might eat and be aware of the different terms used for various foods, such as caseinate for milk or albumin for eggs.
- Parents should ask about ingredients in foods while eating out with the child, bring safe substitutes from home, and bring an allergy kit.
- Parents should be aware of possible cross-contamination, such as when an ice cream scoop is used for Rocky Road ice cream, which contains peanuts, and then for vanilla ice cream.
- School kitchen personnel should be notified of the childs condition.
- The child should avoid eating foods that might cross-react with foods that the child is allergic to, for example, if the child is allergic to shrimp, the child may also be allergic to crab or lobster.
- When traveling to other countries, parents should learn the appropriate words for foods that trigger their childs allergy; in addition, parents can request that air carriers serve peanut-free snacks to all passengers when their child is traveling; also the child should avoid eating airline meals.

Latex:

- The child should avoid all latex rubber products.
- If the child has to be hospitalized, the parents should alert the hospital personnel to the childs allergy to latex.
- A child with a latex allergy may also have allergies to kiwi fruit, passion fruit, papayas, bananas, avocados, figs, peaches, nectarines, plums, tomatoes, celery, and chestnuts.

In addition, children with a history of allergic reactions should carry an emergency kit containing injectable epinephrine and chewable antihistamine and be instructed in its use. A child who is not prepared to deal with an anaphylactic reaction is at an increased risk of dying. The allergy kit should include simple instructions on when and how to use the kit; sterilizing swabs to cleanse the skin before and after the injection; epinephrine in a preloaded syringe, as prescribed by the childs doctor in doses appropriate for children; and antihistamine tablets. The expiration date on the medications in the allergy kit should be checked and medications replaced as needed. Also, the epinephrine solution should be clear; if it is pinkish brown, it should be discarded and replaced.

There are many brands of allergy kits. The simplest kit to use is the Ana-kit, which contains a sterile syringe preloaded with two doses of epinephrine with a stop between. Another commonly used kit is the Epi-Pen, which carries a single self-injecting, spring-loaded syringe of epinephrine. Two Epi-Pen kits should be carried, so that two doses are available. Allergy kits should be kept at home, school, and **day care**; and the school administrator, teachers, and friends should be made aware of the childs allergies. Adults associated with the child should be trained in giving an injection and have a plan to transport the child to the hospital. Older children should be taught to give self-injections. Children at risk for anaphylaxis should also wear a Medic Alert bracelet or necklace or carry a medical emergency card with them at all times that clearly describes their allergy.

A consultation with an allergist can help to identify the substances that trigger the reaction; the allergist can also provide information on how to best avoid the triggering substance. The allergist may also be able to give allergy shots to children with wasp, yellow jacket, hornet, honey bee, or fire ant allergies. These shots provide 90 percent protection against the first four insect reactions, but less protection against fire ant reactions. Premedication is also helpful in preventing anaphylaxis from x-ray dyes; also there may be alternative dyes available for use that are less likely to cause reactions. Desensitization to medications has also been successful in some cases. The process involves gradually increasing the amount of medication given under controlled conditions. The procedure has worked for sensitivities to penicillin, sulfa drugs, and insulin.

The risk of anaphylaxis sometimes diminishes over time if there are no repeated exposures or reactions. However, the child at risk should also expect the worst and be prepared with preventive medication.

Parental concerns

Parents caring for children who are at risk for life-threatening anaphylactic reactions may experience high stress levels, for they have to maintain vigilance in order to protect the child while creating a sense of normalcy as

KEY TERMS

Allergen—A foreign substance that provokes an immune reaction or allergic response in some sensitive people but not in most others.

Allergy—A hypersensitivity reaction in response to exposure to a specific substance.

Epinephrine—A hormone produced by the adrenal medulla. It is important in the response to stress and partially regulates heart rate and metabolism. It is also called adrenaline.

Immunoglobulin E (IgE)—A type of protein in blood plasma that acts as an antibody to activate allergic reactions. About 50% of patients with allergic disorders have increased IgE levels in their blood serum.

the child grows up. Parents can reduce their stress by using social support groups, accepting their childs condition, and maintaining a positive attitude.

See also Allergies.

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Judith Sims

Anatomical age

Definition

Anatomical age is the numerical **assessment** of a child's physical growth in relation to the statistical average based on the child's chronological age.

Description

Using statistical data, the American Academy of Pediatrics and the National Institutes of Health in the United States have developed tables to illustrate the growth patterns of children. These tables describe the population of all children of a certain age, with ranges for weight, height, and other physical characteristics. For most children, anatomical age—based on weight and height measurements—is the same as chronological age—based on the number of months or years since birth. However, when a child's physical growth falls outside the range of his chronological age, the child's age is determined by his growth. For example, if a six-year-old's height and weight falls within the range for five-year-olds, his anatomical age will be given as five, not six years.

Anatomical age has been used as a determination of chronological age by immigration and **adoption** agencies when no birth certificates have been available. This has been crucial for refugee children in particular. However, prolonged **malnutrition** can cloud actual age assessments. That is why radiographs, or **x rays**, of a child's bones have been taken and read by osteologists (doctors who special in the skeletal system) and radiologists.

William Walter Greulich and S. Idell Pyle's atlas of **skeletal development** and Tanner and White's method are the two major assessment tools for wrist radiography. X rays of a child's left wrist are compared visually with a series of x rays of wrists of children of various ages. A computerized system has been developed based on Tanner and Whitehouse's model, which uses pattern recognition like finger print databases. This technique greatly speeds up the diagnostic process.

In addition, an ultrasound version of the Greulich-Pyle Atlas has proven to be as effective as wrist x rays. This method does not expose children to radiation.

Common problems

Standardized growth charts and wrist radiographs can assess normal skeletal growth and determine whether there are problems concerning growth that is too fast or

KEY TERMS

Osteologist—A doctor who specializes in the skeletal system.

Radiograph—The actual picture or film produced by an x-ray study.

too slow. Anatomical age assessment can also reveal other conditions. Because bone growth is affected by calcium regulating hormones, sex steroids, and thyroid hormones, disturbances in the endocrine system, which controls hormones, can be detected.

Parental concerns

Determining anatomical age can give parents an indication of their children's future growth and can help them work with their doctors to determine treatment if there seem to be growth problems.

When to call the doctor

Usually, the child sees the doctor for immunizations, school physical exams, or childhood illnesses. At these times, the doctor may discuss findings about anatomical age with parents. If parents have concerns about their children's growth, they can bring them up at these visits.

See also X rays.

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Janie Franz

Anemias

Definition

Anemia is a blood disorder characterized by abnormally low levels of healthy red blood cells (RBCs) or reduced hemoglobin (Hgb), the iron-bearing protein in red blood cells that delivers oxygen to tissues throughout the body. Reduced blood cell volume (hematocrit) is also considered anemia. The reduction of any or all of the three blood parameters reduces the oxygen-carrying capability of the blood, causing reduced oxygenation of body tissues, a condition called hypoxia.

Description

All tissues in the human body need a regular supply of oxygen to stay healthy and perform their functions. RBCs contain Hgb, a protein pigment that allows the cells to carry oxygen (oxygenate) tissues throughout the body. RBCs live about 120 days and are normally replaced in an orderly way by the bone marrow, spleen, and liver. As RBCs break down, they release Hgb into the blood stream, which is normally filtered out by the kidneys and excreted. The iron released from the RBCs is returned to the bone marrow to help create new cells. Anemia develops when either blood loss, a slow-down in the production of new RBCs (erythropoiesis), or an increase in red cell destruction (hemolysis) causes significant reductions in RBCs, Hgb, iron levels, and the essential delivery of oxygen to body tissues.

Anemia can be mild, moderate, or severe enough to lead to life-threatening complications. More than 400 different types of anemia have been identified. Many of them are rare. Most are caused by ongoing or sudden blood loss. Other causes include vitamin and mineral deficiencies, inherited conditions, and certain diseases that affect red cell production or destruction.

Anemia in newborn infants is noted when hemoglobin levels are lower than expected for the birth weight and postnatal age. Premature or low birth-weight infants may have lower hemoglobin levels. The normal newborn Hgb is 16.8 dL, which may be 1 to 2 dL lower if birth weight is abnormally low. Anemia may be the first sign of certain disorders in the newborn, such as blood loss that has occurred from transplacental hemorrhage, a condition in which the infant's blood bleeds back into the mother's circulation; bleeding from ruptures in the liver, spleen, adrenals, or kidneys; or hemorrhage within the brain (intracranial hemorrhage). Anemia can also be caused by the destruction of red blood cells or reduced red blood cell production. Newborns may also have low red blood cell volume (hematocrit or Hct) if they were born by **cesarean section**. It must be noted, however, that hemoglobin decreases naturally (physiologic decrease) in infants by eight to 12 weeks of age, leveling at a normal value of 11 g/dL or better.

Iron-deficiency anemia

Iron deficiency anemia is the most common form of anemia worldwide. In the United States, it affects thousands of toddlers between one and two years of age and more than 3 million women of childbearing age. This condition is less common in older children and in adults over 50 and rarely occurs in teenage boys and young men.

The onset of iron deficiency anemia is gradual and may not have early symptoms. The deficiency begins when the body loses more iron than it derives from food and other sources. Because depleted iron stores cannot meet the red blood cell's needs, fewer red blood cells develop. In this early stage of anemia, the red blood cells look normal, but they are reduced in number. Then the body tries to compensate for the iron deficiency by producing more red blood cells, which are characteristically small in size (spherocytosis). Symptoms of anemia, especially weakness and fatigue, develop at this stage. Individuals may be given iron preparations by injection or advised to take oral iron supplements. It sometimes helps to take vitamin C along with oral iron supplementation to encourage better absorption of the iron. Taking iron supplements can result in **diarrhea**, cramps, or **vomiting**.

Folic acid deficiency anemia

Folic acid deficiency anemia is the most common type of megaloblastic anemia, arising from a problem with the synthesis of deoxyribonucleic acid (DNA) within the cells of the body. It is characterized by RBCs that are larger than normal and is caused by a deficiency of folic acid, a vitamin that the body needs to produce normal cells and normal DNA.

Folic acid anemia is especially common in infants and teenagers. This condition usually results from a dietary deficiency but may also be due to an inability to absorb (malabsorption) folic acid. Folic acid is available in many foods, such as cheese, eggs, fish, green vegetables, meat, milk, mushrooms, and yeast. **Smoking** raises the risk of developing this condition by interfering with the absorption of vitamin C, which the body needs to absorb folic acid. Folic acid anemia can be a complication of pregnancy, when a woman's body needs eight times more folic acid than it does otherwise. Folic acid deficiency in pregnant women may lead to birth defects in their children. Supplementation of folic acid is recommended during pregnancy.

Vitamin B_{12} deficiency anemia

Less common in the United States than folic acid anemia, vitamin B_{12} deficiency anemia is another type of megaloblastic anemia that develops when the body does not absorb enough of this nutrient. Necessary for the creation of healthy RBCs, B_{12} is found in meat, eggs, whole grains, and most vegetables. Large amounts of B_{12} are stored in the body, so this condition may not become apparent until up to four years after B_{12} absorption stops or slows down. The resulting drop in RBC production can cause loss of muscle control; loss of sensation in the legs, hands, and feet; soreness, slickness, or burning of the tongue; weight loss; or yellow-blue **color blindness**. Confusion, depression, and memory loss may also be associated with the deficiency.

Pernicious anemia is the most common form of B_{12} deficiency. Since most people who eat meat or eggs get enough B_{12} in their diets, a deficiency of this vitamin usually means that the body is not absorbing it properly. This condition can be found in those who do not produce adequate amounts of a chemical secreted by the stomach lining that combines with B_{12} to help its absorption in the small intestine. Pernicious anemia is diagnosed more often in adults between ages 50 and 60 than in children or young people, although there is the possibility of inheriting the condition, with symptoms not appearing until later in life.

Vitamin C deficiency anemia

Anemia due to vitamin C deficiency is a rare disorder that causes the bone marrow to manufacture abnormally small red blood cells. Vitamin C deficiency anemia results from a severe, long-standing dietary deficiency or malabsorption of this essential vitamin. It is usually easily corrected with supplementation.

Hemolytic anemia

Hemolytic anemia can be present at birth (congenital hemolytic anemia or spherocytosis) or acquired later in life. It is the result of either infection or the presence of antibodies that destroy RBCs more rapidly than bone marrow can replace them. Hemolytic anemia can enlarge the spleen, an organ that also produces red blood cells when necessary. Production of cells by the spleen will increase to meet the demands of accelerated RBC destruction (hemolysis). Complications of hemolytic anemia in older children or adults include **pain**, gallstones, and other serious health problems.

Hemolytic disease of the newborn is a specific variation of hemolytic anemia in which an incompatibility exists between antigens on the cells of the mother and baby, causing antibodies to develop in the mother's circulation. The antibodies are produced as an immune response to what the body views as foreign antigens on the surface of the infant's RBCs. Several specific anti-

gens are responsible for the incompatibilities: Rh type incompatibility, ABO blood group incompatibility, and other incompatibilities involving antigens known as Kell, Duffy, M, N, and P, among many others. Hemolytic disease of the newborn and the anemia that results is detectable within the first few days after birth. Depending on the strength of the antibody, the anemia may clear up on its own or exchange transfusions may be necessary to replace the newborn's blood.

Thalassemia

An inherited form of hemolytic anemia, **thalassemia** comes from the production of abnormal hemoglobin. It is characterized by low hemoglobin and unusually small and fragile RBCs (microcytosis), although the RBC count may be normal. Thalassemia has several types that involve imbalances in the four chains of amino acids that comprise hemoglobin (alpha- and beta-globins). In thalassemia minor or thalassemia trait (heterozygous thalassemia), also called alpha-thalassemia, there is an imbalance in the production of the alpha chain of amino acids. In thalassemia minor, fetal hemoglobin (HbF), the hemoglobin form that circulates in the fetus, does not decrease normally after birth and may remain high in later life. A child may inherit thalassemia trait when only one parent has the genes responsible for it. It is usually not treated and does not have serious consequences. Thalassemia major (homozygous thalassemia or Cooley's anemia) occurs in children in whom both parents pass on the genes responsible. It is known as beta-thalassemia, because of an imbalance in the beta chain amino acids of hemoglobin. It also involves the persistence of HbF with larger than normal amounts appearing in the child's circulation. Alpha-thalassemias occur most commonly in African Americans; beta-thalassemias most commonly affect people of Mediterranean or middle-Eastern ancestry and Southeast Asians. Hemoglobin H disease is another form of thalassemia in which three of the four beta-globin genes are missing.

Sickle cell anemia

Sickle cell anemia is an inherited, chronic, incurable blood disorder that causes the body to produce defective hemoglobin, the abnormal HgbS, which occurs primarily in African Americans. The condition is characterized by abnormal, crescent-shaped RBCs. Unlike normal oval cells, fragile sickle cells cannot hold enough hemoglobin to nourish body tissues. The deformed shape makes it hard for sickle cells to pass through narrow blood vessels. When capillaries become obstructed, a life-threatening condition called sickle cell crisis is likely to occur. A child who inherits the sickle cell gene from each parent will have the disease. A child who inherits the sickle cell gene from only one parent carries the sickle cell trait but does not have the disease.

Aplastic anemia

Sometimes curable by bone marrow transplant, but potentially fatal, aplastic anemia is characterized by decreased production of red and white blood cells and platelets (disc-shaped cells that are a key component of blood coagulation). This disorder may be inherited or acquired as a result of the following:

- recent severe illness
- long-term exposure to industrial chemicals
- chemotherapy, use of anticancer drugs, and certain other medications

Anemia of chronic disease

Cancer, chronic infection or inflammation, and kidney and liver disease often cause mild or moderate anemia. Chronic liver failure generally produces the most severe symptoms because the production of RBCs is directly affected.

Causes and symptoms

Anemias do not all stem from the same causes. Anemia can be the result of injuries, chronic or acute illnesses, complications of surgery or **childbirth**, metabolic disturbances or deficiencies, and adverse response to drug therapy administered for other conditions. Causes may include sudden or ongoing loss of blood, nutritional deficiencies, decreased red blood cell production, or increased red blood cell destruction. **Malnutrition** or malabsorption of nutrients can contribute to vitamin deficiency anemia and iron deficiency anemias. Although red cell destruction and replacement is an ongoing process in the body, hereditary disorders and certain diseases can accelerate blood cell destruction, resulting in anemia. However, excessive bleeding is the most common cause of severe anemia, and the speed with which blood loss occurs has a significant effect on the severity of symptoms. Chronic blood loss may be a consequence of the following:

- cancer
- gastrointestinal tumors
- diverticulosis
- polyposis
- heavy or frequent menstrual flow
- hemorrhoids
- nosebleeds

- stomach ulcers
- long-standing alcohol abuse

Acute blood loss may occur as a result of injury, a ruptured blood vessel, or a complication of surgery or childbirth. When a lot of blood is lost within a short time, blood pressure and the amount of oxygen in the body drop suddenly, sometimes leading to heart failure or death. Loss of even one third of the body's blood volume in the space of several hours can be fatal. Gradual blood loss is less threatening, because the body has time to replace RBCs and blood volume.

Symptoms

Weakness, fatigue, and a run-down feeling may be the first signs of anemia. Pasty or sallow skin color, or the absence of color in the gums, nail beds, creases of the palm, or lining of the eyelids are other signs of anemia. Individuals who appear to be weak, easily tired, often out of breath, and who may feel faint or dizzy on movement may be severely anemic.

Other symptoms of anemia may include the following:

- unusual cravings for ice (chewing on ice cubes), paint, or earth (actually eating dirt)
- headache
- inability to concentrate, memory loss
- inflammation of the mouth (**stomatitis**) or tongue (glossitis)
- insomnia
- irregular heartbeat
- loss of appetite
- dry, brittle, or ridged nails
- rapid breathing
- sores in the mouth, throat, or rectum
- perspiration, especially around the head and neck
- swelling of hands and feet
- constant thirst
- ringing in the ears (tinnitus)
- unexplained bleeding or bruising
- angina pectoris, i.e., chest pain accompanied by a **choking** sensation that may provoke anxiety

Demographics

Acquired anemias affect about 4 million individuals in the United States, and over 50 percent of these are under age 45, although less than 10 percent of cases occur in children and adolescents. In the United States, iron deficiency anemia is the most prevalent type of anemia, affecting about 240,000 toddlers between one and two years of age and 3.3 million women of childbearing age. Anemia due to gradual blood loss is more common in women than in men, particularly pregnant women or women of menstruating age. Pernicious anemia is more common in women and in African Americans and is less common in other racial groups. Folate deficiency is not common in young people who eat an adequate diet and is usually associated with malnutrition, pregnancy, and **alcoholism**. Sickle cell anemia is more frequently diagnosed than thalassemias and occurs most often among African Americans. Thalassemia occurs in four out of 100,000 individuals in the United States, particularly among those of Mediterranean, Asian, or middle Eastern descent.

When to call the doctor

When a child exhibits weakness, **dizziness**, listlessness, or fatigue, it may be the first sign of anemia. The pediatrician should be consulted if the child is also extremely pale or has little or no color in the gums, nail beds, creases of the palm, or lining of the eyelids. Any prolonged bleeding or sudden blood loss requires examination by a physician and testing for anemia.

Diagnosis

The child's medical history will be taken, including the child's age, symptoms, illnesses, and general state of health, and a **family** history of ancestry and known inherited anemias will be noted. Symptoms noticed in children by their parents may include fatigue, weight loss, inability to concentrate, loss of appetite, and light-headedness when standing up. The physical examination may reveal paleness, lack of color in the creases of the palm, gums, and the linings of the eyelids. The child's breathing rate may be increased and, in advanced cases, the spleen or liver may be enlarged when palpated. If anemia is due to chronic disease, there may be evidence of infection or inflammation. Urine output may be reduced in severe anemia.

Diagnostic testing begins with a complete blood count (CBC) and differential to reveal the RBC count, white blood cell (WBC) count, hemoglobin (Hgb), and hematocrit (Hct); any of these counts can be altered, and in most anemias the RBC and hemoglobin will be reduced. The mean corpuscular volume (MCV) will be measured to compare the size of RBCs with normal RBCs. A reticulocyte (young RBCs) count will help determine if anemia is caused by impaired RBC production or increased RBC destruction. Iron, vitamin C, vitamin B_{12}, and folate levels will be measured to evaluate

and identify possible deficiencies. Diagnosing thalassemia and sickle cell anemia, both of which involve disorders of hemoglobin, will require measuring the different types of hemoglobin through a laboratory testing method called hemoglobin electrophoresis. In some anemias, a bone marrow sample will be removed (bone marrow biopsy) for microscopic examination, especially to confirm iron deficiency anemia or the megaloblastic anemias. Kidney function tests, coagulation tests, and stool examinations for occult blood may also be performed.

Treatment

Surgery may be necessary to correct blood losses caused by injury or hemorrhage (nose bleeds, aneurysm, cerebral hemorrhage, bleeding ulcer) or childbirth. Transfusions of packed red blood cells or whole blood may also be used to replace blood volume and to stimulate the body's own production of red blood cells. Medication or surgery may also be necessary to control heavy menstrual flow or to remove polyps (growths or nodules) from the bowels.

Anemia due to nutritional deficiencies can usually be treated with iron replacement therapy, specific vitamin supplements, or self-administered injections of vitamin B_{12}. People with folic acid anemia may be advised to take oral folic acid.

Vitamin B_{12} deficiency anemia requires a life-long regimen of B_{12} shots to maintain vitamin levels and control symptoms of pernicious anemia. The patient may be advised to limit physical activity until treatment restores strength and balance.

Anemia resulting from chronic disease is typically corrected by treating the underlying illness. This type of anemia rarely becomes severe. If it does, transfusions or hormone treatments to stimulate red blood cell production may be given.

Thalassemia minor is typically not treated. Thalassemia major may be treated with regular transfusions, surgical resection of the spleen to avoid its removal of RBCs from circulation, and sometimes iron chelation therapy. Symptoms are treated as they occur. Children or young adults with thalassemia major may require periodic **hospitalization** to receive blood transfusions or, in some cases, bone marrow transplants.

Sickle cell anemia will be monitored by regular eye examinations and diagnostic blood work. Immunizations for **pneumonia** and infectious diseases are part of treatment along with prompt treatment for sickle cell crises and infections of any kind. Psychotherapy or counseling may help older children deal with the emotional symptoms characteristic of this condition.

Children with aplastic anemia are especially susceptible to infection. Treatment for aplastic anemia may involve blood transfusions and bone marrow transplantation to replace malfunctioning cells with healthy ones.

Hemolytic anemia of the warm-antibody type may be treated with large doses of intravenous and oral corticocosteroids (cortisone). Individuals who do not respond to medical therapy, may undergo surgery to remove the spleen, which controls the anemia in some individuals by helping to add more RBCs to the circulation. Immune-system suppressants are prescribed when surgery is not successful. There is no specific treatment for cold-antibody hemolytic anemia.

Treatment of newborn anemia depends on the severity of symptoms, the level of Hgb, and the presence of any other diseases that may affect oxygen delivery, such as lung or heart disease or hyaline membrane disease. Transfusions may be given in certain situations or exchange transfusions if hemolytic disease of the newborn is not quickly resolved. The risk of transfusion (such as transfusion reactions, potential toxins, and infections such as HIV or hepatitis) are carefully weighed against the severity of the anemia in the infant.

Alternative treatment

Vitamin C is noted for helping to absorb iron and folate supplements. Cooking in a cast iron skillet may leach small amounts of absorbable iron into the diet. Folic acid can be readily absorbed from raw salad greens such as lettuce, spinach, arugula, alfalfa sprouts, and others. Blackstrap molasses is a good source of iron and B **vitamins**. Herbal supplements that will benefit individuals who have anemia include bilberry, dandelion, goldenseal, mullein, nettle, Oregon grape root, red raspberry, and yellow dock. Herbs are available as tinctures and teas or in capsules.

Nutritional concerns

The diet is a ready source of nutrients that prevent and treat anemia. Children with anemia can include more of these nutrients in their diet by eating a broad variety of whole grains, fruits and vegetables, beans, lean meat, poultry and fish, and supplementing the diet regularly with vitamins, **minerals**, and iron (as recommended). Pediatricians should be consulted before iron supplements are taken, however, because of the difficulty in absorbing non-food sources of iron. Vitamin C can stimulate iron absorption. Good food sources of iron

KEY TERMS

Erythropoiesis—The process through which new red blood cells are created; it begins in the bone marrow.

Hematocrit—A measure of the percentage of red blood cells in the total volume of blood in the human body.

Hemoglobin—An iron-containing pigment of red blood cells composed of four amino acid chains (alpha, beta, gamma, delta) that delivers oxygen from the lungs to the cells of the body and carries carbon dioxide from the cells to the lungs.

Hemolysis—The process of breaking down of red blood cells. As the cells are destroyed, hemoglobin, the component of red blood cells which carries the oxygen, is liberated.

Hypoxia—A condition characterized by insufficient oxygen in the cells of the body

Megaloblast—A large erythroblast (a red marrow cell that synthesizes hemoglobin).

Reticulocyte—An early, immature form of a red blood cell. Over time, the reticulocyte develops to become a mature, oxygen-carrying red blood cell.

include: almonds, broccoli, dried beans, raisins, dried apricots, seaweed (as soup stock), whole-grain breads and cereals, brown rice, lean red meat, liver, potatoes, poultry, and shellfish.

Because light and heat destroy folic acid, fruits and vegetables should be eaten raw or cooked as little as possible to help assimilation of folic acid. Folic acid can also be taken as a supplement.

Prognosis

Most anemias can be treated or managed. The prognosis for anemias generally depends upon the severity of the anemia, the type of anemia, and the response to treatment. The hereditary anemias, such as the thalassemias and sickle cell anemia, may require life-long treatment and monitoring whereas other types of anemia, once treated, are apt not to recur. Thalassemia major may cause deformities and may shorten life expectancy. Severe anemia may lead to other serious conditions, particularly if oxygen delivery is compromised for long periods of time or RBC destruction is more rapid than can be controlled by normal RBC replacement or specific treatment. Severe blood loss or prolonged anemia can result in life-threatening complications.

Prevention

Safety is the primary preventive measure for blood loss by injury. A wholesome, balanced diet rich in nutrients can help prevent dietary deficiencies that lead to anemia. Hereditary anemias cannot be prevented; parents can seek genetic testing and counseling if they are concerned about inherited anemias noted in their families or ethnic background.

Nutritional concerns

Sources of iron such as liver, red meat, whole grains, and poultry may help maintain hemoglobin levels and reduce the likelihood of deficiency-related anemias. Vitamin C is noted for helping to improve assimilation of iron taken as supplements.

Parental concerns

Parents may be particularly concerned about the possibility of inherited anemias. Genetic testing is available to address their doubts. **Nutrition** education is readily available from public health sources, books, and the reliable Internet sources for parents who are concerned about providing essential nutrients for children who may be susceptible to deficiency anemias. Regular physical examinations can help evaluate a child's overall health and reveal possible signs or symptoms of anemia.

Resources

BOOKS

"Blood Disorders." *The Merck Manual of Medical Information*, 2nd Home ed. Edited by Mark H. Beers et al. White House Station, NJ: Merck & Co., 2003.

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Ross, Allison J. *Everything You Need to Know about Anemia*. New York: Rosen Publishing Group, 2001.

Wick, M., et al. *Iron Metabolism, Anemias, Clinical Aspects and Laboratory*. New York: Springer, 2003.

ORGANIZATIONS

National Heart, Lung, and Blood Institute (NHLBI). 6701 Rockledge Drive, PO Box 30105, Bethesda, MD 20824–0105. Web site: <http://www.nhlbi.nih.gov>.

WEB SITES

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"Understanding Anemia: Your Life May Depend on It." *Anemia Lifeline*. Available online at <www.anemia.com> (accessed October 10, 2004).

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KEY TERMS

Ataxia—A condition marked by impaired muscular coordination, most frequently resulting from disorders in the brain or spinal cord.

Developmental delay—The failure of a child to meet certain developmental milestones, such as sitting, walking, and talking, at the average age. Developmental delay may indicate a problem in development of the central nervous system.

Angelman's syndrome

Definition

Angelman's syndrome is a relatively rare genetic disorder that causes a variety of neurological problems, including **developmental delay**, seizures, speech impairment, and problems with movement and balance.

Description

Angelman's syndrome was first described in 1965 by Harold Angelman, who noted that a group of children in his medical practice had flat heads, made jerky movements, held their tongues in a protruding way, and had curious bouts of laughter.

Demographics

Angelman's syndrome is relatively rare. As of the early 2000s there were only about 1,000 to 5,000 known cases of the syndrome in the United States. There is no predilection for either sex or for any particular ethnicity.

Causes and symptoms

Most cases of Angelman's syndrome can be traced to a genetic abnormality inherited from a maternal chromosome (15). A particular area of genes that should control the production and function of a protein called ubiquitin is either absent or ineffective. A minority of cases of Angelman's syndrome are due to new mutations in this same area of genes.

Children with Angelman's syndrome have an abnormally small, flat appearance to their skull. By one to two months of age, infants with the syndrome develop feeding difficulties. By six to 12 months, developmental delay is usually noted. Most children develop seizures by three years of age. Other characteristics of the syndrome include abnormally decreased muscle tone, fair skin and hair, protruding jaw, hyperactivity, episodes of uncontrollable laughter, difficulty sleeping, and severe problems with movement and balance. The disorder is sometimes called "happy puppet syndrome," because many children with the disorder have jerky, flapping movements of the arms; a stiff, jerky style of walking (gait); a happy, excited demeanor; and regular episodes of uncontrollable laughter.

Diagnosis

Diagnosis is made by noting the characteristic cluster of symptoms. Careful chromosomal study can reveal abnormalities on chromosome 15 that are consistent with those identified in Angelman's syndrome.

Treatment

As of 2004 there is no cure for Angelman's syndrome. Treatments attempt to ameliorate the symptoms in order to improve the quality of life. Treatments may include anti-seizure medications, physical and occupational therapy, and speech and language therapy.

Prognosis

Most children with Angelman syndrome are severely developmentally delayed. They never acquire normal speech, and they require care and supervision throughout their lives.

Prevention

There are no methods to prevent Angelman syndrome. However, if the disorder is known to run in a **family**, genetic counseling may help parents evaluate their level of risk for having a child with this disorder. Specialized testing of chromosome 15 will be required; the usual tests done during **amniocentesis** or chorionic villi sampling will not reveal the specific, small genetic flaw that causes Angelman syndrome.

Parental concerns

Caring for a child with Angelman syndrome constitutes a complex challenge. Parents should be encouraged to seek out parental and sibling support groups and respite care in order to help them face these challenges.

Resources

BOOKS

Hall, Judith G. "Chromosomal Clinical Abnormalities." In *Nelson Textbook of Pediatrics*. Edited by Richard E. Behrman, et al. Philadelphia: Saunders, 2004.

Jankovic, Joseph. "Movement Disorders." In *Textbook of Clinical Neurology*. Edited by Christopher G. Goetz. Philadelphia: Saunders, 2003.

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ORGANIZATIONS

American Academy of Pediatrics. 141 Northwest Point Blvd., Elk Grove Village, IL 60007-1098. Web site: <www.aap.org>.

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Animal bite infections

Definition

Animal bite infections develop in humans when an animal's teeth break the skin and introduce saliva containing disease organisms below the skin surface. The saliva of dogs, cats, ferrets, and rabbits is known to contain a wide variety of bacteria. According to one study, bacteria or other pathogens show up in about 85 percent of animal **bites**. These microorganisms may grow within the wound and cause an infection. The consequences of infection from these bites range from mild discomfort to life-threatening complications.

Description

Animal bites may occur in a variety of circumstances, ranging from unprovoked attacks in the wild by rabid or naturally aggressive animals to injuries inflicted by household pets who do not feel well, are frightened, are interrupted during their meal, or are annoyed by a child's teasing or overly rough play. The bite may be a simple warning to back off (as in most household cats), an assertion of dominance and control (as in many dogs), or an intention to seriously injure or kill (as in a few breeds of dogs and some wild animals). Animal bites can range from small injuries that barely break the skin to severe **wounds** that can cause a person to lose the use of a hand, eye, or foot or even bleed to death.

Demographics

The number of animal bites that occur in the United States each year is difficult to estimate because many of these injuries are treated successfully at home. Still, U.S. figures range from 1 million to 4.5 million animal bites each year. About 1 percent of these bites requires hospital inpatient treatment. Cat and dog bites result in 334,000 emergency room visits per year, which represents approximately 1 percent of all emergency hospital visits, at an annual cost of $100 million dollars in healthcare expenses and lost income. Children are the most frequent victims of dog bites, with five to nine year-old boys having the highest incidence. The average age of a dog bite victim is 13, whereas the average age of a cat bite victim is 19 or 20. Men are more often bitten by dogs than are women (3:1), whereas women are more often bitten by cats (3:1).

Children are more likely than adults to suffer dog bites on the face and neck, partly because they are shorter than adults. Cat bites in children as well as adults are far more likely to injure the hands or lower arms rather than other parts of the body.

Dog bites make up 80 to 85 percent of all reported animal bites in the United States and Canada. Cats account for about 10 percent of reported bites, and other animals (including rats, hamsters, ferrets, rabbits, horses, sheep, raccoons, bats, skunks, and monkeys) make up the remaining 5 to 10 percent. Cat bites, however, become infected more frequently than dog bites. A dog's mouth is rich in bacteria, but only 15 to 20 percent of dog bites become infected. In contrast, approximately

30 to 50 percent of cat bites become infected because a cat's teeth can penetrate more deeply than a dog's and carry bacteria deeper into a wound.

Figures on bite injuries from animals other than cats and dogs are difficult to obtain, although bites from pet hamsters and ferrets have been reported more frequently since the late 1990s. Rat bites are becoming more common, particularly in large cities where the rat population has been increasing in the early 2000s. Bites from such wild animals as mountain lions and bears are also reported more frequently as humans explore or move into their natural habitats.

Causes and symptoms

Causes

Many factors contribute to the risk of infection from an animal bite, including the type of wound inflicted, the location of the wound, pre-existing health conditions in the bitten person, the extent of delay before treatment, compliance with treatment, and the presence of a foreign body in the wound. Dogs usually inflict crush injuries because they have rounded teeth and strong jaws; thus, the bite of an adult dog can exert up to 200 pounds per square inch of pressure. This pressure usually results in a crushing injury, causing damage to such deep structures as bones, blood vessels, tendons, muscles, and nerves. The canine teeth are sharp and strong, often inflicting lacerations. Cats, with their needle-like incisors and carnassial teeth, typically cause puncture wounds. Puncture wounds appear innocuous on the surface, but the underlying injury goes deep. The teeth of a cat essentially inject bacteria deep within the bite, and the deep, narrow wound is difficult to clean. Persons with impaired immune systems—for example, individuals with HIV infection—are especially vulnerable to infection from cat bites.

The bacterial species most commonly found in animal bite wounds include *Pasteurella multocida*, *Staphylococcus aureus*, *Pseudomonas sp.*, and *Streptococcus sp.*, *P. multocida*, the root cause of pasteurellosis, is especially prominent in cat bite infections. Other infectious diseases from animal bites include **cat-scratch disease**, **tetanus**, and **rabies**.

Doctors are increasingly aware of the importance of checking animal bite wounds for anaerobic organisms, which are microbes that can live and multiply in the absence of air or oxygen. A study published in 2003 reported that about two-thirds of animal bite wounds contain anaerobes. These organisms can produce such complications as septic arthritis, tenosynovitis, **meningitis**, and infections of the lymphatic system.

With regard to the most common types of domestic pets, it is useful to note that biting and other **aggressive behavior** has different causes in dogs and cats. To some extent these differences are rooted in divergent evolutionary pathways, but they have also been influenced by human interference through selective breeding. Dogs were first domesticated by humans as early as 10,000 B.C. for hunting and as guard or attack dogs. Many species travel in packs or groups in the wild, and many human fatalities resulting from dog bites involve a large group of dogs attacking one or two persons. In addition, dogs typically relate to humans according to a hierarchical model of dominance and submission, and many of the techniques of dog training are intended to teach the dog to respect human authority. Certain breeds of dogs are much more likely to attack humans than others; those most often involved in fatal attacks are pit bulls, Rottweilers, German shepherds, huskies, and mastiffs. According to the Centers for Disease Control (CDC), there are between 15 and 20 fatal dog attacks on humans in the United States each year. There are several assessment or evaluation scales that veterinarians or animal trainers can use to score individual dogs and screen them for dominant or aggressive behavior.

Unlike dogs, cats were not domesticated until about 3000 B.C., and were important to ancient civilizations as rodent catchers and household companions rather than as protectors or hunters of wild game. Biologists classify cats as solitary predators rather than as pack or herd animals; as a result, cats do not relate to humans as authority figures in the same way that dogs do, and they do not form groups that attack humans when threatened or provoked. In addition, domestic cats have been selectively bred for appearance rather than for fierceness or aggression. Most cat bites are the result of **fear** on the cat's part (as when being placed in a carrier for a trip to the vet) or a phenomenon known as petting-induced aggression. Petting-induced aggression is a behavior in which a cat that has been apparently enjoying contact with a human suddenly turns on the human and bites. This behavior appears to be more common in cats that had no contact with humans during their first seven weeks of life. In other cats, this type of aggression appears to be related to a hypersensitive nervous system; petting or cuddling that was pleasurable to the cat for a few seconds or minutes becomes irritating, and the cat bites as a way of indicating that it has had enough. In older cats, petting-induced aggression is often a sign that the cat feels **pain** from touching or pressure on arthritic joints in its neck or back.

General signs of infection

The most common sign of infection from an animal bite is inflammation, which usually develops within

eight to 24 hours following the bite. The skin around the wound is red and feels warm, and the wound may ooze pus or a whitish discharge. Nearby lymph glands may be swollen, and there may be red streaks running up the arm or leg from the wound toward the center of the body. Complications can arise if the infection is not treated and spreads into deeper structures or into the bloodstream. Complications are more likely to develop if the bite is deep or occurs on the hand or at a joint.

Live disease-causing bacteria within the bloodstream and tissues may cause complications far from the wound site. Such complications include meningitis, brain abscesses, **pneumonia** and lung abscesses, and heart infections, among others. These complications can be fatal. Deep bites or bites near joints can damage joints and bones, causing inflammation of the bone and bone marrow or septic arthritis.

Cat-scratch fever

Cat-scratch disease is caused by *Bartonella henselae*, a bacterium that is carried in cat saliva; infection may be transmitted by a bite or scratch. Approximately 22,000 cases are reported each year in the United States; worldwide, nine out of every 100,000 individuals become infected. More than 80 percent of reported cases occur in persons under the age of 21. The disease is not normally severe in individuals with healthy immune systems. Symptoms may become serious, however, in immunocompromised individuals, such as those with acquired immune deficiency syndrome (**AIDS**) or those undergoing **chemotherapy**. Common symptoms include an inflamed sore in the area of the bite or scratch, swollen lymph nodes, **fever**, fatigue, and rash.

Rabies

Rabies is caused by a virus that is transmitted through the bite of an animal that is already infected. It is classified as a zoonosis, which is a term that refers to any disease of animals that can be transmitted to humans. More than 90 percent of animal rabies cases occur in such wild animals as skunks, bats, and raccoons, with such domestic animals as dogs and cats accounting for fewer than 10 percent of cases. The World Health Organization (WHO) estimates that between 35,000 and 50,000 individuals worldwide die each year as a result of rabies. The highest incidence of rabies occurs in Asia where, in 1997, over 33,000 deaths were noted, most occurring in India. Rabies is in the early 2000s rare in the United States, as a result of good animal control practices. Onset is delayed, usually weeks to months after the person has been bitten. Early symptoms of rabies include fever, **headache**, and flu-like symptoms. These conditions progress to **anxiety**, hallucinations, **muscle spasms**, partial paralysis, fear of water (hydrophobia), and other neurological symptoms as the virus spreads to the central nervous system. Medical treatment must be sought soon after exposure because death invariably follows once the infection becomes established.

Most deaths from rabies in the United States in the late twentieth and early twenty-first centuries have resulted from bat rather than dog bites; one victim was a man in Iowa who died in September 2002.

When to call the doctor

Minor animal bites and scratches (those that just break the surface of the skin) can be treated satisfactorily at home. The American Academy of Family Practice (AAFP) recommends the following steps:

- Wash the bite with soap and water, and rinse well.
- Apply pressure with a clean towel or cloth to stop the bleeding.
- Cover the wound with a sterile dressing or bandage.
- Hold the injured area above the level of the heart to lower the risk of tissue swelling and infection.
- Apply antibiotic ointment to the bite twice a day until it heals.
- Report the incident to the local animal control authority or police.

Parents should, however, take their child to a doctor or the emergency room in any of the following circumstances:

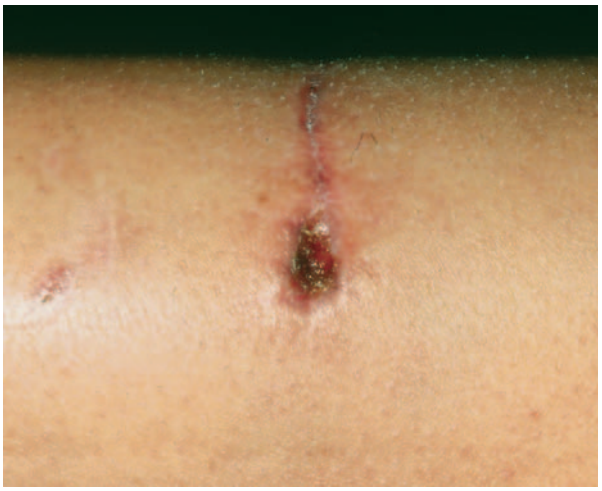
- The bite was inflicted by a cat.
- The child was bitten by a dog on the hand, foot, head, or neck.
- The wound is deep or gaping.
- The child has an **immunodeficiency** disease, **cancer**, diabetes, a kidney or liver disorder, or any other condition that affects the body's ability to fight off infection.
- The wound already shows signs of infection.
- The bleeding does not stop after 15 minutes of pressure.
- The child has a broken bone, nerve damage, injury to the skull, or other serious injury in addition to the bite.
- The child has not had a tetanus shot or booster within the past five years.

Diagnosis

Most animal bites that cannot be treated at home are examined by a doctor in the emergency room of a

hospital. The medical examination involves taking the history of the injury and assessing the type of wound and damage. The child's record of tetanus immunization and general health status are checked. An x ray may be ordered to assess bone damage and to check for **foreign objects** in the wound. Wound cultures are done for infected bites if the victim is at high risk for complications or if the infection does not respond to treatment. If the child was bitten severely on the head, the emergency room doctor will call in a neurologist for consultation, particularly if the eyes, ears, or neck were injured or the skull was penetrated. The doctor may also consult a plastic surgeon if the bites are extensive, if large pieces of tissue have been lost, or if the functioning or appearance of the affected part of the body is likely to be affected.

Evaluation of possible exposure to rabies is also important. A biting animal suspected of having rabies is usually caught and restrained, tested, and observed for a period of time for evidence of pre-existing infection.



View of an infected animal bite. *(Photograph by Dr. P. Marazzi. Science Photo Library/Custom Medical Stock Photo, Inc.)*

Treatment

Treatment depends on the wound type, its site, and risk factors for infection. All wounds from animal bites are cleaned and disinfected as thoroughly as possible. The doctor begins by injecting a local anesthetic in order to examine the wound thoroughly without causing additional pain to the child. The next step is to remove dead tissue, foreign matter, and blood clots, all of which can become sources of infection. This removal is called debridement. After debriding the wound, the doctor will cut away the edges of the tissue, as clean edges heal faster and are less likely to form scar tissue. The doctor then irrigates, or flushes, the wound with saline solution forced through a syringe under pressure. Irrigation is highly effective in lowering the risk of infection from animal bites; in one study of 45 children with a total of 145 dog bite wounds on the face, pressure irrigation combined with trimming the edges of the wounds resulted in an infection rate of 0.4 percent.

Bites to the head and face usually receive sutures, as do severe lacerations elsewhere. Puncture wounds are left open. If an abscess forms, the physician may perform an incision in order to drain the abscess.

If infection does occur, the doctor will prescribe antibiotic medications. **Antibiotics** may also be used for infection prevention. Since a single bite wound may contain many different types of bacteria, no single antibiotic is always effective. Commonly prescribed antibiotics are penicillin or a combination of amoxicillin and clavulanate potassium (Augmentin). Aztreonam (Azactam) has been reported to be effective in treating infections caused by *P. multocida*. In most cases, antibiotics taken by mouth are sufficient; however, some deep bites may require treatment with intravenous antibiotics.

Because rabies is caused by a virus, antibiotics are not effective against it. In addition, there is no known cure for the disease as of 2004 once symptoms become apparent. People with a high risk of contracting the disease should receive preexposure **vaccination**. Individuals bitten by an unknown or potentially rabid animal should receive postexposure vaccination, also called postexposure prophylaxis (PEP). The PEP regimen consists of one dose of vaccine given at the initial visit as well as one dose of human immune globulin. Additional doses of vaccine are given on days three, seven, 14, and 28.

Prognosis

Once a bacterial infection is halted, the bite victim usually recovers fully. There is no known cure for rabies once symptoms become evident and death is almost certain. WHO reports that 114 rabies deaths occurred in the Americas in 1997, with only four deaths occurring that year in the United States, thus emphasizing the importance of good animal control practice and postexposure prophylaxis.

The prognosis for restoring the function or appearance of a hand or other body part following a severe bite depends on the location of the bite, the promptness of treatment, and the availability of specialized surgical repair.

Prevention

Preventing bites obviously prevents subsequent infections. With regard to domestic pets, parents should inform themselves about the aggression level and other characteristics of a particular breed before bringing a purebred pet dog into the family and consider having a specific dog evaluated by a veterinarian or animal behaviorist before adopting it. In addition, parents should make sure that the dog has been neutered or spayed, since intact dogs of either sex are more likely to bite than those that have been altered. Cat bites can often be prevented by learning about a cat's body language and recognizing the signs of petting-induced aggression. These include dilating pupils, a low growl, stiffening of the body, twitching of the tail, and flattening the ears backward against the head.

Children under 12 years of age are at a higher risk for bites due to their small size and their inexperience with animals; therefore, they should be supervised with animals and taught to act appropriately around them. In particular, children should be taught not to tease a dog by pulling its fur or tail; to leave a dog alone while it is eating; and to avoid running or screaming in the presence of a dog, as the animal is more likely to chase a moving object. Direct eye contact with a threatening dog should be avoided, as the dog may interpret that as aggression. It is best to stand still if at all possible, with feet together and arms against the chest; most dogs will lose interest in an object that is not moving and will eventually go away.

A wild animal that is unusually aggressive or behaving strangely (e.g. a raccoon or bat that is active during the daytime or is physically uncoordinated) should be avoided and reported to the local animal control authorities; it may be infected with the rabies virus. Wild animals should not be taken in as pets, and garbage or pet food that might attract wild animals should not be left outside the home or campsite. People should also avoid trying to break up fights between animals and should as a rule approach unknown cats and dogs very cautiously, especially on their territory. Finally, animals should not be trained to fight.

Domestic pets should be vaccinated against rabies; people should consult a veterinarian for advice about the frequency of booster vaccinations for the area in which they live. In addition, families planning to travel to countries where rabies is endemic should consider vaccination before leaving the United States. The AAFP frequently posts updated travel advisories for rabies immunizations.

KEY TERMS

Anaerobic—An organism that grows and thrives in an oxygen-free environment.

Canines—The two sharp teeth located next to the front incisor teeth in mammals that are used to grip and tear. Also called cuspids.

Carnassials—The last upper premolar teeth in the mouths of cats and other carnivores, adapted to shear or puncture food. Carnassial teeth often cause puncture wounds when a cat bites a human.

Culture—A test in which a sample of body fluid is placed on materials specially formulated to grow microorganisms. A culture is used to learn what type of bacterium is causing infection.

Debridement—The surgical removal of dead tissue and/or foreign bodies from a wound or cut.

Irrigation—Cleansing a wound with large amounts of water and/or an antiseptic solution. Also refers to the technique of removing wax (cerumen) from the ear canal by flushing it with water.

Microorganism—An organism that is too small to be seen with the naked eye, such as a bacterium, virus, or fungus.

Pasteurellosis—A bacterial wound infection caused by *Pasteurella multocida*. Pasteurellosis is characterized by inflammation around the wound site and may be accompanied by bacteria in the bloodstream and infection in tissues and organs.

Pathogen—Any disease-producing microorganism.

Postexposure prophylaxis—Any treatment given after exposure to a disease to try to prevent the disease from occurring. In the case of rabies, postexposure prophylaxis involves a series of vaccines given to an individual who has been bitten by an unknown animal or one that is potentially infected with the rabies virus.

Tenosynovitis—Inflammation of a tendon and its enveloping sheath, usually resulting from overuse injury.

Zoonosis—Any disease of animals that can be transmitted to humans. Rabies is an example of a zoonosis.

Parental concerns

Given prompt treatment, most animal bites are not cause for major concern; as has been mentioned, minor bites can be treated at home without a visit to the doctor.

Some children bitten by large dogs, however, may become extremely fearful of dogs in general and may require counseling, particularly if the bite was severe or had long-term effects on the child's health. Fear of cats following a bite is much less common. Parents may also need to talk with or comfort their child if it is necessary to give up a family pet that cannot be retrained. In many cases, however, a qualified animal behaviorist can assess the reasons for a pet's biting or other aggressive behavior and suggest appropriate treatments.

See also Cat-scratch disease; Human bite infections; Rabies.

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American Veterinary Medical Association (AVMA). 1931 North Meacham Road, Suite 100, Schaumburg, IL 60173–4360. Web site: <www.avma.org>.

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Anorexia nervosa

Definition

Anorexia nervosa is an eating disorder characterized by self-starvation, unrealistic **fear** of weight gain, and conspicuous distortion of body image.

Description

The term anorexia nervosa comes from two Latin words that mean "nervous inability to eat." Anorexics have the following characteristics in common:

- inability to maintain weight at or above what is normally expected for age or height
- intense fear of becoming fat
- distorted body image
- in females who have begun to menstruate, the absence of at least three menstrual periods in a row, a condition called **amenorrhea**

There are two subtypes of anorexia nervosa: a restricting type, characterized by strict dieting and **exercise** without binge eating; and a binge-eating/purging type, marked by episodes of compulsive eating with or without self-induced **vomiting** and/or the use of **laxatives** or enemas. A binge is defined as a time-limited (usually under two hours) episode of compulsive eating in which the individual consumes a significantly larger amount of food than most people would eat in similar circumstances.

Demographics

Anorexia nervosa was not officially classified as a psychiatric disorder until the third edition of *Diagnostic and Statistical Manual of Mental Disorders (DSM)* in 1980. It is, however, a growing problem in the early 2000s among adolescent females. Its incidence in the United States has doubled since 1970. The rise in the number of reported cases reflects a genuine increase in the number of persons affected by the disorder and not simply earlier or more accurate diagnosis. Estimates of the incidence of anorexia range between 0.5 percent and 1 percent of Caucasian female adolescents. Over 90 percent of patients diagnosed with the disorder as of 2001 are female. The peak age range for onset of the disorder is 14 to 18 years. In the 1970s and 1980s, anorexia was regarded as a disorder of upper- and middle-class women, but that generalization is as of 2004 also changing. Studies indicate that anorexia is increasingly common among females of all races and social classes in the United States.

Causes and symptoms

While the precise cause of the disease is not known, anorexia is a disorder that results from the interaction of cultural and interpersonal as well as biological factors.

Social influences

The rising incidence of anorexia is thought to reflect the present idealization of thinness as a badge of upper-class status as well as of female beauty. In addition, the increase in cases of anorexia includes "copycat" behavior, with some patients developing the disorder from imitating other girls.

The onset of anorexia in **adolescence** is attributed to a developmental crisis caused by girls' changing bodies coupled with society's overemphasis on female appearance. The increasing influence of the mass media in spreading and reinforcing gender stereotypes has also been noted.

Occupational goals

The risk of developing anorexia is higher among adolescents preparing for careers that require attention to weight and/or appearance. These high-risk groups include dancers, fashion models, professional athletes (including gymnasts, skaters, long-distance runners, and jockeys), and actresses.

Genetic and biological influences

Girls whose biological mothers or sisters have or have had anorexia nervosa appear to be at increased risk of developing the disorder.

Psychological factors

A number of theories have been advanced to explain the psychological aspects of the disorder. No single explanation covers all cases. Anorexia nervosa has been given the following interpretations:

- Overemphasis on control, autonomy, and independence: Some anorexics come from achievement-oriented families that stress physical fitness and dieting. Many anorexics are perfectionists who are driven about schoolwork and other matters in addition to weight control.
- Evidence of **family** dysfunction: In some families, a daughter's eating disorder serves as a distraction from marital discord or other family tensions.

- A rejection of female sexual maturity: This rejection is variously interpreted as a desire to remain a child or as a desire to resemble males.
- A reaction to sexual abuse or assault.
- A desire to appear as fragile and nonthreatening as possible: This hypothesis reflects the idea that female passivity and weakness are attractive to males.
- Inability to interpret the body's hunger signals accurately due to early experiences of inappropriate feeding.

Male anorexics

Although anorexia nervosa largely affects females, its incidence in the male population is rising in the early 2000s. Less is known about the causes of anorexia in males, but some risk factors are the same as for females. These include certain occupational goals and increasing media emphasis on external appearance in men. Homosexual males are under pressure to conform to an ideal body weight that is about 20 pounds lighter than the standard attractive weight for heterosexual males.

When to call the doctor

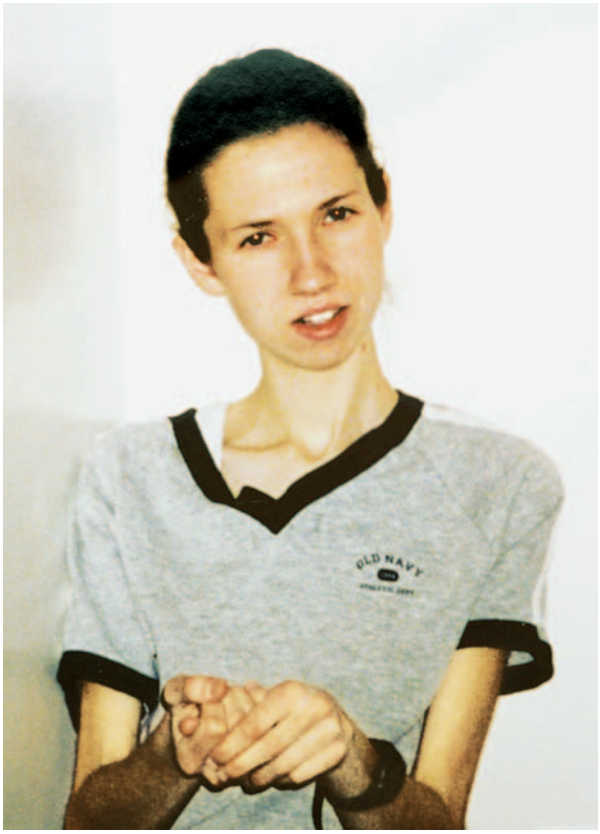
A healthcare professional should be contacted if a child or adolescent is suspected of having anorexia nervosa or displays early signs of the disorder, such as the following:

- fear of gaining weight
- distorted body image
- recent weight loss
- restrictive or abnormal eating patterns such as skipping meals or eliminating once-liked foods
- preoccupation with food and dieting
- compulsive exercising
- purging behaviors such as vomiting or using laxatives
- withdrawal from friends and family
- wearing baggy clothes to hide weight loss

Diagnosis

Diagnosis of anorexia nervosa is complicated by a number of factors. One is that the disorder varies somewhat in severity from patient to patient. A second factor is denial, which is regarded as an early sign of the disorder. Many anorexics deny that they are ill and are usually brought to treatment by a family member.

Anorexia nervosa is a serious public health problem not only because of its rising incidence, but also because it has one of the highest mortality rates of any psychiatric disorder. Moreover, the disorder may cause serious long-term health complications, including congestive heart failure, sudden death, growth retardation, dental problems, **constipation**, stomach rupture, swelling of the salivary glands, anemia and other abnormalities of the blood, loss of kidney function, and osteoporosis.



Extreme weight loss in an anorexic adolescent. *(© Ed Quinn/ Corbis.)*

Most anorexics are diagnosed by pediatricians or family practitioners. Anorexics develop emaciated bodies, dry or yellowish skin, and abnormally low blood pressure. There is usually a history of amenorrhea in female patients, and sometimes of abdominal **pain**, constipation, or lack of energy. The patient may feel chilly or have developed lanugo, a growth of downy body hair. If the patient has been self-inducing vomiting, she may have eroded tooth enamel or Russell's sign (scars on the back of the hand). The second step in diagnosis is measurement of the patient's weight loss. *DSM-IV* specifies a weight loss leading to a body weight 15 percent below normal, with some allowance for body build and weight history.

Anorexia nervosa

Criteria

1. Refusal to maintain body weight at or above a minimally normal weight for age and height. Body weight is less than 85 percent of what is expected.
2. Intense fear of gaining weight or becoming fat, even though patient is underweight.
3. Undue influence of body weight or shape on self-evaluation, or denial of the seriousness of current underweight condition.
4. Absence of at least three consecutive menstrual cycles in previously menstruating females.

Restricting type: No regular episodes of binge-eating or purging (self-induced vomiting or misuse of laxatives, diuretics, or enemas).

Binge-eating/purging type: Regular episodes of binge-eating or purging (self-induced vomiting or misuse of laxatives, diuretics, or enemas).

SOURCE: *Diagnostic and Statistical Manual of Mental Disorders IV*.

(Table by GGS Information Services.)

The doctor will need to rule out other physical conditions that can cause weight loss or vomiting after eating, including metabolic disorders, brain tumors (especially hypothalamus and pituitary gland lesions), diseases of the digestive tract, and a condition called superior mesenteric artery syndrome. Persons with this condition sometimes vomit after meals because the blood supply to the intestine is blocked. The doctor will usually order blood tests, an electrocardiogram, urinalysis, and bone densitometry (bone density test) in order to exclude other diseases and to assess the patient's nutritional status.

The doctor will also need to distinguish between anorexia and other psychiatric disorders, including depression, **schizophrenia**, social phobia, **obsessive-compulsive disorder**, and body dysmorphic disorder. Two diagnostic tests that are often used are the Eating Attitudes Test (EAT) and the Eating Disorder Inventory (EDI).

Treatment

Treatment of anorexia nervosa includes both short- and long-term measures and requires **assessment** by dietitians and psychiatrists as well as medical specialists. Therapy is often complicated by the patient's resistance or failure to carry out a treatment plan.

Hospital treatment

Hospitalization is recommended for anorexics with any of the following characteristics:

- weight of 40 percent or more below normal or weight loss over a three-month period of more than 30 lbs (13.6 kg)
- severely disturbed metabolism
- severe binging and purging
- signs of psychosis
- severe depression or risk of **suicide**
- family in crisis

Hospital treatment includes individual and group therapy as well as refeeding and monitoring of the patient's physical condition. Treatment usually requires two to four months in the hospital. In extreme cases, hospitalized patients may be force-fed through a tube inserted in the nose (nasogastric tube) or into a vein (hyperalimentation).

Outpatient treatment

Anorexics who are not severely malnourished can be treated by outpatient psychotherapy. The types of treatment recommended are supportive rather than insight-oriented and include behavioral approaches as well as individual or group therapy. **Family therapy** is often recommended when the patient's eating disorder is closely tied to family dysfunction. Self-help groups are often useful in helping anorexics find social support and encouragement. Psychotherapy with anorexics is a slow and difficult process; about 50 percent of patients continue to have serious psychiatric problems after their weight has stabilized.

Medications

Anorexics have been treated with a variety of medications, including **antidepressants**, antianxiety drugs, selective serotonin reuptake inhibitors, and lithium carbonate. The effectiveness of medications in treatment regimens is as of 2004 debated. However, at least one study of fluoxetine (Prozac) showed it helped the patient maintain weight gained while in the hospital.

Nutritional concerns

A key focus of treatment for anorexia nervosa is teaching the principles of healthy eating and improving disordered eating behaviors. A dietician or nutritionist plays an important role in forming a **nutrition** plan for the patient; such plans are individualized and ensure that the patient is consuming enough food to gain or maintain weight as needed and stabilize medically. The anorexic's weight and food intake are closely monitored to ensure that the plan is being followed.

KEY TERMS

Amenorrhea—The absence or abnormal stoppage of menstrual periods.

Binge—A pattern of eating marked by episodes of rapid consumption of large amounts of food; usually food that is high in calories.

Body dysmorphic disorder—A psychiatric disorder marked by preoccupation with an imagined physical defect.

Hyperalimentation—A method of refeeding anorexics by infusing liquid nutrients and electrolytes directly into central veins through a catheter.

Lanugo—A soft, downy body hair that covers a normal fetus beginning in the fifth month and usually shed by the ninth month. Also refers to the fine, soft hair that develops on the chest and arms of anorexic women. Also called vellus hair.

Purging—The use of vomiting, diuretics, or laxatives to clear the stomach and intestines after a binge.

Russell's sign—A scraped or raw area on the patient's knuckles, caused by self-induced vomiting.

Superior mesenteric artery syndrome—A condition in which a person vomits after meals due to blockage of the blood supply to the intestine.

Prognosis

Figures for long-term recovery vary from study to study, but reliable estimates are that 40 to 60 percent of anorexics make a good physical and social recovery, and 75 percent gain weight. The long-term mortality rate for anorexia is estimated at around 10 percent, although some studies give a lower figure of 3 to 4 percent. The most frequent causes of death associated with anorexia are starvation, electrolyte imbalance, heart failure, and suicide.

Prevention

Short of major long-term changes in the larger society, the best strategy for prevention of anorexia is the cultivation of healthy attitudes toward food, weight control, and beauty (or body image) within families. Early treatment such as counseling may help to prevent early signs of disordered eating from progressing into more serious behaviors.

Parental concerns

There are many strategies that parents can undertake to help encourage healthy attitudes toward weight, food, and exercise in their children. These include the following:

- teaching children the importance of healthy eating and exercise
- avoiding using food as a punishment or reward
- instilling healthy eating and exercise habits by example
- being a good role model by promoting healthy body image and encouraging children and adolescents to find role models in the media who do the same
- encouraging children or teens who wish to diet to talk to a healthcare professional about healthy strategies to lose weight

See also Binge eating disorder; Bulimia nervosa.

Resources

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American Anorexia/Bulimia Association. 418 East 76th St., New York, NY 10021. Telephone: 212/734–1114.

National Association of Anorexia Nervosa and Associated Disorders. Web site: <www.anad.org>.

National Institute of Mental Health Eating Disorders Program. Building 10, Room 3S231. 9000 Rockville Pike, Bethesda, MD 20892. Telephone: 301/496–1891.

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Antenatal testing

Definition

Antenatal testing describes procedures performed during pregnancy to detect health problems in the growing fetus; establish characteristics such as fetal age, sex, or weight; or diagnose any material conditions that may affect fetal development.

Purpose

Antenatal tests and exams are important tools for protecting the health of a pregnant woman and her developing child. Various tests are administered over the course of pregnancy to determine if the mother has any health conditions that may interfere with normal development of the fetus or if the fetus has any health conditions that may affect the baby's quality of life. Often, families will use information provided by the tests to prepare for the baby's birth and make arrangements for special care if needed or make the decision to terminate the pregnancy. Physicians also use antenatal tests to determine various characteristics of the fetus, such as gestational age, size, and position in the uterus, or to verify the presence of multiple fetuses.

Description

Women who become pregnant may undergo tests at any stage in their pregnancy: during the first trimester (weeks one through 12), second trimester (weeks 13–26), or third trimester (weeks 27–40+). What tests are ordered depends on the stage of pregnancy, the age and health of the mother, the medical history of both parents, and the family's background or ethnicity. There are two distinct types of antenatal tests:

- Screening tests tend to be less invasive and indicate the possibility of a certain genetic disorder or birth defect but do not determine with certainty that the abnormality exists.
- Diagnostic tests tend to more invasive but are able to determine with more certainty that a fetus will be born with a certain condition.

Blood and urine tests

In the early stages of pregnancy, physicians may order blood or urine tests to screen for possible disorders or infections that could affect the growing fetus. The tests may also be ordered in later stages if the pregnant woman comes in contact with an infectious agent or develops symptoms of infection. In many cases, complications can be avoided if early diagnosis is made and treatment initiated. Examples of conditions that are commonly screened for with blood and/or urine tests include:

- Rh factor: About 15 percent of people lack a certain blood protein called Rh factor and are called Rh negative. Complications may arise if an Rh-negative mother is carrying an Rh-positive child. These can be avoided if the mother is given a substance called Rh immune globulin (RhIg) at approximately 28 weeks into the pregnancy and again within 72 hours after the baby is born.
- Anemia: If there is too little of a substance called hemoglobin in a pregnant woman's red blood cells, a condition called anemia may result. If a blood test reveals low hemoglobin, supplementation with iron may be recommended.
- Human **immunodeficiency** virus: HIV can be transmitted from mother to child, although treatment of the disease during pregnancy can greatly reduce the risk of transmission. Because it is possible for individuals to be infected without exhibiting symptoms and because unprotected sex is a major risk factor for contracting the virus, the American Academy of Pediatrics recommended in 1999 that a screening test for HIV be routinely offered. A second test later in pregnancy may be recommended if the pregnant woman is considered to be at high risk of becoming infected with HIV.

- Syphilis: If transmitted from mother to child during pregnancy, syphilis leads to death of the fetus or newborn in approximately 40 percent of cases. The goal of syphilis screening is to diagnose and treat infections before transmission occurs. Syphilis is treated with **antibiotics**.
- **Rubella** (German **measles**): Although the majority of women in the United States are immune to rubella because of prior immunization or infection, serious complications to the fetus (such as deafness, blindness, or heart defects) can arise if a woman becomes infected during pregnancy. If a woman is found to not have immunity, it will be recommended that she avoid contact with infected individuals during her pregnancy and receive a **vaccination** against rubella after she gives birth.
- Group beta strep (GBS): GBS is a type of bacteria commonly found in the vagina and rectum. GBS can be present in a person's body without causing any symptoms, so many women do not realize they are infected with it. Newborns who are exposed to GBS, however, can develop serious complications such as **meningitis**, **pneumonia**, blindness, deafness, and death is possible. Doctors test for the presence of GBS in urine or in samples collected from the vagina or rectum. This test is usually performed late in pregnancy, at 35 to 37 weeks of gestation. If a woman is found to be infected with GBS, physicians generally administer antibiotics to the mother so the baby is not born with the infection.

A multiple marker test or triple screen is used to determine if a fetus is at an increased risk of having certain congenital abnormalities. The test has a high rate of false-positives; as few as 10 percent of women with abnormal results go on to have babies with congenital defects. The purpose of the test is to determine if further testing (such as ultrasound or **amniocentesis**) is warranted. The test requires a sample of maternal blood, typically taken during the fifteenth and twentieth week of pregnancy, and measures the level of certain pregnancy hormones.

Ultrasound

Ultrasound is a device that records sound waves as they bounce off the developing fetus and create an image that is projected onto a large computer screen. A physician may order an ultrasound scan to listen for a fetal heartbeat, determine a woman's precise due date, or check for **twins**, among other uses. Also known as a sonogram, the procedure takes only a few minutes, is safe and painless, and usually is covered by health insurance.

During the procedure, an ultrasound technician asks the pregnant woman to remove her clothes and change into a gown. The technician may rub some gel on the woman's fundus (lower abdomen), which helps the hand-held device pick up sound waves. In certain cases, the technician may insert a plastic probe into the woman's vaginal canal to get a clearer picture of the fetus. Early in pregnancy, the test may need to be done with a full bladder.

Pregnant women will often have their first ultrasound between eight and 12 weeks of gestation. In normal cases, the technician is able to identify a fetal heartbeat, which appears as a flashing light on the screen. Closer to the due date, physicians use ultrasound to make sure the fetus is in the correct head-first position to exit the birth canal, to assess the fetus for certain birth defects, and to determine the sex of the fetus if the parents desire.

Between ten and 14 weeks of gestation, ultrasonography may be used to measure a small collection of fluid beneath the skin at the back of the neck. Called nuchal translucency, the measurement tends to be larger in fetuses with genetic abnormalities such as **Down syndrome**, trisomy 13, trisomy 18, **Turner syndrome**, and triploidy. A particular neck measurement combined with maternal age as an indicator (e.g. the incidence of the disorder increases in proportion to the age of the mother) has been shown to correctly diagnose Down syndrome in 75 to 80 percent of cases; this number increases to 90 percent if the procedure is combined with the multiple marker test.

Amniocentesis

Amniocentesis is a more invasive test that carries a higher risk of complications than blood tests or ultrasonography, but is able to determine more precisely the presence of certain birth defects. It is also used to determine the level of maturity of the baby's lungs, of particular interest if the baby will be delivered prematurely. During amniocentesis, a doctor inserts a thin needle through a woman's abdomen and into the uterus. Using ultrasound as a guide, the doctor uses the needle to withdraw a sample of fluid from the amniotic sac. Afterward, tiny cells shed by the fetus can be studied in the laboratory; scientists can analyze the samples to determine if the fetus has certain genetic conditions. Amniocentesis is typically performed during the second trimester of pregnancy and particularly in mothers over the age of 40.

Chorionic villus sampling

Chorionic villus sampling (CVS) is a procedure that allows for prenatal diagnosis during the first trimester (generally between ten and 12 weeks of gestation, during the embryonic stage of development). The test involves

KEY TERMS

Alpha fetoprotein test—A screening blood test that can be done after the sixteenth week of pregnancy to evaluate the possibility of spina bifida and other neural tube defects in the fetus.

Amniocentesis—A procedure performed at 16–18 weeks of pregnancy in which a needle is inserted through a woman's abdomen into her uterus to draw out a small sample of the amniotic fluid from around the baby for analysis. Either the fluid itself or cells from the fluid can be used for a variety of tests to obtain information about genetic disorders and other medical conditions in the fetus.

Breech position—The foot-down or bottom-down position of a fetus just before delivery.

Group B streptococcus—A serotype of streptococcus, *Streptococcus agalactiae*, which is beta hemolytic and can cause neonatal sepsis, pneumonia, or meningitis if present in the birth canal at the time of delivery especially when the delivery is difficult.

Hemoglobin—An iron-containing pigment of red blood cells composed of four amino acid chains (alpha, beta, gamma, delta) that delivers oxygen from the lungs to the cells of the body and carries carbon dioxide from the cells to the lungs.

Meningitis—An infection or inflammation of the membranes that cover the brain and spinal cord. It is usually caused by bacteria or a virus.

Ultrasonography—A medical test in which sound waves are directed against internal structures in the body. As sound waves bounce off the internal structure, they create an image on a video screen. Ultrasonography is often used to diagnose fetal abnormalities, gallstones, heart defects, and tumors. Also called ultrasound imaging.

taking a small sample of cells from the placenta with a needle through the abdominal wall or a small tube (catheter) through the cervix. The procedure is guided by ultrasound. The cells are then analyzed in a laboratory for the presence of genetic abnormalities.

Risks

The risks associated with antenatal testing depend on the specific procedure being performed:

- Blood tests: The risks associated with blood testing are minimal and include discomfort, bleeding from the puncture site, hematoma (collection of blood under the skin), and infection.
- Ultrasound: As of 2004 there is no recognized risk for ultrasonography.
- Amniocentesis: The risk of miscarriage associated with amniocenteses performed between 15 and 20 weeks is less than 0.5 percent. The other risks are maternal infection, injuries to the fetus, and premature labor.
- Chorionic villus sampling: The risk of miscarriage associated with CVS performed between nine and 11 weeks is less than 2 percent. The risks are similar to those associated with amniocentesis.

Parental concerns

Impending parenthood is often accompanied by concerns that the baby may be born with defects or other health problems that would affect the quality of life of the baby and the **family**. That worry is often weighed against the **anxiety** associated with antenatal tests and their risks. Parents should be encouraged to educate themselves on the benefits and risks associated with the various antenatal screening procedures and work with their obstetrician to formulate an individualized prenatal care plan.

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Antepartum testing

Definition

Antepartum testing involves the use of **electronic fetal monitoring** (EFM) or ultrasound (US) to assess fetal well-being as determined by the fetal heart rate (FHR) and other characteristics during the antepartal period, which is the period spanning from conception to labor. Antepartum tests include the nonstress test (NST), modified biophysical profile (MBPP), contraction stress test (CST), oxytocin contraction stress test (OCT), biophysical profile (BPP), doppler flow studies, **amniocentesis**, cordocentesis, and fetal echocardiography.

Purpose

Antepartum testing can start as early as 24 weeks but usually begins after 32 weeks of pregnancy depending on the mother's physical status. It provides a means for the physician and pregnant woman to identify any problems and be alert to any changes that may necessitate additional testing or interventions. The testing results reflect how well the placenta is functioning in its ability to adequately supply blood and, therefore, oxygen to the fetus.

The testing is done for pregnancies at risk for maternal and/or fetal complications. Some of these risks include:

- any chronic illness in the mother, such as high blood pressure, diabetes, or autoimmune diseases, including systemic lupus erythematosus (SLE)
- problems with previous pregnancies, such as a history of unexplained stillbirth
- fetal complications, such as intrauterine growth restriction (IUGR) (growth in the fetus below the tenth percentile), birth defects, **twins**, or other multiple gestations in which a growth discrepancy occurs, such as twin-to-twin transfusion syndrome
- problems in current pregnancy, including pregnancy-induced **hypertension** (frequently referred to as pre-eclampsia); gestational diabetes (diabetes caused by pregnancy) requiring the use of insulin; premature rupture of the membranes (PROM); too little or too much amniotic fluid (the liquid surrounding the fetus) called oligohydramnios and polyhydramnios, respectively; vaginal bleeding; placental abnormalities, i.e., partial abruption (a portion of the placenta pulls away from the wall of the uterus), or placenta previa (a condition in which the placenta is covering the cervix instead of near the top of the uterus)

Antepartum testing is also used in low-risk pregnancies to evaluate decreased fetal activity, a lag in fundal height (as measured from top of the pubic area to the highest point in the midline at the top of the uterus), and postdates or post-term pregnancy. A normal pregnancy is 40 weeks and testing should begin at 41 weeks to assess the status of the placenta, which may no longer be capable of meeting the baby's needs. This can be indicated by the FHR pattern, amniotic fluid status, and fetal movement patterns.

Description

The spectrum of fetal **assessment** includes fetal movement (FM) counting, nonstress test (NST), modified biophysical profile (MBPP), contraction stress test (CST), oxytocin contraction stress test (OCT), biophysical profile (BPP), doppler flow studies, amniocentesis, cordocentesis, and fetal echocardiography. Fetal movement should be determined on a daily basis by all pregnant women regardless of risk status. The woman should be instructed to monitor fetal movement by selecting a consistent time of day to document how long it takes to feel 10 fetal movements. She should call her healthcare provider if there are fewer than 10 movements in a 10-hour period or immediately if there are no movements in any 10-hour period. She should also be instructed to report significant decreases in fetal activity from the baby's normal pattern. This daily monitoring of FM by all pregnant women is the least expensive and easiest of all antepartum tests to perform.

Non-stress test (NST)

The NST is performed with an electronic fetal monitor (EFM) that traces the fetal heart rate (FHR) and the presence of any contractions on a monitor strip. The mother reclines with a slight pelvic tilt to prevent compression of the large blood vessels by the pregnant uterus. The EFM is applied to her abdomen by two straps: one is to listen to the FHR by means of a transducer, and the other is to pick up any contractions by pres-

sure on a tocodynameter. The NST indirectly provides information about fetal status by the observation of FHR accelerations that occur with fetal movement. If a fetus is not receiving adequate oxygen from the placenta, the FHR will not accelerate, but if the oxygen supply is sufficient, accelerations will be noted. If it is difficult to obtain fetal movements, a vibroacoustic stimulator (VAS) is sometimes used to provide a loud noise to awaken the fetus and produce the desired results. The minimum amount of time required for an NST is 20 minutes. During those 20 minutes, there must be two accelerations in the FHR that are 15 beats above the baseline FHR and last for 15 seconds, often called the 15 by 15 rule. Depending upon the conditions, however, it may sometimes take up to 60 to 90 minutes to obtain definitive results.

Modified biophysical profile (MBPP)

The MBPP is performed in the same manner as the NST with a limited ultrasound (US) performed to assess the amount of amniotic fluid, which is reported as the amniotic fluid index (AFI). Following the NST, an US is done to observe the amount of amniotic fluid present in four quadrants, which are divided along the umbilicus midline and perpendicular to the midline. There must be no fetal parts or any umbilical cord present in any of these pockets of fluid in order to be counted.

Contraction stress test (CST) and oxytocin contraction stress test (OCT)

The contraction stress test (CST) is similar to the NST except the FHR is evaluated for accelerations, 15 beats higher than baseline lasting 15 seconds, and in response to contractions as well. A CST requires the presence of three uterine contractions (UCs) within a 10-minute period lasting at least 40 seconds and of moderate intensity. During a contraction, the blood flow to the baby is temporarily restricted, which provides a form of ''stress'' to the baby. The baby's response to this stressor reveals significant information regarding available oxygen stores. If contractions are not spontaneously present, the pregnant woman will be instructed on the use of nipple stimulation to produce contractions through the release of natural oxytocin, or oxytocin can be administered through an intravenous infusion (IV) called pitocin to produce contractions. When oxytocin is administered IV, it is called an oxytocin contraction stress test (OCT). The CST/OCT is generally used after an abnormal NST is obtained in order to verify if there are problems present. Many clinicians require their diabetic patients to have at least one CST/OCT a week to assure fetal well-being. Maternal blood pressure is taken with each test.

Biophysical profile (BPP)

The biophysical profile (BPP) is performed by an ultrasound exam over a 30-minute period. The ultrasonographer/examiner looks for gross fetal movement, i.e., kicking and moving around; fetal tone, i.e., making a fist; breathing movements (which the mother can often perceive as hiccoughs); and amniotic fluid volume. A score of 0 or 2 points is assigned to each observation with the results of the NST also adding 2 points for a total possible score of 10 points.

Doppler flow studies, amniocentesis, cordocentesis, and fetal echocardiography

A physician or specially trained ultrasonographer performs Doppler flow studies, which examines the blood flow in the umbilical artery and the baby's middle cerebral artery. An experienced obstetrician/perinatologist performs the amniocentesis or the cordocentesis. For the amniocentesis, an US is used to determine an appropriate place to insert a needle and withdraw amniotic fluid for testing. In a similar manner, US is used with cordocentesis, but in this procedure a needle is guided into the umbilical cord to withdraw fetal blood for testing. Fetal echocardiography is a specialized ultrasound of a baby's heart. Since it detects most congenital heart defects, it is recommended if a baby is at a higher risk for a defect than the risk in the general population. The majority of health insurance companies do cover a portion, if not all, of the tests' costs.

Precautions

Clinicians should only prescribe these tests if they are ready to intervene when faced with worrying results. A fetus is considered viable at 24 weeks since that is the minimum gestational age for sufficient lung development. There are no significant risks to the mother or the fetus from the nonstress test (NST), modified biophysical profile (MBPP), or the biophysical profile (BPP). Ultrasound waves utilized in detecting the FHR and for the BPP are painless and safe because this method employs no harmful radiation. There is no evidence that sound waves cause any harm to the mother or the baby.

Aftercare

If the test results are acceptable, the pregnant woman is instructed to continue following her current medical regimen and return for additional testing on the dates prescribed. For NSTs/MBPPs/CSTs, the time period between tests should be no longer than three to four days under high-risk conditions with fetal movement counting taking place in between testing dates. Ultrasounds should be rescheduled as the need dictates per the physician.

Electronic fetal monitoring occurs after an amniocentesis or cordocentesis to assure fetal well-being.

Risks

There are no complications per se from the tests themselves with the exception of unfavorable test results or supine (lying horizontality on the back) hypotension secondary to a pregnant woman lying on her back for an ultrasound. When a pregnant woman lies on her back, the vena cava (one of two large veins that return blood from peripheral circulation to the heart) can become compressed from the pressure of the pregnant uterus such that blood flow to the heart is significantly reduced. There are potential complications from an amniocentesis, i.e., preterm labor, spontaneous rupture of membranes, fetal or placental injury; and the clinician performing the procedure should explain what these are prior to the procedure. There are similar risks and potential complications from cordocentesis as well which should be explained.

Normal results

Usually, a report of normal results for NSTs provides reassurance that the fetus is healthy and should remain so for three to four days, at which time repeat testing will be necessary. A normal NST is reported as being reactive, which means the fetal heart is "reacting" to movement such that the FHR is accelerating 15 beats per minute above the baseline FHR for 15 seconds twice within a 20-minute period of time. A non-reactive NST is one that fails to meet this criterion within an 80 to 90 minute period of time. For an extremely preterm fetus, a normal NST is reported as being reactive for gestational age, which indicates the FHR demonstrated two accelerations of 10 beats per minute above baseline for 10 seconds over a 20–30 minute period. Typically, the central nervous system is not completely mature until approximately 32 weeks gestational age, and a report of reactive for gestational age takes this into consideration. It is important to remember that a normal result does not guarantee that no problems are present. Although very rare in occurrence, false normal results can be observed.

The CST results are reported as reactive/negative, reactive/suspicious, reactive/positive (a very unlikely result), or non-reactive/negative, non-reactive/suspicious, non-reactive/positive. The reactive/non-reactive part of the test report refers to the presence or absence of accelerations. The negative part refers to no decelerations being present with uterine contractions (UCs). "Suspicious" refers to the presence of some decelerations with UCs, and "positive" refers to the presence of decelerations more than 50 percent of the time with UCs. A suspicious or positive result requires further evaluation, i.e., prolonged EFM monitoring or a BPP. A normal BPP report without an NST is 8 points and 10 points with a reactive NST. Six points is suspicious and requires either a CST or a repeat BPP within 24 hours. A total of 4 points is not reassuring and requires immediate evaluation by prolonged EFM.

All results are given to the primary physician who must then make a decision as to the appropriate course of action. Abnormal CST results generally indicate the baby is not receiving sufficient oxygen and may not be capable of withstanding the stress of labor and subsequent vaginal delivery. If this is the case, a **cesarean section** may be performed. The final outcome depends on the mother's individual circumstances. Severe pregnancy-induced hypertension may require immediate delivery via cesarean section. In some cases, medications such as betamethasone may be given to the mother to speed up the lung maturity of the baby. If the mother's cervix is favorable for induction, labor may be induced.

Parental concerns

The healthcare provider should give a complete explanation to the pregnant woman and her partner regarding the tests, i.e., what to expect, how long the test may take, what it means, and why it is being done. It fre-

KEY TERMS

Amniotic fluid—The liquid in the amniotic sac that cushions the fetus and regulates temperature in the placental environment. Amniotic fluid also contains fetal cells.

Deceleration—A decrease in the fetal heart rate that can indicate inadequate blood flow through the placenta.

Fundal height—Measured by a tape measure from the top of the symphysis pubis, over the arch of the growing uterus, to the top of the fundus.

Oxytocin—A hormone that stimulates the uterus to contract during child birth and the breasts to release milk.

Pitocin—A synthetic hormone that produces uterine contractions.

Vibroacoustic stimulation—In the biophysical profile, use of an artificial larynx to produce a loud noise to "awaken" the fetus.

quently helps if the pregnant woman has eaten prior to undergoing the test. Pregnant women should know that every test is not compulsory, and that if the results of the test do not matter to the parents, it may not have to be performed.

See also High-risk pregnancy.

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American College of Obstetricians and Gynecologists. 409 12th Street, SW, PO Box 96920, Washington, DC 20090. Web site: <www.acog.org>.

Association of Women's Health, Obstetric, and Neonatal Nursing. 2000 L Street, NW, Suite 740, Washington, DC 20036. Web site: <www.awhonn.org/>.

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Antiacne drugs

Definition

Acne is a skin disorder that leads to an outbreak of lesions called pimples or "zits." The most common form of the disease in adolescents is called acne vulgaris. Antiacne drugs are the medicines that help clear up the pimples, blackheads, whiteheads, and more severe forms of lesions that occur when a teen has acne.

Different types of antiacne drugs are used for different treatment purposes, depending on the severity of the condition. For example, lotions, soaps, gels, and creams containing substances called benzoyl peroxide or tretinoin may be used to clear up mild to moderately severe acne. Isotretinoin (Accutane) is an oral drug that is prescribed only for very severe, disfiguring acne.

Acne is caused by the overproduction of sebum during **puberty** when high levels of the male hormone androgen cause excess sebum to form. Sebum is an oily substance that forms in glands just under the surface of the skin called sebaceous glands. Sebum normally flows out hair follicles onto the skin to act as a natural skin moisturizer. The glands are connected to hair follicles that allow the sebum, or oil, to empty onto the skin through a pore.

Sometimes the sebum combines with dead, sticky skin cells and bacteria called *Propionibacterium acnes (P. acnes)* that normally live on the skin. The mixture of oil and cells allows the bacteria to grow in the follicles. When this happens, a hard plug called a comedo can form. A comedo is an enlarged hair follicle. It can appear on the skin as a blackhead, which is a comedo that reaches the skin's surface and looks black, or as a whitehead, which is a comedo that is sealed by keratin, the fibrous protein produced by the skin cells and looks like a white bump.

In addition, pimples can form on the skin. Types of pimples include:

- papules, which are small, red bumps that may be tender to touch
- pustules, which are pus-filled lesions that are often red at the base
- nodules, which are large, painful lesions deep in the skin
- cysts, which are painful pus-filled lesions deep in the skin that can cause scarring

Pimples form when the follicle is invaded by the *P. acnes* bacteria. The damaged follicle weakens and bursts open, releasing sebum, bacteria, skin cells, and white blood cells into surrounding tissues. Scarring happens when new skin cells are created to replace the damaged cells. The most severe type of acne includes both nodules and cysts.

Description

Acne cannot be cured, but antiacne drugs can help clear the skin and reduce the chance of scarring. The goal of treating moderate acne is to decrease inflammation and prevent new comedones from forming. Benzoyl peroxide and tretinoin work by mildly irritating the skin. This encourages skin cells to slough off, which helps open blocked pores. Benzoyl peroxide also kills bacteria,

which helps prevent whiteheads and blackheads from turning into pimples. Isotretinoin shrinks the glands that produce sebum. It is used for severe acne lesions and must be carefully monitored because of its side effects. **Antibiotics** also may be prescribed to kill bacteria and reduce inflammation.

General use

Benzoyl peroxide is found in many over-the-counter acne products that are applied to the skin, such as Benoxyl, Neutrogena Acne, PanOxyl, and some formulations of Clean & Clear, Clearasil, and Oxy. Some benzoyl peroxide products are available without a physician's prescription; others require a prescription. Acne treatments that can dry the skin should be used with caution by people with skin of color.

Tretinoin (Retin-A) is available only with a physician's prescription. It comes in liquid, cream, and gel forms, which are applied to the skin. Isotretinoin (Accutane), which is taken by mouth in capsule form, is available only with a physician's prescription. Only physicians experienced in diagnosing and treating severe acne, such as dermatologists, should prescribe isotretinoin.

Recommended dosages

The recommended dosage depends on the type of antiacne drug. These drugs usually come with written directions for patients and should be used only as directed by the prescribing physician. Teens who have questions about how to use the medicine should check with their physician or pharmacist.

Patients who use isotretinoin usually take the medicine for a few months, then stop for at least two months. Their acne may continue to improve even after they stop taking the medicine. If the condition is still severe after several months of treatment and a two-month break, the physician may prescribe a second course of treatment.

Precautions

Isotretinoin

Isotretinoin can cause serious birth defects, including **mental retardation** and physical deformities. This medicine should not be used during pregnancy. Females who are able to bear children should not use isotretinoin unless they have very severe acne that has not cleared up with the use of other antiacne drugs. In that case, a woman who uses this drug must have a pregnancy test two weeks before beginning treatment and each month she is taking the drug. Another pregnancy test must be done one month after treatment ends. The woman must use an effective birth control method for one month before treatment begins and must continue using it throughout treatment and for one month after treatment ends. Females who are able to bear children and who want to use this medicine should discuss this information with their healthcare providers. Before using the medicine, they will be asked to sign a consent form stating that they understand the danger of taking isotretinoin during pregnancy and that they agree to use effective birth control.

People using this drug should not donate blood to a blood bank while taking isotretinoin or for 30 days after treatment with the drug ends. This will help reduce the chance of a pregnant woman receiving blood containing isotretinoin, which could cause birth defects.

Isotretinoin may cause a sudden decrease in night vision. If this happens, users should not drive or do anything else that could be dangerous until vision returns to normal. They should also let the physician know about the problem.

This medicine may also make the eyes, nose, and mouth dry. Ask the physician about using special eye drops to relieve eye dryness. To temporarily relieve the dry mouth, chew sugarless gum, suck on sugarless candy or ice chips, or use saliva substitutes, which come in liquid and tablet forms and are available without a prescription. If the problem continues for more than two weeks, check with a physician or dentist. Mouth dryness that continues over a long time may contribute to **tooth decay** and other dental problems.

Isotretinoin may increase sensitivity to sunlight. Patients being treated with this medicine should avoid exposure to the sun and should not use tanning beds, tanning booths, or sunlamps until they know how the drug affects them.

In the early stages of treatment with isotretinoin, some people's acne seems to get worse before it starts getting better. If the condition becomes much worse or if the skin is very irritated, they should check with the physician who prescribed the medicine.

Benzoyl peroxide and tretinoin

When applying antiacne drugs to the skin, people should be careful not to get the medicine in the eyes, mouth, or inside the nose. They should not put the medicine on skin that is wind burned, sunburned, or irritated, and not apply it to open **wounds**.

Because antiacne drugs such as benzoyl peroxide and tretinoin irritate the skin slightly, users should avoid

doing anything that might cause further irritation. They should wash the face with mild soap and water only two or three times a day, unless the physician says to wash it more often. They should also avoid using abrasive soaps or cleansers and products that might dry the skin or make it peel, such as medicated cosmetics, cleansers that contain alcohol, or other acne products that contain resorcinol, sulfur, or salicylic acid.

If benzoyl peroxide or tretinoin make the skin too red or too dry or cause too much peeling, the user should check with a physician. Using the medicine less often or using a weaker strength may be necessary. Benzoyl peroxide can irritate the skin of people with skin of color and cause darkened spots called hyperpigmentation on the skin. Benzoyl peroxide may discolor hair or colored fabrics.

ORAL DRUGS Oral antibiotics are taken daily for two to four months. The drugs used include tetracycline, erythromycin, minocycline (Minocin), doxycycline, clindamycin (Cleocin), and trimethoprim-sulfamethoxazole (Bactrim, Septra). Possible side effects include allergic reactions, stomach upset, vaginal yeast infections, **dizziness**, and tooth discoloration.

The goal of treating moderate acne is to decrease inflammation and prevent new comedones from forming. One effective treatment is topical tretinoin, used along with a topical or oral antibiotic. A combination of topical benzoyl peroxide and erythromycin is also very effective. Improvement is normally seen within four to six weeks, but treatment is maintained for at least two to four months.

Special conditions

People who have certain medical conditions or who are taking certain other medicines may have problems if they use antiacne drugs. Before using these products, the physician should be informed about any of the following conditions.

ALLERGIES Anyone who has had unusual reactions to etretinate, isotretinoin, tretinoin, vitamin A preparations, or benzoyl peroxide in the past should let the physician know before using an antiacne drug. The physician should also be told about any **allergies** to foods, dyes, preservatives, or other substances.

PREGNANCY Teens who are pregnant or who may become pregnant should check with a physician before using tretinoin or benzoyl peroxide. Isotretinoin causes birth defects in humans and must not be used during pregnancy.

OTHER MEDICAL CONDITIONS Before using antiacne drugs applied to the skin, people with any of these medical problems should make sure their physicians are aware of their conditions:

- Eczema. Antiacne drugs that are applied to the skin may make this condition worse.
- Sunburn or raw skin. Antiacne drugs that are applied to the skin may increase the **pain** and irritation of these conditions.

In people with certain medical conditions, isotretinoin may increase the amount of triglyceride (a fatty-substance) in the blood. This may lead to heart or blood vessel problems. Before using isotretinoin, adolescents with any of the following medical problems should make sure their physicians are aware of their conditions:

- alcoholism or heavy drinking, currently or in the past
- diabetes or **family** history of diabetes (Isotretinoin may change blood sugar levels.)
- family history of high triglyceride levels in the blood
- severe weight problems

Using antiacne drugs with certain other drugs may affect the way the drugs work or may increase the chance of side effects.

Side effects

Conditions caused by isotretinoin

Minor discomforts such as dry mouth or nose, dry eyes, dry skin, or **itching** usually go away as the body adjusts to the drug and do not require medical attention unless they continue or are bothersome.

Other side effects should be brought to a physician's attention. These include:

- burning, redness, or itching of the eyes
- nosebleeds
- signs of inflammation of the lips, such as peeling, burning, redness or pain

Bowel inflammation is not a common side effect, but it may occur. If any of the following signs of bowel inflammation occur, stop taking isotretinoin immediately and check with a physician:

- pain in the abdomen
- bleeding from the rectum
- severe diarrhea

Conditions caused by benzoyl peroxide and tretinoin

The most common side effects of antiacne drugs applied to the skin are slight redness, dryness, peeling, and stinging, and a warm feeling to the skin. These problems usually go away as the body adjusts to the drug and do not require medical treatment.

Other side effects should be brought to a physician's attention. Check with a physician as soon as possible if any of the following side effects occur:

- blistering, crusting, or swelling of the skin
- severe burning or redness of the skin
- darkening or lightening of the skin (This effect will eventually go away after treatment with an antiacne drug ends.)
- skin rash

Other side effects are possible with any type of antiacne drug. Anyone who has unusual symptoms while using antiacne drugs should get in touch with his or her physician.

Interactions

Patients using antiacne drugs on their skin should tell their physicians if they are using any other prescription or nonprescription (over-the-counter) medicine that they apply to the skin in the same area as the antiacne drug.

Isotretinoin may interact with other medicines. When this happens, the effects of one or both drugs may change or the risk of side effects may be greater. Anyone who takes isotretinoin should let the physician know about all other medicines being used and should ask whether the possible interactions can interfere with drug therapy. Among the drugs that may interact with isotretinoin are listed below:

- Etretinate (Tegison), used to treat severe **psoriasis**. Using this medicine with isotretinoin increases side effects.
- Tretinoin (Retin-A, Renova). Using this medicine with isotretinoin increases side effects.
- Vitamin A or any medicine containing vitamin A. Using any vitamin A preparations with isotretinoin increases side effects. Do not take vitamin supplements containing vitamin A while taking isotretinoin.
- Tetracyclines (used to treat infections). Using these medicines with isotretinoin increases the chance of swelling of the brain. Make sure the physician knows if tetracycline is being used to treat acne or another infection.

KEY TERMS

Acne—A chronic inflammation of the sebaceous glands that manifests as blackheads, whiteheads, and/or pustules on the face or trunk.

Bacteria—Singular, bacterium; tiny, one-celled forms of life that cause many diseases and infections.

Bowel—The intestine; a tube-like structure that extends from the stomach to the anus. Some digestive processes are carried out in the bowel before food passes out of the body as waste.

Cyst—An abnormal sac or enclosed cavity in the body filled with liquid or partially solid material. Also refers to a protective, walled-off capsule in which an organism lies dormant.

Eczema—A superficial type of inflammation of the skin that may be very itchy and weeping in the early stages; later, the affected skin becomes crusted, scaly, and thick.

Noncomedogenic—A substance that does not contribute to the formation of blackheads or pimples on the skin. Jojoba oil is noncomedogenic.

Pimple—A small, red swelling of the skin.

Psoriasis—A chronic, noncontagious skin disease that is marked by dry, scaly, and silvery patches of skin that appear in a variety of sizes and locations on the body.

Pus—A thick, yellowish or greenish fluid composed of the remains of dead white blood cells, pathogens, and decomposed cellular debris. It is most often associated with bacterial infection.

Triglyceride—A substance formed in the body from fat in the diet. Triglycerides are the main fatty materials in the blood. Bound to protein, they make up high- and low-density lipoproteins (HDLs and LDLs). Triglyceride levels are important in the diagnosis and treatment of many diseases including high blood pressure, diabetes, and heart disease.

Parental concerns

Acne comes at a difficult time, the adolescent years. While mild acne can be treated with over-the-counter medications, more severe acne needs medical attention. Experts advise against a wait-and-see attitude. Treatment options can help control acne and avoid scarring.

Isotretinoin can cause serious birth defects, including mental retardation and physical deformities. This medicine should not be used during pregnancy. Sexually active adolescent females who are able to bear children should not use isotretinoin unless they have very severe acne that has not cleared up with the use of other antiacne drugs. In addition, acne treatments that can dry the skin should be used with caution by people with skin of color.

See also Acne.

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Antiasthmatic drugs

Definition

Antiasthmatic drugs are medicines that treat or prevent **asthma** attacks.

Description

Three types of drugs are used in treating and preventing asthma attacks:

- Bronchodilators relax the smooth muscles that line the airway. This makes the airways open wider, letting more air pass through them. These drugs are used mainly to relieve sudden asthma attacks or to prevent attacks that might come on after **exercise**. They may be taken by mouth, injected, or inhaled.
- Corticosteroids block the inflammation that narrows the airways. Used regularly, these drugs help prevent asthma attacks. Those attacks that do occur will be less severe. However, corticosteroids cannot stop an attack that is already underway. These drugs may be taken by mouth, injected, or inhaled.
- Cromolyn also is taken regularly to prevent asthma attacks and may be used alone or with other asthma medicines. It cannot stop an attack that already has started. The drug works by preventing certain cells in the body from releasing substances that cause allergic reactions or asthma symptoms. One brand of this drug, Nasalcrom, comes in capsule and nasal spray forms and is used to treat hay fever and other **allergies**. The inhalation form of the drug, Intal, is used for asthma. It comes in aerosol canisters, in capsules that are inserted into an inhaler, and in liquid form that is used in a nebulizer.

General use

All three types of drugs may be used in combination with each other.

Cromolyn is a common but not invariable first choice for children who have asthma. It reduces the frequency of asthmatic attacks and is suitable for long-term use. Cromolyn may not be needed when attacks are mild and infrequent.

Bronchodilators should be used to treat attacks once they begin. They may also be taken on a regular basis to prevent attacks.

Corticosteroids are valuable, but some have serious long-term side effects. Except in patients whose conditions cannot be managed with cromolyn and brochodilators, corticosteroids should be reserved for emergency room use. In patients who require ongoing use of steroids, alternate day dosing or inhalation of some of the newer corticosteroids may minimize the adverse effects of this class of drugs.

A young girl suffering from asthma uses an inhaler to assist her breathing. *(© Alan Towse; Ecoscene/Corbis.)*

KEY TERMS

Asthma—A disease in which the air passages of the lungs become inflamed and narrowed, causing wheezing, coughing, and shortness of breath.

Inflammation—Pain, redness, swelling, and heat that develop in response to tissue irritation or injury. It usually is caused by the immune system's response to the body's contact with a foreign substance, such as an allergen or pathogen.

Inhalant—Medication that is breathed into the lungs.

Mucus—The thick fluid produced by the mucous membranes that line many body cavities and structures. It contains mucin, white blood cells, water, inorganic salts, and shed cells, and it serve to lubricate body parts and to trap particles of dirt or other contaminants.

Nebulizer—A device that turns liquid forms of medication into a fine spray that can be inhaled.

Precautions

Using antiasthmatic drugs properly is important. Because bronchodilators provide quick relief, some people may be tempted to overuse them. However, with some kinds of bronchodilators, doing so can lead to serious and possibly life-threatening complications. In the long run, patients are better off using bronchodilators only as directed and also using corticosteroids, which eventually will reduce their need for bronchodilators.

Parents whose children are using their antiasthmatic drugs correctly but feel their asthma is not under control should see consult their child's physicians. The physician can either increase the dose, switch to another medicine, or add another medicine to the regimen.

Corticosteroids are powerful drugs that may cause serious side effects when used over the long term. However, these problems are much less likely with the inhalant forms than with the oral and injected forms. While the oral and injected forms generally should be used only for one to two weeks, the inhalant forms may be used for long periods.

When used to prevent asthma attacks, cromolyn must be taken as directed every day. The drug may take as long as four weeks to start working. Unless told to do so by a physician, patients should not stop taking the drug just because it does not seem to be working. When symptoms do begin to improve, patients should continue taking all medicines that have been prescribed, unless a physician directs otherwise.

Side effects

Inhalant forms of antiasthmatic drugs may cause dryness or irritation in the throat, dry mouth, or an unpleasant taste in the mouth. To help prevent these problems, patients can gargle and rinse the mouth or take a sip of water after each dose.

More serious side effects are not common when these medicines are used properly. However, parents whose children have unusual or bothersome symptoms after taking an antiasthmatic drug should get in touch with the child's physician.

Interactions

There are many drugs that are used in treatment of asthma. Interactions should be reviewed on an individual basis.

Drugs which decrease blood levels of aminophylline (which opens bronchial passages) and may require a dose increase are:

- carbamazepine
- isoprenolol
- phenobarbital
- phenytoin
- rifampin

Parental concerns

All health professionals with responsibility for an asthmatic's drug therapy should have an up-to-date list of the drugs and doses being used by the child. Asthmatic children should wear a suitable identification bracelet with a list of drugs being used, in case of an emergency room admission. Children using inhalers should be knowledgeable on the use of these devices.

See also Asthma.

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Antibiotics

Definition

Antibiotics are used for treatment or prevention of bacterial infection. They may be informally defined as the subgroup of anti-infectives that are derived from bacterial sources and are used to treat bacterial infections. Other classes of drugs, most notably the **sulfonamides**, may be effective antibacterials. Similarly, some antibiotics may have secondary uses, such as the use of demeclocycline (Declomycin, a tetracycline derivative) to treat the syndrome of inappropriate antidiuretic hormone (SIADH) secretion. Other antibiotics may be useful in treating protozoal infections.

Description

Classifications

Although there are several classification schemes for antibiotics, based on bacterial spectrum (broad versus narrow) or route of administration (injectable versus oral versus topical), or type of activity (bactericidal versus bacteriostatic), the most useful is based on chemical structure. Antibiotics within a structural class will generally show similar patterns of effectiveness, toxicity, and allergic potential.

PENICILLINS The **penicillins** are the oldest class of antibiotics and have a common chemical structure that they share with the cephalosporins. Classed as the beta-lactam antibiotics, the two groups are generally bactericidal, which means that they kill bacteria rather than simply inhibit its growth. The penicillins can be further subdivided. The natural penicillins are based on the original penicillin G structure; penicillinase-resistant penicillins, notably methicillin and oxacillin, are active even in the presence of the bacterial enzyme that inactivates most natural penicillins. Aminopenicillins such as ampicillin and amoxicillin have an extended spectrum of action compared with the natural penicillins; extended spectrum penicillins are effective against a wider range of bacteria. These generally include coverage for *Pseudomonas aeruginosa*.

CEPHALOSPORINS Cephalosporins and the closely related cephamycins and carbapenems, like the penicillins, contain a beta-lactam chemical structure. Consequently, there are patterns of cross-resistance and cross-allergenicity among the drugs in these classes. The ''cepha'' drugs are among the most diverse classes of antibiotics and are themselves subdivided into first, second, and third generations. Each generation has a broader spectrum of activity than the one before. In addition, cefoxitin, a cephamycin, is highly active against anaerobic bacteria, which offers utility in treatment of abdominal infections. The third generation drugs, cefotaxime, ceftizoxime, ceftriaxone, and others, cross the blood-brain barrier and may be used to treat **meningitis** and **encephalitis**. Cephalosporins are the usually preferred agents for surgical prophylaxis.

FLUOROQUINOLONES The fluoroquinolones are synthetic antibacterial agents and not derived from bacteria. They are included here because they can be readily interchanged with traditional antibiotics. An earlier, related class of antibacterial agents, the quinolones, drugs that were not well absorbed, could be used only to treat urinary tract infections. The fluoroquinolones, which are based on the older group, are broad-spectrum bacteriocidal drugs that are chemically unrelated to the penicillins or the cephalosporins. They are well distributed into bone tissue and so well absorbed that in general they are as effective by the oral route as by intravenous infusion.

TETRACYCLINES **Tetracyclines** got their name from the fact that they share a chemical structure that has four rings. They are derived from a species of *Streptomyces* bacteria. Broad-spectrum bacteriostatic agents, the tetracyclines may be effective against a wide variety of microorganisms, including rickettsia and amoebic parasites.

MACROLIDES The macrolide antibiotics are derived from *Streptomyces* bacteria. Erythromycin, the prototype of this class, has a spectrum and use similar to penicillin. Newer members of the group, azithromycin and clarithromycin, are particularly useful for their high level of lung penetration. Clarithromycin has been widely used to treat *Helicobacter pylori* infections, the cause of stomach ulcers.

OTHERS Other classes of antibiotics include the aminoglycosides, which are particularly useful for their effectiveness in treating *Pseudomonas aeruginosa* infections, and the lincosamide drugs clindamycin and lincomycin, which are highly active against anaerobic pathogens. There are other, individual drugs which may have utility in specific infections.

General use

Antibiotics are used for treatment or prevention of bacterial infections. In most cases, they are prescribed for a short period of time to treat a specific infection. This period may range from three days to 10 days or more. More serious infections may require longer periods of treatment, up to several months or longer. Lower doses may be used over a long period of time to prevent the return of a serious infection.

Precautions

All antibiotics should be used as prescribed. These drugs will degrade over time and lose their potency. Not completing a prescribed course of treatment increases the probability that drug-resistant strains of organisms will develop.

Side effects

All antibiotics cause risk of overgrowth by non-susceptible bacteria. Manufacturers list other major hazards by class; however, the healthcare provider should review each drug individually to assess the degree of risk. Generally, breastfeeding may be continued while taking antibiotics, but nursing mothers should always check with their physician first. Excessive or inappropriate use may promote growth of resistant pathogens.

Hypersensitivity to penicillins may be common, and cross allergenicity with cephalosporins has been reported. (That is, those who are allergic to penicillin may also be allergic to cephalosporins.) Penicillins are classed as category B during pregnancy.

Several cephalosporins and related compounds have been associated with seizures. Cefmetazole, cefoperazone, cefotetan, and ceftriaxone may be associated with problems in poor blood clotting. Pseudomembranous colitis (an intestinal disorder) has been reported with cephalosporins and other broad spectrum antibiotics. Some drugs in this class may cause kidney toxicity. Cephalosporins are classed as category B during pregnancy.

Regarding fluoroquinolones, lomefloxacin has been associated with increased sensitivity to light. All drugs in this class have been associated with convulsions. Fluoroquinolones are classed as category C during pregnancy.

Of the tetracyclines, demeclocycline may cause increased photosensitivity. Minocycline may cause **dizziness**. Healthcare providers do not prescribe tetracyclines in children under the age of eight, and they specifically avoid doing so during periods of tooth

KEY TERMS

Bacteria—Singular, bacterium; tiny, one-celled forms of life that cause many diseases and infections.

Bacterial spectrum—The number of bacteria an antibiotic is effective against. Broad-spectrum antibiotics treat many different kinds of bacteria. Narrow-spectrum antibiotics treat fewer kinds.

Inflammation—Pain, redness, swelling, and heat that develop in response to tissue irritation or injury. It usually is caused by the immune system's response to the body's contact with a foreign substance, such as an allergen or pathogen.

Meningitis—An infection or inflammation of the membranes that cover the brain and spinal cord. It is usually caused by bacteria or a virus.

Microorganism—An organism that is too small to be seen with the naked eye, such as a bacterium, virus, or fungus.

Organism—A single, independent unit of life, such as a bacterium, a plant, or an animal.

Pregnancy category—A system of classifying drugs according to their established risks for use during pregnancy. Category A: Controlled human studies have demonstrated no fetal risk. Category B: Animal studies indicate no fetal risk, but no human studies, or adverse effects in animals, but not in well-controlled human studies. Category C: No adequate human or animal studies, or adverse fetal effects in animal studies, but no available human data. Category D: Evidence of fetal risk, but benefits outweigh risks. Category X: Evidence of fetal risk. Risks outweigh any benefits.

development. Oral tetracyclines bind to anions such as calcium and iron. Although doxycycline and minocycline may be taken with meals, people must be advised to take other tetracycline antibiotics on an empty stomach and not to take the drugs with milk or other calcium-rich foods. Expired tetracycline should never be administered. These drugs have a pregnancy category D. Use during pregnancy may cause alterations in fetal bone development.

Of the macrolides, erythromycin may aggravate the weakness of people with myasthenia gravis. Azithromycin has, rarely, been associated with allergic reactions, including angioedema (swelling), **anaphylaxis**, and severe skin reactions. Oral erythromycin may be highly irritating to the stomach and when given by injection may cause severe phlebitis (inflammation of the veins). These drugs should be used with caution in people with liver dysfunction. Azithromycin and erythromycin are pregnancy category B. Clarithromycin, dirithromycin, and troleandomycin are pregnancy category C.

The aminoglycosides class of drugs causes kidney and ear problems. These problems can occur even with normal doses. Dosing should be based on kidney function, with periodic testing of both kidney function and hearing. These drugs are pregnancy category D.

Parental concerns

Parents should be sure to follow all dosage and label directions. This includes using all of a prescription at the time it is prescribed. Parents should also ensure that children cannot ingest any prescription medications by accident.

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Antibiotics, topical

Definition

Topical **antibiotics** are medicines applied to the skin to kill bacteria.

Description

Some topical antibiotics are available without a prescription and are sold in many forms, including creams, ointments, powders, and sprays. Some widely used topical antibiotics are bacitracin, neomycin, mupirocin, and polymyxin B. Among the products that contain one or more of these ingredients are Bactroban (a prescription item), Neosporin, Polysporin, and triple antibiotic ointment or cream.

General use

Topical antibiotics help prevent infections caused by bacteria that get into minor cuts, scrapes, and **burns**. Treating minor **wounds** with antibiotics allows quicker healing. If the wounds are left untreated, the bacteria will multiply, causing **pain**, redness, swelling, **itching**, and oozing. Untreated infections can eventually spread and become much more serious.

Different kinds of topical antibiotics kill different kinds of bacteria. Many antibiotic first-aid products contain combinations of antibiotics to make them effective against a broad range of bacteria.

When treating a wound, it is not enough to simply apply a topical antibiotic. The wound must first be cleaned with soap and water and patted dry. After the antibiotic is applied, the wound should be covered with a dressing, such as a bandage or a protective gel or spray. It is best to keep wounds clean and moist while they heal. The covering should still allow some air to reach the wound, however.

Precautions

The recommended dosage depends on the type of topical antibiotic. Parents should follow the directions on the package label or ask a pharmacist for directions before dressing their child's wound.

In general, topical antibiotics should be applied within four hours after injury. More than the recommended amount should not be used, and the antibiotic should not be applied more often than three times a day.

In the early 2000s many people are concerned about antibiotic resistance, a problem that can develop when antibiotics are overused. Over time, bacteria develop new defenses against antibiotics that once were effective against them. Because bacteria reproduce so quickly, these defenses can be rapidly passed on through generations of bacteria until almost all are immune to the effects of a particular antibiotic. The process happens faster than new antibiotics can be developed. To help control this development, many experts advise people to use topical antibiotics only for short periods, that is, until the wound heals, and only as directed. For the topical antibiotic to work best, it should be used only to prevent infection in a fresh wound, not to treat an infection that has already started. Wounds that are not fresh may need the attention of a physician in order to prevent complications such as blood poisoning.

Topical antibiotics are meant to be used only on the skin and only for only a few days at a time. If the wound has not healed in five days, the antibiotic should be discontinued and a doctor called.

Topical antibiotics should not be used on large areas of skin or on open wounds. These products should not be used to treat **diaper rash** in infants or incontinence rash in adults.

Only minor cuts, scrapes, and burns should be treated with topical antibiotics. Certain kinds of injuries may need medical care and should not be self-treated with topical antibiotics. These include:

- large wounds
- deep cuts

KEY TERMS

Bacteria—Singular, bacterium; tiny, one-celled forms of life that cause many diseases and infections.

Conception—The union of egg and sperm to form a fetus.

Fungal—Caused by a fungus.

Fungus—A member of a group of simple organisms that are related to yeast and molds.

Incontinence—A condition characterized by the inability to control urination or bowel functions.

Inflammation—Pain, redness, swelling, and heat that develop in response to tissue irritation or injury. It usually is caused by the immune system's response to the body's contact with a foreign substance, such as an allergen or pathogen.

- cuts that continue bleeding
- cuts that may need stitches
- burns any larger than a few inches in diameter
- scrapes imbedded with particles that will not wash away
- animal bites
- deep puncture wounds
- eye injuries

Regular topical antibiotics should never be used in the eyes. Special prescription antibiotic products are available for treating eye infections.

Although topical antibiotics control infections caused by bacteria, they may allow fungal infections to develop. The use of other medicines to treat the fungal infections may be necessary. Parents should check with the physician or pharmacist.

Some people may be allergic to one or more ingredients in a topical antibiotic product. If an allergic reaction develops, the product should be discontinued immediately and a physician called.

As of 2004, no harmful or abnormal effects had been reported in babies whose mothers used topical antibiotics while pregnant or nursing. However, pregnant women generally are advised not to use any drugs during the first three months after conception. A woman who is pregnant or breastfeeding or who plans to become pregnant should check with her physician before using a topical antibiotic.

Unless a parent is so advised by the childs' physician, topical antibiotics should not be used on children under two months of age.

Side effects

The most common minor side effects of topical antibiotics are itching or burning. These problems usually do not require medical treatment unless they do not go away or they interfere with normal activities.

If any of the following side effects occur, a doctor should be called as soon as possible:

- rash
- swelling of the lips and face
- sweating
- tightness or discomfort in the chest
- breathing problems
- fainting or dizziness
- low blood pressure
- nausea
- diarrhea
- hearing loss or ringing in the ears

Other rare side effects may occur. Anyone who has unusual symptoms after using a topical antibiotic should get in touch with the physician who prescribed or the pharmacist who recommended the medication.

Parental concerns

Using certain topical antibiotics at the same time as hydrocortisone (a topical corticosteroid used to treat inflammation) may hide signs of infection or allergic reaction. People should not use these two medicines at the same time unless told to do so by a healthcare provider.

Anyone who is using any other type of prescription or nonprescription (over-the-counter) medicine on the skin should check with a doctor before using a topical antibiotic.

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American Academy of Family Physicians. 11400 Tomahawk Creek Parkway, Leawood, KS 66211-2672. Web site: <www.aafp.org/>.

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Antidepressants

Definition

An antidepressant is a medication used primarily in the treatment of depression. Depression can occur if some of the chemicals called neurotransmitters in the brain are not functioning effectively. There are three specific chemicals that can affect a person's mood: serotonin, norepinephrine, or dopamine. Antidepressants affect one or more of these chemicals in different ways to help stabilize the chemical imbalance often seen in depression. Antidepressant drugs are not happy pills, and they are not a panacea. They are prescription-only drugs that come with risks as well as benefits and should only be taken under a doctor's supervision. Because children and adolescents experience depression just as adults do, they are sometimes prescribed antidepressants by their physician.

Description

Antidepressants are medicines used to help people who have depression. Antidepressant medications may be indicated for those children and adolescents with bipolar depression, psychotic depression, depression with severe symptoms that prevent effective psychotherapy or counseling, and depression that does not respond to psychotherapy. However, given the psychosocial dynamics that often coexist with depression, antidepressants are usually insufficient as the only treatment for children who have the disorder. Psychotherapy is often recommended as an adjunct treatment along with the prescribed antidepressant. The use of antidepressants among children has been growing steadily since the late 1980s.

All antidepressant medications have a slow onset of action, typically three to five weeks. Although side effects may be observed as early as the first dose, significant therapeutic improvement is always delayed. Most antidepressants are believed to work by slowing the removal of certain chemicals from the brain. These chemicals are called neurotransmitters, which are needed for normal brain function. Antidepressants help people with depression by making these natural chemicals more available to the brain. There are many different kinds of antidepressants, including the ones listed below.

Monoamine oxidase (MAO) inhibitors

MAO inhibitors work by blocking the action of a chemical substance known as monoamine oxidase in the nervous system. Studies done in animals suggest that MAO inhibitors may slow growth in children. Little information on the use of MAO inhibitors in children under 16 years old was available as of 2004.

Tricyclics

Tricyclics have been used to treat depression for a long time. They include amitriptyline, desipramine, imipramine, nortriptyline, and trimipramine. Tricyclic antidepressants work by shoring up the brain's supply of norepinephrine and serotonin, chemicals that are abnormally low in depressed patients. This effect allows the flow of nerve impulses to return to normal. The tricyclics do not act by stimulating the central nervous system or by blocking monoamine oxidase.

Selective serotonin reuptake inhibitors (SSRIs)

SSRIs are a group of antidepressants that includes drugs such as citalopram (Celexa), fluoxetine (Prozac), paroxetine (Paxil), sertraline (Zoloft), and escitalopram (Lexapro). In the early 2000s SSRIs have replaced tricyclic antidepressants as the drugs of choice in the treatment of **depressive disorders**, primarily because of their improved tolerability and safety if taken in overdose. These medicines tend to have fewer side effects than the tricyclics.

Others

There are several antidepressants available as of 2004 that, because they are not chemically structured like the other types of antidepressants, are grouped into the category "other" or miscellaneous. Bupropion (Wellbutrin), mirtazapine (Remeron), and venlafaxine (Effexor) are among those in this category.

General use

SSRIs

Selective serotonin reuptake inhibitors (SSRIs) are considered an improvement over older antidepressants because they are better tolerated and are safer if taken in an overdose. The prescription of SSRIs has risen dramatically in the past several years in children and adolescents age 10 to 19. Some research points out that this increase has coincided with a significant decrease in **suicide** rates in this age group, but it is unknown if SSRIs are directly responsible for this improvement. As of 2004, fluoxetine (Prozac) was the only SSRI that the Food and Drug Administration (FDA) has approved for the treatment of children's depression. Fluoxetine (Prozac), sertraline (Zoloft), and fluvoxamine (Luvox) are approved by the FDA for the treatment of **obsessive-compulsive disorder** because studies have shown they are safe and effective medicines for adolescents with this disorder. An early 2000s study showed that citalopram (Celexa) significantly reduced symptoms of major depression in children and adolescents. Sertraline (Zoloft) was also found in studies to be effective with youths, slightly more so for adolescents than younger children. Physicians may frequently prescribe many of the SSRI antidepressants besides fluoxetine (Prozac) for children to treat depression, even though they have not been approved for this use by the FDA. This is called "off-label" use. Off-label refers to the use by doctors of FDA-approved drugs for purposes other than those approved by the agency.

Tricyclics

Tricyclic antidepressants (TCAs) are primarily used to treat depression in adults. The most commonly used ones are nortriptyline (Pamelor), desipramine (Elavil), and imipramine (Tofranil). They function similarly and have similar risks and side effects. They are not as effective in treating depression in children who have not reached **puberty**, and for these children should only be used as a second line agent. There is marginal evidence to support the use of tricyclics in the treatment of depression in adolescents, but the effect is likely to be moderate. Although they are actually not very effective as antidepressants with children, they can be quite helpful for a variety of other problems, including attention deficit disorder, enuresis (**bed-wetting**), and obsessive-compulsive disorder. The American Academy of Child and Adolescent Psychiatry (AACAP) does not recommend TCAs as a first-line treatment for youths requiring medicine for depressive disorders. However, the AACAP acknowledges that some young people with depression may respond better to TCAs than to other antidepressants.

MAO inhibitors

Studies on MAO inhibitors have only been performed on adult patients, and there is as of 2004 no specific information comparing the use of MAO inhibitors in children with use in other age groups. However, animal studies have shown that these medicines may slow growth in young children and are therefore not generally recommended for use in children. Parents should be sure to speak with the doctor regarding whether the use of these medicines is appropriate before giving a monoamine oxidase inhibitor to their child.

Others

Bupropion (Wellbutrin) seems to be a better antidepressant for children than the tricyclic antidepressants. Again, as of 2004 bupropion has not been approved for this use by the FDA. It has also proven to be an effective treatment for children diagnosed with attention deficit disorder. The manufacturer of venlafaxine (Effexor) has issued a statement that the drug is not effective in

treating depression in children and teenagers and is recommending that venlafaxine (Effexor) not be used in pediatric patients. Early 2000s studies have found increased reports of thinking about suicide and self-harm, among children and teens taking venlafaxine (Effexor). Mirtazapine (Remeron) must be used with caution in children with depression. Studies have shown occurrences of children thinking about suicide or attempting suicide in clinical trials for this medicine.

Precautions

In 2004, the FDA issued a health advisory recommending close observation for worsening depression in both adults and children treated with certain antidepressants. The FDA requested that a warning of a possible association between the use of SSRIs and **suicidal behavior** be inserted in the labeling of these medications. Studies have found no direct link between these antidepressants and worsening depression or increased suicide in children. In fact, no suicide has been reported among the more than 4,100 people studied who take SSRIs. However, the FDA continues to study this issue. Some believe the increased risk of suicide is not related to the SSRIs themselves, but a phenomenon seen when the symptoms of depression first begin to improve. This phenomenon occurs when the depressed person starts to gain more energy but is not yet fully relieved of the depressive symptoms. The drugs under review include bupropion (Wellbutrin), citalopram (Celexa), fluoxetine (Prozac), mirtazapine (Remeron), nefazodone (Serzone), paroxetine (Paxil), sertraline (Zoloft), escitalopram (Lexapro) and venlafaxine (Effexor). It should be again noted that the only drug that has received approval for use in children with major depressive disorder is fluoxetine (Prozac). Several of these drugs, including sertraline (Zoloft) and fluoxetine (Prozac) are approved for the treatment of obsessive-compulsive disorder in pediatric patients. The drug escitalopram (Lexapro) does not appear to help depressed children and adolescents, according to one clinical study.

Side effects

MAO inhibitors

MAO inhibitors have largely been supplanted in therapy because of their high risk of significant side effects, most notably severe, possibly fatal high blood pressure, if foods or alcoholic beverages containing tyramine are consumed. Other side effects include **dizziness**, fainting, **headache**, tremors, muscle twitching, confusion, memory impairment, **anxiety**, agitation, insomnia, weakness, drowsiness, chills, blurred vision, and heart palpitations. Treatment with MAO inhibitors should never be halted abruptly, and should not be stopped without first consulting a physician.

Tricyclics

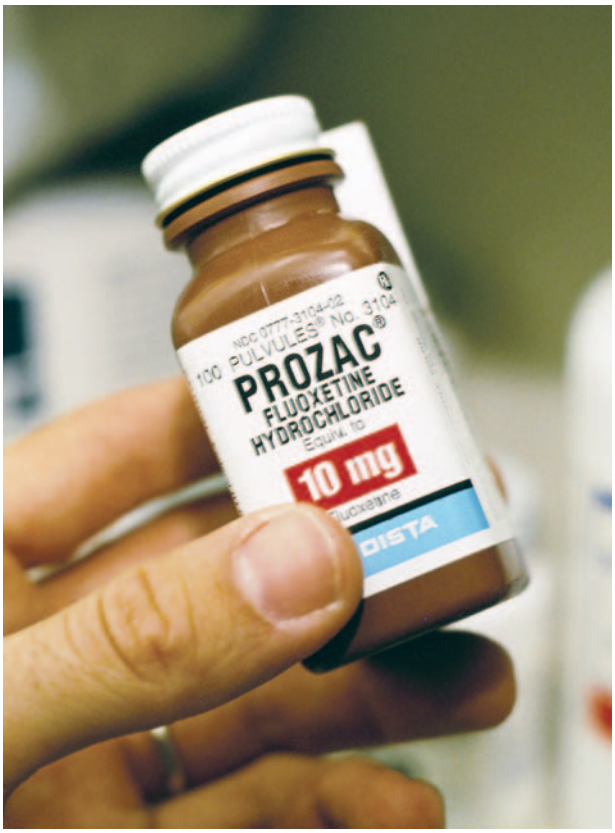
Although TCAs have been shown to be effective in many clinical situations, their use is associated with potentially serious side effects. The most important of these is the potential for an irregular heartbeat, which can at times (though rarely) be fatal. The vast majority of TCA-related deaths happen when an overdose is taken. Physician will likely monitor blood levels, as well as perform echocardiograms to monitor heart functioning. Other side effects include dry mouth, **constipation**, difficulty urinating, blurred vision, sedation, weight gain, central nervous system and cardiovascular toxicity, delirium, and risk of suicide by overdose. The risk of side effects can be reduced with careful prescribing practices.

SSRIs

Several side effects are possible with SSRIs. Special care should be paid in the first few weeks of taking the prescribed drug. Should nervousness, agitation, irritability, mood instability, or sleeplessness emerge or worsen during treatment with SSRIs, parents should obtain a prompt evaluation by their doctor. Some of the side effects that can be caused by SSRIs include dry mouth, **nausea**, nervousness, insomnia, and headache. Those taking fluoxetine (Prozac) might also have a feeling of being unable to sit still. Children already on any of the SSRIs should remain on the drug if it has been helpful, but they should also be carefully monitored by a physician for evidence of side effects. Once begun, treatment with these medications should not be abruptly stopped, because the child may experience further agitation and restlessness. Families should not discontinue treatment without consulting their physician.

Others

Bupropion (Wellbutrin) has several side effects, including drowsiness, lightheadedness, headache, constipation, dry mouth, nausea, and **vomiting**. Occasionally patients may experience tiredness, muscle twitching, weight loss, blurred vision, and trouble sleeping. The main side effect is appetite suppression. In some children this may also lead to **hypoglycemia** (low blood sugar). It is recommended that children on Wellbutrin should eat mid-morning, mid-afternoon, and bedtime snacks in addition to the usual three meals in a manner similar to that of diabetics. The main risk of Wellbutrin is that it increases the likelihood of seizures, though the incidence is rare. Some of these seizures may be related to hypoglycemia and so may be prevented by sticking to the diet as described



The antidepressant Prozac is used to treat depressive disorders. *(© David Butow/Corbis Saba.)*

above. The drug should not be used when there is a past history of seizures or a **family** history of epilepsy.

Interactions

MAO inhibitors

MAO inhibitors have many dietary restrictions, and people taking them need to follow the dietary guidelines and physician's instructions very carefully. A rapid, potentially fatal increase in blood pressure can occur if foods or alcoholic beverages containing tyramine are ingested by a person already taking MAO inhibitors. Foods containing tyramine include sour cream; parmesan, mozzarella, cheddar and other cheeses; beef or chicken liver; cured meats; game meat; caviar; dried fish; bananas; avocados; raisins; soy sauce; fava beans; and caffeine-containing products like colas, coffee and tea, and chocolate. Beverages to be avoided include beer, red wine, other alcoholic beverages, non-alcoholic and reduced alcohol beer, and red wine products.

SSRIs

SSRIs should not be used with any drug that increases serotonin concentrations, including MAO inhibitors, tramadol, sibutramine, meperidine, sumatriptan, lithium, St. John's wort, ginkgo biloba, and some antipsychotic agents. A "serotonin syndrome" may occur, where mental status changes and where agitation, sweating, shivering, tremors, **diarrhea**, and uncoordination, and **fever** may develop. This syndrome may be life-threatening. SSRIs interact with a number of other drugs that act on the central nervous system. Care should be used in combining SSRIs with major or minor tranquilizers or with anti-epileptic agents such as phenytoin (Dilantin) or carbamazepine (Tegretol).

Tricyclics

Tricylic antidepressants should not be taken with the gastric acid inhibitor cimetidine (Tagamet), since this increases the blood levels of the tricyclic compound. TCAs have many interactions, and specialized references should be consulted. Specifically, it is best to avoid other drugs with anticholinergic effects. Tricyclics should not be taken with the **antibiotics** grepafloxacin and sprafloxacin, since the combination may cause serious heart arrythmias.

Others

Alcohol, phenothiazines, and benzodiazepines may all increase the likelihood of seizures if consumed with bupropion (Wellbutrin).

Parental concerns

Major depression in children and adolescents is a serious condition that should be treated in a way that includes careful follow-up and monitoring. If the physi-

KEY TERMS

Monoamine oxidase (MAO) inhibitors—A type of antidepressant that works by blocking the action of a chemical substance known as monoamine oxidase in the nervous system.

Selective serotonin reuptake inhibitors (SSRIs)—A class of antidepressants that work by blocking the reabsorption of serotonin in the brain, thus raising the levels of serotonin. SSRIs include fluoxetine (Prozac), sertraline (Zoloft), and paroxetine (Paxil).

Tricyclic antidepressant—A class of antidepressants, named for their three-ring structure, that increase the levels of serotonin and other brain chemicals. They are used to treat depression and anxiety disorders, but have more side effects than the newer class of antidepressants called selective serotonin reuptake inhibitors (SSRIs).

cian determines that medication is indicated, parents should ensure their child continues to receive ongoing assessment. Selection of an antidepressant for their child is done on an individual basis, as drugs may work differently for different people. What is effective for some may not be effective for others. If one antidepressant is ineffective, then there is probably another one that can be tried. All potentially effective treatments can be associated with side effects. A careful weighing of risks and benefits, with appropriate follow-up to help reduce risks, is the best that can be recommended.

See also Depression.

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Antiepileptics

Definition

Antiepileptic drugs are medicines that reduce the frequency of epileptic seizures.

This class of drugs includes some drugs that have other uses as well. Phenobarbital is a barbiturate. Barbiturates were once widely used as sleeping pills and are still used in anesthesia for surgery. Clonazepam, clorazepate, and diazepam are members of the benzodiazepine group of drugs and are best known for their use as tranquilizers. Phenytoin is used both to control epileptic seizures and to control irregular heart beats.

Description

There are several different types of epilepsy. Different drugs work best on different types of epilepsy. Following is a list of some of the drugs and their uses:

- topamax (Topiramate), keppra, and ACTH: for infantile spasms (IS)
- ativan: for status epilepticas
- phenobarbital: widely used for tonic-clonic but effective in all forms of epilepsy
- phenytoin (Dilantin): used in tonic-clonic and psychomotor epilepsy
- ethosuximide (Zarontin): used to treat absence seizures
- methsuximide (Celontin): used to treat absence seizures
- zonisamide (Zonegran): used to treat partial seizures
- clonazepam (Klonopin): a benzodiazepine, used to treat absence, myoclonic, and akinetic seizures
- clorazepate (Tranxene): a benzodiazepine used to treat partial seizures
- diazepam (Valium): a benzodiazepine used for treatment of status epilepticus but effective against all forms of epilepsy
- primidone (Mysoline, Myidone, Sertan): useful for tonic-clonic, psychomotor, and focal epilepsy
- valproic acid and sodium valproate (Depakene, Depakote): used to treat all types of generalized seizures
- carbamazepine (Carbatrol, Tegretol): used in treatment of tonic-clonic, mixed, and psychomotor seizures
- felbamate (Felbatol): used primarily in adults but may be used to treat seizures associated with Lennox-Gastaut syndrome
- oxcarbazepine (Trileptal): for treatment of complex-partial, simple-partial, and focal seizures
- lamotrigine (Lamictal): used primarily in adults to treat simple and complex partial seizures but may be used to treat seizures associated with Lennox-Gastaut syndrome in children

General use

Although epilepsy is a collective term for a variety of different types of seizures, all forms of epilepsy start with a random discharge of nerve impulses into the brain. Antiepileptic drugs act by either raising the seizure threshold or by limiting the spread of impulses from one

nerve to another inside the brain. As of 2004 the exact mechanism of action is not understood, but there are theories about how some of these drugs work.

Phenobarbital appears to act by slowing down all parts of the brain.

Hydantoins, the class that includes phenytoin, mephenytoin, and ethotoin, seem to work by reducing the flow of sodium into and out of nerve cells. This makes the cells less likely to send out spontaneous impulses, which are the beginning of an epileptic seizure.

Succinimides (ethosuximide, methsuximide, phensuximide) elevate the seizure threshold and make it harder for a nerve impulse to spread from one nerve to another.

Zonisamide may work in a manner similar to the hydantoins, by restricting sodium flow, but some studies contradict this theory.

Benzodiazepines may work by stimulating some brain chemicals that normally slow down nerve function, but the exact mechanism is not known.

Felbamate is similar to the tranquilizer meprobamate (Equanil, Miltown) and may work by blocking the effects of some of the brain chemicals that stimulate the nervous system.

Precautions

Antiepileptic drugs have a large number of side effects and possible adverse effects. To work best, the blood levels of drugs must be kept within a fairly narrow range. Patients should be seen by a qualified physician on a regular basis and, if required, have their blood tested routinely. Too high a blood level of these drugs is likely to cause toxic reactions, while a level that is too low may lead to seizures.

Side effects

Most anticonvulsant medications cause some drowsiness and stomach upset. The following list gives some of the common adverse effects of the various classes of drugs. Parents should consult specific references for more comprehensive lists.

Barbiturates cause the following side effects:

- clumsiness
- dizziness
- constipation
- depression
- faintness

Benzodiazepines cause the following side effects:

- fatigue
- abnormal behavior which can include hallucinations and agitation
- slowed breathing and slowed heart rate
- increased or decreased appetite
- rash and itching

Hydantoins cause the following side effects:

- confusion
- dizziness
- growth of the gums
- severe skin reactions
- stuttering and trembling

Succinimides cause the following side effects:

- dizziness and loss of balance
- severe skin reactions
- depression
- headache
- aggressive behavior

Valproic acid and sodium valproate cause the following side effects:

- stomach and intestinal discomfort
- weight gain or loss
- hair loss
- menstrual bleeding changes
- trembling

Interactions

These drugs have many interactions. People should consult specific references for full information regarding the interactions of all drugs that may be used to treat epilepsy.

Phenobarbital and the hydantoins have a large number of other interactions, but most of the drugs involved are not commonly used in patients under the age of 18 years. The succinimides have no significant drug interactions.

Valproic acid and sodium valproate interact with many of the other drugs used to treat epilepsy. If valproates are added to an existing anti-epileptic drug regimen or other drugs are added to a regimen that contains valproate, additional dose adjustments will usually be needed.

KEY TERMS

Absence seizure—A brief seizure with an accompanying loss of awareness or alertness. Also known as a petit mal seizure.

Lennox-Gastaut syndrome—A severe form of epilepsy that is characterized by the onset in early childhood of frequent seizures of multiple types and by developmental delay.

Seizure threshold—The amount of stimulation required to induce a seizure.

Sodium—An element; sodium is the most common electrolyte found in animal blood serum.

Tonic-clonic seizure—This is the most common type of seizure among all age groups and is categorized into several phases beginning with vague symptoms hours or days before an attack. These seizures are sometimes called grand mal seizures.

Cimetidine (Tagamet) increases the effects of diazepam, clonazepam, and clorazepate.

Interaction prevention

Because antiepileptic drugs have large numbers of adverse effects and drug interactions, they should be prescribed only by physicians who are experienced in their use. Parents should consult specific references for complete information on the drugs related to their child's case.

Best effects and lowest toxicity are achieved when the blood levels of these drugs are kept constant. Maintaining that constant level requires taking the drugs at the same time each day.

Excessive gum growth associated with the hydantoins can normally be prevented or minimized by good dental care.

Patients taking antiepileptic drugs should not receive additional medications without checking with a physician or pharmacist for possible drug interactions. This precaution includes over-the-counter remedies.

Parental concerns

For liquid dosage forms, parents should always use a medicinal teaspoon or calibrated teaspoon. These are designed to deliver an exact amount of medication. Household teaspoons vary in size and should not be used for measuring medication.

Different dosage forms of the same drug may vary in their onset and duration of action. This fact is particularly relevant to phenytoin, which comes in liquid, chewable tablets, short-acting capsules and long-acting capsules. Patients should not switch from one dosage form to another without consulting their physician.

Because children may not be able to describe some of the symptoms associated with some of the adverse effects of these drugs, any evidence of change in behavior or activity should be discussed with the physician who prescribed the drug.

See also Seizure disorder.

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Antihistamines

Definition

Antihistamines are drugs used to treat the symptoms of **allergies** and **allergic rhinitis** by blocking the action of histamine, a chemical released by the immune system in allergic reactions.

Description

Antihistamines are used to treat the sneezing, runny nose, and itchy eyes of allergies and allergic **rhinitis**, as well as allergic skin reactions and anaphylactic reactions to insect **stings** and certain foods. Antihistamines are available as prescription and over-the-counter tablets, topical preparations, nasal sprays, and eye drops.

Antihistamines work by blocking the effects of histamine, a chemical released by mast cells during an allergic response to an allergen. Histamine irritates and inflames the airways to produce sneezing and mucus production. Antihistamines attach to the areas on cells that histamines attach to, thereby blocking the allergic response.

Antihistamines are most effective when taken before exposure to an allergen. When used over time as an allergy treatment, antihistamines reduce the amount of histamine released by cells and decrease the likelihood that an allergic reaction will occur.

General use

Antihistamines are prescribed or recommended for infants, children, and adolescents with allergies and allergic rhinitis. Depending on the type of allergy, oral antihistamines may be taken regularly or seasonally to combat responses to allergens. Common allergens include dog and cat hair, dust mites, grass and tree pollen, and molds and mildew. For allergies that produce nasal symptoms, an antihistamine nasal spray may be used. For itchy eyes, antihistamine eye drops may be used.

Antihistamine tablets and topical creams, gels, sprays, or ointments are used to treat skin **hives** related to **food allergies** and **itching** and hives associated with allergic **contact dermatitis** and insect **bites and stings**.

In addition to treating allergies, some antihistamines have side effects that are used to treat other conditions. The strong sedating effect of some antihistamines is used to treat insomnia and difficulties in falling asleep. Some antihistamines also help inhibit **nausea and vomiting** and reduce **motion sickness**.

Commonly used antihistamines include the following:

- diphenhydramine (Benadryl)
- loratadine (Claritin)
- cetirizine (Zyrtec)
- fexofenadine (Allegra)
- clemastine fumarate (Tavist)
- chlorpheniramine (Chlor Trimeton)
- brompheniramine (Dimetapp)

Precautions

Some antihistamines produce drowsiness, although clinical studies have shown that children are less susceptible to antihistamine-induced drowsiness than adults. Some nonsedating antihistamines can act as stimulants in children and produce hyperactivity and sleeplessness.

Children with certain medical conditions may not be able to take antihistamines. The following are absolute or relative contraindications to use of antihistamines. The significance of the contraindication will vary with the drug and dose.

- glaucoma
- hyperthyroidism (overactive thyroid)
- high blood pressure
- heart disease
- ulcers or other stomach problems
- stomach or intestinal blockage
- liver disease
- kidney disease
- bladder obstruction
- diabetes

Side effects

The frequency and severity of adverse effects will vary depending on the antihistamine.

Central nervous system reactions include drowsiness, sedation, **dizziness**, faintness, disturbed coordination, lassitude, confusion, restlessness, excitation, tremor, seizures, **headache**, insomnia, euphoria, blurred vision, hallucinations, disorientation, disturbing dreams/nightmares, schizophrenic-like reactions, weakness, vertigo, nerve **pain**, and convulsions.

Gastrointestinal problems include increased appetite, decreased appetite, **nausea**, **vomiting**, **diarrhea**, and **constipation**.

Hematologic reactions are rare but may be severe. These include anemia, or breakdown of red blood cells; reduced platelets; reduced white cells; and bone marrow failure.

A large number of additional reactions have been reported. Not all apply to every drug, and some reactions may not be drug related. Some of the other adverse effects are chest tightness; wheezing; nasal stuffiness;

KEY TERMS

Allergen—A foreign substance that provokes an immune reaction or allergic response in some sensitive people but not in most others.

Anaphylaxis—Also called anaphylactic shock; a severe allergic reaction characterized by airway constriction, tissue swelling, and lowered blood pressure.

Histamine—A substance released by immune system cells in response to the presence of an allergen. It stimulates widening of blood vessels and increased porousness of blood vessel walls so that fluid and protein leak out from the blood into the surrounding tissue, causing localised inflammation of the tissue.

Mast cells—A type of immune system cell that is found in the lining of the nasal passages and eyelids. It displays a type of antibody called immunoglobulin type E (IgE) on its cell surface and participates in the allergic response by releasing histamine from intracellular granules.

dry mouth, nose, and throat; **sore throat**; respiratory depression; sneezing; and a burning sensation in the nose.

Interactions

Drug interactions vary with the chemical class of antihistamine. In general, antihistamines increase the effects of other sedatives, including alcohol.

Monoamine oxidase inhibitor **antidepressants** may prolong and increase the effects of some antihistamines.

Parental concerns

For children who resist taking pills, many antihistamines are available as flavored chewable tablets, tablets that easily dissolve on the tongue, and in flavored syrups. Because many over-the-counter allergy medicines contain multiple drugs, parents should be sure to read the prescribing and dosage information for any antihistamine their children are taking to ensure safe use.

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Jennifer E. Sisk, MA

Antisocial behavior

Definition

Antisocial behaviors are disruptive acts characterized by covert and overt hostility and intentional aggression toward others. Antisocial behaviors exist along a severity continuum and include repeated violations of social rules, defiance of authority and of the rights of others, deceitfulness, theft, and reckless disregard for self and others. Antisocial behavior can be identified in children as young as three or four years of age. If left unchecked these coercive behavior patterns will persist and escalate in severity over time, becoming a chronic behavioral disorder.

Description

Antisocial behavior may be overt, involving aggressive actions against siblings, peers, parents, teachers, or other adults, such as verbal abuse, bullying and hitting; or covert, involving aggressive actions against property, such as theft, vandalism, and fire-setting. Covert antisocial behaviors in early childhood may include noncompliance, sneaking, **lying**, or secretly destroying another's property. Antisocial behaviors also include drug and alcohol abuse and high-risk activities involving self and others.

Demographics

Between 4 and 6 million American children have been identified with antisocial behavior problems. These disruptive behaviors are one of the most common forms

of psychopathology, accounting for half of all childhood mental health referrals.

Gender differences in antisocial behavior patterns are evident as early as age three or four. There has been far less research into the nature and development pattern of antisocial behavior in girls. Pre-adolescent boys are far more likely to engage in overtly aggressive antisocial behaviors than girls. Boys exhibit more physical and verbal aggression, whereas antisocial behavior in girls is more indirect and relational, involving harmful social manipulation of others. The gender differences in the way antisocial behavior is expressed may be related to the differing rate of maturity between girls and boys. Physical aggression is expressed at the earliest stages of development, then direct verbal threats, and, last, indirect strategies for manipulating the existing social structure.

Antisocial behaviors may have an early onset, identifiable as soon as age four, or late onset, manifesting in middle or late **adolescence**. Some research indicates that girls are more likely than boys to exhibit late onset antisocial behavior. Late onset antisocial behaviors are less persistent and more likely to be discarded as a behavioral strategy than those that first appear in early childhood.

As many as half of all elementary school children who demonstrate antisocial behavior patterns continue these behaviors into adolescence, and as many as 75 percent of adolescents who demonstrate antisocial behaviors continue to do so into early adulthood.

Causes and symptoms

Antisocial behavior develops and is shaped in the context of coercive social interactions within the **family**, community, and educational environment. It is also influenced by the child's **temperament** and irritability, cognitive ability, the level of involvement with deviant peers, exposure to violence, and deficit of cooperative problem-solving skills. Antisocial behavior is frequently accompanied by other behavioral and developmental problems such as hyperactivity, depression, learning disabilities, and impulsivity.

Multiple risk factors for development and persistence of antisocial behaviors include genetic, neurobiological, and environmental stressors beginning at the prenatal stage and often continuing throughout the childhood years.

Genetic factors are thought to contribute substantially to the development of antisocial behaviors. Genetic factors, including abnormalities in the structure of the prefrontal cortex of the brain, may play a role in an inherited predisposition to antisocial behaviors.

Neurobiological risks include maternal drug use during pregnancy, birth complications, low birth weight, prenatal brain damage, traumatic **head injury**, and chronic illness.

High-risk factors in the family setting include the following:

- parental history of antisocial behaviors
- parental alcohol and drug abuse
- chaotic and unstable home life
- absence of good parenting skills
- use of coercive and corporal punishment
- parental disruption due to **divorce**, death, or other separation
- parental psychiatric disorders, especially maternal depression
- economic distress due to poverty and unemployment

Heavy exposure to media violence through television, movies, Internet sites, **video games**, and even cartoons has long been associated with an increase in the likelihood that a child will become desensitized to violence and behave in aggressive and antisocial ways. However, research relating the use of violent video games with antisocial behavior is inconsistent and varies in design and quality, with findings of both increased and decreased aggression after exposure to violent video games.

Companions and peers are influential in the development of antisocial behaviors. Some studies of boys with antisocial behaviors have found that companions are mutually reinforcing with their talk of rule breaking in ways that predict later delinquency and substance abuse.

When to call the doctor

Parents and teachers who notice a pattern of repeated lying, cheating, **stealing**, bullying, hitting, noncompliance, and other disruptive behaviors should not ignore these symptoms. Early screening of at-risk children is critical to deterring development of a persistent pattern of antisocial behavior. Early detection and appropriate intervention, particularly during the **preschool** years and middleschool years, is the best means of interrupting the developmental trajectory of antisocial behavior patterns. Serious childhood antisocial behaviors can lead to diagnoses of **conduct disorder** (CD) or **oppositional defiant disorder** (ODD). Children who exhibit

antisocial behaviors are at an increased risk for alcohol use disorders (AUDs).

Diagnosis

Systematic diagnostic interviews with parents and children provide opportunity for a thorough assessment of individual risk factors and family and societal dynamics. Such assessment should include parent-adolescent relationships; peer characteristics; school, home, and community environment; and overall health of the individual.

Various diagnostic instruments have been developed for evidence-based identification of antisocial behavior in children. The onset, frequency, and severity of antisocial behaviors such as stealing, lying, cheating, sneaking, peer rejection, low academic achievement, negative attitude, and aggressive behaviors are accessed to determine appropriate intervention and treatment.

Treatment

Enhanced parent-teacher communications and the availability of school psychologists and counselors trained in family intervention within the school setting are basic requirements for successful intervention and treatment of childhood antisocial behaviors.

School-based programs from early childhood onward that teach conflict resolution, emotional literacy, and anger management skills have been shown to interrupt the development of antisocial behavior in low-risk students. Students who may be at higher risk because of difficult family and environmental circumstances will benefit from more individualized prevention efforts, including counseling, academic support, social-skills training, and behavior contracting.

Academic settings with the capacity to deliver professional parental support and provide feedback in a motivating way can help parents to develop and hone effective parenting skills that may interrupt further progression of antisocial behavior patterns in their children. Access to written and video information on parenting skills and information about community family resources, as well as promotion of parent-support groups, are effective intervention strategies for changing family dynamics that shape antisocial behavior in the children.

Older students who already exhibit a persistent pattern of antisocial behavior can be helped with intensive individualized services that may involve community mental health agencies and other outside intervention.

KEY TERMS

Alcohol use disorder (AUD)—The repetitive, long-term ingestion of alcohol in ways that impair psychosocial functioning and health, leading to problems with personal relationships, school, or work. Alcohol use disorders include alcohol dependence, alcohol abuse, alcohol intoxication, and alcohol withdrawal.

Coercive behavior—Maladaptive behaviors engaged in as a means of avoiding or escaping aversive events. Coercive behavior may include whining, noncompliance, and lying.

Conduct disorder—A behavioral and emotional disorder of childhood and adolescence. Children with a conduct disorder act inappropriately, infringe on the rights of others, and violate societal norms.

Oppositional defiant disorder (ODD)—A persistent disruptive behavior that includes three or more of the following types of antisocial behaviors occurring frequently over a six-month period: loss of temper; arguments with adults; defiance or refusal to comply with adult's requests/rules; annoying others deliberately and being easily annoyed;, blaming others with unwillingness to accept responsibility for mistakes or behavior; angry, resentful, spiteful, and vindictive behaviors.

Community-based programs, including youth centers and recreational programs with trained therapists, can provide additional support for at-risk children.

Prognosis

The longer antisocial behavior patterns persist, the more intractable they become. Early-onset conduct problems left untreated are more likely to result in the development of chronic antisocial behavior than if the disruptive behavior begins in adolescence. Though it is never too late to intervene, researchers warn that if by age eight a child has not learned ways other than coercion to meet his social goals, he has a high chance of continuing with antisocial behavior throughout his lifetime.

Longitudinal studies have found that as many as 71 percent of chronic juvenile offenders had progressed from childhood antisocial behaviors through a history of early arrests to a pattern of chronic law breaking.

Prevention

Healthy **nutrition** and prenatal care, a safe and secure family and social environment, early **bonding** with an emotionally mature and healthy parent, role models for prosocial behaviors, non-coercive methods of parenting, peer relationships with prosocial individuals, and early intervention when problems first appear are all excellent means of assuring development of prosocial behaviors and reducing and extinguishing antisocial behaviors in children.

Parental concerns

Parents may hesitate to seek help for children with antisocial behavior patterns out of **fear** of the child being negatively labeled or misdiagnosed. Almost all children will engage in some form of antisocial behavior at various stages of development. Skilled parents will be able to lovingly confront the child and help the child recognize that certain behaviors are unacceptable. However if these conduct disturbances persist and worsen, they should be taken seriously as precursors to more serious problems. Early intervention is important for the sake of the child and the entire family system.

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Clare Hanrahan

Antisocial personality disorder

Definition

Antisocial behavior is that which is verbally or physically harmful to other people, animals, or property, including behavior that severely violates social expectations for a given environment. Antisocial personality disorder in adults is also referred to as sociopathy or psychopathy.

Description

Antisocial behavior can be broken down into two components: the presence of antisocial (i.e., angry, aggressive, or disobedient) behavior and the absence of prosocial (i.e., communicative, affirming, or cooperative) behavior. Most children exhibit some antisocial behavior during their development, and different children demonstrate varying levels of prosocial and antisocial behavior. Some children—for example, the popular but rebellious child—may exhibit high levels of both antisocial and prosocial behaviors. Others—for example, the withdrawn, thoughtful child—may exhibit low levels of both types of behaviors.

High levels of antisocial behavior are considered a clinical disorder. Young children may exhibit hostility towards authority, and be diagnosed with oppositional-defiant disorder. Older children may lie, steal, or engage in violent behaviors, and be diagnosed with **conduct disorder**. A minority of children with conduct disorder whose behavior does not improve as they mature will go on to develop adult antisocial personality disorder.

A salient characteristic of antisocial children and adolescents is that they appear to have no feelings. They demonstrate no care for others' feelings or remorse for hurting others, and tend not to show their own feelings except for anger and hostility, and even these are communicated through aggressive acts and are not necessarily expressed through affect. One analysis of antisocial behavior is that it is a defense mechanism that helps children avoid painful feelings, or avoid the **anxiety** caused by lack of control over the environment.

Antisocial behavior may also be a direct attempt to alter the environment. Social learning theory suggests that negative behaviors are reinforced during childhood by parents, caregivers, or peers. In one formulation, a child's negative behavior (e.g., whining, hitting) initially serves to stop the parent from behaving in ways that are aversive to the child (the parent may be fighting with a partner, yelling at a sibling, or even crying). The child will apply the learned behavior at school, and a vicious

cycle sets in: he or she is rejected, becomes angry and attempts to force his will or assert his pride, and is then further rejected by the very peers from whom he might learn more positive behaviors. As the child matures, "mutual avoidance" sets in with the parent(s), as each party avoids the negative behaviors of the other. Consequently, the child receives little care or supervision and, especially during **adolescence**, is free to join peers who have similarly learned antisocial means of expression.

Demographics

Mental health professionals agree, and rising rates of serious school disciplinary problems, delinquency, and violent crime indicate, that antisocial behavior in general is increasing. Thirty to 70% of childhood psychiatric admissions are for disruptive behavior disorders, and diagnoses of behavior disorders are increasing overall. A small percentage of antisocial children (about 3% of males and 1% of females) grow up to become adults with antisocial personality disorder, and a greater proportion suffer from the social, academic, and occupational failures resulting from their antisocial behavior.

Causes and symptoms

Factors that contribute to a particular child's antisocial behavior vary, but they usually include some form of **family** problems (e.g., marital discord, harsh or inconsistent disciplinary practices or actual **child abuse**, frequent changes in primary caregiver or in housing, learning or cognitive disabilities, or health problems). Attention deficit/hyperactivity disorder is highly correlated with antisocial behavior.

A child may exhibit antisocial behavior in response to a specific stressor (such as the death of a parent or a **divorce**) for a limited period of time, but this is not considered a psychiatric condition. Children and adolescents with antisocial behavior problems have an increased risk of accidents, school failure, early alcohol and substance use, **suicide**, and criminal behavior. The elements of a moderate to severely antisocial personality are established as early as kindergarten. Antisocial children score high on traits of impulsiveness, but low on anxiety and reward-dependence—the degree to which they value, and are motivated by, approval from others. Yet underneath their tough exterior, antisocial children have low **self-esteem**.

Although antisocial personality disorder is only diagnosed in people over age 18, the symptoms are similar to those of conduct disorder, and the criteria for diagnosis include the onset of conduct disorder before the age of 15. According to the *Diagnostic and Statistical Manual of Mental Disorders, 4th Edition Text Revision (DSM-IV-TR)*, people with antisocial personality disorder demonstrate a pattern of antisocial behavior since age 15.

The adult with antisocial personality disorder displays at least three of the following behaviors:

- fails to conform to social norms, as indicated by frequently performing illegal acts, and pursuing illegal occupations
- is deceitful and manipulative of others, often in order to obtain money, sex, or drugs
- is impulsive, holding a succession of jobs or residences
- is irritable or aggressive, engaging in physical fights
- exhibits reckless disregard for the **safety** of self or others, misusing motor vehicles, or playing with fire
- is consistently irresponsible, failing to find or sustain work or to pay bills and debts
- demonstrates lack of remorse for the harm his or her behavior causes others

An adult diagnosed with antisocial personality disorder will demonstrate few of his or her own feelings beyond contempt for others. Authorities have linked antisocial personality disorder with abuse, either physical or sexual, during childhood, neurological disorders (which are often undiagnosed), and low IQ. Those with a parent with an antisocial personality disorder or substance abuse problem are more likely to develop the disorder. The antisocially disordered person may be poverty-stricken, homeless, a substance abuser, or have an extensive criminal record. Antisocial personality disorder is associated with low socioeconomic status and urban settings.

When to call the doctor

When symptoms of antisocial behavior appear, a child should be taken to his or her health care provider as soon as possible for evaluation and possible referral to a mental health care professional. If a child or teen reveals at any time that he/she has had recent thoughts of self-injury or suicide, or if he/she demonstrates behavior that compromises personal safety or the safety of others, professional assistance from a mental health care provider or care facility should be sought immediately.

Diagnosis

Antisocial behavior and childhood antisocial disorders such as conduct disorder may be diagnosed by a family physician or pediatrician, social worker, school counselor, psychiatrist, or psychologist. A comprehen-

sive evaluation of the child should ideally include interviews with the child and parents, a full social and medical history, review of educational records, a cognitive evaluation, and a psychiatric exam.

One or more clinical inventories or scales may be used to assess the child, including the Youth Self-Report, the School Social Behavior Scales (SSBS), the Overt Aggression Scale (OAS), Behavioral **Assessment** System for Children (BASC), Child Behavior Checklist (CBCL), the Nisonger Child Behavior Rating Form (N-CBRF), Clinical Global Impressions scale (CGI), and Diagnostic Interview Schedule for Children (DISC). The tests are verbal and/or written and are administered in both hospital and outpatient settings.

Treatment

The most important goals of treating antisocial behavior are to measure and describe the individual child's or adolescent's actual problem behaviors and to effectively teach him or her the positive behaviors that should be adopted instead. In severe cases, medication will be administered to control behavior, but it should not be used as a substitute for therapy. A child who experiences explosive rage may respond well to medication. Ideally, an interdisciplinary team of teachers, social workers, and guidance counselors will work with parents or caregivers to provide services to help the child in all aspects of his or her life: home, school, work, and social contexts. In many cases, parents themselves need intensive training on modeling and reinforcing appropriate behaviors in their child, as well as in providing appropriate **discipline** to prevent inappropriate behavior.

A variety of methods may be employed to deliver social skills training, but especially with diagnosed antisocial disorders, the most effective methods are systemic therapies which address **communication skills** among the whole family or within a peer group of other antisocial children or adolescents. These probably work best because they entail actually developing (or redeveloping) positive relationships between the child or adolescent and other people. Methods used in social skills training include modeling, role-playing, corrective feedback, and token reinforcement systems. Regardless of the method used, the child's level of cognitive and emotional development often determines the success of treatment. Adolescents capable of learning communication and problem-solving skills are more likely to improve their relations with others.

Unfortunately, conduct disorders, which are the primary form of diagnosed antisocial behavior, are highly resistant to treatment. Few institutions can afford the comprehensiveness and intensity of services required to support and change a child's whole system of behavior. In most cases, for various reasons, treatment is terminated (usually by the client) long before it is completed. Often, the child may be fortunate to be diagnosed at all. Schools are frequently the first to address behavior problems, and regular classroom teachers only spend a limited amount of time with individual students. **Special education** teachers and counselors have a better chance at instituting long-term treatment programs—if the student stays in the same school for a period of years. One study showed teenage boys with conduct disorder had had an average of nine years of treatment by 15 different institutions. Treatments averaged seven months each.

Studies show that children who are given social skills instruction decrease their antisocial behavior, especially when the instruction is combined with some form of supportive peer group or **family therapy**. But the long-term effectiveness of any form of therapy for antisocial behavior has not been demonstrated. The fact that peer groups have such a strong influence on behavior suggests that schools that employ collaborative learning and the mainstreaming of antisocial students with regular students may prove most beneficial to the antisocial child. Because the classroom is a natural environment, learned skills do not need to be transferred. By dividing the classroom into groups and explicitly stating procedures for group interactions, teachers can create opportunities for positive interaction between antisocial and other students.

Prognosis

Early and intensive intervention is the best hope for children exhibiting antisocial behaviors or diagnosed conduct disorder. For those who grow into adults with antisocial personality disorder, the prognosis is not promising; the condition is difficult to treat and tends to be chronic. Although there are medications available that could quell some of the symptoms of antisocial personality disorder, noncompliance or abuse of the drugs prevents their widespread use. The most successful treatment programs are long-term, structured residential settings in which the patient systematically earns privileges as he or she modifies behavior.

Prevention

A supportive, nurturing, and structured home environment is believed to be the best defense against antisocial behavioral problems. Children with learning disabilities and/or difficulties in school should get appropriate academic assistance. Addressing these problems

KEY TERMS

Attention deficit hyperactivity disorder (ADHD)—A condition in which a person (usually a child) has an unusually high activity level and a short attention span. People with the disorder may act impulsively and may have learning and behavioral problems.

Major depressive disorder—A mood disorder characterized by profound feelings of sadness or despair.

Modeling—A type of teaching method used in social skills training. Therapists who use this method may offer positive and negative examples of the behaviors that make up a social skill.

Prosocial behavior—Actions that promote communication, cooperation, and other positive interactions with peers and family members.

when they first appear helps to prevent the frustration and low self-esteem that may lead to antisocial issues later.

Parental concerns

A child with antisocial behavioral problems can have a tremendous impact on the home environment and on the physical and emotional welfare of siblings and others sharing the household, as well as their peers at school. While seeking help for their child, parents must remain sensitive to the needs of their other children. This may mean avoiding leaving siblings alone together, getting assistance with childcare, or even seeking residential or hospital treatment for the child if the safety and well-being of other family members is in jeopardy. Parents should also maintain an open dialog with their child's teachers to ensure that their child receives appropriate educational assistance and that classmates are not put at risk.

See also Aggression; Conduct disorder; Oppositional defiant disorder.

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Paula Ford-Martin

Antiviral drugs

Definition

Antiviral drugs act against diseases caused by viruses.

Description

Viruses represent a large group of infective agents that are composed of a core of nucleic acids, either RNA or DNA, surrounded by a layer of protein. They are not really living organisms according to general understanding, since they lack the cell membrane that is associated with living cells. Viruses can reproduce only inside a living cell, and they cause many diseases. Viruses are not normally affected by **antibiotics** but a small number of viruses can either be destroyed or have their growth stopped by drugs.

The drugs as of 2004 available for treatment of viral diseases in children are:

- Acyclovir (Zovirax), used for treatment of diseases caused by the **herpes simplex** virus and herpes zoster virus. Although it is approved only for children over the age of six months, the drug has been used for newborn infants with **encephalitis**. This drug is most reliable when given intravenously.

- Amantidine (Symmetrel), used to prevent or treat infections of the **influenza** virus type A. It is recommended for patients who cannot or should not receive influenza virus vaccine. As of 2004 it has not been studied in children below the age of one year.
- Foscarnet (Foscavir), is not recommended for young children but may be given to adolescents. It is used to treat cytomegalovirus infections of the eye, and for herpes simplex infections that are resistant to other drugs.
- Ganciclovir (Cytovene), used to treat cytomegalovirus infections of the eye. Although the manufacturer does not recommend use of ganciclovir in patients below the age of 12 years, the drug is recommended by standard pediatric references for children as young as three months.
- Oseltamivir (Tamiflu), used for treatment of influenza virus infections of children over the age of 13 years. In adults, oseltamivir has also been used for prevention if influenza, but this use has not been studied in children.
- Ribavirin (Rebetol, Virazol), used for treatment of hospitalized infants and young children with severe lower respiratory tract infections caused by respiratory syncytial virus (RSV), but its value is controversial.
- Rimantidine (Flumadine), used to protect against the influenza virus type A.
- Valacyclovir (Valtrex), used for treatment of diseases caused by the herpes simplex virus and herpes zoster virus. This drug is converted to acyclovir inside the body and is more reliable for oral use. Although the manufacturer says that safety and efficacy in children have not been established, valacyclovir is recommended for use in standard pediatric resources.
- Vidarabine (Vira-A), used to treat severe herpes infections in the newborn, but its primary value is in the form of an eye ointment to treat herpes infections of the eye.
- Zanamivir (relenza), used to treat influenza infections caused by viruses types A and B in adults and children over the age of seven.

In addition to the above drugs, there are drugs which treat retrovirus infections. Retroviruses are composed of RNA molecules instead of DNA, and the only treatable one is the one that causes acquired immune deficiency syndrome (**AIDS**). The drugs in this group that are appropriate for treatment of children are as follows:

- abacavir (Ziagen)
- amprenavir (Agenerase), for children above the age of four
- didanosine (Videx)
- efavirenz (Sustiva), for children over the age of three
- indinavir (Crixavan), according to the manufacturer safety and efficacy of which in children has not been established, but the drug has been recommended in standard pediatric references
- lamivudine (Epivir), for treatment of **hepatitis B** as well as for AIDS
- lopinavir/Ritonavir fixed combination (Kaletra), used in children as young as six months
- stavudine (Zerit)
- nelfinavir (Viracept), the manufacturer of which does not recommend use of this drug for children younger than two, but it has been studied with some success in children as young as newborns
- ritonavir (Norvir)
- saquinavir (Fortovase, Invirase)
- zalcitabine (Hivid)
- zidovudine (Retrovir)

Other drugs for treatment of HIV disease are marketed, but there have been neither sufficient studies not clinical experience to recommend their use in children.

General use

The antiviral drugs are used to prevent or treat the diseases listed above. These drugs are specific for individual viruses and offer no benefit for conditions caused by other viruses.

Precautions

Each of the drugs listed has specific warnings. See specific drugs references or ask a pediatrician.

Side effects

Each of the drugs listed has its own side effects. See specific drugs references or ask a pediatrician.

Indinavir (Crixivan) has the unique adverse effects of causing changes in patterns of fat distribution. This has been called Crix belly and may be more distressing to the patient than more serious side effects caused by other drugs since these effects are clearly visible. As of 2004 it is not clear whether this effect can be reversed when the drug is discontinued. Antiretroviral drugs should not be discontinued unless there is an alternative antiretroviral regimen to adopt.

KEY TERMS

Herpes virus—A family of viruses including herpes simplex types 1 and 2, and herpes zoster (also called varicella zoster). Herpes viruses cause several infections, all characterized by blisters and ulcers, including chickenpox, shingles, genital herpes, and cold sores or fever blisters.

Influenza virus type—The nature of the proteins in the outer coat of an influenza virus. Depending on the proteins, influenza viruses may be classified as A, B, or C.

Retrovirus—A family of RNA viruses containing a reverse transcriptase enzyme that allows the viruses' genetic information to become part of the genetic information of the host cell upon replication. Human immunodeficiency virus (HIV) is a retrovirus.

Virus—A small infectious agent consisting of a core of genetic material (DNA or RNA) surrounded by a shell of protein. A virus needs a living cell to reproduce.

Interactions

See specific drugs references or ask a pediatrician about interactions for an antiviral drug that has been prescribed.

Patients should use these drugs exactly as directed. With regard to the AIDS drugs in particular, the drugs should not be discontinued without consultation with the prescriber. AIDS drugs are normally prescribed in combinations of two and three drugs used together, and discontinuing any single drug may lead to the virus developing resistance to the other agents.

Parental concerns

Liquid dosage forms must always be measured with a calibrated teaspoon or dropper, never with a household teaspoon. Household teaspoons vary in the volume they deliver and may result in inadvertent overdose or under dose.

Anti-influenza drugs should be used only for patients who cannot receive vaccinations. Annual **vaccination** remains the preferred method of preventing influenza.

Antiretroviral drugs are routinely given in combinations of three to four drugs at a time. In some cases, fixed combinations of medications are the most practical way to administer these drugs, since they require the lowest number of doses each day.

Some antiviral drugs, particularly the antiretroviral agents, have potentially severe adverse effects. They should be prescribed only by qualified professionals experienced in their use. These drugs must be routinely monitored. Regular laboratory testing is essential for safe and effective use. Adverse effects and side effects must be reported to the prescriber as soon as they are observed.

Antiherpetic drugs may have only a limited value in reducing the severity or duration of herpes attacks. They are more important for their effect in reducing the period of viral shedding, the period of time in which a person infected with herpes virus can infect other people. For this reason, continued use of the drugs is important to **family** members and those in close proximity to the patient. The drugs should not be discontinued, even if there is no observed benefit.

See also Herpes simplex; HIV infection and AIDS; Influenza.

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Samuel Uretsky, PharmD

Anxiety

Definition

Anxiety is a condition of persistent and uncontrollable nervousness, stress, and worry that is triggered by anticipation of future events, memories of past events, or ruminations over day-to-day events, both trivial and major, with disproportionate fears of catastrophic consequences.

Description

Stimulated by real or imagined dangers, anxiety affects people of all ages and social backgrounds. When it occurs in unrealistic situations or with unusual intensity, it can disrupt everyday life. Some researchers believe anxiety is synonymous with **fear**, occurring in varying degrees and in situations in which people feel threatened by some danger. Others describe anxiety as an unpleasant emotion caused by unidentifiable dangers or dangers that, in reality, pose no threat. Unlike fear, which is caused by realistic, known dangers, anxiety can be more difficult to identify and alleviate.

A small amount of anxiety is normal in the developing child, especially among adolescents and teens. Anxiety is often a realistic response to new roles and responsibilities, as well as to sexual and identity development. When symptoms become extreme, disabling, and/or when children or adolescents experience several symptoms over a period of a month or more, these symptoms may be a sign of an anxiety disorder, and professional intervention may be necessary. Two common forms of childhood anxiety are general anxiety disorder (GAD) and **separation anxiety** disorder (SAD), although many physicians and psychologists also include panic disorder and **obsessive-compulsive disorder**, which tend to occur more frequently in adults. Anxiety that is the result of experiencing a violent event, disaster, or physical abuse is identified as post-traumatic stress disorder (PTSD). Most adult anxiety disorders begin in **adolescence** or young adulthood and are more common among women than men.

Demographics

According to the U.S. surgeon general, 13 percent, or over 6 million children, suffer from anxiety, making it the most common emotional problem in children. Among adolescents, more girls than boys are affected. About half of the children and adolescents with anxiety disorders also have a second anxiety disorder or other mental or behavioral disorder, such as depression.

Causes and symptoms

A child's genetics, biochemistry, environment, history, and psychological profile all seem to contribute to the development of anxiety disorders. Most children with these disorders seem to have a biological vulnerability to stress, making them more susceptible to environmental stimuli than the rest of the population.

Emotional and behavioral symptoms of anxiety disorders include tension; self-consciousness; new or recurring fears (such as fear of the dark, fear of being alone, or fear of strangers); self-doubt and questioning; crying and whining; worries; constant need for reassurance (clinging to parent and unwilling to let the parent out of sight); distractibility; decreased appetite or other changes in eating habits; inability to control emotions; feeling as if one is about to have a heart attack, die, or go insane;

nightmares; irritability, stubbornness, and anger; regression to behaviors that are typical of an earlier developmental stage; and unwillingness to participate in **family** and school activities. Physical symptoms include rapid heartbeat; sweating; trembling; muscle aches (from tension); dry mouth; **headache**; stomach distress; **diarrhea**; **constipation**; frequent urination; new or recurrent bedwetting; **stuttering**; hot flashes or chills; throat constriction (lump in the throat); **sleep** disturbances; and fatigue. Many of these anxiety symptoms are very similar to those of depression, and as many as 50 percent of children with anxiety also suffer from depression. Generally, physiological hyperarousal (excitedness, shortness of breath, the fight or flight response) characterizes anxiety disorders, whereas underarousal (lack of pleasure and feelings of guilt) characterizes depression. Other signs of anxiety problems are poor school performance, loss of interest in previously enjoyed activities, obsession about appearance or weight, social **phobias** (e.g., fear of walking into a room full of people), and the persistence of imaginary fears after ages six to eight. Children with anxiety disorders are often perfectionists and are concerned about "getting everything right," but rarely feel that their work is satisfactory.

Shyness does not necessarily indicate a disorder, unless it interferes with normal activities and occurs with other symptoms. A small proportion of children do experience social anxiety, incapacitating shyness that persists for months or more, which should be treated. Similarly, performance anxiety experienced before athletic, academic, or theatrical events does not indicate a disorder, unless it significantly interferes with the activity.

Separation anxiety disorder (SAD) is the most common anxiety disorder among children, affecting 2 to 3 percent of school-aged children. SAD involves extreme and disproportionate distress over day-to-day separation from parents or home and unrealistic fears of harm to self or loved ones. Approximately 75 to 85 percent of children who refuse to go to school have separation anxiety. Normal separation fears are outgrown by children by the ages of five or six, but SAD usually starts between the ages of seven and 11.

When to call the doctor

A qualified mental health professional should be consulted if a child's anxiety begins to affect his or her ability to perform the three main responsibilities of childhood: to learn, to make friends, and to have fun. Often fears and anxieties come and go with time and age. However, in some children, anxiety becomes severe, excessive, unreasonable, and long-lasting (usually considered as long-lasting if the child experiences the elevated level of anxiety for a month or more), interferes with the child's ability to function normally, and causes the child to be distraught and easily upset, thus necessitating professional intervention.

Diagnosis

Diagnosing children with an anxiety disorder can be very difficult, since anxiety often results in disruptive behaviors that overlap with other disorders such as attention-deficit hyperactivity. Children showing signs of an anxiety disorder should first get a physical exam to rule out any possible illness or physical problem. Diagnosis of normal versus abnormal anxiety depends largely upon the degree of distress and its effect on a child's functioning. The degree of abnormality must be gauged within the context of the child's age and developmental level. The specific anxiety disorder is diagnosed by the pattern and intensity of symptoms using various psychological diagnostic tools.

Treatment

Depending on the severity of the problem, treatments for anxiety include school counseling, **family therapy**, and cognitive-behavioral or dynamic psychotherapy, sometimes combined with antianxiety drugs. Therapies generally aim for support by providing a positive, entirely accepting, pressure-free environment in which to explore problems; by providing insight through discovering and working with the child or adolescent's underlying thoughts and beliefs; and by exposure through gradually reintroducing the anxiety-producing thoughts, people, situations, or events in a manner so as to confront them calmly. Relaxation techniques, including meditation, may be employed in order to control the symptoms of physiological arousal and provide a tool the child can use to control his or her response.

Creative visualization, sometimes called rehearsal imagery by actors and athletes, may also be used. In this technique, the child writes down (or draws pictures of) each detail of the anxiety-producing event or situation and imagines his or her movements in performing the activity. The child also learns to perform these techniques in new, unanticipated situations.

In severe cases of diagnosed anxiety disorders, antianxiety and/or antidepressant drugs may be prescribed in order to enable therapy and normal daily activities to continue. Previously, narcotics and other sedatives, drugs that are highly addictive and interfere with cognitive capacity, were prescribed. With pharmacological advances and the development of synthetic drugs, which act in specific ways on brain chemicals, a more refined

set of antianxiety drugs became available. Studies have found that generalized anxiety responds well to these drugs (benxodiazepines are the most common), which serve to quell the physiological symptoms of anxiety. Other forms of anxiety such as panic attacks, in which the symptoms occur in isolated episodes and are predominantly physical (and the object of fear is vague, fantastic, or unknown), respond best to the antidepressant drugs. Childhood separation anxiety is thought to be included in this category. Psychoactive drugs should only be considered as a last treatment alternative, and extra caution should be used when they are prescribed for children.

Prognosis

Studies consistently report that anxiety disorders can be debilitating and impinge seriously on a person's quality of life. Despite their common occurrence, little is understood about the natural course of anxiety disorder. Adults experiencing anxiety disorders often report that they have felt anxious all of their lives, with one half of adults with general anxiety disorder reporting that the onset of the condition occurred during childhood or adolescence. Anxiety disorders can be chronic, and the severity of symptoms can fluctuate significantly, with symptoms being more severe when stressors are present. Without treatment, extended periods of remission are not likely.

Prevention

Parents can help their child respond to stress by taking the following steps:

- providing a safe, secure, familiar, and consistent home life
- being selective in the types of television programs that children watch (including news shows), which can produce fears and anxieties
- spending calm and relaxed time with their child
- encouraging questions and expressions of fears, worries, or concerns
- listening to the child with encouragement and affection and without being critical
- rewarding (and not punishing) the child for effort rather than success
- providing the child with opportunities to make choices; with more control over situations, the child has a better response to stress
- involving the child in activities in which he or she can succeed and limiting events and situations that are stressful for the child

KEY TERMS

Psychological—Pertaining to the mind, its mental processes, and its emotional makeup.

Psychotherapy—Psychological counseling that seeks to determine the underlying causes of a patient's depression. The form of this counseling may be cognitive/behavioral, interpersonal, or psychodynamic.

Shyness—The feeling of insecurity when among people, talking with people, or asking somebody a favor.

Stress—A physical and psychological response that results from being exposed to a demand or pressure.

- developing an awareness of the situations and activities that are stressful for the child and recognizing signs of stress in the child
- keeping the child informed of necessary and anticipated changes (e.g., moving, change of school) that may cause the child to be stressed
- seeking professional help or advice when the symptoms of stress do not decrease or disappear

The child should also be encouraged to use various techniques to reduce stress, including the following strategies:

- talking about problems to parents or others whom the child trusts
- relaxing by listening to music, taking a warm bath, meditating, practicing breathing exercises, or participating in a favorite hobby or activity
- exercising
- respecting themselves and others
- avoiding the use of drugs and alcohol
- feeling free to ask for help if he or she is having difficulties with stress management

Parental concerns

Parenting an anxious child is difficult and can create stress within the entire family. Parents need to help the child learn and apply techniques to manage his or her anxiety. The use of support groups and professional assistance is recommended.

Parents of children with anxiety disorders may exhibit anxiety symptoms themselves and should also seek professional assistance.

See also Fear; Separation anxiety.

Resources

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Apgar testing

Definition

The Apgar scoring system evaluates the physical condition of the newborn at one minute after birth and again at five minutes after birth. The newborn receives a total score (Apgar score) that ranges from 0 to 10 based on rating color, heart rate, respiratory effort, muscle tone, and reflex irritability.

Purpose

Virginia Apgar specialized in anesthesiology and **childbirth**. She developed the Newborn Scoring System, later called the Apgar score, in 1949 for practitioners to use in deciding whether or not a newborn needed resuscitation. This score provides a uniform method of observation and evaluation of a newborn infant's need for resuscitation immediately after delivery at one minute and again at five minutes. The score is significant because one person in the delivery room evaluates the infant using five signs in an objective, standard and measurable manner. Research published in *The New England Journal of Medicine* in 2001 concluded that the Apgar scoring system remains as relevant for the prediction of neonatal survival in the early 2000s as it was in 1949.

Description

Five factors are considered in the evaluation of a newborn and the word Apgar can be used as a mnemonic to remember them, i.e., A = Activity (or muscle tone); P = Pulse; G = Grimace (or reflexes to stimuli); A = Appearance (or skin color), and R = Respiration. Scores are given as follows:

- Activity: Limpness, no movement at all = 0; some flexion of the limbs = 1; active movement, vigorous movements of arms and legs = 2.
- Pulse: No pulse = 0; pulse below 100 beats per minute (bpm) = 1; pulse over 100 per minute = 2. This is the most important assessment and can be determined by auscultation with a stethoscope or palpation at the junction of the umbilical cord and skin. A newborn heart rate of less than 100 bpm indicates the need for immediate resuscitation.
- Grimace: No response to stimuli = 0; some response, a slight cry or grimace = 1; active response, coughing, sneezing, or vigorously crying = 2. The stimuli used to evoke a response can be the use of nasal suctioning, stroking the back to assess for spinal abnormalities, having the foot tapped.
- Appearance: The whole body is blue, gray, or very pale = 0; acrocyanosis, i.e., trunk and head have a pink skin color and hands and feet are blue = 1; pink all over = 2. Newborns with naturally darker skin color will not be pink, but pallor is still noticeable and especially in the soles of the feet and palms of the hands. Skin color is related to the

newborn's ability to oxygenate its body and extremities and is dependent on heart rate and respirations.

- Respiration: No breathing, apnea = 0; slow and irregular respiration = 1; good regular respiration, especially accompanied by crying = 2. Respirations are best assessed by watching the rise and fall of the neonate's abdomen since infants are diaphragmatic breathers.

Preparation

Essentially no preparation is needed to determine an Apgar score. Clinicians have suctioning equipment available and may use it during the birth process for nasal and oral suctioning to remove mucus and amniotic fluid. This is usually performed when the head of the newborn is safely delivered while the mother rests for her final push. The Apgar score should not be performed by the individual doing the delivery, but by the labor and delivery nurse or nursery nurse.

Aftercare

The Apgar score is primarily observational in nature and its only purpose is to alert the healthcare provider that the baby may need immediate assistance or prolonged observation in the nursery. It provides a means of monitoring the effectiveness of interventions and a process of determining which interventions are valuable.

Normal results

It is important to note that an Apgar score is strictly used to determine a newborn's immediate condition at birth and that it does not necessarily reflect the future health of a baby. The maximum obtainable score is 10 and the minimum is zero. It is quite rare to receive a true 10 as some acrocyanosis is considered normal and not a cause for concern. A score of 7 to 10 is considered normal, and these infants are expected to have an excellent outcome. A score of 4, 5, or 6 requires immediate intervention, usually in the form of oxygen and respiratory assistance or in the form of suctioning if breathing has been obstructed by mucus. A source of oxygen referred to as "blow-by" may be placed near but not directly over the nose and mouth of the neonate during suctioning. A score in the 4–6 range indicates that the neonate is having difficulty adapting to extrauterine life, which in some cases may be related to medications given to the mother during labor, **prematurity**, or a rapid delivery.

A low Apgar score provides a warning signal that the baby may have hidden health problems, such as breathing difficulties or internal bleeding. With a score of 0–3, the newborn is unresponsive, pale, limp, and may not have a pulse; therefore, an infant with a score of 0—3 needs immediate resuscitation. An ongoing evaluation is continued during resuscitation and documented again at five minutes. In the event of a difficult resuscitation, the Apgar score is done at 10, 15, and 20 minutes as well. A newborn with an Apgar score in this range generally requires advanced medical care and emergency measures, such as assisted breathing, administration of fluids or medications, and observation in a neonatal intensive care unit (NICU) by a neonatologist. An Apgar score of 0–3 at 20 minutes of age, for example, is indicative of high rates of morbidity (disease) and mortality (death).

KEY TERMS

Acrocyanosis—A condition characterized by blueness, coldness, and sweating of the extremities. A slight cyanosis, or blueness, of the hands and feet of the neonate is considered normal. This impaired ability to fully oxygenate the extremities is due to an immature circulatory system which is still in flux.

Amniotic fluid—The liquid in the amniotic sac that cushions the fetus and regulates temperature in the placental environment. Amniotic fluid also contains fetal cells.

Apnea—The temporary absence of breathing. Sleep apnea consists of repeated episodes of temporary suspension of breathing during sleep.

Extrauterine—Occurring outside the uterus.

Neonate—A newborn infant, from birth until 28 days of age.

Neonatologist—A physician (pediatrician) who has special training in the care of newborn infants.

Pallor—Extreme paleness in the color of the skin.

Risks

There are no risks involved with the Apgar scoring process. It is an evaluation of the baby at birth to determine if any resuscitation procedures are needed.

Parental concerns

Parental concerns may be addressed if the Apgar score is low at five minutes and then again at 10 minutes. The healthcare provider should address the possible risks associated with a low score and advise the parents as to follow-up care. A persistently low Apgar score could indicate neurological problems and the parents would

want to obtain additional treatment for the baby to ensure appropriate development. Children with **cerebral palsy** often have neurological damage at birth and the use of physical therapy or speech therapy enhances their outcome.

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Apnea of infancy

Definition

Apnea is a temporary cessation of breathing. Among children, this is most common in newborns.

Description

Babies born prematurely (before 34 weeks of gestation) usually lack a fully developed central nervous system. A component of this temporary deficit is inadequate control of their breathing reflex. The more premature a baby is born, the greater is the likelihood of apnea. Episodes of apnea are also more problematic for smaller than for larger babies. A small baby stores a smaller amount of oxygen, so the effects of oxygen deprivation are more severe. Apnea usually appears within the first several days after the baby is born.

Mild apnea causes no ill effects. The breathing pause is short (10–15 seconds), and the baby starts breathing again on his or her own. In a severe episode, though, breathing may cease for 20 seconds or longer. The infant begins to turn blue (cyanosis) because of the lack of oxygen in the blood. The baby retains carbon dioxide and may lapse into unconsciousness unless stimulated to breathe. Rubbing the infant with a finger or striking the soles of the feet may be all that is needed to end a short episode of apnea. If the baby has become unconscious, however, he or she may need to be revived with an oxygen mask. If apnea is frequent or severe, the baby's doctor may decide to treat it by altering conditions in the incubator, such as lowering the temperature, increasing oxygen, or placing the infant in a rocking incubator. Blood transfusions and medication may also be necessary.

Premature babies are also at higher risk for "late apnea," which occurs when the infant is older than six weeks. Late apnea can also affect full-term babies and may be a sign of an underlying problem such as **congenital heart disease**, infection, anemia, **meningitis**, or seizures. The baby usually recovers from apnea as the underlying disease is treated. Even if no underlying cause is found, late apnea is usually outgrown by the time the baby turns one year old.

Demographics

Apnea usually occurs during **sleep** and is primarily a disorder of premature infants. Sleep apnea before the early 2000s was thought to be a disease of older adults, but it can occur in children as well.

Causes and symptoms

In apnea of newborns, breathing stops and begins again automatically after a few seconds; it can also cause a prolonged pause which requires that the baby be resuscitated. Babies born before 34 weeks of gestation do not have a fully developed central nervous system, and they often do not have adequate control of the breathing reflex.

There are no specific measures for preventing apnea. It seems to be a sign of developmental immaturity, and it subsides as the baby grows older. Usually a

premature baby in an incubator is continually monitored, and hospital staff can easily detect apnea. With late apnea, parents may not notice that a child has stopped breathing while sleeping. If apnea is suspected or diagnosed, parents may install a home monitor until the condition is outgrown. Undiagnosed late apnea can be fatal and is associated with **sudden infant death syndrome** (SIDS). Parents of premature babies need to be apprised of the possibility of apnea and should be instructed on how to resuscitate their infant if it occurs. Those particularly worried about late apnea may also wish to be trained in infant first aid. Since apnea usually occurs during sleep, parents may decide to sleep near the baby.

When to call the doctor

Parents of a newborn who have taken the baby home should call the child's pediatrician if they notice the baby has episodes of not breathing during sleep. This especially true if the child was born prematurely or has other medical conditions.

Diagnosis

Diagnosis of sleep apnea of newborns is made by observation of the baby by a physician. Premature babies who are still in the hospital under neonatal care are monitored by machines that will alert staff when a baby stops breathing.

Treatment

Sleep apnea in infants is treated by gently stimulating the children by stroking their bodies. Touching them this way induces them to resume breathing. In severe cases, giving the baby oxygen or medication may be necessary.

Prognosis

If apnea is diagnosed, it will probably recur, but most premature babies outgrow the condition by the time they reach their normal due date.

Prevention

Premature babies are usually kept in an incubator, where their breathing and heart rate are monitored. A drop in the baby's heart rate or respiratory rate will sound an alarm, and a nurse can stimulate the baby to resume breathing, if necessary.

KEY TERMS

Cyanosis—A bluish tinge to the skin that can occur when the blood oxygen level drops too low.

Sudden infant death syndrome (SIDS)—The general term given to "crib deaths" of unknown causes.

Parental concerns

Parents should monitor the breathing patterns of infants who are born prematurely. Doing so is especially important during the first few weeks of life or until the infant reaches an age commensurate with full-term gestation.

Resources

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Appendicitis

Definition

Appendicitis is an inflammation of the appendix, which is the small, finger-shaped pouch attached to the beginning of the large intestine on the lower-right side of the abdomen. Appendicitis is a medical emergency, and if left untreated, the appendix may rupture and cause a potentially fatal infection.

Description

In children, appendicitis is the most common abdominal medical emergency and most common pediatric emergency surgical procedure. Although the appendix has no known function, it can become inflamed and diseased. This condition, called appendicitis, can rapidly evolve into a life-threatening or fatal infection of the abdominal cavity (peritonitis) if not treated immediately. Appendicitis usually involves emergency consultation with a physician and evaluation in a hospital emergency department.

Demographics

Appendicitis is the most common abdominal emergency found in children and young adults. One person in 15 develops appendicitis in his or her lifetime. The incidence is highest among males aged 10 to 14, and among females aged 15 to 19. More males than females develop appendicitis between **puberty** and age 25. It is rare in infants and children under the age of two. In the United States, appendicitis occurs in four out of 1,000 children.

Causes and symptoms

Appendicitis is usually caused by a blockage of the inside of the appendix, which is called the lumen. Most often, the lumen is blocked by fecal material. Lymphoid tissue, which is present in mucosal lining of the appendix and intestines to help fight bacterial and viral infections, can swell and lead to obstruction of the appendix. This condition, called lymphoid hyperplasia, may also be associated with a variety of inflammatory and infectious diseases, such as Crohn's disease, **gastroenteritis**, respiratory infections, mononucleosis, and **measles**. Appendicitis can also be caused by foreign bodies (e.g., intrauterine device or something swallowed), traumatic abdominal injury, or tumors. In addition, genetics may play a role in appendicitis; some children may inherit genes that make them more susceptible to blockage of the appendiceal lumen. Having **cystic fibrosis** also increases a child's risk for appendicitis.

Blockage of the appendix then causes inflammation, increased pressure, and restricted blood flow, leading to abdominal **pain** and tenderness in the right lower quadrant of the abdomen. If the appendix is not removed, bacteria and inflammation within the appendix rapidly expand, the wall of the appendix stretches, and perforation can occur. Once the appendix is perforated, bacteria-filled fluid is released into the abdominal cavity and peritonitis then develops. Perforation is more common in younger children. Perforation can occur as soon as 48 to 72 hours after symptoms first begin and can become life-threatening.

Classic symptoms of appendicitis include the following:

- abdominal pain, first around the navel then moving to the lower right quadrant of the abdomen
- **nausea**
- **vomiting**
- loss of appetite
- **diarrhea**, **constipation**, and/or inability to pass gas
- **fever** beginning after other symptoms
- abdominal swelling and tenderness

Other possible symptoms are pain on urination, inability to urinate, or frequent urge to urinate if the swollen appendix is near the urinary tract and bladder. When perforation occurs, abdominal pain becomes more

intense and involves the whole abdominal area, and fever may be very high.

Symptoms of appendicitis vary, and not every child will have all the symptoms. In children younger than age two years, the most common symptoms are vomiting and a bloated or swollen abdomen. Toddlers with appendicitis may have difficulty eating and may seem very tired. Children may have constipation, but may also have small stools that contain mucus. Although infants and children younger than two years may also have abdominal pain and other symptoms, they are too young to effectively communicate their symptoms to adults, who may then miss the symptoms of appendicitis.

When to call the doctor

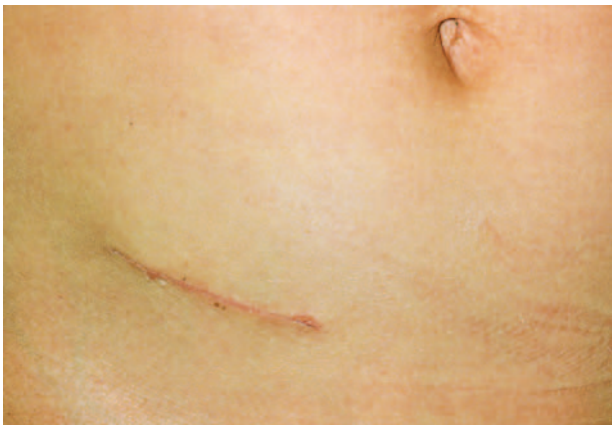
Appendicitis is a medical emergency. A doctor should be called immediately if appendicitis is suspected so that children can receive prompt medical treatment before perforation occurs. Parents who suspect that their child has appendicitis should not give the child any pain medication because it may interfere with the results of a doctor's physical examination for appendicitis. In addition, parents should not give their child anything to eat or drink in case surgery is required immediately.

Symptoms in combination that require a doctor's immediate attention include significant abdominal pain, fever, diarrhea, **nausea and vomiting**, swollen or bloated abdomen, and loss of appetite. If abdominal pain begins before nausea and vomiting, rather than after, appendicitis rather than intestinal infection is more likely.

Diagnosis

Appendicitis is diagnosed by physical examination, laboratory tests, and imaging tests. During a physical examination, the doctor palpates the abdomen to find tender and painful spots. A physical examination can also include a rectal examination, examination of the genitals in boys, and a gynecologic examination in girls, because other conditions, such as **testicular torsion** and ectopic pregnancy may have symptoms similar to appendicitis. Laboratory tests involve an analysis of white blood cell count to determine whether infection is present, urinalysis to rule out urinary tract or kidney infection, and other tests, such as pregnancy and liver function tests, to rule out other causes of abdominal pain. Imaging tests can include abdominal x rays, ultrasound, and **computed tomography** (CT).

In 2004, a new imaging technique that uses nuclear medicine imaging and an injection of an imaging agent called NeutroSpec was introduced for the diagnosis of appendicitis. This technique provides images of infected areas and may help physicians decide which children are candidates for surgery to remove the appendix. Up to 20 percent of appendectomies are performed on infants and children with a normal appendix.



Appendectomy scar on the right side of the patient's abdomen. *(© Dr. P. Marazzi/Science Photo Library/Photo Researchers, Inc.)*

Abdominal pain is a common complaint in children, and making a timely diagnosis of appendicitis before perforation is often difficult. Up to 30 percent of children with appendicitis are misdiagnosed, even by experienced physicians. In infants, diagnosis is often not possible and not made until after perforation. Appendicitis is most often misdiagnosed as gastroenteritis or respiratory infection.

Treatment

Appendicitis is treated by immediate surgery to remove the appendix, called an appendectomy. Appendectomy is the most common emergency surgery performed by pediatric surgeons. In an open appendectomy, the appendix is removed through a standard abdominal incision. In laparoscopic appendectomy, surgeons insert a small scope through tiny abdominal incisions to remove the appendix. A laparoscopic appendectomy results in less postoperative pain and fewer surgical incision infections. However, the procedure is longer and requires specialized surgical experience in operating on pediatric patients. In female teen patients, laparascopy has the added benefit of being able to diagnose and treat gynecologic conditions and ectopic pregnancy during the appendectomy if the appendix is found to be normal.

Preoperative **antibiotics** are given to children with suspected appendicitis and stopped after surgery if there is no perforation. Antibiotic treatment kills bacteria, and

KEY TERMS

Appendectomy—Surgical removal of the appendix.

Appendix—The worm-shaped pouch attached to the cecum, the beginning of the large intestine.

Computed tomography (CT)—An imaging technique in which cross-sectional x rays of the body are compiled to create a three-dimensional image of the body's internal structures; also called computed axial tomography.

Ectopic pregnancy—A pregnancy that develops outside of the mother's uterus, such as in the fallopian tube. Ectopic pregnancies often cause severe pain in the lower abdomen and are potentially life-threatening because of the massive blood loss that may occur as the developing embryo/fetus ruptures and damages the tissues in which it has implanted.

Laparoscopy—A surgical procedure in which a small incision is made, usually in the navel, through which a viewing tube (laparoscope) is inserted. This allows the doctor to examine abdominal and pelvic organs. Other small incisions can be mad to insert instruments to perform procedures. Laparoscopy is done to diagnose conditions or to perform certain types of surgeries.

Peritonitis—Inflammation of the peritoneum. It is most often due to bacterial infection, but can also be caused by a chemical irritant (such as spillage of acid from the stomach or bile from the gall bladder).

Testicular torsion—A condition involving the twisting of the spermatic cord inside the testicle that shuts off its blood supply and can seriously damage the testicle.

Ultrasonography—A medical test in which sound waves are directed against internal structures in the body. As sound waves bounce off the internal structure, they create an image on a video screen. Ultrasonography is often used to diagnose fetal abnormalities, gallstones, heart defects, and tumors. Also called ultrasound imaging.

stronger and longer courses of antibiotics are required if peritonitis occurs.

If the appendix is removed before perforation occurs, the hospital stay is usually two to three days. A child with a perforated appendix and peritonitis must remain in the hospital up to a week.

Prognosis

Appendicitis is usually treated successfully by appendectomy, and unless there are complications, children should recover without further problems. The mortality rate in cases without complications is less than 0.1 percent. Perforated and ruptured appendix, as well as peritonitis, occur at higher rates among children. When the appendix has ruptured or a severe infection has developed, the likelihood for developing complications is higher, and recovery is longer. Peritonitis is a life-threatening condition, and death occurs in about 1 percent of cases.

Prevention

In general, appendicitis cannot be prevented. The incidence of appendicitis is lower in cultures where people eat more daily dietary fiber, which is thought to decrease the viscosity of feces, decrease bowel transit time, and discourage formation of fecaliths, which predispose individuals to obstructions within the appendix.

Parental concerns

Because the appendix is more likely to perforate in children than adults, parents should not hesitate to call the doctor if their child develops symptoms that may indicate appendicitis. Parents should feel free to ask their doctor and other medical staff questions about any medical tests or treatments their child receives.

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Arachnodactyly *see* **Marfan syndrome**

Arnold-Chiari malformation *see* **Chiari malformation**

Art *see* **Drawings**

Arteriovenous fistula

Definition

An arteriovenous fistula is an abnormal channel or passage between an artery and a vein.

Description

An arteriovenous fistula is a disruption of the normal blood flow pattern. Normally, oxygenated blood flows to the tissue through arteries and capillaries. Following the release of oxygen in the tissues, the blood returns in veins to the heart. An arteriovenous fistula is an abnormal connection of an artery and a vein. The blood bypasses the capillaries and tissues and returns to the heart. Arterial blood has a higher blood pressure than blood in veins. Although both the artery and the vein retain their normal connections, the new opening between the two causes some arterial blood to shunt (be diverted) into the vein because of the blood pressure difference. As a result, the vein swells.

Demographics

Congenital arteriovenous fistula is rare. Acquired arteriovenous fistula is also uncommon in children. When it occurs, it is most likely to be found in the arms or legs.

Causes and symptoms

There are two types of arteriovenous fistulas, congenital and acquired. A congenital arteriovenous fistula is a rare birth defect that formed during fetal development. In congenital fistulas, blood vessels of the lower extremity are more frequently involved than other areas of the body. An acquired arteriovenous fistula is one that develops after a person is born. It usually occurs when an artery and vein that are side-by-side are damaged, and the healing process results in the two becoming linked. For example, after catheterizations, arteriovenous fistulas may occur as a complication of the arterial puncture in the leg or arm. Fistulas also form without obvious cause. In the case of patients on hemodialysis, physicians perform surgery to create a fistula. These patients receive many needle sticks to flush their blood through dialysis machines and for routine blood analysis testing. The veins used may scar and become difficult to access. Surgery is used to connect an artery and vein so that arterial blood pressure and flow rate widens the vein and decreases the chance of blood clots forming inside the vein.

The main symptoms of arteriovenous fistulas near the surface of the skin are bulging and discolored veins. In some cases, the bulging veins can be mistaken for varicose veins. Other fistulas can cause more serious problems without obvious symptoms, depending on their location and the blood vessels involved.

When to call the doctor

If the child has veins that appear to be varicose, the doctor should be consulted.

Diagnosis

Using a stethoscope, a physician can detect the sound of a pulse in the affected vein (bruit). The sound is a distinctive to-and-fro sound. Dye injected into the blood vessels can be tracked by x ray to confirm the presence of a fistula.

Treatment

Small arteriovenous fistulas can be corrected by surgery. Fistulas in the brain or eye are very difficult to

KEY TERMS

Congenital—Present at birth.

Sclerotherapy—Injection of an irritating chemical into a blood vessel so that it forms a scar to repair itself.

treat. If surgery is not possible or is very difficult, injection therapy may be used. Injection therapy, also called sclerotherapy, is the injection of an irritating chemical that causes scaring at the site of the injection. In the case of an arteriovenous fistula, this procedure should stop the passage of blood from the artery to the vein. Surgery is the most common method of treating acquired fistulas.

Prognosis

The prognosis for treated acquired arteriovenous fistula is usually very good. Congenital arteriovenous fistula is not usually treated quite as successfully, but it can also be treated in such a way as to minimize further problems.

Prevention

As of 2004, there is no known way to prevent arteriovenous fistula.

Parental concerns

If not treated, arteriovenous fistulas can be very dangerous. Tissues below the fistula may not get enough blood and may die. If too much blood is diverted through the fistula, heart complications may occur.

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Tish Davidson, A.M.
John T. Lohr, PhD

Arthritis *see* **Juvenile arthritis**

"Asleep" body parts *see* **Numbness and tingling**

Asperger's syndrome *see* **Pervasive developmental disorders**

Asphyxia neonatorum

Definition

Asphyxia neonatorum is respiratory failure in the newborn, a condition caused by the inadequate intake of oxygen before, during, or just after birth.

Description

Asphyxia neonatorum, also called birth or newborn asphyxia, is defined as a failure to start regular respiration within a minute of birth. Asphyxia neonatorum is a neonatal emergency as it may lead to hypoxia (lowering of oxygen supply to the brain and tissues) and possible brain damage or death if not correctly managed. Newborn infants normally start to breathe without assistance and usually cry after delivery. By one minute after birth most infants are breathing well. If an infant fails to establish sustained respiration after birth, the infant is diagnosed with asphyxia neonatorum. Normal infants have good muscle tone at birth and move their arms and legs actively, while asphyxia neonatorum infants are completely limp and do not move at all. If not correctly managed, asphyxia neonatorum will lead to hypoxia and possible brain damage or death.

Demographics

According to the National Center for Health Statistics (NCHS), in 2002, **infant mortality** caused by asphyxia neonatorum amounted to 14.4 deaths per 100,000 live births in the United States, representing the tenth leading cause of infant mortality. Worldwide, more than 1 million babies die annually from complications of birth asphyxia. According to the World Health Organization, asphyxia neonatorum is one of the leading causes of newborn deaths in developing countries, in which 4 to 9 million cases of newborn asphyxia occur each year, accounting for about 20 percent of the infant mortality rate.

Causes and symptoms

There are many causes of asphyxia neonatorum, the most common of which include the following: prenatal hypoxia (a condition resulting from a reduction of the oxygen supply to tissue below physiological levels despite adequate perfusion of the tissue by blood), umbilical cord compression during **childbirth**, occurrence of a preterm or difficult delivery, and maternal anesthesia (both the intravenous drugs and the anesthetic gases cross the placenta and may sedate the fetus). High-risk pregnancies for asphyxia neonatorum include:

- maternal age of less than 16 years old or over 40 years old
- low socioeconomic status
- maternal illnesses, such as diabetes, **hypertension**, Rh-sensitization, severe anemia
- mothers with previous abortions, stillbirths, early neonatal deaths, or preterm births
- lack of prenatal care
- abnormal fetal presentation or position
- alcohol abuse and **smoking** by the mother
- severe fetal growth retardation
- preterm labor

The symptoms of asphyxia neonatorum are bluish or gray skin color (cyanosis), slow heartbeat (bradycardia), stiff or limp limbs (**hypotonia**), and a poor response to stimulation.

When to call the doctor

Pregnant women who are at high risk of delivering newborns with asphyxia neonatorum should arrange for a close follow-up of their pregnancy with their obstetrician.

Diagnosis

Diagnosis can be objectively assessed using the Apgar score—a recording of the physical health of a newborn infant, determined after examination of the adequacy of respiration, heart action, muscle tone, skin color, and reflexes. Normally, the Apgar score is of 7 to 10. Infants with a score between 4 and 6 have moderate depression of their vital signs while infants with a score of 0 to 3 have severely depressed vital signs and are at great risk of dying unless actively resuscitated.

Treatment

The treatment for asphyxia neonatorum is resuscitation of the newborn. All medical delivery rooms have adequate resuscitation equipment should an infant not breathe well at delivery. Between 1970 and 2000, neonatal resuscitation has evolved from disparate teaching methods to organized programs. The most widely used procedure is the Neonatal Resucitation Program, supported by the American Academy of Pediatrics (AAP) and the American Heart Association (AHA).

If stimulation fails to initiate regular respiration in the newborn, the attending physician attempts resuscitation. He may decide first to gently suction the oropharynx—the area of the throat at the back of the mouth, with a soft catheter. When stimulation and a clear airway do not result in adequate respiration, the physician may give 100 percent oxygen via a face mask. If the infant is still not breathing, some form of artificial ventilation is then required. The usual method is to use mask ventilation with a resuscitator. The mask is applied tightly to the infant's face. If this procedure fails, the infant can be intubated with a endotracheal tube to which the resuscitator can then be connected. The more severe the fetal asphyxia, the longer it will take before the infant starts to breathe spontaneously. If the infant does not breathe despite adequate ventilation, or if the heart rate remains below 80 beats per minute, the physician can give an external cardiac massage using two fingers to depress the lower sternum at approximately 100 times a minute while continuing with respiratory assistance. Adrenaline may also be administered to increase cardiac output. Once the infant starts breathing, he or she is transferred to a nursery for observation and further assessment. Temperature, pulse and respiratory rate, color, and activity are recorded, and blood glucose levels checked for at least four hours.

Treatment may also include the following:

- giving the mother extra amounts of oxygen before delivery

KEY TERMS

Adrenaline—Another name for epinephrine, the hormone released by the adrenal glands in response to stress. It is the principal blood-pressure raising hormone and a bronchial and intestinal smooth muscles relaxant.

Anemia—A condition in which there is an abnormally low number of red blood cells in the bloodstream. It may be due to loss of blood, an increase in red blood cell destruction, or a decrease in red blood cell production. Major symptoms are paleness, shortness of breath, unusually fast or strong heart beats, and tiredness.

Anesthesia—Treatment with medicine that causes a loss of feeling, especially pain. Local anesthesia numbs only part of the body; general anesthesia causes loss of consciousness.

Apgar score—The results of an evaluation of a newborn's physical status, including heart rate, respiratory effort, muscle tone, response to stimulation, and color of skin.

Asphyxia—Lack of oxygen.

Asphyxia neonatorum—Respiratory failure in the newborn.

Bradycardia—A slow heart rate, usually under 60 beats per minute.

Cyanosis—A bluish tinge to the skin that can occur when the blood oxygen level drops too low.

Hemoglobin—An iron-containing pigment of red blood cells composed of four amino acid chains (alpha, beta, gamma, delta) that delivers oxygen from the lungs to the cells of the body and carries carbon dioxide from the cells to the lungs.

Hypotonia—Having reduced or diminished muscle tone or strength.

Hypoxia—A condition characterized by insufficient oxygen in the cells of the body

Neonatal—Refers to the first 28 days of an infant's life.

Oropharynx—One of the three regions of the pharynx, the oropharynx is the region behind the mouth.

Respiratory failure—Inability to rid the body of CO_2 or establish an adequate blood oxygen level.

Resuscitation—Bringing a person back to life or consciousness after he or she was apparently dead.

- medications to support the baby's breathing and sustain blood pressure
- extracorporeal membrane oxygenation (ECMO)

ECMO is a technique similar to a heart-lung bypass machine, which assists the infant's heart and lung functions with use of an external pump and oxygenator.

Alternative treatment

If an inadequate supply of oxygen from the placenta is detected during labor, the infant is at high risk for asphyxia, and an emergency delivery may be attempted either using forceps or by cesarean section.

Prognosis

The prognosis for asphyxia neonatorum depends on how long the newborn is unable to breathe. For example, clinical studies show that the outcome of babies with low five-minute Apgar scores is significantly better than those with the same scores at 10 minutes. With prolonged asphyxia, brain, heart, kidney, and lung damage can result and also death, if the asphyxiation lasts longer than 10 minutes.

Prevention

Anticipation is the key to preventing asphyxia neonatorum. It is important to identify fetuses that are likely to be at risk of asphyxia and to closely monitor such high-risk pregnancies. High-risk mothers should always give birth in hospitals with neonatal intensive care units where appropriate facilities are available to treat asphyxia neonatorum. During labor, the medical team must be ready to intervene appropriately and to be adequately prepared for resuscitation.

Parental concerns

Women at risk for asphyxia neonatorum pregnancies should receive focused prenatal care from an obstetrician skilled at preventing and detecting problems such as anemia that may contribute to asphyxia neonatorum. While prenatal care will not necessarily prevent newborn asphyxia, it can help ensure that both the mother and her baby are as healthy as possible at the time of birth.

See also Hypotonia.

Resources

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Assessment

Definition

Assessment is a process of gathering and documenting information about the achievement, skills, abilities, and personality variables of an individual.

Description

Assessment is used in both an educational and psychological setting by teachers, psychologists, and counselors to accomplish a range of objectives. These include the following:

- to learn more about the competencies and deficiencies of the individual being tested
- to identify specific problem areas and/or needs
- to evaluate the individual's performance in relation to others
- to evaluate the individual's performance in relation to a set of standards or goals
- to provide teachers with feedback on effectiveness of instruction
- to evaluate the impact of psychological or neurological abnormalities on learning and behavior
- to predict an individual's aptitudes or future capabilities

In the early 2000s standardized tests are increasingly used to evaluate performance in U.S. schools. Faced with declining test scores by American students when compared to others around the world, state governments and the federal government have sought ways to measure the performance of schools and bring a measurable accountability to the educational process. Thus, states and the federal government have adopted standardized tests for evaluating knowledge and skills on the assumption that testing is an effective way to measure outcomes of education. One prominent program has been the No Child Left Behind Act that requires schools to meet certain performance standards annually, for their students as a group and also for individual ethnic and racial subgroups. The use of this type of standardized tests is controversial. Many educators feel that it limits the **creativity** and effectiveness of the classroom teacher and produces an environment of "teaching to the test."

Educational assessments

The choice of an assessment tool depends on the purpose or goal of the assessment. Assessments might be made to establish rankings among individual students, to determine the amount of information students have retained, to provide feedback to students on their levels of achievement, to motivate students by recognizing and rewarding good performances, to assess the need for remedial education, and to evaluate students for class placement or ability grouping. The goal of the assessment should be understood by all stakeholders in the process: students, parents, teachers, counselors, and outside experts. An assessment tool that is appropriate for one goal is often inappropriate for another, leading to misuse of data.

Assessment tools fall broadly into two groups. Traditional assessments rely on specific, structured procedures and instructions given to all test-takers by the test administrator (or to be read by the test-takers themselves). These tests are either norm-referenced or criterion-referenced tests. Standardized tests allow researchers to compare data from large numbers of students or subgroups of students. Alternative assessments are often handled on an individual basis and offer students the opportunity to be more closely involved with the recognition of their progress and to discover what steps they can take to improve.

NORM-REFERENCED ASSESSMENTS In norm-referenced assessments, one person's performance is interpreted in relation to the performance of others. A norm-referenced test is designed to discriminate among individuals in the area being measured and to give each individual a rank or relative measure regarding how he or she performs compared to others of the same age, grade, or other subgroup. Often the mean, or average score, is the reference point, and individuals are scored on how much above or below the average they fall. These tests are usually timed. Norm-referenced tests are often used to tell how a school or school district is doing in comparison to others in the state or nation.

CRITERION-REFERENCED ASSESSMENTS A criterion-referenced assessment allows interpretation of a test-taker's score in relation to a specific standard or criterion. Criterion-referenced tests are designed to help evaluate whether a child has met a specific level of performance. The individual's score is based not on how he or she does in comparison to how others perform, but on how the individual does in relation to absolute expectations about what he or she is supposed to know. An example of a criterion-referenced test is a timed arithmetic test that is scored for the number of problems answered correctly. Criterion-referenced tests measure what information an individual has retained and they give teachers feedback on the effectiveness of their teaching particular concepts.

PERFORMANCE ASSESSMENT Performance assessment can be used to evaluate any learning that is skill-based or behavioral. Performance assessment requires the test-taker to perform a complex task that has to do with producing a certain product or performing a specific task. Performance assessments can be either individual or group-oriented and may involve application of real-life or workplace skills (for example, making a piece of furniture in wood shop).

AUTHENTIC ASSESSMENT Authentic assessment derives its name from the idea that it tests students in skills and knowledge needed to succeed in the real world. Authentic assessment focuses on student task performance and is often used to improve learning in practical areas. An advantage of authentic assessment is that students may be able to see how they would perform in a practical, non-educational setting and thus may be motivated to work to improve.

PORTFOLIO ASSESSMENT Portfolio assessment uses a collection of examples of the actual student's work. It is designed to advance through each grade of school with the student, providing a way for teachers and others to evaluate progress. One of the hallmarks of portfolio assessment is that the student is responsible for selecting examples of his or her own work to be placed in the portfolio. The portfolio may be used by an individual classroom teacher as a repository for work in progress or for accomplishments. Portfolios allow the teacher to evaluate each student in relation to his or her own abilities and learning style. The student controls the assessment samples, helping to reinforce the idea that he or she is responsible for learning and should have a role in choosing the data upon which he or she is judged. Portfolios are often shared by the student and teacher with parents during parent-teacher conferences.

INTERVIEW ASSESSMENT The assessment interview involves a one-on-one or small group discussion between the teacher and student, who may be joined by parents or other teachers. Standardized tests reveal little about the test-taker's thought process during testing. An interview allows the teacher or other administrator to gain an understanding of how the test-taker reached his or her answer. Individual interviews require a much greater time commitment on the part of the teacher than the administration of a standardized test to the entire class at one time. Thus, interviews are most effective when used to evaluate the achievements and needs of specific students. To be successful, interviews require both the teacher and the student to be motivated, open to discussion, and focused on the purpose of the assessment.

JOURNALS Journals have been used as part of the English curriculum since at least the 1980s. In assessment, the journal allows the student to share his or her thoughts on the learning process. A journal may substitute for or supplement a portfolio in providing a student-directed assessment of achievement and goals.

ATTITUDE INVENTORY Attitude is one component of academic success that is rarely measured objectively. An attitude inventory is designed to reveal both positive and negative (or productive and unproductive) aspects of a student's outlook toward school and learning. However, this type of assessment may be of limited use if the student's negative attitude makes him or her unwilling to actively participate in the assessment. By demonstrating a sincere interest in addressing student concerns that affect attitude, a school can improve the effectiveness of attitude inventory assessments.

COMPUTER-AIDED ASSESSMENT Computer-aided assessment is increasingly employed as a supplement to other forms of assessment. A key advantage in the use of computers is the capability of an interactive assessment to provide immediate feedback on responses. Students must be comfortable with computers and reading on a computer screen for these assessments to be successful.

Psychological assessments

Psychological assessment of children is used for a variety of purposes, including diagnosing learning disabilities and behavioral and attention problems. Psychologists can obtain information about a child in three general ways: observation, verbal questioning or written questionnaires, and assignment of tasks. The child's pediatrician, parents, or teacher may ask for psychological assessment to gain a greater understanding of the child's development and needs. There are many different **psychological tests**, and the psychologist must choose the ones that will provide the most relevant and reliable information in each situation. Often multiple tests are performed. However, most psychological assessments fall into one of three categories: observational methods, personality inventories, or projective techniques.

OBSERVATIONAL ASSESSMENT Observations are made by a trained professional either in a familiar setting (such as a classroom or playroom), an experimental setting, or during an office interview. **Toys**, dolls, or other items are often included in the setting to provide stimuli. The child may be influenced by the presence of an observer. However, researchers report that younger children often become engrossed in their activities and thus are relatively unaffected by the presence of an observer. Sometimes, for example, if attention deficit is suspected, several people are asked to observe the child under different circumstances: the teacher at school, the parent at home, and the psychologist in an office setting. Observational assessments are usually combined with other types of educational or psychological assessments when learning needs and behavioral problems are being evaluated.

PERSONALITY INVENTORIES A personality inventory is a questionnaire used with older children and adults that contains questions related to the subject's feelings or reactions to certain scenarios. One of the best-known personality inventories for people over age 16 is the **Minnesota Multiphasic Personality Inventory** (MMPI), a series of over 500 questions used to assess personality traits and psychological disturbances. Interviews or verbal questionnaires for personality assessment may be structured with a specific series of questions or be unstructured, allowing the subject to direct the discussion. Interviewers often use rating scales to record information during interviews.

PROJECTIVE TESTS A projective test asks the test-taker to interpret ambiguous situations. It requires a skilled, trained examiner to administer and interpret a projective test. The reliability of these tests with children is difficult to establish due to their subjective nature, with results varying widely among different examiners. One well-known projective test is the Rorschach Psychodiagnostic Test, or inkblot test, first devised by the Swiss psychologist Hermann Rorschach in the 1920s. Another widely used projective test for people ages 14 to 40 is the **Thematic Apperception Test** (TAT), developed at Harvard University in the 1930s. In this test, the subject is shown a series of pictures, each of which can be interpreted in a variety of ways, and asked to construct a story based on each one. An adaptation administered to children aged three to ten is the **Children's Apperception Test** (CAT). Apperception tests are administered to children individually by a trained psychologist to assess personality, maturity, and psychological health.

ASSIGNMENT OF TASK ASSESSMENT Assignment of tasks is an assessment method involving the performance of a specific task or function. These tests are designed to inform the test administrator about attributes such as the test-taker's abilities, perceptions, and motor coordination. They can be especially helpful in assessing if there is a physical or neurological component that needs to be addressed medically or with occupational, speech, or physical therapy.

Common problems

Assessment of children is challenging given the rapid changes in growth they experience during childhood. In childhood, it is difficult to ensure that the test-taker's responses will be stable for even a short time. Thus, psychologists, educators, and other test administrators are careful to take the stage of childhood into account when interpreting a child's test scores.

Traditional standardized tests rely on specific, structured procedures, which with young children presents some problems. Young children (**preschool** and early elementary years) do not have past experience and familiarity with tests and have limited understanding of the expectations of testing procedures. With young test-takers, the test administrator represents a significant factor that influences success. The child must feel comfortable with the test administrator and feel motivated to complete the test exercise. The administrator helps support the test-taker's attention to the test requirements. The testing environment affects all test-takers but may represent a more significant variable for the youngest test-takers.

One shortcoming of standardized testing is that it assumes that the same instrument can evaluate all students. Because most standardized tests are norm-referenced and measure a student's test performance against the performance of other test-takers, students and educators focus their efforts on the test scores, and schools develop curricula to prepare students to take the test.

KEY TERMS

Authentic task assessment—Evaluation of a task performed by a student that is similar to tasks performed in the outside world.

Criterion-referenced test—An assessment that measures the achievement of specific information or skills against a standard as opposed to being measured against how others perform.

Halo effect—An observer bias in which the observer interprets a child's actions in a way that confirm the observer's preconceived ideas about the child.

Norm-referenced test—A test that measures the performance of a student against the performance of a group of other individuals.

Portfolio—A student-controlled collection of student work products that indicates progress over time.

Standardized test—A test that follows a regimented structure, and each individuals scores may be compared with those of groups of people. In the case of the Cognistat, test taker's scores can be compared to groups of young adults, middle-aged adults, the geriatric, and people who have undergone neurosurgery.

Task—A goal directed activity used in assessment.

Other criticisms of standardized tests are that they are culturally insensitive and that they may not accurately represent the abilities of children in the United States for whom English is not their first language or who are not a part of mainstream American culture. Finally, in middle and high school settings, disgruntled students may inconspicuously sabotage their tests since these scores do not affect the students' own grades but reflect rather upon the competency of the teacher and the school administration.

Alternative assessments are subject to other concerns. Observer biases and inconsistencies have been identified through study of the assessment procedures. In the halo effect, the observer evaluates the child's behavior in a way that confirms his general previous impression of the child. For example, the observer believes a particular child is happy and loving. If, when the observer assesses that child, the child lays a doll face down on the table, the observer interprets this act as parenting behavior. On the other hand, if the observer believes the child is angry and hostile, when this child is observed laying the doll face down on the table, the observer may interpret the action as aggression. The expectations of the observer conveyed directly or through body language and other subtle cues may also influence how the child performs and how the observer records and interprets his or her observations. This observer bias can influence the outcome of an assessment.

Parental concerns

Parents are justifiably concerned that their child be evaluated fairly and appropriately. They have the right to understand the purpose of the assessment, how it will be performed, how the information will be used, who will see the assessment results, and how the privacy of their child will be protected. Any professional performing an educational or psychological assessment should be willing discuss these concerns and to share the results of the assessment and their implications with the parent. Parents should be willing to share with examiners any information that might alter interpretation of the assessment results (for example, medical problems, cultural concerns).

When to ask for an assessment

Parents should request an assessment from the teacher whenever necessary to understand their child's progress, both in relation to expected grade-level expectations and performance in relation to other children in the class. Most schools and teachers offer parents many opportunities to discuss the assessment of their child. When teacher assessment indicates that a child has special needs or problems, the parent should request an evaluation by the school's child study team or an outside expert. Parents may also want to discuss appropriate assessments with their child's pediatrician and ask for an referral to a child psychologist or psychiatrist.

See also California Achievement Tests (CAT); Children's Apperception Test (CAT); Development tests.

Resources

BOOKS

Carter, Phillip, and Ken Russell. *Psychometric Testing: 1000 Ways to Assess Your Personality, Creativity, Intelligence, and Lateral Thinking*. New York: Wiley & Sons, 2001.

Groth-Marnat, Gary. *Handbook of Psychological Assessment*, 4th ed. New York: Wiley & Sons, 2003.

Joint Committee on Standards for Educational Evaluation. *The Student Evaluation Standards*. Thousand Oaks, CA: Corwin Press, 2003.

ORGANIZATIONS

Evaluation Center. 4405 Ellsworth Hall, Western Michigan University, Kalamazoo, MI 49008–5237. Web site: <www.wmich.edu/evalctr/jc>.

National Association for the Education of Young Children. 1509 16th Street, NW Washington, DC 20036. Web site: <www.naeyc.org>.

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Asthma

Definition

Asthma is a chronic (long-lasting) inflammatory disease of the airways. In people susceptible to asthma, this inflammation causes the airways to narrow periodically. This narrowing, in turn, produces wheezing and breathlessness that sometimes causes the patient to gasp for air. Obstruction to air flow either stops spontaneously or responds to a wide range of treatments, but continuing inflammation makes the airways hyper-responsive to stimuli such as cold air, **exercise**, dust mites, pollutants in the air, and even stress and **anxiety**.

Description

The changes that take place in the lungs of people with asthma make the airways (the "breathing tubes," or bronchi and the smaller bronchioles) hyper-reactive to many different types of stimuli that do not affect healthy lungs. In an asthma attack, the muscle tissues in the walls of the bronchi go into spasm, and the cells lining the airways swell and secrete mucus into the air spaces. These two actions cause the bronchi to become narrowed (bronchoconstriction). As a result, a person with asthma has to make a much greater effort to breathe.

Cells in the bronchial walls, called mast cells, release certain substances that cause the bronchial muscles to contract and stimulate mucus formation. These substances, including histamine and a group of chemicals called leukotrienes, also bring white blood cells into the area, which play a key role in the inflammatory response. Many patients with asthma are prone to react to such "foreign" substances as pollen, house dust mites, or animal dander. These are called allergens. An acute asthma attack can begin immediately after exposure to a trigger or several days or weeks later.

When asthma begins in childhood, it often affects a child who is likely, for genetic reasons, to become sensitized to common "allergens" in the environment (atopic person). When these children are exposed to house dust mites, animal proteins, fungi, or other potential allergens, they produce a type of antibody that is intended to engulf and destroy the foreign materials. This makes the airway cells sensitive to particular materials. Further exposure can rapidly lead to an asthmatic response.

Demographics

Asthma affects about 17 million Americans, including nearly five million children. Asthma usually begins in childhood or **adolescence**, but it also may first appear in adulthood. Asthma is the leading cause of chronic illness in children, accounting for 14 million missed school days annually. It is the third-ranking cause of **hospitalization** among children under age 15.

Asthma affects as many as 10–12 percent of children in the United States and the number has been steadily increasing. Since 1980, asthma has increased by 160 percent among children at least four years of age. Asthma is becoming more frequent, and—despite modern drug treatments—it is more severe than in the past. Some experts suggest this is due to increased exposure to allergens such as dust, air pollution, second-hand smoke, and industrial components.

Asthma can begin at any age, but most children experience their first symptoms by the time they are five years old. Boys have a higher incidence of asthma than girls, and the disease is more prevalent in African American children. Children living in inner cities, low-income populations, and minorities have disproportionately higher morbidity and mortality due to asthma.

Causes and symptoms

Causes

About 80 percent of childhood asthma cases are caused by **allergies**. In most cases, inhaling an allergen sets off the chain of biochemical and tissue changes leading to airway inflammation, bronchoconstriction, and wheezing characteristic of asthma. Because avoiding (or at least minimizing) exposure is the most effective way of treating asthma, it is vital to identify the allergen or irritant that is causing symptoms in a particular child.

Once asthma is present, symptoms can be triggered or made worse if the child also has **rhinitis** (inflammation of the lining of the nose) or **sinusitis**. **Gastroesophageal reflux disease** (GERD), a condition that causes stomach acid to pass back up the esophagus, can worsen asthma. Many pulmonary infections in early childhood, including those due to *Chlamydia pneumoniae,*

Mycoplasma pneumoniae, and respiratory syncytial virus, have been linked with an increased risk for wheezing and asthma. Aspirin and a class of drugs called beta-blockers (often used to treat high blood pressure) can also worsen the symptoms of asthma. Foggy and cloudy environments have been noted to aggravate asthma, and **obesity** facilitates asthma, but does not cause it.

The most important inhaled allergens and triggers contributing to attacks of asthma are:

- animal dander
- mites in house dust
- fungi (molds) that grow indoors
- mold spores that grow outdoors
- cockroach allergens
- tree, grass, and weed pollen
- occupational exposure to chemicals, fumes, or particles of industrial materials in the air
- strong odors, such as from perfume
- wood smoke

Inhaling tobacco smoke (from secondhand smoke or **smoking**) can irritate the airways and trigger an asthmatic attack. Air pollutants can have a similar effect.

There are three important factors that regularly produce attacks in certain patients with asthma, and they may sometimes be the sole cause of symptoms. They are:

- humidity and temperature changes, especially inhaling cold air
- exercise (in certain children, asthma is caused simply by exercising, and is called exercise-induced asthma)
- stress, strong emotions, or a high level of anxiety

Risk factors

There are many risk factors for childhood asthma, including:

- presence of allergies
- **family** history of asthma and/or allergies
- frequent respiratory infections
- low birth weight
- mother's exposure to tobacco smoke during pregnancy and/or child's exposure after birth
- wheezing with upper respiratory infections

Symptoms

Wheezing is often very obvious, but mild asthmatic attacks may be confirmed when the physician listens to the patient's chest with a stethoscope. Wheezing is often loudest when the child breathes out, in an attempt to expel used air through the narrowed airways. Besides wheezing and shortness of breath, the child may **cough** and experience **pain** or pressure in the chest. The child may have **itching** on the back or neck at the start of an attack. Infants may have feeding problems and may grunt while sucking or feeding. Tiring easily or becoming irritated are other common symptoms.

Some children with asthma are free of symptoms most of the time, but may occasionally experience brief periods during which they are short of breath. Others spend much of their days (and nights) coughing and wheezing, until the asthma is properly treated. Crying or even laughing may bring on an attack. Severe episodes, which are less common, may be seen when the patient has a viral respiratory tract infection or is exposed to a heavy load of an allergen or irritant. Asthmatic attacks may last only a few minutes or can go on for hours or even days (a condition called status asthmaticus).

Asthma symptoms can be classified as:

- Mild intermittent: Symptoms occur twice a week or less; nighttime symptoms occur twice a month or less; symptoms are brief and last a few hours to a few days; no symptoms occur between more severe episodes.
- Mild persistent: Symptoms occur more than twice a week but not every day; nighttime symptoms occur more than twice a month; episodes are severe and sometimes affect activity.
- Moderate persistent: Symptoms occur daily; nighttime symptoms occur more than once a week; quick-relief medication is used daily; symptoms affect daily activities; severe episodes occur twice a week or more and last for days.
- Severe persistent: Symptoms occur continually throughout the day and frequently at night; symptoms affect daily activities and cause the patient to limit activities.

Shortness of breath may cause a patient to become very anxious, sit upright, lean forward, and use the neck or chest wall muscles to help with breathing. These symptoms require emergency attention. In a severe attack that lasts for some time, some of the air sacs in the lung may rupture so that air collects within the chest. This makes it even harder to breathe in adequate amounts of air.

Almost always, even patients with the most severe attacks will recover completely.

When to call the doctor

If a child has the following symptoms, the parent should contact the child's pediatrician:

- inability to participate in normal activities
- missed school due to asthma symptoms
- symptoms that do not improve about 15 minutes after initial treatment with medication
- signs of infection such as increased fatigue or weakness, **fever** or chills, **sore throat**, coughing up mucus, yellow or green mucus, sinus drainage, nasal congestion, headaches, or tenderness along the cheekbones

If the parent is unsure about what action to take to treat the child's symptoms, he or she should call the child's doctor.

The parent or caregiver should seek emergency care by calling 911 in most areas when the child has these symptoms or conditions:

- bluish skin tone
- bluish coloration around the lips, fingernail beds, and tongue
- severe wheezing
- uncontrolled coughing
- very rapid breathing
- inability to catch his or her breath
- tightened neck and chest muscles due to breathing difficulty
- inability to perform a peak expiratory flow
- feelings of anxiety or panic
- pale, sweaty face
- difficulty talking
- difficulty walking
- confusion
- dizziness or fainting
- chest pain or pressure

Diagnosis

Early diagnosis is critical to proper asthma treatment and management. Asthma may be diagnosed by the child's primary pediatrician or an asthma specialist, such as an allergist.

The diagnosis of asthma may be strongly suggested when the typical symptoms and signs are present, including coughing, wheezing, shortness of breath, rapid breathing, or chest tightness. The physician will question the child (if old enough to provide an accurate history of symptoms) or parent about his or her physical health (the medical history), perform a physical examination, and perform or order certain tests to rule out other conditions.

The medical and family history help the physician determine if the child has any conditions or disorders that might be the cause of asthma. A family history of asthma or allergies can be a valuable indicator of asthma and may suggest a genetic predisposition to the condition. The physician will ask detailed questions about the child's symptoms, including when they first occurred, what seems to cause them, the frequency and severity, and how they are being managed.

During the physical exam, the pediatrician will listen to the patient's chest with a stethoscope to evaluate distinctive breathing sounds. He or she also will look for maximum chest expansion during inhalation. Hunched shoulders and contracting neck muscles are signs of narrowed airways. Nasal polyps or increased amounts of nasal secretions are often noted in patients with asthma. Skin changes, like **atopic dermatitis** or eczema, may demonstrate that the patient has allergic problems.

When asthma is suspected, the diagnosis can be confirmed using certain respiratory tests. Spirometry is a test that measures how rapidly air is exhaled and how much air is retained in the lungs. Usually the child should be at least five years of age for this test to be successful. During the test, the child exhales and the spirometer measures the airflow, comparing lung capacity to the normal range for the child's age and race. The child then inhales a drug that widens the air passages (a short-acting bronchodilator) and the doctor takes another measurement of the lung capacity. An increase in lung capacity after taking this medication often indicates the asthma symptoms are reversible (a very typical finding in asthma). The spirometer is similar to the peak flow meter that patients use to keep track of asthma severity at home.

Often, it is difficult to determine what is triggering asthma attacks. Allergy skin testing may be performed, especially if the doctor suspects the child's symptoms are persistent. An allergic skin response does not always mean that the allergen being tested is causing the asthma. Also, the body's immune system produces an antibody to fight off the allergen. The amount of antibody can be measured by a blood test that will show how sensitive the patient is to a particular allergen. If the diagnosis is

still in doubt, the patient can inhale a suspect allergen while using a spirometer to detect airway narrowing. Spirometry can also be repeated after a bout of exercise if exercise-induced asthma is a possibility. A chest x ray will help rule out other disorders.

Treatment

Once asthma is diagnosed, a treatment plan should be initiated as quickly as possible to manage asthma symptoms.

In most cases, asthma treatment is managed by the child's pediatrician. Referral to an asthma specialist should be considered if:

- There has been a life-threatening asthma attack or severe, persistent asthma.
- Treatment for three to six months has not met its goals.
- Some other condition, such as nasal polyps or chronic lung disease, complicates the asthma.
- Special tests, such as allergy skin testing or an allergen challenge, are needed.
- Intensive steroid therapy has been necessary.

The first step in bringing asthma under control is to reduce or avoid exposure to known allergens or triggers as much as possible. Treatment goals for all patients with asthma are to prevent troublesome symptoms, maintain lung function as close to normal as possible, avoid emergency room visits or hospitalizations, allow participation in normal activities—including exercise and those requiring exertion—and improve the quality of life.

Medications

The best drug treatment plan will control asthmatic symptoms while causing few or no side effects. The child's doctor will work with the parent to determine the drugs that are most appropriate and may be the most effective, based on the severity of symptoms. Age and the presence of other medical conditions may affect the drugs selected.

Two types of asthma medications include short-acting, quick relief, medications and long-acting, controller, medications. Quick relief medications are used to treat asthma symptoms when they occur. They relieve symptoms rapidly and are usually taken only when needed. Long-acting medications are preventative and are taken daily to help a patient achieve and maintain control of asthma symptoms.

Asthma treatment guidelines may be based on these symptom classifications:

- Mild intermittent: No daily medication is needed but a short-acting beta2 agonist may be used when needed to treat symptoms.
- Mild persistent: Daily long-term medication may be prescribed.
- Moderate persistent: Two medications may be prescribed, including a long-term medication to control inflammation and a short-acting medication to use when symptoms are more severe.
- Severe persistent: Multiple long-term control medications are required.

When asthma symptoms worsen, medication is increased. When asthma symptoms are controlled, less medication is needed. It is very important to discuss any desired changes to the medication schedule with the doctor. The medication dose should never be changed without the doctor's approval. The condition can worsen if certain medications are not taken.

Inhaled medications have a special inhaler that meters the dose. The inhaler may have a spacer that holds the burst of medication until it is inhaled. Patients will be instructed on how to properly use an inhaler to ensure that it will deliver the right amount of medication.

A home nebulizer, also known as a breathing machine, may be used to deliver asthma medications at home. The nebulizer changes medication from liquid form to a mist. The child wears a face mask to breathe in the medications. Nebulizer treatments generally take seven to 10 minutes.

Quick relief medications include short-acting, inhaled beta2 agonists and anticholinergics. Long-acting medications include leukotriene modifiers, mast cell stabilizers, inhaled and oral corticosteroids, long-acting beta2 agonists, and methylxanthines.

SHORT-ACTING BETA-2 AGONISTS These drugs, which are bronchodilators, open the airways by relaxing the muscles around the airways that have tightened (bronchospasm). The short-acting forms of beta-receptor agonists are the best choice for relieving sudden attacks of asthma and for preventing attacks triggered by exercise. These drugs generally start acting within minutes, but their effects last only four to six hours (although longer-acting forms are being developed). They may be taken by mouth, inhaled, or injected.

ANTICHOLINERGICS Anticholinergics are medications that open the airways by relaxing the muscle bands that tighten around the airways. They also suppress mucus production. They do not provide immediate relief, but can be used to control severe attacks when added to an inhaled beta-receptor agonist.

LEUKOTRIENE MODIFIERS Leukotriene modifiers, also called antileukotrienes, can be used in place of steroids for older children who have a mild degree of asthma that persists. They work by counteracting leukotrienes, substances released by white blood cells in the lung that cause the air passages to constrict and promote mucus secretion.

MAST CELL STABILIZERS Available only in inhaled form, mast cell stabilizers, such as cromolyn and nedocromil, prevent asthma symptoms. These anti-inflammatory drugs are often given to children as the initial treatment to prevent asthmatic attacks over the long term. They can also prevent attacks when given before exercise or when exposure to an allergen cannot be avoided. They are not effective until three to four weeks after therapy is started. These medications need to be taken two to four times a day.

STEROIDS These drugs, which resemble natural body hormones, block inflammation. Steroids are extremely effective in relieving asthma symptoms and can control even severe cases over the long term while maintaining good lung function. When steroids are taken by inhalation for a long period, asthma attacks become less frequent as the airways become less sensitive to allergens. Besides being inhaled, steroids may be taken by mouth or injected, to rapidly control severe asthma. Steroids are the strongest class of asthma medications and can cause numerous side-effects, including bleeding from the stomach, loss of calcium from bones, cataracts in the eye, and a diabetes-like state. Patients using steroids for lengthy periods also may have problems with wound healing, weight gain, and mental disorders. In children, growth may be slowed. To prevent serious side effects, the child will have periodic monitoring tests.

LONG-ACTING BETA-2 AGONISTS Long-acting beta-2 agonists are used for better control—not relief—of asthma symptoms. The medications take longer to work and the effects last longer, up to 12 hours.

METHYLXANTHINES Theophylline is the chief methylxanthine drug. It may exert some anti-inflammatory effect, and is especially helpful in controlling nighttime symptoms of asthma. If a patient cannot use an inhaler to maintain long-term control, sustained-release theophylline is a good alternative. The blood levels of the drug must be measured periodically, as too high of a dose can cause an abnormal heart rhythm or convulsions.

OTHER DRUGS Some inhalers contain a combination of two different medications that can be delivered together to shorten treatment times and decrease the number of inhalers that need to be purchased. Clinical trials are continuously evaluating new asthma medications.

IMMUNOTHERAPY If a patient's asthma is caused by an allergen that cannot be avoided, or if medications have not been effective in controlling symptoms, immunotherapy (also called **allergy shots**) may be considered. Immunotherapy is helpful when symptoms tend to occur throughout all or most of the year. Typically, increasing amounts of the allergen are injected over a period of three to five years, so that the body can build up an effective immune response. There is a risk that this treatment may cause the airways to become narrowed and bring on an asthmatic attack.

An international conference, Immunotherapy in Allergic Asthma, hosted by the American College of Allergy, Asthma, and Immunology (ACAII) in 2000 concluded that immunotherapy is an effective treatment for allergic asthma and can prevent the onset of asthma in children with **allergic rhinitis**. The Preventive Allergy Treatment study, published in 2002, confirmed the ACAII conference conclusions, documenting that immunotherapy reduces the risk of developing asthma and reduces lung airway inflammation in children with hay fever, a condition that predisposes them to asthma.

Managing asthmatic attacks

Urgent measures to control asthma attacks and ongoing treatment to prevent attacks are equally important. No matter how severe a person's asthma, quick-relief medications must be readily available to treat acute symptoms. If the patient's asthma symptoms are present most of the time, an anti-inflammatory medication should be used regularly.

A severe asthma attack should be treated as quickly as possible. It is most important for a patient suffering an acute attack to be given extra oxygen. Rarely, it may be necessary to use a mechanical ventilator to help the patient breathe. A beta-receptor agonist is inhaled repeatedly or continuously. A steroid is given if the patient's symptoms do not improve promptly and completely. Steroids also may help if a viral infection caused severe asthmatic symptoms. A course of steroid therapy, given after the attack is over, will make a recurrence less likely.

Starting treatment at home, rather than in a hospital, minimizes delays and helps the patient gain a sense of control over the disease. When deciding whether a patient should be hospitalized, the past history of acute attacks, severity of symptoms, current medication, and availability of adequate support at home must be taken into account.

Maintaining control

Children with asthma should follow up with their doctor every one to six months, depending on the frequency of attacks. During the follow-up visits, the child's lung function should be measured by spirometry to make sure treatment goals are being met. Once asthma has been controlled for several weeks or months, the child's physician may adjust the medication dosage. If there is no clear improvement with the current treatment plan, another treatment plan should be established.

All patients with asthma should learn how to monitor their symptoms so that they will know when an attack is starting. Symptoms can be monitored with a peak flow meter (also called a peak expiratory flow meter). To effectively follow the instructions for using a peak flow meter, the child should be at least five years old. The peak flow meter measures the child's airflow when he or she blows into it quickly and forcefully. The peak flow meter can be used to determine when to call the doctor or seek emergency care.

Knowing the child's allergens or triggers will help parents reduce exposure by making improvements in the home environment. Specific guidelines may include reducing indoor humidity, using allergen-impermeable bedding covers, minimizing the use of carpet and upholstered furniture, and minimizing pet exposure. For more information, see the Prevention section.

All patients with asthma should have a written action plan to follow if symptoms suddenly become worse, including how to adjust medication and when to seek medical help. A Northwestern University study indicates that asthma symptoms and the need for emergency medications in children can be greatly reduced by using a planned-care method. This method involves regularly scheduled visits with specially trained nurses to help the patient and family learn how to anticipate and improve the management of asthma symptoms.

The health care provider should write out an asthma treatment plan for the child's school personnel or care providers. The plan should detail the early warning signs of an asthma attack, what medications the student uses and how they are taken, and when to contact the doctor or seek emergency care. Children with asthma often need medication at school to control acute symptoms or to prevent exercise-induced attacks. Proper management will usually allow a child to take part in play activities. Only as a last resort should activities be limited.

Alternative treatment

Alternative and complementary therapies include approaches considered to be outside the mainstream of traditional health care. Alternative treatments for asthma include **yoga** to control breathing and relieve stress and acupuncture to reduce asthma attacks and improve lung function. Biofeedback, which teaches patients how to direct mental thoughts to influence physical functions, may be helpful for some patients. For example, learning to increase the amount of air inhaled may help some patients reduce **fear** and anxiety. Some Chinese traditional herbs, such as *ding-chan tang*, have been thought to help decrease inflammation and relieve bronchospasm.

Before learning or practicing any particular technique, it is important for the parent or caregiver and child to learn about the therapy, its safety and effectiveness, potential side effects, and the expertise and qualifications of the practitioner. Although some practices are beneficial, others may be harmful to certain patients.

Relaxation techniques and dietary supplements should not be used as a substitute for medical therapies prescribed by a doctor. Parents should discuss these alternative treatments with the child's doctor to determine the techniques and remedies that may be beneficial.

Nutritional concerns

Some children have reportedly experienced improved symptoms by limiting dairy products and sugar in the diet. Some studies show that vitamin C helps improve asthma symptoms.

Food additives may trigger asthma symptoms in some children, although this is rare. If the parent suspects that certain foods trigger asthma symptoms in the child, the pediatrician may recommend keeping a food diary for a few weeks to identify problematic foods. Allergy skin testing may be recommended to rule out foods that may trigger asthma symptoms.

Prognosis

Although there is no cure for asthma, it can be treated and managed. Most patients with asthma respond well and are able to lead relatively normal lives when the best drug or combination of drugs is found. Asthma should not be a progressive, disabling disease; a child with asthma can have normal or near-normal lung function with the proper treatment.

Some children stop having attacks as they grow and their airways get bigger. About 50 percent of children have less frequent and less severe attacks as they grow older. However, symptoms can recur when the child reaches his or her thirties or forties.

KEY TERMS

Acute—Refers to a disease or symptom that has a sudden onset and lasts a relatively short period of time.

Allergen—A foreign substance that provokes an immune reaction or allergic response in some sensitive people but not in most others.

Allergy—A hypersensitivity reaction in response to exposure to a specific substance.

Alveoli—The tiny air sacs clustered at the ends of the bronchioles in the lungs in which oxygen-carbon dioxide exchange takes place.

Anti-inflammatory—A class of drugs, including nonsteroidal anti-inflammatory drugs (NSAIDs) and corticosteroids, used to relieve swelling, pain, and other symptoms of inflammation.

Atopy—A state that makes persons more likely to develop allergic reactions of any type, including the inflammation and airway narrowing typical of asthma.

Bronchial tubes—The major airways to the lungs and their main branches.

Bronchioles—Small airways extending from the bronchi into the lobes of the lungs.

Bronchospasm—The tightening of the muscle bands that surround the airways, causing the airways to narrow.

Dander—Loose scales shed from the fur or feathers of household pets and other animals. Dander can cause allergic reactions in susceptible people.

Dust mites—Tiny insects, unable to be seen without a microscope, that are present in carpet, stuffed animals, upholstered furniture, and bedding, including pillows, mattresses, quilts, and other bed covers. Dust mites are one of the most common asthma triggers. They grow best in areas with high humidity.

Hypersensitivity—A condition characterized by an excessive response by the body to a foreign substance. In hypersensitive individuals even a tiny amount of allergen can cause a severe allergic reaction.

Inflammation—Pain, redness, swelling, and heat that develop in response to tissue irritation or injury. It usually is caused by the immune system's response to the body's contact with a foreign substance, such as an allergen or pathogen.

Peak flow measurement—Measurement of the maximum rate of airflow attained during a forced vital capacity determination.

Pollen—A fine, powdery substance released by plants and trees; an allergen.

Spirometry—A test using an instrument called a spirometer that measures how much and how fast the air is moving in and out of a patient's lungs. Spirometry can help a physician diagnose a range of respiratory diseases, monitor the progress of a disease, or assess a patient's response to treatment.

Trigger—Any situation or substance that causes asthma symptoms to start or become worse.

A small number of patients will have progressively more difficulty breathing. These patients have an increased risk of respiratory failure, and they must receive intensive treatment. Asthma can be a deadly disease if it is not managed properly; an estimated 5,000 people die each year from asthma or its complications.

Prevention

Prolonged breastfeeding in infants for six to 12 months has been shown to reduce the child's likelihood for developing persistent asthma.

Minimizing exposure to allergens

There are a number of ways parents can reduce or prevent a child's exposure to the common allergens and irritants that provoke asthmatic attacks:

- If the child is sensitive to a family pet, the pet should be removed or kept out of the child's bedroom (with the bedroom door closed). The pet should be kept away from carpets and upholstered furniture. All products made from feathers should be removed. An air filter should be used on air ducts in the child's room.
- To reduce exposure to house dust mites, wall-to-wall carpeting should be removed, humidity should be kept down, and special pillow and mattress covers should be used. The number of stuffed **toys** should be reduced, and they should be washed in hot water weekly. Bedding should also be washing weekly in hot water, and dried in a dryer on the hot setting. The child should not be allowed to **sleep** on upholstered furniture. Carpets should be removed from the child's bedroom.
- If cockroach allergen is causing asthma attacks, the roaches should be killed (using poison, traps, or boric

acid rather than chemicals). Food or garbage should not be exposed.

- Indoor air may be kept clean by vacuuming carpets once or twice a week (with the child absent), avoiding humidifiers, and using air conditioning during warm weather (so that windows remain closed).
- To reduce exposure to mold, indoor humidity should be decreased to less than 50 percent, leaky faucets and pipes should be repaired, and vaporizers avoided.
- Family members should quit smoking and others should not be allowed to smoke in the house or near the child.
- The child should not exercise outdoors when air pollution levels are high.

Parental concerns

Parents should take an open and honest approach when explaining asthma to their child. They should explain that asthma does not define or limit the child. The success of the child's treatment plan will depend on parental guidance and support. As a child ages, the responsibility for personal asthma management can be increased. For example, toddlers can mimic treatment on a toy or doll; preschoolers can help parents in peak flow monitoring and discuss symptoms with them; school-aged children can begin to take medications on their own (while supervised); and adolescents can be nearly independent in following the structured management plan.

Parents should stress the consequences of improper symptom management with their child. The main concern with older children is **peer pressure** and the desire to fit in; therefore, symptoms may not be reported accurately and medications may not be taken to avoid comments from peers or appearing different. Parents may want to counteract peer pressure by offering a contract that outlines the management plan and lists specific rewards and consequences.

Parents should work with school personnel to foster a supportive environment that so the child's symptoms can be managed properly. A specific action plan can be developed for school by the child's doctor. Parents should inform school personnel about the child's specific allergens and asthma triggers so steps can be taken to help the child avoid them at school. Students who are able to recognize symptoms requiring medication and know how to use their inhaler properly should be permitted to keep the medication with them. For younger children, parents must ensure that school personnel know how to administer the child's medications.

Asthma should not be used as an excuse to avoid exercise. Sometimes children with asthma avoid school activities because they are afraid of being embarrassed if symptoms occur. Parents should encourage athletic or physical activity participation and talk to gym teachers or coaches to ensure they understand the child's symptoms and treatment protocol. They should make sure the child knows what to do if exercise causes symptoms. Swimming is generally well-tolerated by many people with asthma because it is usually performed in a warm, moist environment. Other activities that involve brief, intermittent periods of exertion, such as volleyball, gymnastics, baseball, walking, and wrestling are usually well-tolerated. Cold-weather **sports**, such as skiing, ice skating, or hockey, may be not be tolerated as well. The child's doctor can provide specific exercise recommendations and guidelines.

See also Allergy shots.

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Allergy and Asthma Network/Mothers of Asthmatics America, Inc. 2751 Prosperity Ave., Suite 150, Fairfax, VA 22031. (800) 878-4403. Web site: <www.aanma.org.>.

American Academy of Allergy, Asthma and Immunology (AAAAI). 611 E. Wells St., Milwaukee, WI 53202. (800) 822-ASTHMA or (414) 272-6071. Web site: <www.aaaai.org>.

American College of Asthma, Allergy and Immunology (AACI). 85 W. Algonquin Rd., Suite 550, Arlington Hts., IL 60005. (800) 842-7777. Web site: <www. aaci.org.>.

American Lung Association. 1740 Broadway, New York, NY 10019. (800) 586-4872. Web site: <www.lungusa.org.>.

Asthma and Allergy Foundation of America. 1233 20th Street, NW, Suite 402, Washington, DC 20036. (800) 727-8462 or (202) 466-7643. Web site: <www.aafa.org>.

National Asthma Education Program. National Heart, Lung and Blood Institute Information Center. P.O. Box 30105, Bethesda, MD 20824-0105. (301) 592-8573. Web site: <www.nhlbi.nih.gov/about/naepp/>.

National Institute of Allergy and Infectious Diseases. NIAID Office of Communications and Public Liaison, Building 31, Room 7A-50, 31 Center Dr., MSC 2520, Bethesda, MD 20892-2520. Web site: <www.niaid.nih.gov>.

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Ataxia *see* **Movement disorders**

Ataxia telangiectasia/chromosome breakage disorders

Definition

Ataxia telangiectasia (A-T), also called Louis-Bar syndrome or cerebello-oculocutaneous telangiectasia, is a rare, inherited disease that attacks the neurological and immune systems of children. A-T is a recessive disorder, meaning that it affects children who carry two copies of a defective (mutated) A-T gene, one copy from each parent. A-T affects the brain and many parts of the body and causes a wide range of severe disabilities.

Description

Ataxia means poor coordination, and the telangiectasia are tiny, red spider blood vessels which develop in A-T patients, especially on the whites of the eyes and on the surface of the ears. A-T is a progressive disease that affects the cerebellum (the body's motor control center) and, in about 70 percent of cases, weakens the immune system as well, leading to respiratory disorders. The weakening of the immune system (**immunodeficiency**) resulting from A-T has been traced to defects in both B-cells and T-cells, the specialized white blood cells (lymphocytes) that defend the body against infection, disease, and foreign substances. In A-T children, B-cell responses are very weak, and levels of immunoglobulins, the proteins that B-cells make to fight infection by specific recognition of invading organisms, may also be low. T-cells are few and weak, and the thymus gland is immature. This is why A-T is also considered an immunodeficiency disease. A-T first shows itself in early childhood, usually at the toddler stage. The characteristic symptoms are lack of balance, slurred speech, and perhaps a higher-than-normal number of infections. All children at this age take a little while to develop good walking skills, coherent speech, and an effective immune system, so it often takes a few years before A-T is correctly diagnosed. Other features of the disease may include mild diabetes, premature graying of the hair, difficulty swallowing, and delayed physical and sexual development. Children with A-T usually have normal or above normal **intelligence**, but some cases of **mental retardation** have been reported.

Transmission

A-T is genetically transmitted by parents who are carriers of the gene responsible for A-T. The A-T mode of inheritance is autosomal recessive (AR) and requires two copies of the predisposing gene—one from each parent—for the child to have the disease. Parents do not exhibit symptoms, but they each carry a recessive gene that may cause A-T in their offspring. In AR families, there is one chance in four that each child born to the parents will have the disorder. Every healthy sibling of an A-T patient has a 66 percent chance of being a carrier, like the parents.

Demographics

According to the National **Cancer** Institute, the incidence of A-T is between one out of 40,000 and one out of 100,000 persons worldwide, and for Caucasians it is about three per million, so the disorder is very rare. In the United States, there are about 500 children with A-T with both males and females equally affected. An estimated 1 percent (2.5 million) of the general population carries one of the defective A-T genes. Carriers of one copy of this gene do not develop A-T but have a significantly increased risk of cancer (over 38 percent of children with A-T develop cancer).

Causes and symptoms

A-T is a genetic disorder, meaning that it is caused by a defect in a gene that is present in a person at birth. All people have genes that contain a few mistakes or variations that do not result in a disorder. Disorders result when the gene variations are significant enough to affect the function a gene controls. Variations that cause disease are called mutations and A-T results from a defective gene, the ATM gene (for ataxia telangiectasia, mutated), first identified in 1995. The ATM gene is located on the long arm of chromosome 11 at position 11q22-23. It encodes for (controls) the production of a protein that plays a role in regulating cell division following DNA damage. The various symptoms seen in A-T reflect the main role of this protein, which is to induce several cellular responses to DNA damage. The protein made by the ATM gene is located in the nucleus of the cell and normally functions to control the rate at which the cell grows. The ATM protein does this by sending

signals and modifying other proteins in the cell, which then changes the function of the proteins. The ATM protein also interacts with other special proteins when DNA is damaged as a result of exposure to some type of radiation. If the strands of DNA are broken, the ATM protein coordinates DNA repair by activating repair proteins, which helps to maintain the stability of cells. Mutations in ATM prevent cells from repairing DNA damage, which may lead to cancer. Mutations can also signal cells in the brain to die inappropriately, causing the movement and coordination problems associated with A-T.

A-T affects several different organs in the body. The most important symptoms are as follows:

- Neurologic abnormalities resulting in poor coordination and an unsteady gait (ataxia). Shortly after learning to walk, children with A-T begin to stagger. They tend to sway when they stand or sit and wobble when they walk. Jerking and tremors are present in about 25 percent of patients. This symptom results from neurologic abnormalities affecting the cerebellum that controls balance. Writing is affected by seven or eight years of age.
- Dilated blood vessels (telangiectasia). Telangiectasias usually occur on the white portion of the eye or on the ears, neck and extremities.
- Variable immunodeficiency resulting in increased vulnerability to infections. This symptom is a major feature in some individuals. The infections most commonly involve the lungs and sinuses and are usually of bacterial or viral origin. About 10 percent of patients have severe immunodeficiency.
- Predisposition to certain types of cancer. At least 10 percent of all A-T patients, including adults, develop cancer. Most of these are cancers of the lymphoid tissues (leukemias and lymphomas), but one fifth of the cancers occur in the stomach, brain, ovary, skin, liver, larynx, parotid gland, and breast.

Additional clinical symptoms include the following:

- autosomal recessive inheritance of the ATM gene
- involuntary, rapid, rhythmic movement of the eyeball (nystagmus)
- impaired ability to coordinate certain types of eye movements (oculomotor apraxia)
- squint of ocular muscles
- speech defect (dysarthria)
- slow, writhing motions (choreoathetosis)
- lack of T-lymphocytes (thymic aplasia)
- albinism of hair
- decreased to absent deep tendon reflexes
- multiple skin changes including eczema and "coffee-with-milk"x colored spots
- incomplete development of tonsils, lymph nodes, and spleen (hypoplasia)
- seizures (any type)
- abnormal ovaries
- small testes
- high blood sugar levels (hyperglycemia)

When to call the doctor

A-T children appear normal as infants. The decreased coordination of movements (ataxia) associated with A-T first becomes apparent when a child begins to walk, typically between 12 and 18 months of age. Toddlers with A-T are usually wobbly walkers. In their **preschool** years, children with A-T begin to stumble and fall, and drooling is frequent. Parents should contact their pediatrician if they observe any A-T signs or symptoms in their child. Telangiectasias are another typical warning sign. They become apparent after the onset of the ataxia, often between two and eight years of age.

Diagnosis

Establishing a diagnosis for ataxia telangiectasia is most difficult in very young children, primarily because the full-blown syndrome is not yet apparent. As of 2004, the A-T diagnosis is usually based on the characteristic clinical findings and supported by laboratory tests that point to a defect of DNA (genes and chromosomes) and to an inability to repair some types of damage to DNA. Laboratory tests are helpful but not as important as the individual patient's symptoms and signs, **family** history, and complete neurological evaluation including a **magnetic resonance imaging** (MRI) scan of the brain. The cerebellum atrophies early in the disease, being visibly smaller on MRI examination by seven or eight years of age. Diagnosis is more difficult before the disorder has fully developed, when the child is still uncertain on his/her feet. The most difficult time to diagnose A-T is during the period when neurologic symptoms start to appear (early childhood) and the typical telangiectasias have not yet appeared. During this period, a history of recurrent infections and typical immunologic findings can suggest the diagnosis. Four tests are used to help establish the A-T diagnosis:

- Increased alpha-fetoprotein levels in blood. Alpha-fetoproteins are fetal proteins that are usually produced during fetal development but may persist at high blood levels after birth. The vast majority of A-T patients

(more than 95 percent) have elevated levels of serum alpha-fetoprotein. This test is considered good but yields similar results for other conditions.

- Decreased immunoglobulin levels (Iga, IgG, IgM). Approximately 30 percent of patients with A-T have immunodeficiency. The drawback of this test is that immunoglobulin levels are not always low for A-T, and they are also low in other conditions.
- Sequence analysis. Sequence analysis of the ATM coding region is available on a clinical basis. Sequencing detects more than 95 percent of ATM sequence alterations but significant difficulties exist in distinguishing normal variations from A-T-causing mutations.
- Increased chromosome breaks and rearrangements. Individuals with A-T have an increased frequency of spontaneous breaks in their chromosomes as well as an increased frequency of chromosomal rearrangements. These chromosomal abnormalities often occur close to genes that control the function of white blood cells, such as immunoglobulins and T-lymphocytes. The frequency of chromosomal breaks is increased when T-lymphocytes are exposed to **x rays** in the laboratory, and this sensitivity to ionizing irradiation forms the basis for a specialized A-T diagnostic test.

The ionizing irradiation sensitivity test is the most useful test for diagnosing A-T. However, it can only be carried out in specialized centers and takes much longer than the other tests.

Because of its variable symptoms, A-T is often misdiagnosed as a form of **cerebral palsy** or as slow development.

Treatment

As of 2004, there is no cure for ataxia telangiectasia, thus specific therapy is not available, and treatment is largely supportive. Patients are encouraged to participate in as many activities as possible. Children are encouraged to attend school on a regular basis and receive support to maintain as normal a lifestyle as possible. The following are some types of interventions have been shown to help those with the disorder:

- Exercise and physiotherapy. These programs help prevent the development of stiffness in muscles and help maintain functional mobility, showing A-T patients how to best use muscle control and stretch muscles and ligaments.
- Antibiotics, immunoglobulins, vaccinations. For patients who have normal levels of immunoglobulins and normal antibody responses to vaccines, immunization with **influenza** and pneumococcal vaccines may be helpful. For patients with total IgG deficiencies or patients who have problems making normal antibody responses to vaccines, therapy with gammaglobulin may be indicated.
- Speech therapy. Speech therapy can significantly improve diction, especially in the second decade of the disorder.
- Orthopedic referral/assessment. Corrective procedures can be helpful for joint or postural problems, particularly in the lower limbs or spine.

Because cells from patients with A-T are 30 percent more sensitive to ionizing radiation than the cells of normal individuals, any required radiotherapy or **chemotherapy** should be reduced or monitored carefully; conventional doses are contraindicated and are potentially lethal.

Alternative treatment

No single alternative medicine or herbal remedy can help people with A-T. The use of thymic transplants and hormones has not led to improvement. Similarly, there is no scientific evidence as of 2004 that any specific supplemental nutritional therapy is beneficial.

Concerning drug therapy, most drugs which act on the nervous system can cause problems in A-T. Some people have found Benzhexol beneficial, but others have suffered reactions to it. Drug therapy for A-T remains in 2004 experimental and accordingly requires highly specialized A-T clinical teams.

Since the 1995 isolation of the ATM gene, scientists have worked very hard to understand how the ATM protein is activated or turned on following damage to a cell's DNA. This knowledge is in turn being used to develop A-T treatment approaches. The following are among the most promising:

- Gene therapy: Scientists are starting to test the efficiency of gene therapy protocols in mice and are simultaneously developing a new gene therapy protocol for A-T which would allow for stable, long-term production of the ATM protein.
- Neural stem cells: Researchers have demonstrated a significant therapeutic effect by using neural stem cells in mice that have a pattern of neurodegeneration similar to A-T.
- Bone marrow transplantation: Significant progress has been made in the development of a successful bone marrow transplantation protocol in mice with A-T. Researchers are testing how effectively this protocol prevents immune abnormalities and immune-related cancers in these mice.

KEY TERMS

Albinism—An inherited condition that causes a lack of pigment. People with albinism typically have light skin, white or pale yellow hair, and light blue or gray eyes.

Allele—One of two or more alternate forms of a gene.

Ataxia—A condition marked by impaired muscular coordination, most frequently resulting from disorders in the brain or spinal cord.

Atrophy—The progressive wasting and loss of function of any part of the body.

B-cell (B lymphocyte)—A small white blood cell from bone marrow responsible for producing antibody and serving as a precursor for plasma cells.

Carrier—A person who possesses a gene for an abnormal trait without showing signs of the disorder. The person may pass the abnormal gene on to offspring. Also refers to a person who has a particular disease agent present within his/her body, and can pass this agent on to others, but who displays no symptoms of infection.

Central nervous system—Part of the nervous system consisting of the brain, cranial nerves, and spinal cord. The brain is the center of higher processes, such as thought and emotion and is responsible for the coordination and control of bodily activities and the interpretation of information from the senses. The cranial nerves and spinal cord link the brain to the peripheral nervous system, that is the nerves present in the rest of body.

Cerebellum—The part of the brain involved in the coordination of movement, walking, and balance.

Chromosome—A microscopic thread-like structure found within each cell of the human body and consisting of a complex of proteins and DNA. Humans have 46 chromosomes arranged into 23 pairs. Chromosomes contain the genetic information necessary to direct the development and functioning of all cells and systems in the body. They pass on hereditary traits from parents to child (like eye color) and determine whether the child will be male or female.

Diabetes—A disease characterized by an inability to process sugars in the diet, due to a decrease in or total absence of insulin production.

DNA—Deoxyribonucleic acid; the genetic material in cells that holds the inherited instructions for growth, development, and cellular functioning.

Fetal proteins—Proteins that are usually produced during fetal development but may persist at high blood levels in some conditions (such as A-T) after birth. The vast majority of A-T patients (more than 95%) have elevated levels of serum alpha-fetoprotein.

Gene—A building block of inheritance, which contains the instructions for the production of a particular protein, and is made up of a molecular sequence found on a section of DNA. Each gene is found on a precise location on a chromosome.

Immune response—A physiological response of the body controlled by the immune system that involves the production of antibodies to fight off specific foreign substances or agents (antigens).

Immune system—The system of specialized organs, lymph nodes, and blood cells throughout the body that work together to defend the body against foreign invaders (bacteria, viruses, fungi, etc.).

Immunodeficiency—A condition in which the body's immune response is damaged, weakened, or is not functioning properly.

Immunoglobulin G (IgG)—Immunoglobulin type gamma, the most common type found in the blood and tissue fluids.

Leukemia—A cancer of the blood-forming organs (bone marrow and lymph system) characterized by an abnormal increase in the number of white blood cells in the tissues. There are many types of leukemias and they are classified according to the type of white blood cell involved.

Lymphocyte—A type of white blood cell that participates in the immune response. The two main groups are the B cells that have antibody molecules on their surface and T cells that destroy antigens.

Lymphocytic leukemia—An acute form of childhood leukemia characterized by the development of abnormal cells in the bone marrow.

Lymphoma—A diverse group of cancers of the lymphatic system characterized by abnormal growth of lymphatic cells. Two general types are commonly recognized–Hodgkin's disease and non-Hodgkin's lymphoma.

Magnetic resonance imaging (MRI)—An imaging technique that uses a large circular magnet and radio waves to generate signals from atoms in the body. These signals are used to construct detailed images of internal body structures and organs, including the brain.

KEY TERMS (contd.)

Motor skills—Controlled movements of muscle groups. Fine motor skills involve tasks that require dexterity of small muscles, such as buttoning a shirt. Tasks such as walking or throwing a ball involve the use of gross motor skills.

Neurodegenerative disease—A disease in which the nervous system progressively and irreversibly deteriorates.

Nystagmus—An involuntary, rhythmic movement of the eyes.

Progressive—Advancing, going forward, going from bad to worse, increasing in scope or severity.

Recessive disorder—Disorder that requires two copies of the predisposing gene one from each parent for the child to have the disease.

Stem cell—An undifferentiated cell that retains the ability to develop into any one of a variety of cell types.

T cell—A type of white blood cell that is produced in the bone marrow and matured in the thymus gland. It helps to regulate the immune system's response to infections or malignancy.

Telangiectasia—Abnormal dilation of capillary blood vessels leading to the formation of telangiectases or angiomas.

Thymic aplasia—A lack of T lymphocytes, due to failure of the thymus to develop, resulting in very reduced immunity.

- High throughput drug screening: Testing methods are also being developed to help scientists screen large numbers of already-approved drugs as well as new compounds to see if they are useful for treating A-T.
- Transplants of thymus tissue: The new approaches that medical researchers are testing also include transplants of thymus tissue to boost the immune system.

Clinical trials

Parents may consider enrolling their A-T diagnosed child in a NIH-approved clinical trial. The first-ever A-T clinical treatment study took place at Children's Hospital in Philadelphia, with a second trial that started in 2000. In 2004, the A-T Clinical Center at Johns Hopkins Hospital in Baltimore also started a clinical study. Children who participate in these clinical trials receive complete immunological and neurological evaluations as part of being enrolled in the study. Many patients also receive nutritional evaluations and consultations as well.

Nutritional concerns

Some A-T patients have impaired swallowing function. Patients who aspirate or have food and liquids reaching their lungs have been shown to improve when thin liquids are removed from their diet. In some individuals, a tube from the stomach to the outside of the abdomen (gastrostomy tube) may be required to eliminate the need for swallowing large volumes of liquids and to decrease the risk of aspiration. Vitamin E supplements are often recommended, although the vitamin has not been formally tested for efficacy in patients with A-T.

Prognosis

Generally, the prognosis for individuals with A-T is poor. Those with the disease are frequently wheelchair-bound by their teens and usually die in their teens or early 20s. However, the course of the disease can be quite variable, and it is difficult to predict the outcome for any given individual as A-T varies considerably from patient to patient. Even within families, in which the specific genetic defect should be the same, some children have mostly neurologic problems while others have recurrent infections, and still others have neither neurologic problems nor recurrent infections.

There was no cure for A-T as of 2004. The cloning and sequencing of the ATM gene has opened several avenues of research with the goal of developing better treatment, including gene therapy and the design of drugs for more effective treatments. Research is also leading to a greater understanding of AT, increased awareness, and more genetic counseling.

Prevention

In the past, A-T carriers were identified because they were parents of a child diagnosed with A-T. But the cloning of the ATM gene responsible for A-T as of 2004 allows physicians or cancer genetics professionals to conduct genetic testing, analyzing patients' DNA to look for A-T mutations in the ATM gene. Thus, prenatal diagnosis can be carried out in most families. Genetic counseling is also of benefit to prospective parents with a family history of ataxia-telangiectasia. Parents of a child

diagnosed with A-T may have a slight increased risk of cancer. They should have genetic counseling and more intensive screening for cancer.

Parental concerns

Any family touched by ataxia telangiectasia is forever affected. Old assumptions have to be discarded and new, often very difficult, realities need to be accepted, including the uncertainty of the A-T outcome. Significant adjustments, both physical and psychological, are required, many of them agonizingly difficult. A-T support groups have been organized by all major A-T organizations, such as the Ataxia Telangiectasia Children's Project, the National Ataxia Foundation (NAF), and the Ataxia Telangiectasia Medical Research Foundation. These organizations are dedicated to improving the lives of families affected by A-T. They also provide the latest news on A-T research, information on coping with A-T, and personal accounts of living with A-T.

See also Immunodeficiency; Magnetic resonance imaging.

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Ataxia Telangiectasia (A-T) Medical Research Foundation. 5241 Round Meadow Road, Hidden Hills, CA 91302. Web site: <www.gspartners.com/at/>.

National Ataxia Foundation (NAF). 2600 Fernbrook Lane, Suite 119, Minneapolis, MN 55447–4752. (763) Web site: <www.ataxia.org>.

National Institute of Child Health and Human Development (NICHD). 31 Center Drive, Rm. 2A32, MSC 2425, Bethesda, MD 20892–2425. Web site: <www.nichd.nih.gov>.

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Atopic dermatitis

Definition

Atopic **dermatitis** (AD) is a chronic skin disorder associated with biochemical abnormalities in the patient's body tissues and immune system. It is characterized by inflammation, **itching**, weepy skin lesions, and an individual or **family** history of **asthma**, hay fever, **food allergies**, or similar allergic disorders. Atopic dermatitis is also known as infantile eczema or atopic eczema. The word atopic comes from *atopy*, which is derived from a Greek word that means "out of place." Atopy is a genetic predisposition to type I (immediate) hypersensitivity reactions to various environmental triggers. It includes bronchial asthma and food **allergies** as well as atopic dermatitis.

Description

AD varies in severity but in general is characterized by red, weeping, crusted patches of inflamed skin that itch constantly. The distribution of the skin lesions depends on the child's age. In infants, the skin lesions are usually found on the face, scalp, diaper area, body folds, hands, and feet, and tend to be exudative (oozing fluid that has escaped from blood vessels as a result of inflammation). Infants old enough to crawl may have patches of inflamed skin on the neck and trunk as well. In older children, the affected areas are usually located on the wrists, ankles, back of the neck, insides of the elbows, and the backs of the knees. The skin lesions in older children are more likely to be lichenified than exudative. Lichenification is the medical term for a leather- or bark-like thickening of the outermost layer of skin cells (the epidermis) as a result of long-term scratching or rubbing of itching lesions. In addition, the normal markings of the skin are exaggerated in lichenification.

The lesions of AD are accompanied by intense pruritus, which is the medical term for itching. Children with atopic dermatitis often have a lowered threshold of sensitivity to itching, which means that they feel itching sensations more intensely than children without the disorder. The pruritus often creates a vicious cycle of itching and scratching, which leads to more widespread rash, which leads to more itching. The child may scratch the affected skin only intermittently during the day, however. It is common for children with AD to do more scratching in the early evening and at night; moreover, disruptions of normal **sleep** patterns are common in these children.

Transmission

Atopic dermatitis is not contagious but may affect several members of the same family at the same time.

Demographics

Atopic dermatitis is a very common condition in the general population. According to the National Institute of Arthritis and Musculoskeletal and Skin Diseases (NIAMS), about 15 million people in the United States have one or more symptoms of the disease. It accounts for 15 to 20 percent of all visits to dermatologists (doctors who specialize in treating diseases of the skin). About 20 percent of infants develop symptoms of atopic dermatitis. Moreover, the proportion of people affected by AD is increasing; the American Academy of Allergy, Asthma, and Immunology (AAAAI) began a long-term study in 1999 that indicates that a larger percentage of children are affected by AD than was the case in the 1980s. This rise in prevalence is true of all developed countries, not just the United States and Canada. People who immigrate to Europe or North America from underdeveloped countries have increased rates of atopic dermatitis, which suggests that environmental factors play a role in the development or triggering of the disorder.

Atopic dermatitis begins early in life; about 65 percent of patients with AD develop symptoms during the first 12 months of life, with 90 percent showing symptoms before five years of age. The most common age for the onset of symptoms in infants is between six and 12 weeks of age. It is unusual for adults over the age of 30 to develop AD for the first time.

There is some disagreement among researchers with regard to race or ethnicity as risk factors for atopic dermatitis. Some studies indicate that all races and ethnic groups are equally at risk, while others suggest that Asians and Caucasians have slightly higher rates of AD than African Americans or Native Americans. Some skin lesions typical of AD may be more difficult to evaluate in African Americans because of the underlying skin pigmentation. With regard to sex, males and females appear to be equally at risk.

Atopic dermatitis is a major economic burden on families with children affected by the disorder. One researcher in Australia stated that the stresses on families with children diagnosed with moderate or severe AD are greater than the burdens on families with children with type 1 diabetes. These stresses include loss of sleep, loss of employment for the parents, time taken for direct care of the skin disorder, and the financial costs of treatment. The National Institutes of Health (NIH) estimates that atopic dermatitis costs U.S. health insurance companies more than $1 billion every year.

Causes and symptoms

Causes

The causes of atopic dermatitis were not completely understood as of 2004 but are thought to be a combination of genetic susceptibility, damaged skin barrier function, and abnormal responses of the child's immune system to environmental triggers. With regard to genetic factors, the disorder has been tentatively linked to loci on chromosomes 11 and 13. A child with one parent with AD has a 60 percent chance of developing the disorder; if both parents are affected, the risk rises to 80 percent. Nearly 40 percent of newly diagnosed children have at least one first-degree relative with atopic dermatitis.

In addition to genetic susceptibility, AD is the end result of a complex inflammatory process involving

abnormalities in the child's skin and immune system. Some researchers have noted that the skin of people with AD contains lower levels of fatty acids, which may cause the skin to lose moisture more readily and become more sensitive to chemicals and other irritants. Others point to decreased production of a hormone in the immune system called interferon-gamma that ordinarily helps to regulate the body's response to allergens. People with AD may be hypersensitive to irritants because they have abnormally low levels of interferon-gamma in their systems.

About 80 to 90 percent of children with AD also have unusually high levels of an antibody called IgE in their blood. Antibodies are specialized proteins produced by the immune system that seek out and destroy bacteria, viruses, and other invaders. The high levels of IgE in the blood of AD patients are produced by hyperactive T helper 2 cells reacting against antigens in the environment. Although the role of increased IgE production in the development of atopic dermatitis was not fully understood as of 2004, measuring the level of this antibody in a sample of blood serum may be done to help distinguish AD from other skin diseases with similar symptoms.

Symptoms

The basic symptoms of AD have already been described. Dermatologists classify the lesions of AD into three basic categories:

- Acute lesions: These include extremely itchy reddened papules (small solid eruptions resembling pimples) and vesicles (small blister-like elevations in the skin surface that contain tissue fluid) over erythematous (reddened) skin. Acute lesions produce a watery exudate and are often accompanied by exfoliation (scaling or peeling of layers of skin) and erosion (destruction of the skin surface).
- Subacute lesions: These are characterized by reddening, peeling, and scaling but are less severe than acute lesions and do not produce an exudate.
- Chronic lesions: These are characterized by thickened plaques of skin, lichenification, and fibrous papules.

It is possible for a child or adolescent with chronic atopic dermatitis to have all three types of lesions at the same time.

Associated symptoms and disorders

Children and adolescents with AD frequently develop one or more of the following disorders or problems:

- Asthma: About 50 percent of children diagnosed with AD eventually develop asthma.
- **Allergic rhinitis**: Between 70 and 75 percent of children with AD eventually develop a nasal allergy. Allergic **rhinitis**, which is sometimes called atopic rhinitis, may be either seasonal (hay fever or rose fever) or nonseasonal (caused by dust, mold spores, pet dander, cigarette smoke, and other household allergens).
- Eye complications: These include such disorders as **conjunctivitis** (inflammation of the tissue that lines the eyelid), keratoconus (a cone-shaped distortion of the cornea of the eye), and cataracts. Although cataracts are usually associated with older adults, between 4 and 12 percent of children with AD develop rapidly maturing cataracts that may begin to interfere with vision as early as age 20. About 1 percent develop keratoconus.
- Ichthyosis, xerosis (dry skin), lichenification, and other skin abnormalities not caused by infections: Children with AD are likely to develop other skin problems.
- Secondary skin infections: Children and adolescents with AD frequently develop infections from bacteria that live on the skin and multiply when the child's scratching causes breaks or open sores in the skin. Most of these secondary infections are caused by *Staphylococcus aureus* and *Streptococcus pyogenes*.
- Psychosocial problems: Children with atopic dermatitis may withdraw socially if the lesions are extensive or otherwise noticeable. In addition, children with severe cases may have frequent absences from school. Adolescents may suffer depression or **anxiety** related to concerns about their appearance or the need to avoid participating in **sports** in order to minimize sweating.

When to call the doctor

Atopic dermatitis is rarely a medical emergency and can often be treated by the child's pediatrician. Parents should, however, consider consulting a dermatologist, allergist, or immunologist under any of the following circumstances:

- The child's AD has been diagnosed as severe. This classification means that 20 percent of the body's skin surface has been affected or 10 percent of the skin area in addition to involvement of the eyes, hands, and body folds.
- There is extensive exfoliation (peeling and scaling) of the skin.
- The child has eye complications.
- The child has recurrent secondary bacterial infections.

- The child is frequently absent from school, has developed psychosocial complications, or has impaired quality of life. In many cases the entire family's quality of life is affected by the stresses and frustrations of coping with the disease, and other family members' reactions may in turn upset the child with AD.
- The child has had to be hospitalized for treatment of the AD.
- The child has had to take more than one course of oral steroid drugs.
- The diagnosis is uncertain.

Diagnosis

History and physical examination

Diagnosis of atopic dermatitis begins with a history-taking and physical examination by the child's doctor. In the case of infants or very young children, the doctor will ask the parents for information about a family history of atopic disorders as well as information about the onset of the symptoms. The doctor will then examine the child's skin and assess the following factors:

- physical appearance of the lesions and their distribution on the child's body
- timing, which includes seasonal variations in the severity of the rash as well as its chronic or recurrent nature
- environmental factors, which includes foods as well as such common triggers of AD as dust, pet dander, household cleaning agents, plastics, nail polish remover, and other cosmetics or chemicals
- presence of such other conditions associated with AD as eye complications or bacterial infections of the skin

The doctor will ask older children and adolescents directly whether their skin lesions are affected by such factors as pets in the household; **smoking**; using perfumes, shampoos, deodorants, or other personal care products; taking certain prescription medications; wearing wool or other rough-textured fabrics; using laundry detergents or fabric softeners; being exposed to extremes of temperature or humidity; athletic activity; emotional stress; and (in females past **puberty**) hormonal changes related to **menstruation**.

There are no laboratory tests that can confirm the diagnosis of AD; in some cases, the doctor may need to examine the child more than once in order to distinguish between atopic and **seborrheic dermatitis**. In most cases, the doctor will make the diagnosis on the basis of criteria established by the AAAAI in the 1990s. To be considered atopic dermatitis, the child's symptoms must at total at least three major and three minor symptom criteria.

There are four major criteria for AD:

- pruritus
- typical form and distribution of skin lesions
- chronic or recurrent dermatitis
- a personal or family history of atopic disorders

There are about two dozen minor criteria for atopic dermatitis. The most common minor characteristics are early age of onset, food intolerance, wool intolerance, susceptibility to skin infections, immediate type I response to skin test, elevated total serum IgE, eczema of the nipples, xerosis or dry skin, dermatitis of the hands and feet, recurrent conjunctivitis, sensitivity to emotional stress, and ichthyosis.

Family practitioners often refer patients with AD to an allergist for consultation, particularly if the child has developed asthma or has acute reactions to foods.

Laboratory tests

In addition to a general physical examination, the doctor may order a blood test to look for the presence of elevated IgE levels in the blood serum. The doctor may also test tissue fluid or smears from the child's lesions to rule out skin parasites or infections that mimic atopic dermatitis, such as bacterial infections, **scabies**, or herpesvirus infections.

The doctor may recommend skin prick testing to determine whether certain specific substances or foods trigger the child's AD. These tests are usually given only to children with moderate or severe cases of atopic dermatitis. The child must discontinue taking oral antihistamine medications for one week before the tests and discontinue using topical steroid creams for two weeks. The test is performed by pricking the surface of the skin with a thin needle containing a small amount of a suspected allergen.

Treatment

The AAAAI recommends a four-part approach to the treatment of atopic dermatitis. Children with AD should take the following steps:

- Avoid foods or other factors that trigger symptoms, avoid such irritating fabrics as wool and synthetic fibers, wear 100 percent cotton underwear, trim fingernails short to minimize damage to the skin from scratching, keep the skin moist with proper use of emollient creams or oils after bathing, avoid the use of

fabric softeners or scented detergents when laundering clothes and rinse clothes completely, and try to reduce emotional stress.

- Use appropriate medications as prescribed. The types of medications used vary depending on the severity of the child's symptoms and the presence of other infections. Most children are given both oral and topical (applied to the skin) medications. Topical medications include corticosteroid creams (Aristocort, Kenalog, Halog, Topicort, and many other brand names) and ointments containing immunomodulators, usually tacrolimus (Protopic) or pimecrolimus (Elidel). Corticosteroid creams are used to suppress inflammation, while the immunomodulator creams work by reducing the reactivity of the child's immune system. Although the corticosteroid creams have been used in both prescription-strength and over-the-counter (OTC) formulations for many years to treat AD, they may cause such side effects as thinning of the skin or stretch marks when used for long periods. They may also make skin infections worse. For these reasons, doctors recommend using the least powerful corticosteroid creams that control the symptoms. With regard to oral medications, **antihistamines** are often prescribed to stop itching at night so that the child can sleep. Oral or injected corticosteroids are sometimes used for short-term treatment of severe cases of AD that have not responded to topical medications; however, these drugs often have severe side effects, including stunted growth, thinning or weakening of the bones, high blood sugar levels, infections, and an increased risk of cataracts. Children with skin infections are usually given oral rather than **topical antibiotics**, most commonly penicillin or a cephalosporin.
- Regarding asthma or allergic rhinitis, the child should be evaluated for immunotherapy.
- The child's family and friends need to be educated about the condition, and the child needs to maintain a schedule of regular follow-ups. In addition to follow-up visits with the pediatrician and allergist, the child should have regular eye examinations as a safeguard against cataracts or other eye complications.

Other treatments that are sometimes used for atopic dermatitis are tar preparations and ultraviolet light therapy (phototherapy). Tar preparations are messy but were still as of 2004 considered useful for treating patients with chronic lichenified areas of skin. Phototherapy with ultraviolet A or B light waves, or a combination of both, may be used to treat older children or adolescents with mild or moderate atopic dermatitis; it is not suitable for infants or younger children. Some patients who do not respond to ultraviolet light alone benefit from a combination of phototherapy and an oral medication known as psoralen, which makes the skin more sensitive to the light. Phototherapy has two potential side effects from long-term use: premature aging of the skin and an increased risk of skin **cancer**.

Children or adolescents with AD must use extra care when bathing or showering. The doctor may recommend a non-soap skin cleanser, as standard bath soaps tend to dry and irritate the skin. If soaps are used, they should never be applied directly to broken or eroded areas of skin. The water should be lukewarm rather than hot, and the skin should be allowed to air-dry or be gently patted with a towel; brisk rubbing or the use of bath brushes must be avoided. After the skin has dried, the patient should apply a skin lubricant to seal moisture in the skin and create a barrier against further dryness or irritation.

Children with AD should also avoid unnecessary exposure to extremely hot, cold, moist, or dry outdoor environments. They should take care to avoid getting sunburned and should avoid participating in sports that involve physical contact or cause heavy perspiration.

Alternative treatment

There are a number of different complementary and alternative (CAM) approaches that have been used to treat atopic dermatitis, in part because the disorder is so widespread among children. In fact, infantile eczema is one of the most common conditions for which parents seek help from alternative practitioners. Most alternative therapies for atopic dermatitis fall into one of the following groups.

NATUROPATHY Naturopathy is a commonly used form of alternative treatment for AD; in one British study it was found effective for 19 out of 46 children in the subject group. Naturopaths favor food elimination diets as a way of managing AD, as well as lowering the child's overall intake of animal products. They recommend adding fish oil, flaxseed oil, or evening primrose oil to the child's diet to improve the condition of the skin, as many naturopaths believe that deficient intake of essential fatty acids is a major cause of AD. With regard to botanical products, a naturopath may suggest herbal preparations taken by mouth as well as topical creams made from herbs. Oral preparations may include extracts of hawthorn berry, blackthorn, or licorice root, while topical preparations to relieve itching typically include licorice or German chamomile. One German study found that a cream made with an extract of St. John's wort relieved the symptoms of AD better than a placebo, but the herbal preparation had not as of 2004 been compared to a standard corticosteroid cream.

HOMEOPATHY Homeopathy is the single most common CAM approach to atopic dermatitis in Europe, although it is frequently used in the United States as well. One German study followed a group of 2800 adults and 1130 children diagnosed with AD who were treated by homeopathic practitioners. The researchers found that over 600 different homeopathic remedies were recommended for the patients, although *Sepia*, *Lycopodium*, *Sulphur*, and *Natrum muriaticum* were the remedies most frequently prescribed. Most homeopathic practitioners in the United States as well as Europe consider AD a chronic condition that should be treated by constitutional homeopathic prescribing rather than by what is known as acute prescribing. In constitutional prescribing, the remedy is selected for long-term treatment of the patient's underlying susceptibility or constitutional weakness rather than short-term relief of present symptoms.

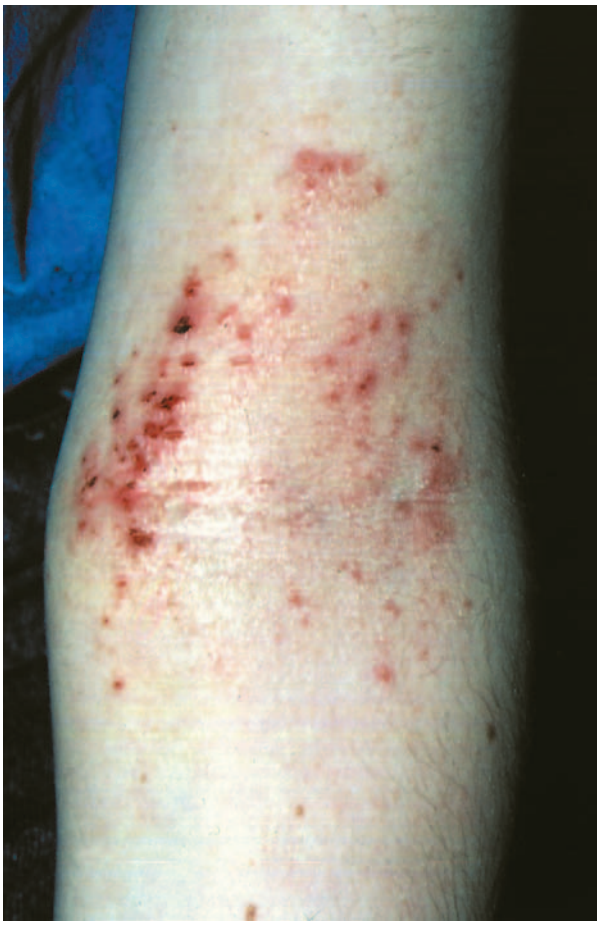
TRADITIONAL SYSTEMS OF MEDICINE According to Kenneth Pelletier, the former director of the alternative medicine program at Stanford University School of Medicine, both traditional Chinese remedies and Ayurvedic medicines benefit some people with atopic dermatitis. The British study of the use of CAM treatments in children with AD found that parents of Indian or Afro-Caribbean background were more likely to use these traditional approaches than Caucasian parents.

MIND/BODY APPROACHES Because flare-ups of AD are often related to increased emotional stress, some researchers have hypothesized that alternative approaches to lowering stress might help in treating the disorder. There is disagreement, however, about the effectiveness of such treatments as hypnosis or autogenic training. While some studies have reported that self-hypnosis, biofeedback, or autogenic training helped children with AD to manage their skin lesions with lower levels of steroid medications, other studies have reported that there is no conclusive evidence of the effectiveness of mind/body approaches in treating atopic dermatitis.

Nutritional concerns

Children and adolescents should avoid foods that trigger their AD. The most common offenders in flare-ups are peanuts and peanut butter, eggs and milk, seafood, soy, and chocolate. Long-term food elimination diets as a strategy for controlling AD are discussed below.

Children with moderate or severe AD often develop eroded areas or open cracks in the skin around the mouth from licking their lips or from allergic reactions to specific foods. They should apply a thin layer of petroleum jelly around the mouth before a meal to avoid irritation from citrus fruits, tomatoes, and other highly acidic foods.



Close-up view of atopic dermatitis in the crook of the elbow of a 12-year-old patient. *(SPL/Custom Medical Stock Photo, Inc.)*

Prognosis

As of the early 2000s, there is no cure for atopic dermatitis. People diagnosed with AD have highly individual combinations of symptoms that may vary greatly in severity over time. A significant percentage of children diagnosed with the condition, however, remain atopic into adulthood; one source states that 20 to 40 percent of children with infantile eczema continue to be affected, while NIAMS gives a figure of 60 percent. Some children included in these figures, however, outgrow the more severe forms of atopic dermatitis and suffer flare-ups in adult life only when they are exposed to high stress levels, chemical irritants, or other triggers in the environment. Other children may have only mild symptoms of AD until **adolescence**, when changes in hormone levels may cause a sudden worsening of symptoms.

Prevention

While atopic dermatitis in children cannot be completely prevented, NIAMS offers the following tips to

KEY TERMS

Allergen—A foreign substance that provokes an immune reaction or allergic response in some sensitive people but not in most others.

Atopy—A state that makes persons more likely to develop allergic reactions of any type, including the inflammation and airway narrowing typical of asthma.

Autogenic training—A form of self-hypnosis developed in Germany that appears to be beneficial to migraine sufferers.

Conjunctivitis—Inflammation of the conjunctiva, the mucous membrane covering the white part of the eye (sclera) and lining the inside of the eyelids also called pinkeye.

Dander—Loose scales shed from the fur or feathers of household pets and other animals. Dander can cause allergic reactions in susceptible people.

Dermatologist—A physician that specializes in diseases and disorders of the skin.

Eczema—A superficial type of inflammation of the skin that may be very itchy and weeping in the early stages; later, the affected skin becomes crusted, scaly, and thick.

Erythema—A diffuse red and inflamed area of the skin.

Exfoliate—To shed skin. In skin care, the term exfoliate describes the process of removing dead skin cells.

Exudation—Leakage of cells, proteins, and fluids through the blood vessel wall into the surrounding tissue.

Ichthyosis—A group of congenital skin disorders of keratinization characterized by dryness and scaling of the skin.

Keratin—A tough, nonwater-soluble protein found in the nails, hair, and the outermost layer of skin. Human hair is made up largely of keratin.

Keratoconus—An eye condition in which the central part of the cornea bulges outward, interfering with normal vision. Usually both eyes are affected.

Lichenification—Thickening of the outer layer of skin cells caused by prolonged scratching or rubbing and resulting in a leathery or bark-like appearance of the skin.

Papule—A solid, raised bump on the skin.

Pruritus—The symptom of itching or an uncontrollable sensation leading to the urge to scratch.

Rhinitis—Inflammation and swelling of the mucous membranes that line the nasal passages.

Scabies—A contagious parasitic skin disease caused by a tiny mite and characterized by intense itching.

Vesicle—A bump on the skin filled with fluid.

Xerosis—The medical term for dry skin. Many children diagnosed with atopic dermatitis have a history of xerosis even as newborns.

parents as they try to help control the severity and frequency of flare-ups:

- Keep the child from scratching or rubbing the affected areas whenever possible.
- Avoid dressing the child in rough or scratchy fabrics and protect his or her skin from high levels of moisture.
- Keep the house at a cool, stable temperature with a consistent humidity level, using a humidifier during the heating season in colder climates.
- Quit smoking and do not allow others to smoke inside the house.
- Limit the child's exposure to dust, pollen, and animal dander. Some doctors recommend installing special filters in the house to remove dust and pollen from the air, removing carpets from the floors, or encasing mattresses and pillows with special covers to control dust mites.
- Recognize when the child is under stress and lower the stress level in the household if possible.

Nutritional concerns

The doctor may suggest a food challenge in order to identify a food or foods that may be triggering the child's skin rash. In a food challenge, a particular food is eliminated from the child's diet for a few weeks and then reintroduced. In some cases, a child with AD may benefit from a longer-term diet that eliminates problem foods entirely. In these cases, however, the child's height and weight should be carefully monitored to make sure that the diet is nutritionally adequate, and the diet itself should be reevaluated every four to six months. The doctor may recommend vitamin supplements or a consultation with a dietitian.

Parental concerns

Parental concerns about atopic dermatitis extend to the possible long-term consequences of the disorder as well as the child's present discomfort and sleeping problems. Depending on the severity and location of the skin rash, the child may withdraw from social activities to avoid teasing or resent restrictions on athletic or other outdoor activities. In addition to such possible complications of AD as eye disorders and skin infections, parents must also be attentive to signs of long-term side effects caused by medications or other forms of treatment for the AD. To cope with the impact of AD on other family members, parents may find counseling and support groups helpful. Because atopic dermatitis is so widespread in the general population, many support groups have been formed, particularly in the larger cities.

See also Allergic rhinitis; Allergies; Asthma.

Resources

BOOKS

"Atopic Dermatitis." Section 10, Chapter 111 in *The Merck Manual of Diagnosis and Therapy*, edited by Mark H. Beers and Robert Berkow. Whitehouse Station, NJ: Merck Research Laboratories, 2002.

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ORGANIZATIONS

American Academy of Allergy, Asthma, and Immunology (AAAAI). 611 East Wells Street, Milwaukee, WI 53202. Web site: <www.aaaai.org>.

American Academy of Dermatology (AAD). PO Box 4014, Schaumburg, IL 60168–4014. Web site: <www.aad.org>.

National Institute of Arthritis and Musculoskeletal and Skin Diseases (NIAMS). 1 AMS Circle, Bethesda, MD 20892–3675. Web site: <www.niams.nih.gov>.

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Krafchik, Bernice R. "Atopic Dermatitis." *eMedicine*, January 23, 2002. Available online at <www.emedicine.com/derm/topic38.htm> (accessed November 22, 2004).

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Rebecca Frey, PhD

Atrial septal defect

Definition

An atrial septal defect (ASD) is an abnormal opening in the muscular wall separating the left and right upper chambers (atria) of the heart.

Description

During normal development of the fetal heart, there is an opening in the wall (septum) separating the left and right upper chambers of the heart. Normally, the opening closes before birth, but if it does not, the child is born with a hole between the left and right atria. This abnormal opening is called an atrial septal defect and causes blood from the left atrium to flow (or "shunt") across the hole into the right atrium.

Different types of atrial septal defects occur, and they are classified according to where in the separating wall they are found. The most commonly found atrial septal defect, called secundum atrial septal defect, occurs

in the middle of the atrial septum and accounts for about 70 percent of all atrial septal defects.

Abnormal openings also form in the upper part of the atrial septum (called sinus venosus ASD) where the superior vena cava and right atrium join, and lower parts of the atrial septum (called primum ASD). A sinus venosus ASD usually involves the right upper pulmonary vein, while a primum ASD often occurs along with an abnormality in the mitral valve and/or tricuspid valve, and causes some blood leakage (regurgitation) back through the valves.

Atrial septal defects can occur alone or in combination with other congenital heart disorders, such as ventricular septal defect. They can be as small as a pinpoint or as large as the space where the entire septum should be located.

Demographics

Atrial and ventricular septal defects are the most common congenital heart defects. Atrial septal defect accounts for 4–10 percent of all cases of **congenital heart disease** in the United States. Abnormal openings in the atrial septum are twice as common in females as in males.

Causes and symptoms

Causes

Abnormal openings in the atrial septum occur during fetal development. These abnormalities can go unnoticed if the opening is small and produces no abnormal symptoms. If the defect is large, oxygen-rich blood from the left atrium flows back into the right atrium and gets pumped back to the lungs again, causing more work for the heart and lungs. The right atrium may swell or enlarge to hold the extra blood.

In some cases, an atrial septal defect can allow blood clots from the body to enter the brain and cause a **stroke**. Untreated atrial septal defect can lead to pulmonary **hypertension**, chest infection, Eisenmenger's syndrome, atrial fibrillation, atrial flutter, stroke, or right-sided heart failure.

Symptoms

A person born with an atrial septal defect may have no symptoms in childhood, and the condition may go undetected into adulthood. Stunted growth may be a symptom of atrial septal defect. Other symptoms that might develop over time include:

- shortness of breath
- fainting
- irregular heart beats or palpitations (abnormal heart beats that feel like fluttering in the chest)
- inability to **exercise** without becoming over-tired
- difficulty breathing with exercise or activity

By age 50, most people with atrial septal defects experience symptoms that interfere with activities of daily living.

When to call the doctor

The parent or caregiver should call the child's pediatrician or cardiologist when the child has these symptoms or conditions:

- swelling in the ankles or feet
- swollen abdomen
- poor exercise tolerance
- recurrent chest colds and respiratory infections
- abnormal blood pressure
- signs of infection, including **sore throat**, general body aches, or **fever**

The parent or caregiver should seek emergency treatment by calling 911 in most areas when the child has these symptoms or conditions:

- breathing difficulties or rapid breathing
- dizziness or fainting
- uncontrolled coughing or coughing with blood
- bluish skin tone or bluish coloration around the lips, fingernail beds, and tongue
- irregular heart beats or palpitations (abnormal heart beats that feel like fluttering in the chest)
- chest **pain** (rare in children)

Diagnosis

The medical and **family** history help the physician determine if the child has any conditions or disorders that might contribute to or cause the heart defect. A family history of heart defects may suggest a genetic predisposition to the condition.

During the physical exam, the child's blood pressure is measured, and a stethoscope is used to listen to sounds made by the heart and blood flowing through the arteries. Some **heart murmurs** (abnormal heart sounds) can indicate an atrial septal defect. The child's pulse, reflexes, height, and weight are checked and recorded. The child's blood oxygen level can be measured using a pulse oxi-

meter, a sensor placed on the fingertip or earlobe. Internal organs are palpated, or felt, to determine if they are enlarged.

A chest x ray, electrocardiogram (ECG, EKG), echocardiogram (echo), or **magnetic resonance imaging** (MRI) can confirm the presence of an atrial septal defect. A chest x ray evaluates the size, shape, and location of the heart and lungs.

An electrocardiogram helps the physician evaluate the electrical activity of the heart. During an EKG, small electrode patches are attached to the skin on the chest. The electrodes are connected to a computer that measures the heart's electrical impulses and records them in a zigzag patter on a moving strip of paper.

An echocardiogram uses ultrasound, or high-frequency sound waves, to display an image of the heart's internal structures. It can detect valve and other heart problems. A Doppler echo uses sound waves to measure blood flow.

Magnetic resonance imaging is a scanning method that uses magnetic fields and radio waves to create three-dimensional images of the heart, which reveal how blood flows through the heart and how the heart is working.

In some cases, cardiac catheterization, a more invasive diagnostic procedure, may be performed to diagnose atrial septal defect. This procedure should be performed by a specially trained physician and diagnostic team in a well-equipped heart center. During the procedure, a long, slender tube called a catheter is inserted into a vein or artery and slowly directed to the heart, using x ray guidance. To better view the heart and blood vessels, contrast material (dye) is injected through the catheter and viewed and recorded on an x ray video as it moves through the heart. This imaging technique is called angiography. The catheter measures the amount of oxygen present in the blood within the heart. If the heart has an opening between the atria, oxygen-rich blood from the left atrium enters the right atrium. The cardiac catheterization can help doctors detect the higher-than-normal amount of oxygen in the heart's right atrium and right ventricle, and in the large blood vessels that carry blood to the lungs, where the blood would normally collect its oxygen.

Treatment

Twenty percent of atrial septal defects in children correct themselves without medical treatments by the time a child is two years old. If the opening does not close on its own, it needs to be repaired to prevent the pulmonary arteries from becoming thickened and blocked due to increased blood flow. If this condition (pulmonary vascular obstructive disease) is left untreated, it can increase the risk of death by 25 percent.

Treatment should be provided by a pediatric cardiologist, a specialist trained to diagnose and treat congenital heart disease. Surgery should be performed by a pediatric cardiovascular surgeon. A catheter-based cardiac implant should be done by an interventional cardiologist skilled in performing this procedure on children.

Surgery

There are two types of surgical repair for atrial septal defects: primary closure in which the opening is repaired with sutures alone if the defect is small; or secondary closure in which a patch closes the opening if the defect is large. The secondary closure may involve sewing a synthetic patch made of Dacron material over the opening, or wrapping the patient's own tissue (often from the fluid-filled sac around the heart called the pericardium) to close the opening.

During traditional atrial septal defect surgery, the heart is exposed through an incision made in the chest or between the ribs. A heart-lung bypass machine pumps blood for the heart while the heart is stopped and the wall defect is being repaired. Recuperation from surgery involves three to five days in the hospital and four to six weeks recovering at home. When possible, minimally-invasive surgical techniques that use smaller incisions (3–4 inches [7–10 cm]) may be performed, depending on the size and location of the defect. Minimally invasive surgery results in a much shorter hospital stay, reduced scarring, and a faster recovery than traditional surgery.

Surgical repair in asymptomatic children is usually recommended before the child begins grade school. Earlier surgical treatment is recommended when the child develops symptoms or has stunted growth.

Catheter-based cardiac implant procedure

A catheter-based cardiac implant procedure is less invasive than surgical repair, requires smaller incisions, does not require a heart-lung bypass machine, and results in a much shorter hospital stay, reduced scarring, and a more rapid recovery. The child usually stays in the hospital less than 24 hours after the procedure and returns to normal activities within one to two weeks.

The catheter-based cardiac implant procedure involves the implantation of a closure device that seals the defect. Closure devices cannot be used to treat all atrial septal defects, especially if the defect is large, if it is not centrally located within the atrial septum, or if there is not enough nearby tissue to adequately support

the closure device. Other situations that prevent the use of a closure device include: very narrow blood vessels that will not allow the catheter-based system to be inserted; abnormalities of the heart valves; venous drainage from the lungs; and the presence of blood clots, bleeding disorders, active infections, or aspirin intolerance.

The procedure starts with a cardiac catheterization to determine the size and location of the defect. If the cardiac catheterization indicates that a closure device would be an effective treatment, an anticoagulant medication, is given intravenously to reduce the risk of blood clot. The closure device is placed through a specially designed catheter and guided to the location of the heart wall defect. The closure device stays in place permanently to stop the abnormal flow of blood between the atria. Over time, the heart tissue grows over the implanted closure device, becoming part of the heart. Although the device remains the same size, the heart tissue covering the device grows with the child.

Within 24 hours after the closure device implant procedure, a chest x ray, electrocardiogram, and echocardiogram are performed to ensure that the device is properly placed.

Medications

Patients who undergo the cardiac implant procedure take a daily anticoagulant medication such as aspirin or warfarin (Coumadin) for three to six months after the procedure. This medication reduces the risk of blood clot formation around the closure device.

Diuretics may be prescribed if the atrial septal defect was diagnosed later in life and is causing fluid build-up. Diuretics aid the excretion of water and salts and help remove excess fluid from tissues. A potassium supplement may be prescribed with some diuretics to remove potassium from the body along with excess fluid. Other medications include Digoxin, which strengthens the contraction of the heart, slows the heartbeat, and removes fluid from tissues, and antihypertensive medications that treat high blood pressure.

Nutritional concerns

Infants and children with atrial septal defects may gain weight more slowly. The most common reason for poor growth is inadequate caloric or nutrient intake. Other factors that may interfere with growth include:

- rapid heart beat and increased breathing rate
- poor appetite
- decreased food intake due to rapid breathing and fatigue
- frequent respiratory infections
- poor absorption of nutrients from the digestive tract
- decreased oxygen in the blood

Babies with atrial septal defects tire quickly when they eat, making frequent feedings necessary. Feedings should be on-demand and may need to be as often as every two hours in the first few months. Some babies have difficulty feeding from a regular bottle nipple; parents may need to try different brands. If medications are prescribed, they should be given before a feeding. Medications should not be mixed in the formula or breast milk unless the doctor advises otherwise.

The pediatrician will advise when to introduce solid foods, usually around six months of age. Fat should not be restricted in the diet, especially in the first two years. High-calorie foods and snacks can play an important role in providing good **nutrition** and helping the child grow at a healthy rate.

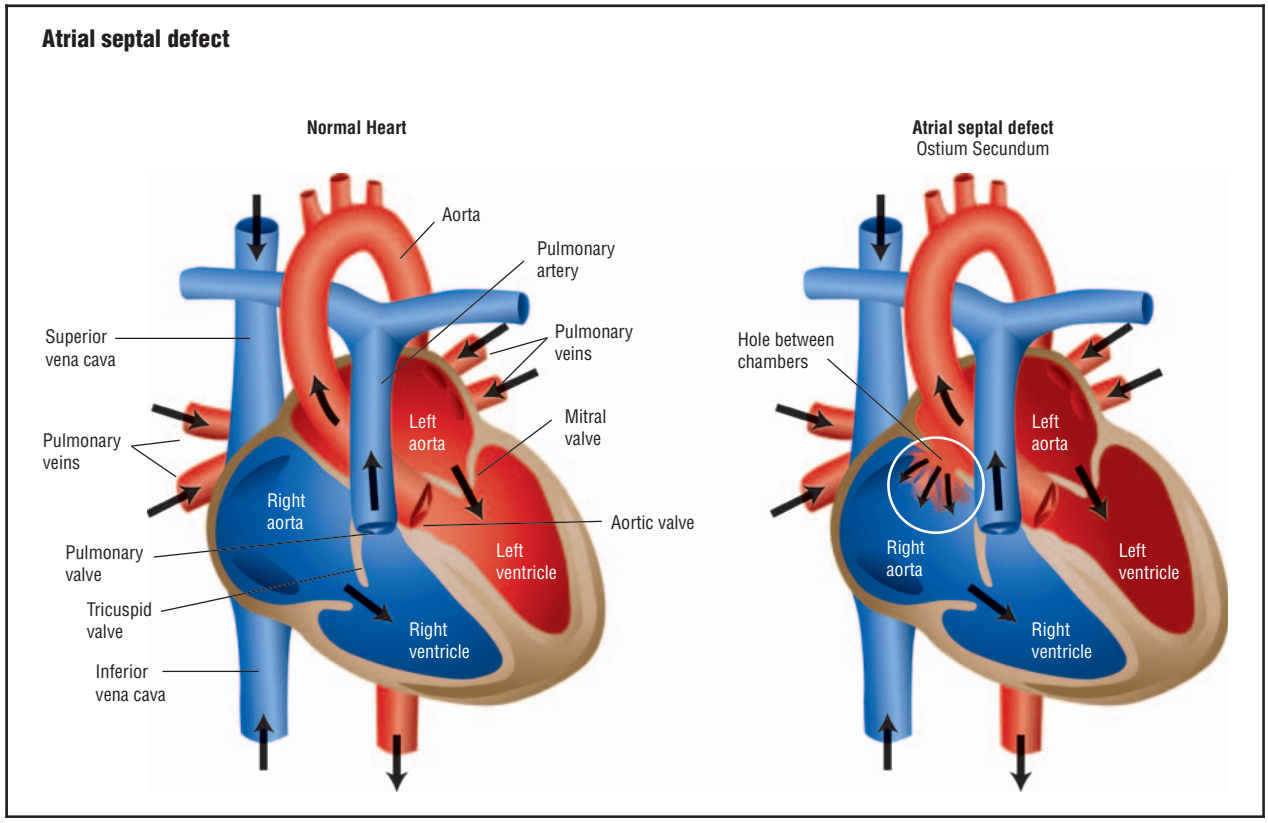
In children older than two years of age, the following low-fat dietary guidelines are recommended:

- total fat intake should comprise 30 percent or less of total calories consumed per day
- calories consumed as saturated fat should equal no more than 8–10 percent of total calories consumed per day
- total cholesterol intake should be less than 300 mg/dl per day

A gradual transition to a heart-healthy diet can help decrease a child's adulthood risk of coronary artery disease and other health conditions. Foods high in fat should be replaced by grains, vegetables, fruits, lean meat, and other foods low in fat and high in complex carbohydrates and protein. Salt should not be added to foods while cooking; highly processed foods, which are usually high in sodium should be avoided. These items include fast foods, canned foods, boxed mixes, and frozen meals.

Follow-up care

Children with atrial septal defects require lifelong monitoring, even after a successful surgery or procedure to close the defect. Along with routine medical care and standard immunizations, periodic heart check-ups are necessary. Usually, heart check-up appointments are scheduled more frequently just after the diagnosis or following the treatment procedure. Additional immunizations, such as the **influenza** vaccine, may be recommended.



A normal heart (left) and one affected by atrial septal defect. The defect is a hole in the wall that separates the chambers of the heart, resulting in the mixing of oxygenated and unoxygenated blood. *(Illustration by GGS Information Services.)*

Medical identification

In case of emergency, a medical identification bracelet or necklace should be worn to alert all health care providers of the child's heart condition.

Prognosis

The outlook for children with atrial septal defects has improved markedly in the past two decades. Individuals with small defects can live a normal life, but larger defects require surgical correction. Less than 1 percent of people younger than age 45 die from corrective surgery. Five to ten percent of patients can die from the surgery if they are older than 40 and have other heart-related problems. There is a 25 percent lifetime risk of death if the atrial septal defect is not repaired. When an atrial septal defect is corrected within the first 20 years of life, there is an excellent chance for the child to live a normal and productive life.

Prevention

Atrial septal defects cannot be prevented. However, to protect patients with atrial septal defects and those with implanted closure devices from heart infections (endocarditis), the American Heart Association recommends regular dental check-ups to prevent infections of the mouth, as well as the preventive use of **antibiotics**. Preventive antibiotics should be taken before surgery, invasive tests or procedures, and all routine dental cleanings and procedures. A 2003 study reported that preventive antibiotics are underused in people with congenital heart conditions, possibly because they do not understand their increased risk of developing bacterial endocarditis.

Parental concerns

If the child needs surgery or a catheter-based cardiac implant, it is important for him or her to be as healthy as possible for the procedure. If the child has a fever, **cough**, or cold, the parent should inform the medical team to determine whether the procedure should be delayed. The medical team can help parents prepare the child for the procedure, and can instruct them on how to explain the procedure based on the child's age, ability to understand, and emotions. Once an atrial septal defect has been closed, it is unlikely that more surgery will be needed. Rarely, a patient may have a residual hole that may require further treatment, depending upon its size.

KEY TERMS

Atrial—Referring to the upper chambers of the heart.

Atrial fibrillation—A type of heart arrhythmia in which the upper chamber of the heart quivers instead of pumping in an organized way. In this condition, the upper chambers (atria) of the heart do not completely empty when the heart beats, which can allow blood clots to form.

Cardiac catheterization—A procedure to passes a catheter through a large vein into the heart and its vessels for the purpose of diagnosing coronary artery disease, assessing injury or disease of the aorta, or evaluating cardiac function.

Congenital—Present at birth.

Dacron—A synthetic polyester fiber used to surgically repair damaged sections of heart muscle and blood vessel walls.

Echocardiogram—A record of the internal structures of the heart obtained from beams of ultrasonic waves directed through the wall of the chest.

Eisenmenger's syndrome—A condition in which high pressures in the pulmonary arteries cause them to thicken. To compensate, the right side of the heart works harder, causing it to stretch and weaken. Eisenmenger's syndrome is a serious condition that leads to heart failure and can result in death by age 40 if left untreated.

Electrocardiagram (ECG, EKG)—A record of the electrical activity of the heart, with each wave being labeled as P, Q, R, S, and T waves. It is often used in the diagnosis of cases of abnormal cardiac rhythm and myocardial damage.

Heart failure—A condition in which the heart is unable to pump enough blood to supply the needs of the body

Pericardium—The thin, sac-like membrane that surrounds the heart and the roots of the great vessels. It has two layers: the inner, serous (or visceral) pericardium and the outer, fibrous (or parietal) pericardium.

Pulmonary hypertension—A disorder in which the pressure in the blood vessels of the lungs is abnormally high.

Septal—Relating to the septum, the thin muscle wall dividing the right and left sides of the heart. Holes in the septum are called septal defects.

Septum—A wall or partition. Often refers to the muscular wall dividing the left and right heart chambers or the partition in the nose that separates the two nostrils. Also refers to an abnormal fold of tissue down that center of the uterus that can cause infertility.

Ventricles—The lower pumping chambers of the heart. The ventricles push blood to the lungs and the rest of the body.

Most children with atrial septal defects can be fully active and are encouraged to exercise. An American Heart Association scientific statement advises children and teens with genetic heart conditions to seek advice from their doctors about the types of physical activities that are safe. The statement was intended to help doctors counsel patients who have an increased risk of sudden cardiac death during physical activity. Certain athletic activities such as competitive **sports** may be limited, depending on the child's type of defect and medical condition. A child with an atrial septal defect may tire more easily than other children; frequent breaks and rest periods should be encouraged as needed during activities. Parents should obtain a doctor's note to explain their child's specific exercise limitations to teachers and coaches.

A child with an atrial septal defect has a greater risk of having a child with a heart defect. The frequency of the condition increases from less than 1 percent in the general population to 2–20 percent when a parent is affected. Genetic counseling and further testing, such as chromosome analysis before pregnancy, or **amniocentesis** during pregnancy, may be recommended in adults with atrial septal defects.

Treatment and care for a child with an atrial septal defect can be costly, and some health insurance plans may not cover all expenses associated with a child's **hospitalization** or surgery. Help is available to cover medical expenses. The parents can discuss financial aid with the hospital. Some organizations, including The Heart of a Child Foundation and Little Hearts on the Mend Fund, provide financial assistance to children in need of heart surgery.

Caring for a child with an atrial septal defect is demanding. Support groups are available to help parents and caregivers cope with the challenges of providing

care for children with special medical needs. It is important for parents to take care of themselves, too, by eating properly, exercising regularly, maintaining personal hygiene, keeping in contact with friends and family members for support, and managing stress by practicing relaxation techniques.

See also Congenital heart disease.

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American College of Cardiology. Heart House. 9111 Old Georgetown Rd., Bethesda, MD 20814-1699. (800) 253-4636 ext. 694 or (301) 897-5400. Web site: <www.acc.org>.

American Heart Association. 7320 Greenville Ave., Dallas, TX 75231-4596. (214) 373-6300 or (800) 242-8721. Web site: <www.americanheart.org/children>.

Children's Heart Services. P.O. Box 8275, Bartlett, IL 60108-8275. (630) 415-0282. Web site: <www.childrensheartservices.org>.

The Cleveland Clinic Heart Center. The Cleveland Clinic Foundation. 9500 Euclid Ave., F25, Cleveland, Ohio, 44195. (800) 223-2273 ext. 46697 or (216) 444-6697. Web site: <www.clevelandclinic.org/heartcenter>.

Congenital Heart Disease Information and Resources. 1561 Clark Dr., Yardley, PA 19067. Web site: <www.tchin.org>.

The Heart of a Child Foundation and Little Hearts on the Mend Fund. Provides financial assistance to children in need of heart surgery. 26710 Fond Du Lac Rd., Rancho Palos Verdes, CA 90275. (310) 375-6617. Web sites: <www.heartofachild.org> and <www.littleheartsonthemend.org>.

Heart Support of America. 4873 N. Broadway, Knoxville, TN 37918. Web site: <www.heartsupport.com>.

International Children's Heart Foundation. 1750 Madison, Suite 100, Memphis, TN 38104. (877) 869-4243. Web site: <www.babyhearts.com>.

Mended Little Hearts. Support program for parents of children with heart defects. (888)-HEART99. Web site: <www.mendedhearts.org/MLH/mlh.htm>.

National Heart, Lung and Blood Institute. P.O. Box 30105, Bethesda, MD 20824-0105. (301) 251-1222. Web site: <www.nhlbi.nih.gov>.

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Heart Information Network. Available online at: <www.heartinfo.org>.

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Attachment between infant and caregiver

Definition

Infant attachment is the deep emotional connection that an infant forms with his or her primary caregiver,

often the mother. It is a tie that binds them together, endures over time, and leads the infant to experience pleasure, joy, **safety**, and comfort in the caregiver's company. The baby feels distress when that person is absent. Soothing, comforting, and providing pleasure are primary elements of the relationship. Attachment theory holds that a consistent primary caregiver is necessary for a child's optimal development.

Description

Attachment theory originated in the early 1950s with John Bowlby, a child psychiatrist, and Mary Ainsworth, a psychologist, who both became interested in young children's responses to experiencing loss. They began studying the realms of attachment and **bonding**. Their theory was developed and integrated over the following 60 years by researchers around the world. (For attachment as it pertains to **adoption**, readers can consult the entry in this encyclopedia on adoption.)

Attachment theory is based on the idea that the bond between an infant and his or her primary caregiver is the crucial and primary influence in infant development and as such forms the basis of coping, the development of relationships, and the formation of personality. If the mother is absent or not available, a primary caregiver serves the role usually assumed by the mother. Attachment refers to a relationship that emerges over time from a history of caregiver-infant interactions. As adults nurture and interact with infants during the first year of life, infants organize their behavior around these caregivers. Attachment is a phenomenon involving physiological, emotional, cognitive, and social processes. The baby displays instinctual attachment behaviors that are activated by cues or signals from the caregiver. Therefore, the process of attachment is defined as a mutual regulatory system, in which the baby and the caregiver have an influence on one another over time. The caregiver's presence provides a feeling of safety and security for the infant. Once this relationship is established, the preference tends to remain stable, and a shift of attachment behavior to a new or strange person becomes more difficult.

Some theorists believe that the attachment system evolved to ensure that infants and caregivers remain physically close, and that the infant is protected. Thus, in order to survive, an infant must become attached to the primary caregiver, who is stronger and wiser regarding the dangers of the world. The caregiver is a safe refuge, a source of comfort and protection, and serves as a secure base from which the infant can explore.

Research has shown that babies and caregivers demonstrate an instinct to attach. Babies instinctively reach out for the safety and security of the safe haven they have with their primary caregiver, while parents usually instinctively protect and nurture their children. Children who start their lives with the essential basis of secure attachment fare better in all aspects of functioning as their development progresses.

Attachment and behavior

From a behavioral perspective, attachment is represented by a group of instinctive infant behaviors that serve to form the attachment bond, protect the child from **fear** and harm, and aid in the infant's protected exploration of the world. These behaviors include:

- reaching
- crying
- grasping
- smiling
- vocalizing
- clinging
- sucking
- moving

All of these behaviors assist in facilitating the maximum physical and emotional development of the child. These particular behaviors may vary from one culture or society to others, but the attachment relationship appears to be universal.

Attachment and emotions

From an emotional perspective, attachment is the development of a mutual bond in which the primary caregiver positively influences infant development through the interactions and relationship that person has with the child. Babies are unable to regulate themselves and become overwhelmed by their emotional states, including those of fear, pleasure, and sadness. Babies are unable to keep themselves in a state of equilibrium, as they lack the skills to control either the intensity or the duration of those emotions. In an attached relationship, babies rely on their primary caregiver to help them navigate the world. The primary caregiver serves as a secure base that is used for exploration and learning. At the same time, the infant forms the necessary skills of self-protection and intimacy.

Other important functions that a secure attachment between an infant and his or her caregiver serves for the developing child include the following:

- learning basic trust, which serves as a basis for all future emotional relationships

- exploring the environment with feelings of safety and security, which leads to healthy intellectual and social development
- developing the ability to control behavior, which results in effective management of impulses and emotions
- creating a foundation for the development of identity, which includes a sense of capability, self-worth, and a balance between dependence and independence
- establishing a moral framework that leads to empathy, compassion, and conscience
- generating a core set of beliefs
- providing a defense against stress and trauma

Children will display distinct attachment styles, which can be loosely defined as either secure or insecure. Secure styles show a child consistently connected to the primary caregiver, with a firmly established sense of trust and a nurturing response; however, insecure styles of attachment have features of instability.

Infancy

Several milestones occur over the course of their first year as infants form an attached relationship with their primary caregiver. These milestones include the following:

- In the first two months of life, even though infants show little observable preference for a particular caregiver, the warm, sensitive, and reliable responses of the caregiver to the child set the stage for the developing attachment relationship.
- From two to seven months, infants tend to interact differently with primary caregivers than they do with strangers but in general still do not display solid preferences.
- By four to six months of age, infants begin to develop expectations of how their primary caregiver will respond to them when they are distressed.
- Between seven months and one year, infants show a definite preference for their primary caregiver. They start to exhibit a wariness of strangers and symptoms of **separation anxiety**.

Toddlerhood

From 12 to 18 months, as they start to walk and crawl, children use their attachment figure as a secure base from which to go out and discover the world and as a safe haven to which to return when frightened or alarmed. Children with secure histories have been shown to be more determined, enthusiastic, and competent in problem-solving as toddlers.

Preschool

During this time, the attachment relationship is characterized by an increased tolerance for separation and an ability to cooperate with others. The child is learning to balance his or her need for independence, self-discipline, and exploration and the need for love and protection from the primary caregiver. However, as **preschool** approaches, children are still susceptible to a variety of dangers. Therefore, attachment behaviors, such as wanting to stay close to the primary caregiver and displaying occasional separation anxiety are adaptive processes, not regressive ones. Western culture has often portrayed this type of behavior as controlling or attention-seeking. Attachment theorists believe this is inaccurate, as these behaviors help serve to ensure the child's survival and socialization.

School age

School-age children with a history of secured attachment histories demonstrate an ability to be more goal-oriented and often display positive leadership skills. Numerous long-term studies have shown that in the following areas securely attached children do better as they grow older:

- self-esteem
- autonomy
- ability to manage impulses and feelings
- long-term friendships
- positive relationships with parents, caregivers, and other authority figures
- effective coping skills
- trust, intimacy, and affection
- positive and hopeful belief systems
- academic success in school

Common problems

Insecure attachment develops when a primary caregiver does not consistently respond in ways that are warm, affectionate, loving, dependable, and sensitive to the infant's needs. The three primary insecure types are resistant attachment, avoidant attachment, and disorganized attachment.

Resistant attachment

This pattern is characterized by an emotional ambivalence in the child and a physical resistance to the primary caregiver. The infant is often hesitant to separate

KEY TERMS

Attachment behavior—Any behavior that an infant uses to seek and maintain contact with and elicit a response from the caregiver. These behaviors include crying, searching, grasping, following, smiling, reaching, and vocalizing.

Insecure attachment—Develops when a primary caregiver does not consistently respond in ways that are warm, affectionate, and sensitive to a baby's needs.

Secure attachment—Usually develops when the primary caregiver is sensitive to the infant's behavior and is emotionally and physically available to the child.

from the caregiver and is quick to display anxiety and distress in an unfamiliar setting. This classification is often referred to as anxious-ambivalent because the child will demonstrate anger towards the caregiver at the same time they are expressing their need for comforting. This type of insecure attachment may be an indicator of risk for the development of emotional, social, and behavioral problems in childhood and later in life.

Avoidant attachment

The key behavior in this type of insecure attachment is an active avoidance of the primary caregiver when the infant is upset. These babies readily separate from their primary caregivers in order to explore and may be more affectionate with strangers than their own mother. They exhibit little preference for and appear emotionally distant from the primary caregiver.

Disorganized attachment

In this type of insecure attachment, infants show a variety of confused and contradictory behaviors. For example, during a reunion with the primary caregiver, the child may look away or even display a blank stare when being held. Other babies may exhibit confusing patterns such as crying unexpectedly after being held or displaying odd, dazed expressions.

Parental concerns

Healthy attachment is the key to healthy babies, and healthy babies are the key to healthy adults. It is crucial for parents, however, to understand that each parent faces times when things do not function flawlessly. What is important in the development of secure attachment is that the primary caregiver is available emotionally to the child and sensitive to the infant's needs.

When to call the doctor

Parents should call their doctor if their child exhibits any of the behaviors of an insecure attachment.

See also Adoption.

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Attempted suicide *see* **Suicide and suicidal behavior**

Attention-deficit/Hyperactivity disorder (AD/HD)

Definition

Attention-deficit/hyperactivity disorder (AD/HD) is a neurobiological disorder characterized by hyperactivity, impulsive behavior, and the inability to remain focused on tasks or activities.

Description

AD/HD, also known as hyperkinetic disorder (HKD) outside of the United States, is estimated to affect 3–7 percent of school-aged children, and seems to afflict boys more often than girls. However, the prevalence in boys may be cited because often girls are not diagnosed until later in age. Although difficult to assess in infancy and toddlerhood, signs of AD/HD may begin to appear as early as age two or three, but visible symptoms change as **adolescence** approaches. Many symptoms, particularly hyperactivity, diminish in early adulthood, while impulsivity and inattention problems often continue.

First documented in 1902, AD/HD has been called minimal brain dysfunction, hyperkinetic reaction, and attention-deficit disorder (ADD). The name AD/HD reflects the various behaviors of inattention, hyperactivity, and impulsiveness that characterize the disorder. Its more precise classification is a result of the *Diagnostic and Statistical Manual, fourth edition (DSM-IV)* system for characterizing and diagnosing mental and behavioral disorders.

Children with AD/HD have difficulties with inattention that can be manifest as a lack of concentration, an easily distracted focus, and an inability to know when and how long to focus. The characteristics of inattention vary with each AD/HD child; however, all most often translate into poor grades and difficulties in school and other social arenas. AD/HD children act impulsively, taking action first and thinking later. They are constantly moving, running, climbing, squirming, and fidgeting. Yet, they often have trouble with gross and **fine motor skills** and, as a result, they may be physically clumsy and awkward. Their clumsiness may also extend to their social skills. They are sometimes shunned by peers due to their impulsive and intrusive behavior.

Demographics

Of the 3–7 percent of school-aged children with AD/HD, some will have a reduction of symptoms as they reach adulthood. However, 65 percent of AD/HD children will continue to display characteristics of AD/HD through adulthood. Until recently, it was believed that boys were three times more likely to have AD/HD; however, that gap has been narrowed. It is more likely that the presence of AD/HD is distributed equally between boys and girls. The reason for the discrepancy was, in part, because young boys tend to more readily and overtly manifest the characteristics of AD/HD, making diagnosis easier. In addition, the inattentive form affects girls more than the hyperactive form; as a result, girls may be less likely to be diagnosed.

Causes and symptoms

The causes of AD/HD are not specifically known. However, it is a neurologically based disease that may be genetic. Children with an AD/HD parent or sibling are more likely to develop the disorder themselves. Although the exact cause of AD/HD is not known, an imbalance or deficiency of certain neurotransmitters—the chemicals in the brain that transmit messages between nerve cells—is believed to be the mechanism behind AD/HD symptoms.

A widely publicized study conducted by Dr. Ben Feingold in the early 1970s suggested that **allergies** to certain foods and food additives caused the characteristic hyperactivity of AD/HD children. By eliminating the food allergen, the premise was that AD/HD characteristics would disappear. Although some children may have adverse reactions to certain foods and food additives that can affect their behavior, carefully controlled follow-up studies have uncovered no link between **food allergies** and AD/HD. Another popularly held misconception about food and AD/HD is that the consumption of sugar causes the hyperactive behavior in an AD/HD child. Again, studies have shown no link between sugar intake and AD/HD. (In a recent study conducted by the National Institute of Mental Health, the level of glucose use in the brain was actually lower in individuals with AD/HD. Since glucose is the main source of fuel for the brain, this is a significant finding.) Finally, parenting style is not a cause for AD/HD. While certain parenting skills and/or deficiencies can affect the environment of an AD/HD child and, as a result, exasperate or help manage the characteristics of AD/HD, it appears that neurological issues are the primary causal agents at play.

In order to diagnose AD/HD, psychologists and other mental health professionals typically use the criteria listed in the *DSM-IV*. *DSM-IV* requires the presence of at least six of the following symptoms of inattention, or six or more symptoms of hyperactivity and impulsivity combined.

Inattention:

- fails to pay close attention to detail or makes careless mistakes in schoolwork or other activities
- has difficulty sustaining attention in tasks or activities
- does not appear to listen when spoken to
- does not follow through on instructions and does not finish tasks
- has difficulty organizing tasks and activities
- avoids or dislikes tasks that require sustained mental effort (e.g., homework)
- is easily distracted
- is forgetful in daily activities

Hyperactivity:

- fidgets with hands or feet or squirms in seat
- does not remain seated when expected to
- runs or climbs excessively when inappropriate (in adolescence and adults, feelings of restlessness)
- has difficulty playing quietly
- is constantly on the move
- talks excessively

Impulsivity:

- blurts out answers before the question has been completed
- has difficulty waiting for his or her turn
- interrupts and/or intrudes on others

Of those symptoms, AD/HD can be categorized further by three subtypes. Each subtype exhibits particular behaviors that make up the general symptoms of a child with AD/HD. They are:

AD/HD predominantly inattentive type (AD/HD-I)

- is disorganized
- is easily distracted
- is forgetful
- has unsustained attention
- has difficulty following instructions
- appears to have poor listening skills
- makes careless mistakes

AD/HD predominantly hyperactive-impulsive type (AD/HD-HI)

- fidgets
- is unable to engage in quiet activity
- is interruptive or intrusive
- cannot remain seated
- speaks out of turn
- climbs or runs about inappropriately
- talks excessively

AD/HD combined type (AD/HD-C) is a combination of the symptoms exhibited by the other two subtypes (inattentive type and hyperactive-impulsive type). Also, for a complete diagnosis, *DSM-IV* requires that some symptoms develop before age seven, and that they significantly impair functioning in two or more settings (e.g., home and school) for a period of at least six months.

Diagnosis

AD/HD cannot be diagnosed with a laboratory test. Diagnosis is difficult and it takes into consideration many aspects of the child's behavior. Often the child's teacher is the one to bring the first signs to the attention of the parents. However, the first step in determining if a child has AD/HD is to consult with a pediatrician. The pediatrician can make an initial evaluation of the child's developmental maturity compared to other children in his or her age group. The physician should also perform a comprehensive physical examination to rule out any organic causes of AD/HD symptoms, such as an overactive thyroid or vision or hearing problems.

If no organic problem can be found, a psychologist, psychiatrist, neurologist, neuropsychologist, or learning specialist is typically consulted to perform a comprehensive AD/HD **assessment**. A complete medical, **family**, social, psychiatric, and educational history is compiled from existing medical and school records and from interviews with parents and teachers. Interviews may also be conducted with the child, depending on his or her age. Along with these interviews, several clinical inventories may also be used, such as the Conners' Rating Scales (Teacher's Questionnaire and Parent's Questionnaire), Child Behavior Checklist (CBCL), and the Achenbach Child Behavior Rating Scales. These inventories provide valuable information on the child's behavior in different settings and situations.

Other disorders such as depression, **anxiety** disorder, and **learning disorders** can cause symptoms similar to AD/HD. A complete and comprehensive psychiatric assessment is critical to differentiate AD/HD from other possible mood and behavioral disorders. **Bipolar disorder**, for example, may be misdiagnosed as AD/HD.

Public schools are required by federal law to offer free AD/HD testing upon request. A pediatrician can also provide a referral to a psychologist or pediatric specialist for AD/HD assessment. Parents should check with their insurance plans to see if these services are covered.

Treatment

Despite similar behavioral characteristics, AD/HD must be treated individually by developing an approach combining various types of treatment. The use of medication in combination with behavioral interventions, classroom accommodations, and proactive parents provide the best treatment option.

Psychostimulants and their effects have been studied in approximately 6,000 children and the positive results of their use have been documented. Such psychostimulants as dextroamphetamine (Dexedrine, Dextrostat), pemoline (Cylert), **methylphenidate** (Ritalin, Concerta, Metadate, Focalin), and mixed salts of a single-entity amphetamine product (Adderall, Adderall XR) are commonly prescribed to control hyperactive and impulsive behavior as well as to increase attention. They work by stimulating the production of certain neurotransmitters in the brain. Generally, short-acting medication lasts for four hours, while long-lasting preparations will last for six to eight hours. Some medication is effective for 10–12 hours. Specific dosages depend upon the patient and that is determined by trial and error in conjunction with close monitoring by a physician in order to find the most beneficial strength. Possible side effects of stimulants include nervous **tics**, irregular heartbeat, loss of appetite, and insomnia. However, the medications are usually tolerated and safe in most cases. In fact, 70–80 percent of AD/HD children respond well to psychostimulants.

In children who do not respond well to stimulant therapy, nonstimulant medications are prescribed. In 2002, the Food and Drug Administration (FDA)approved atomoxetine (Strattera) for the treatment of AD/HD. Unlike the stimulant medications, atomoxetine is not a controlled substance and can be prescribed with refills. (With the use of stimulant medication, the physician must write prescriptions each month of treatment.) Atomoxetine usually takes three to four weeks of use until its effect is evident. In January 2005 the FDA warned that evidence of atleast two cases of liver problems in an adult and teenage patient taking atomoxetine were reported. In both cases, the individuals fully recovered. The manufacturer of atomoxetine (Strattera) planned to notify users of the new FDA warning; however, the company, Eli Lilly & Co., believed that the risk-benefit analysis during trials of the drug was still positive. Such tricyclic **antidepressants** as desipramine (Norpramin, Pertofane) and amitriptyline (Elavil) are frequently recommended as well. Reported side effects of these drugs include persistent dry mouth, sedation, disorientation, and cardiac arrhythmia (particularly with desipramine).

Other medications prescribed for AD/HD therapy include buproprion (Wellbutrin), an antidepressant; fluoxetine (Prozac), an SSRI antidepressant; and carbamazepine (Tegretol, Atretol), an anticonvulsant drug. Clonidine (Catapres), an antihypertensive medication, has also been used to control aggression and hyperactivity in some AD/HD children, although it should not be used in combination with Ritalin.

A child's response to medication will change with age and maturation, so AD/HD symptoms should be monitored closely and prescriptions adjusted accordingly.

Behavior interventions are also crucial to AD/HD treatment. In a Nation Institute of Mental Health (NIMH) study conducted on 579 children over the course of 14 months it was observed that the children receiving AD/HD medication or both medication and behavioral interventions were more likely to see the most relief from their symptoms than those children that only received community aid. The use of a reward system to reinforce good behavior and task completion can be implemented both in the classroom and at home. A chart system may be used to visually illustrate the child's progress and encourage continued success with the use of larger rewards after a certain number of daily rewards are achieved. The reward system stays in place until the appropriate behavior becomes second nature to the child.

A variation of this technique, cognitive-behavioral therapy, works to decrease impulsive behavior by getting the child to recognize the connection between thoughts and behavior, and to change behavior by changing negative thinking patterns.

Individual psychotherapy can help an AD/HD child build **self-esteem**, give them a place to discuss their worries and anxieties, and help them gain insight into their behavior and feelings. **Family therapy** may also be beneficial in helping family members develop coping skills and in working through feelings of guilt or anger parents may be experiencing.

AD/HD children perform better within a familiar, consistent, and structured routine with an emphasis on positive reinforcements for good behavior and minimal use of punishments. When a negative behavior must be acknowledged and corrected, "time outs" give the child with AD/HD an opportunity to regroup without negative reinforcement. Family, friends, and caretakers should all

be educated on the special needs and behaviors of the AD/HD child.

Alternative treatment

A number of alternative treatments exist for AD/HD; however, there are very few studies to prove their efficacy. When choosing a treatment option, it is important to investigate authoritative sources that provide a basis through documented studies for the validity of the treatment. AD/HD is not a disorder that can be cured but rather it is one that is managed by a variety of treatment options. Some of the more popular alternative treatments include:

- EEG (electroencephalograph) biofeedback. By measuring brainwave activity and teaching the AD/HD patient which type of brainwave is associated with attention, EEG biofeedback attempts to train patients to generate the desired brainwave activity. This treatment has been in use for over 25 years and it has had positive response from parents. However, no consistent medical studies are available.
- Chelation therapy focuses on removing excess lead within the body. This treatment is based on the idea that excessive lead in animals causes hyperactivity; yet, not enough medical studies have been done. A physician should be consulted when this approach is considered.
- Intractive metronome training uses a similar instrument as the metronome used by musicians to keep time in order to train individuals to develop their motor and timing skills through repetitively tapping the beat.
- Nutritional supplements claiming to be a cure for AD/HD are not regulated by the Food and Drug Administration (FDA) and should not be considered a treatment option without consultation with a medical doctor.

There are many advertised alternative and complementary treatment options for AD/HD. Only a few are listed here; however, it is always necessary to consult a physician to develop a fine-tuned treatment plan specific to each child's needs.

Nutritional concerns

As mentioned, links between **nutrition** and AD/HD have not been confirmed through medical studies. However, it is important to note that a nutritionally balanced diet is important for normal development in all children.

Prognosis

Untreated, AD/HD negatively affects a child's social and educational performance and can seriously damage his or her self-esteem. Children with AD/HD have impaired relationships with their peers, and may be looked upon as social outcasts. They may be perceived as slow learners or troublemakers in the classroom. Siblings and even parents may develop resentful feelings towards the AD/HD child.

Some AD/HD children also develop a **conduct disorder**. For those adolescents who have both AD/HD and a conduct disorder, up to 25 percent go on to develop **antisocial personality disorder** and the criminal behavior, substance abuse, and high rate of **suicide** attempts that are symptomatic of it. Children diagnosed with AD/HD are also more likely to have a learning disorder, a mood disorder such as depression, or an anxiety disorder.

Approximately 70–80 percent of AD/HD patients treated with stimulant medication experience significant relief from symptoms, at least in the short-term. Approximately half of AD/HD children seem to "outgrow" the disorder in adolescence or early adulthood; the other half will retain some or all symptoms of AD/HD as adults. With early identification and intervention, careful compliance with a treatment program, and a supportive and nurturing home and school environment, children with AD/HD can flourish socially and academically.

Parental concerns

Because AD/HD is often indicated when the AD/HD child is in school, parents are extremely concerned about their child's academic progress. Communication between parents and teachers is especially critical to ensure an AD/HD child has an appropriate learning environment. Educational interventions under Individuals with Disabilities Education Act (IDEA) and Section 504 of the Rehabilitation Act of 1973 mandate that AD/HD children will be served within the public school system. This means that upon request the public school is required to test the child for AD/HD as well as other learning disabilities if they are suspected. In addition, **special education** services are mandated for those children with AD/HD that need extra help and accommodation. It is important that parents assume a positive relationship with their child's educator and school in order to develop the best possible teaching strategies and learning environment for their AD/HD child.

Development of self-esteem is another particular concern for parents of AD/HD children. Because they often have difficulty in school and in social relationships, low self-esteem can be a factor that leads the school aged children toward dangerous or destructive behaviors as they reach adolescence. Finding one activity that the child excels at is essential in fostering a positive self-

KEY TERMS

Conduct disorder—A behavioral and emotional disorder of childhood and adolescence. Children with a conduct disorder act inappropriately, infringe on the rights of others, and violate societal norms.

Nervous tic—A repetitive, involuntary action, such as the twitching of a muscle or repeated blinking.

Oppositional defiant disorder—An emotional and behavioral disorder of children and adolescents characterized by hostile, deliberately argumentative, and defiant behavior towards authority figures that lasts for longer than six months.

image. Often parents look to **sports** as an appropriate outlet. Individual sports such as karate, swimming, tennis, etc. are less socially demanding than team sports; yet they provide an opportunity for the child to thrive in a competitive activity.

AD/HD is a chronic condition. Parents can feel overwhelmed when they have to deal with AD/HD characteristics on a daily basis. Parent should face the issues honestly and directly while fostering a positive relationship with their AD/HD child. The best advocate the AD/HD child has is a parent so it is important that parents be proactive and keep up to date on the latest research. Learning about AD/HD and the various treatment options helps parents cope with their own concerns at the same time they are helping their child.

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Children and Adults with Attention Deficit Disorder (CH.A.D.D.). 8181 Professional Place, Ste. 150, Landover, MD 20785. (800) 233-4050. (305) 306-7070.

National Attention Deficit Disorder Association. (ADDA). 9930 Johnnycake Ridge Road, Suite 3E, Mentor, OH 44060. (800) 487-2282. Web site: <http://www.add.org>

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Audiometry

Definition

Audiometry encompasses those procedures used to measure hearing thresholds.

Purpose

The purpose of audiometry is to establish an individual's range of hearing. It is most often performed when hearing loss is suspected. Audiometry can establish the

extent as well as the type of a hearing loss. Audiometric techniques are also used when an individual has vertigo or **dizziness**, since many hearing and vestibular or balance problems are related. Since those with facial paralysis may also have hearing loss, audiologic testing may be performed on these individuals as well.

Description

The primary purpose of audiometry is to determine the frequency and intensity at which sounds can be heard. Humans can hear sounds in the frequency or pitch range of 20 to 20,000 Hertz (Hz), but most conversations occur between 300 and 3000 Hz. Audiometric testing is done between 125 and 8000 Hz. The intensity levels or degree of loudness at which sounds can be heard for most adults is between 0 and 20 decibels (dB).

Both air conduction and bone conduction of sounds are evaluated by audiometry. Air conduction establishes the extent of sound transmission through the bones of the middle ear. The results of a bone conduction test determine how soft a sound an individual can hear over several frequencies or pitches. Bone conduction audiometry determines the extent to which there is neurosensory hearing loss. An individual with a neurosensory loss may be able to hear sounds but not understand them. Since those with hearing losses often cannot hear sounds at normal decibel levels, intensities as high as 115 dB are used to assess the extent of air conduction loss and as high as 70 dB for bone conduction loss. The difference between bone conduction loss and neurosensory hearing loss is called the air-bone gap.

The most common method of assessing hearing ability is with the audiometer. Audiometric testing with the audiometer is performed while the patient sits in a soundproof booth and the examiner outside the booth communicates to the patient with a microphone. The patient wears headphones when air conduction is tested and a vibrating earpiece behind the ear next to the mastoid bone or along the forehead when bone conduction is tested. One ear is tested at a time, and a technique called masking, in which noise is presented to the ear not being tested, assures the examiner that only one ear is tested at a time. Through the headphones or earpiece pure sounds in both frequency and intensity are transmitted to the patient and the threshold at which the patient can hear for each frequency is established. The patient signals an ability to hear a sound by raising a hand or finger.

When the child is capable of understanding and responding to words, speech discrimination is also assessed as part of audiometry. Speech discrimination establishes one's ability to understand consonant sounds. In speech discrimination testing, two syllable words are read to and then repeated by the patient. This is an important part of audiometry, since much of a child's learning depends on the ability to discriminate speech. Older children of ten to 12 years of age have speech recognition comparable to adults and do well with speech discrimination testing. To insure that speech discrimination only is being assessed, this part of the hearing test is done at decibel levels of 30 to 40 decibels, higher than that of everyday conversation. By age five most children can do some type of speech discrimination testing.

Speech discrimination in the child of three to six years of age may be tested by having the child look at pictures of common objects as a monosyllabic word is read to him or her. The child indicates comprehension of the word by pointing to the corresponding object.

When evaluating infants, rather than testing of threshold levels, the examiner establishes the minimum response level at which the child responds to auditory stimuli. The minimum intensity level at which a neonate responds to sound is 25 dBs. This minimum level gradually decreases through infancy and at 36 months most children respond to sound intensities of less than 10 dBs.

For the young infant under four months of age, audiologists employ behavioral observation audiometry (BOA). The audiologist observes startle responses and motor reflex changes in the child as various noisemakers are employed to elicit these responses. The difficulty with this test is that the noises used are not standardized in frequency or intensity.

Visual reinforcement audiology (VRA) testing evaluates the hearing of infants from six months to two years. Sounds of varying intensity are presented to one of two speakers as the child sits on a parent's lap. If a sound is heard by the child, then he or she turns toward the appropriate speaker and is rewarded by a visual stimulus, such as an animated toy or a flashing light, although video images have been used for older children.

As the child gets older, condition play audiometry (CPA) is useful. The child is instructed to listen for a sound and to respond when a sound is heard by doing varying tasks, such as placing a ball in a cup or placing a peg in a pegboard, when the auditory stimulus is heard. Headphones may be worn by the child for this type of testing.

Because a reliable subjective response is difficult or impossible in a young patient electrophysiological testing is often performed. Electrophysiological testing is a reliable and nonbehavioral method to assess hearing loss in infants and young children and can be done while the child is either sleeping or under sedation. Some electro-

Technician testing a young girl's hearing with an audiometer. *(Photograph by Jon Meyer. Custom Medical Stock Photo, Inc.)*

physiological tests are the auditory brainstem response (ABR) test, auditory steady-state response (ASSR) testing, electroencephalic audiometry (EEG) test, and otoacoustic emission testing (OAE).

To perform the auditory brainstem response (ABR) test, headphones are placed on the infant or child and electrophysiological responses from the scalp and ears are recorded in response to tones sent through the headphones. A computer compiles the findings into a waveform that gives the examiner information about the location of a hearing problem anywhere along this pathway from the ear canal to the brainstem. This test is also called the brainstem auditory evoked response.

Auditory steady-state response (ASSR) testing also involves monitoring recorded responses from the scalp of tones at varying frequencies. This test is a more sensitive test than the ABR and can also measure residual hearing better. The EEG or electroencephalic audiometry test measures tone loss but cannot locate the site of a hearing loss. Otoacoustic emission testing (OAE) records spontaneous emissions from the ear and can detect middle ear problems. It is simpler than ABR, and it can be used to screen infants for severe hearing losses, since if hearing loss of greater than 40 dBs exist, no emission will be recorded.

An adjunct test of audiometry is acoustic immitance testing which assesses the facility with which sound can travel from the external ear to the cochlea inside the ear. The most familiar of this type of testing is the tympanogram, which determines if fluid has built up behind the eardrum.

KEY TERMS

Audiologist—A person with a degree and/or certification in the areas of identification and measurement of hearing impairments and rehabilitation of those with hearing problems.

Cochlear implantation—A surgical procedure in which a small electronic device is placed under the skin behind the ear and is attached to a wire that stimulates the inner ear, allowing people who have hearing loss to hear useful sounds.

Decibel—A unit of the intensity of sound or a measure of loudness. Normal speech is typically spoken in the range of about 20–50 decibels.

Frequency—Sound, whether traveling through air or the human body, produces vibrations—molecules bouncing into each other—as the sound wave travels along. The frequency of a sound is the number of vibrations per second. Within the audible range, frequency means pitch—the higher the frequency, the higher a sound's pitch.

Precautions

Audiometry is a safe procedure to which there are rarely contraindications.

Preparation

For most audiometric testing no special preparation is required, although the first time that hearing testing is done on a child the procedure should be explained as clearly as possible. If ABR or ASSR testing is done under sedation, then the child may not eat for several hours prior to administration of the drugs.

Aftercare

Audiometric testing, except when sedation is involved, requires no special aftercare.

Risks

If the ABR is used under sedation then the side effects of sedatives must be considered. Otherwise there are no risks associated with audiometry.

Parental concerns

Audiometry should be performed on all infants and children since unidentified hearing loss can delay speech and language skills. The earlier that a child with a

hearing problem can be identified, the sooner the child's **communication skills** will develop. The audiometry available as of 2004 can determine the type and extent of a hearing loss as well as identify the location of the hearing problem. The results of audiometric testing can help determine if a hearing aid or cochlear implant may help a child. Audiometric testing can also be an adjunct to diagnosis of more serious problems related to hearing loss such a related syndrome or a tumor.

Parents of a child diagnosed with a hearing loss must be prepared to bring the child back for follow-up evaluations to monitor the hearing loss every three months for the first year after diagnosis and at least annually through the remainder of childhood. As the child gets older, more extensive audiometry testing can be performed.

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Auditory discrimination test

Definition

An auditory discrimination test is a screening or diagnostic **assessment** tool designed to identify and diagnose deficits in auditory discrimination.

Purpose

Auditory discrimination is a central auditory processing skill that involves the ability to differentiate among phonemes—the smallest significant units of sound in a language. Phonemes are combined into words. For example the word "goes" is made up of three phonemes: "g," "oh," and "zzz." Auditory discrimination is part of phonology which, in turn, is one of the five components of language.

Auditory discrimination tests (ADTs) are one type of auditory analysis tests, which are used to measure how well a child understands speech and the spoken word. ADTs are designed to measure a child's phonological awareness—the ability to focus on and manipulate phonemes within spoken words. Phonological awareness skills include the ability to do the following:

- compare and contrast speech sounds
- separate and blend phonemes
- identify phonemes within spoken words
- combine phonemes into spoken words

ADTs measure a child's ability to detect subtle similarities and differences between speech sounds. Two of the most commonly used ADTs are Wepman's Auditory Discrimination Test (WADT) and the Goldman-Fristoe-Woodcock Test of Auditory Discrimination.

Auditory discrimination skills are very important in the classroom. Activities that require auditory discrimination skills include the following:

- following directions
- reading
- writing
- spelling

Auditory discrimination ability or phonological awareness skills have long been correlated with reading ability. Some specialists believe that ADTs should be a component of all reading programs and that poor auditory discrimination can be a major factor in children's failure to reach reading targets. The WADT is used to evaluate **communication skills** in general, as well to identify potential reading difficulties and to predict certain types of speech defects. Because it requires a child to recognize small differences between phonemes, the WADT is widely used to measure a child's readiness for reading instruction using a phonic method.

Some underachieving but gifted children have learning disabilities that are caused by deficits in central auditory skills, including auditory discrimination. The WADT commonly is used to test for an auditory discri-

mination deficit in such children. Deficits in auditory discrimination are also believed to be one of the causes of central auditory processing disorder (CAPD). There are various methods for addressing auditory discrimination problems in children.

Description

Auditory discrimination is one component of central auditory processing skills or auditory perception. The other components are as follows:

- auditory memory: the ability to recall a sequence of auditory stimuli or phonemes
- auditory blending: the ability to perceive separate phenomes, divide a word into phenomes, and combine phenomes into words
- auditory comprehension: the ability to comprehend and interpret information that is presented orally

The WADT

The WADT, first published in 1958 and revised in 1973, is designed to measure the ability of children aged four to eight to recognize small differences between English phonemes. The test consists of 40 pairs of words. The words in a pair are of equal length. In ten of the pairs the words are identical. In the remaining 30 pairs the words differ by a single phoneme. The test requires the child to differentiate between the following:

- 13 word-pairs differing in their initial consonant, such as "coast" and "toast"
- four word-pairs differing in their medial vowels, such as "pat" and "pet"
- 13 word-pairs differing their final consonant, such as "lease" and "leash"
- 10 identical word-pairs or false choices, such as "jam" and "jam"

Often the WADT is administered by a **special education** teacher or a speech/language pathologist. The test is administered orally to an individual child who is seated such that neither the examiner's mouth nor the words on the test form are visible to the child. The examiner reads each word-pair only once, and the child indicates whether the word-pair consists of different or identical words. The test requires about five to 10 minutes to administer. The performance rating scale ranges from "very good development" for the child's age to "below adequate" for the child's age. Two equivalent forms of the test are provided so that children can be retested if their initial scores are questionable or if the test is needed for evaluating the effectiveness of subsequent remedial instruction. The WADT is widely considered to be both reliable and valid, with norms based on the scores of 2,000 children.

The WADT is considered to be a fast, inexpensive means of screening children for auditory discrimination deficits and for identifying children who are slower than average in developing auditory discrimination skills. It also is used to identify children who may have difficulty learning the phonics that are necessary for learning to read. The WADT often is used as a component of formal reading assessments.

Other ADTs

Other ADTs include the following tests:

- Goldman-Fristoe-Woodcock (G-F-W) Test of Auditory Discrimination, which includes visual stimuli
- Goldman-Fristoe-Woodcock (G-F-W) Diagnostic Auditory Discrimination Test
- auditory word discrimination subtest of the Test of Auditory Perceptual Skills (TAPS); the subtest uses only auditory stimuli
- Auditory Discrimination and Attention Test
- Schonell Auditory Discrimination Test which, like the WADT, is a component of some formal reading assessments

In one type of ADT the test administrator says a word and the child is asked to repeat the word, leaving out a syllable or sound. For example the examiner says "outdoor" and tells the child to say the word but to not say "out." The correct answer is "door." Children's responses are graded according to the following:

- The child gives the correct answer quickly.
- The child takes more than five seconds to give the correct answer.
- The child answers incorrectly.

The child's auditory discrimination skill is assigned a grade level:

- a kindergartner told to repeat the word "cucumber" without the "cu (q)" should easily answer "cumber"
- a first-grader told to repeat "please" without the "zzz" sound should easily answer "plea"
- a second-grader told to repeat "clay" without the "k" should easily answer "lay"
- a third-grader told to repeat "smock" without the "mmm" sound should be able to easily answer "sock"

The Sheshore Measures of Musical Talent is a widely used standardized test for measuring musical abilities in students applying to music programs, conservatories, and colleges and universities. It tests the listener's auditory discrimination abilities with regard to the following:

- pitch
- volume
- rhythm
- sound duration
- tonal quality or timbre
- tonal memory

Electrophysiological tests

Sometimes electrophysiological techniques are used to assess various types of central auditory processing including auditory discrimination. These techniques measure auditory evoked potentials (AEPs), which are changes in the brain's neural-electrical activity in response to the reception of auditory signals. AEPs are recorded via electrodes on the child's scalp. During auditory discrimination decisions, which involve various processes including attention and recognition, a large positive peak called P300 appears at about 200 milliseconds after the presentation of the word or other auditory stimulus. The electrical signals that contribute to P300 come from various parts of the brain. The most common way of measuring auditory discrimination with P300 is the oddball paradigm, in which a series of low-frequency auditory stimuli is randomly interspersed with high-frequency stimuli. The child attempts to count the number of high-frequency pitches. Significant differences in the appearance of the P300 peak have been found between poorly achieving gifted children and highly achieving gifted children.

Precautions

ADTs can give confusing or false negative results. Many children do well on the auditory word discrimination subtest of TAPS, which uses auditory stimuli, but perform poorly on the G-F-W Test of Auditory Discrimination, which uses visual stimuli. Such children may have good auditory discrimination skills but poor auditory-visual integration discrimination.

Risks

In the early 2000s research suggests that auditory discrimination and other perceptual processes may not be primary factors in predicting reading ability and learning disabilities. Thus some children may be falsely labeled with a learning disability because of their results on ADTs. Other children might fail to be identified as candidates for early intervention for reading or other learning difficulties on the basis of their ADT scores.

KEY TERMS

Auditory discrimination—The ability to detect small similarities and differences between sounds.

Auditory evoked potential (AEP)—A change in the neural-electrical activity in the brain in response to auditory signals.

Auditory perception—The ability to comprehend and interpret auditory signals.

Central auditory processing skills—The skills needed for auditory perception, including auditory discrimination, auditory memory, auditory blending, and auditory comprehension.

Phonemes—The basic units of sound in a language.

Phonological awareness—The ability to hear and manipulate the sounds that make up words.

Phonology—The science of speech sounds and sound patterns.

Wepman's Auditory Discrimination Test (WADT)—A commonly used test for evaluating auditory discrimination skills.

Normal results

ADTs are standardized by testing large numbers of children to determine the normal range of scores for children of a given age. The vast majority of children have ADT scores within the normal range. Children who score significantly below the normal range may be referred for additional assessment. Early intervention for children with low ADT scores may include exercises and activities designed to improve auditory discrimination.

Parental concerns

ADTs are short, simple tests that do not require preparation on the part of the child. However parents should be aware of the normal developmental milestones of speech and **language development**. Although no two children reach these milestones at precisely the same age, a significant lag may indicate the need for assessment of auditory discrimination and/or other components central auditory processing. Typical milestones include:

- producing vowel sounds within the first six months of life
- understanding certain words by six to 12 months of age
- speaking first words at 12–18 months
- combining words by 18–24 months of age
- understandable speech and the use of consonant sounds by two to three years
- speaking faster and with longer and more varied sentences by three to four years
- a vocabulary of more than 1,500 words, sentences averaging five words, and the ability to modify speech by four to five years of age

Resources

ORGANIZATIONS

American Speech-Language-Hearing Association. 10801 Rockville Pike, Rockville, MD 20852. Web site: <http://asha.org>.

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Autism

Definition

Autism is a severely incapacitating developmental disorder of brain function characterized by three major types of symptoms: impaired social interaction, problems with verbal and nonverbal communication, and unusual or severely limited activities and interests.

Description

Autism is a complex developmental disability with symptoms that typically appear during the first three years of childhood and continue throughout life. It is the most severe disorder within a group of developmental disorders called autism spectrum disorders (ASDs) or **pervasive developmental disorders** (PDDs) that cover a wide range of behaviors and symptoms, all related to a lesser or greater extent to impaired social and **communication skills**.

In its most severe form, autism may include extreme self-injurious, repetitive, highly unusual, and aggressive behaviors.

Demographics

According to the Center for Disease Control and Prevention, PDDs were estimated to occur in two to six per 1,000 births in 2003 with autism being the most common PDD, affecting an estimated one in 250 births. As of 2004, as many as 1.5 million Americans are were believed to have some form of autism. The disorder is four times more prevalent in boys than girls and is not associated with any specific racial or ethnic background. **Family** income, lifestyle, and educational levels also do not affect the chance of the disorder's occurrence.

The Autism Society of America (ASA) warns that autism is on the rise. Based on statistics from the U.S. Department of Education and other governmental agencies, the ASA estimates that the disorder is growing at a rate of 10 to 17 percent per year, which could lead to 4 million Americans being affected by autism between 2005 and 2015.

Causes and symptoms

While understanding of autism grew tremendously since it was first described by Leo Kanner in 1943, no known single cause for autism as of 2004 was yet identified, although research has shown that it results from specific abnormalities in brain structure or function. For example, brain scans show that the shape and structure of the brain in autistic children are different from those of non-autistic children. Researchers investigated several theories and established a firm link between heredity, genetics, and medical problems, while also establishing that no known psychological factors in the development of the child have been shown to cause autism.

The genetic link is supported by observations showing that, in many families, there seems to be a pattern of autism or ASDs. While no one gene was identified as causing autism as of 2004, researchers are searching for irregular segments of genetic code that autistic children may have inherited.

Autism has also been shown to occur more frequently among individuals who have certain medical

conditions, including **fragile X syndrome**, **tuberous sclerosis**, congenital **rubella** syndrome, and untreated **phenylketonuria**.

Toxins and pollution in the environment have also been associated with autism. The Center for the Study of Autism and other agencies documented a high prevalence of autism in certain communities, for example, in the small town of Leomenster, Massachusetts, and in Brick Township, New Jersey, and attempted to uncover the reason.

The symptoms of autism occur in a wide variety of combinations, from mild to severe and are caused by physical disorders of the brain. According to the ASA, they may include any combination of the following in varying degrees of severity:

- insistence on sameness; resistance to change
- difficulty in expressing needs; using gestures or pointing instead of words
- repeating words or sentences instead of using normal, responsive language (echolalia)
- laughing, crying, showing distress for reasons not apparent to others
- aloof behavior, seeking solitude
- tantrums
- refusal to cuddle or be cuddled
- little or no eye contact
- unresponsiveness to normal teaching methods
- sustained odd **play**
- inappropriate attachments to objects
- apparent over-sensitivity or under-sensitivity to **pain**
- no **fear** of danger
- uneven gross/fine motor skills
- not responsive to verbal cues; acts as if deaf although hearing tests in normal range

When to call the doctor

The characteristic behaviors of autism may or may not be apparent in infancy (18 to 24 months) but usually become obvious during early childhood (two to six years).

The National Institute of Child Health and Human Development (NICHD) lists the five following behaviors as signals that medical evaluation is needed:

- does not babble or coo by 12 months
- does not gesture (point, wave, grasp) by 12 months
- does not say single words by 16 months
- does not say two-word phrases on his or her own by 24 months
- loss of any language or social skills at any age

The presence of any of these five behaviors does not mean that a child has autism, but because the characteristics of the disorder vary so much, a child should be evaluated by a multidisciplinary team that may include a neurologist, psychologist, developmental pediatrician, speech/language therapist, learning consultant, or other professionals knowledgeable about autism.

Diagnosis

There are no medical tests for diagnosing autism. An accurate diagnosis must be based on observation of the individual's communication, behavior, and developmental level. A diagnosis of autistic disorder is usually made when an individual displays six or more of 12 symptoms listed across three major areas: social interaction, communication, and behavior. Several screening procedures have been developed for use in diagnosing autism, among which are the following:

- Childhood Autism Rating Scale (CARS). CARS is based on observed behavior. Using a 15-point scale, professionals evaluate a child's relationship to people, body use, adaptation to change, listening response, and verbal communication.
- Checklist for Autism in Toddlers (CHAT). CHAT is used to screen for autism at 18 months of age. The screening tool uses a short questionnaire with two sections, one prepared by the parents, the other by the child's family doctor or pediatrician.
- Autism Screening Questionnaire (ASQ). The ASQ is a 40-item screening scale used with children four and older to help evaluate communication skills and social functioning.
- Screening Test for Autism in Two-Year Olds. This test uses direct observations to study behavioral features in children under two. It is focused on three skills areas, play, motor imitation, and joint attention, that are associated with autism.

Treatment

There is as of 2004 no cure for autism, but appropriate treatment may promote relatively normal development and lower the incidence of undesirable behaviors. Doctors also may prescribe a variety of drugs to reduce the symptoms of autism, such as **antidepressants** and tranquilizers. Educational/behavioral therapies emphasize highly structured and often intensive skill-oriented

training, and they are comparatively the most effective treatments available.

The importance of early treatment is well established among professionals. Researchers have proposed that there is a critical period during which the young, developing brain is highly modifiable. For some children with autism, the repeated, active interaction provided by intensive educational/behavioral therapy may modify their neural circuitry before it goes too much awry, correcting it before autism becomes permanent.

A wide spectrum of educational/behavioral therapies were developed during the last decades of the twentieth century under the umbrella of applied behavior analysis (ABA), the science of human behavior. ABA is the process of systematically applying interventions based upon the principles of learning theory to improve socially significant behaviors to a meaningful degree. ABA methods treat autism with particular strategies: using reinforcement procedures to increase on-task behavior and social interactions; teaching new skills (functional life skills, communication skills, or social skills); maintaining desirable behaviors (teaching self-control and self-monitoring procedures to maintain social skills); transferring behavior from one situation or response to another (from completing assignments in the resource room to performing as well in the mainstream classroom); reducing interfering behaviors (e.g., self-injury).

Specific educational/behavioral therapy programs for the treatment of autism include, for example, the following:

- The Miller Method. Developed at the Language and **Cognitive Development** Center (LCDC) in Boston, MA. The LCDC is a Massachusetts Chapter 766-approved day school, serving students with autism or PDD ages three to 14. The LCDC specializes in a particular approach to teaching children with autism. The Miller Method extensively uses adaptive equipment, including platforms (to elevate the child so as to help increase eye contact), large swinging balls (to expand the child's reality system) and Swiss cheese boards (to teach motor planning, as well as to increase the child's understanding of his or her relation to environment and space).
- Discrete Trial Training (DTT). DTT methodology has been likened to controlling the river of information and interaction that typically confronts the child with autism such that it is presented one drop at a time. This control manages learning opportunities so that skills are more easily mastered by the child. Learning occurs in small steps. Simple skills must be mastered before new learning opportunities are presented, in which the child then builds upon the mastered skill toward a more complex one.
- Treatment and Education of Autistic and Communication Handicapped Children (TEACCH). TEACCH is a statewide program in North Carolina that tries to respond to the needs of autistic people by using the best available approaches and methods. The TEACCH approach includes a focus on the person with autism and development of a program around this person's skills, interests, and needs. The major priorities include centering on the individual, understanding autism, adopting appropriate adaptations, and a broadly based intervention strategy building on existing skills and interests.

Therapy for autistic children may include working on jigsaw puzzles. *(© Michael Macor/San Francisco Chronicle/Corbis.)*

Alternative treatment

Some alternative treatments have been proposed for autism. They include:

- The Son-Rise program. The Son-Rise program was created by Barry and Samahria Lyte Kaufman in the 1970s, as a means to teach their own son, who was diagnosed with autism and **mental retardation**. The program ranges from one week to six months and is designed to

KEY TERMS

Antidepressant drug—A medication prescribed to relieve major depression. Classes of antidepressants include selective serotonin reuptake inhibitors (fluoxetine/Prozac, sertraline/Zoloft), tricyclics (amitriptyline/Elavil), MAOIs (phenelzine/Nardil), and heterocyclics (bupropion/Wellbutrin, trazodone/Desyrel).

Asperger syndrome—A developmental disorder of childhood characterized by autistic behavior but without the same difficulties acquiring language that children with autism have.

Congenital rubella syndrome (CRS)—Viral illness caused by a togavirus of the genus *Rubivirus*. When rubella infection occurs during pregnancy, fetal infection is likely and often causes congenital rubella syndrome (CRS), resulting in miscarriages, stillbirths, and severe birth defects. Up to 20 percent of the infants born to mothers infected during the first half of pregnancy have CRS. The most common congenital defects are cataracts, heart disease, deafness, and mental retardation.

Echolalia—Involuntary echoing of the last word, phrase, or sentence spoken by someone else.

Fragile X syndrome—A genetic condition related to the X chromosome that affects mental, physical, and sensory development. It is the most common form of inherited mental retardation.

Pervasive developmental disorder—A category of childhood disorder that includes Asperger syndrome and Rett's disorder. The PDDs are sometimes referred to collectively as autistic spectrum disorders.

Phenylketonuria (PKU)—A rare, inherited, metabolic disorder in which the enzyme necessary to break down and use phenylalanine, an amino acid necessary for normal growth and development, is lacking. As a result, phenylalanine builds up in the body causing mental retardation and other neurological problems.

Tranquilizer—A medication that has a calming effect and is used to treat anxiety and mental tension.

Tuberous sclerosis—A genetic condition that affects many organ systems including the brain, skin, heart, eyes, and lungs. Benign (non-cancerous) growths or tumors called hamartomas form in various parts of the body, disrupting their normal function.

teach parents, professionals, and support staff of children with a wide range of disabilities how to implement home-based programs based upon the Kaufmans' theories of learning. There have been no studies of the Son-Rise Program's effectiveness, and the method has not been subjected to scientific evaluation.

- Megavitamin therapy. Some studies have shown that vitamin B_6 improves eye contact and speech and lessens tantrum behavior. Vitamin B_6 causes fewer side effects than other medications and is considered safe when used in appropriate doses. However, not many health practitioners advocate its use in the treatment of autism, citing that the studies showing its benefit were flawed.

Nutritional concerns

Dimethylglycine (DMG) is a compound available in many health food stores, that is legally classified as a food, not a vitamin or drug. Some researchers claim that it improves speech in children with autism. Those who respond to this treatment usually do so within a week. Many doctors, however, do not feel that the studies are adequate to promote DMG in the diet of autistic individuals.

Prognosis

People with autism have normal life expectancies. Symptoms in many children improve with treatment, or as the children grow up, some eventually are able to lead normal or near-normal lives. **Adolescence** can worsen behavior problems in some children, and treatment should be adjusted for the child's changing needs. According to the National Institute of Neurological Disorders and **Stroke** (NINDS), about one third of children with ASDs eventually develop epilepsy. The risk is highest in children with severe cognitive impairment and motor deficits.

Prevention

Since the cause of the brain anomalies associated with autism is not known, prevention is not possible.

Parental concerns

Following a diagnosis of autism, parents need to work with health and education professionals for the child's benefit. Specifically, they need to take the following steps:

- Be informed. Parents should learn as much as they can about autism so that they can be involved in determining care.
- Be prepared. Parents should prepare for meetings with doctors, therapists, and school personnel. They should ask questions and communicate their concerns regarding treatment issues and the impact of the diagnosis on the family.

- Be organized. Many parents find it useful to keep a notebook detailing their child's diagnosis, treatment, and the meetings they have with professionals.
- Communicate effectively. Open communication is very important. If parents disagree with a professional's recommendation, for example, they should communicate specifically why they disagree.

See also Fragile X syndrome; Pervasive developmental disorders; Phenylketonuria.

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Autism Network International (ANI). PO Box 35448, Syracuse, NY 13235–5448. Web site: <http://ani.autistics.org>.

Autism Research Institute (ARI). 4182 Adams Ave., San Diego, CA 92116. Web site: <www.autismresearchinstitute.com>.

Autism Society of America. 7910 Woodmont Avenue, Suite 300, Bethesda, MD 20814–3067. Web site: <www.autism-society.org>.

Families for Early Autism Treatment. PO Box 255722, Sacramento, CA 95865–5722. Web site: <www.feat.org>.

MAAP Services for Autism, Asperger's, and PDD. PO Box 524, Crown Point, IN 46308. Web site: <www.maapservices.org>.

National Alliance for Autism Research (NAAR). 99 Wall Street, Research Park, Princeton, NJ 08540. Web site: <www.naar.org>.

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Monique Laberge, Ph.D.

Autoeroticism *see* Masturbation

B

Babysitters

Definition

A babysitter is someone who provides occasional child care for a few hours at a time. Teenage babysitters often provide babysitting services for a few hours at a time. However, in-home sitters range from nannies who may have training in child development and first aid to women who, although not trained formally, have had many years of experience caring for children, including their own. Parents might prefer an au pair, a young person, usually a woman in her early twenties, often from abroad, who lives with the **family** to provide child care.

Purpose

In order to give themselves time off from parenting, parents hire babysitters. However, time away from home is enjoyable only when parents are secure in knowing that their child is well cared for. Some parents join a babysitters' club, for example, a group of mothers may agree to take turns caring for each others' children so that each of them can have some time away from their children and know their children are in excellent hands.

Babysitter course

The American Red Cross provides a certification course for babysitters. Young people over age 11 are eligible to enroll in the eight-hour training course, offered at various community organizations and schools. Some organizations underwrite the cost of the course and offer it free to participants; others charge a fee. The course provides instruction in supervision of children, planning activities for children of all ages, accident prevention, emergency response techniques (including what to do in a **choking** emergency), and job-hunting strategies.

Description

Babysitters should understand that parents often have ambivalent feelings about leaving their children with a babysitter. The primary goal of parents is to provide safe and competent babysitting without feeling a need for substitute parents. No matter how conscientious the sitter, she or he will not care for the child in precisely the same way as a parent, and it is unreasonable to expect a carbon-copy parent. Once convinced the sitter is a decent and kind individual, parents will allow the young person to be herself (within the outline of the family's needs and rules) and to use her own judgments.

Most families provide the babysitter with general guidelines about bedtime, acceptable activity and behavior during the parents' absence, and instructions on who to contact if case of an emergency. In addition, young people who want to be babysitters may take the American Red Cross course. Books and videotapes also outline techniques and strategies for safe and successful babysitting. The teenage babysitter must be able to answer lots of questions in an extensive interview with conscientious parents.

Finding a sitter

Parents advertise for a nanny or sitter in the local paper, on the church or community bulletin boards, or with an agency. Word-of-mouth is often the best source. Parents ask other parents for recommendations. Good sources for sitters who are no longer needed by a family are local preschools and nursery schools. Many parents post their names and numbers in these locations as soon as they anticipate needing a sitter.

Interviewing the sitter

Parents interview interested applicants and request and examine references. This meeting provides an

opportunity for direct exchange of ideas between the sitter and the parents. Questions pertain to rules regarding food and methods for handling **discipline** problems and fees the parents will pay. The sitter and the child(ren) should meet to get to know each other a bit.

Potential babysitters need to prepare for interviews with parents. The sitters should be ready to answer specific questions. They need to provide proof of identity (such as a driver's license or social security card) and supply names, addresses, and phone numbers for three to five references.

During the interview parents may want to address the following concerns:

- Is the teenage babysitter mature, well-groomed, and disciplined?
- What childcare experience does she/he have? What were the sitter's best and worst experiences?
- How does the sitter handle issues of discipline? What would the sitter do if the baby cried for an hour or more? What if the toddler was defiant or inattentive to the sitter's directions? What if the child broke the sitter's watch or destroyed other prized possessions?
- How does the sitter feel about TV? Would the sitter watch TV while the child was playing or napping? What kinds of programs do children and teenagers watch in her home? Would the sitter offer television as regular entertainment for the child?
- How much time does the sitter spend with other sitters and friends? Does the sitter enjoy taking the children outside to **play** in the backyard or to the neighborhood playground?
- What does she know about good **nutrition**? Does she limit snacks to healthy foods?
- Does the sitter drive a car, have a cellular telephone, or have a need to spend time talking on the phone while working?
- What would the teenager do in an emergency such as a sudden illness for the child or if the sitter became ill suddenly? What would the sitter do if there was a fire and other emergency?

The interview is the proper time to discuss hourly or evening rates. Some sitters request a higher rate after midnight. This is also the time to discuss travel arrangements. Does the sitter drive and have a car or does the sitter need to be picked up and returned home?

Paying the babysitter

Babysitter pay rates consider the age and number of children, the age of the babysitter, the type of care expected from the sitter, and the time of day (or night), and whether the sitter drives or not. The distance traveled to the job also dictates the pay rate. For teenagers the base rate is $4.00 to $5.00 an hour for a baby sitter who cares for two children and drives to and from the job. The pay increases with the number of children to perhaps $10.00 an hour for three children.

The going rate varies by geographical location and over time in any one area. Urban rates may range from $3.50 to $10.00 depending on age and experience, the number of children, and the lateness of the hour when the parents return home. In general, fees are higher for younger children.

Babysitter checklist

Parents should leave emergency information for the sitter, most often near the telephone or on the refrigerator, where it can be found easily. The information may include the following:

- 911, as the emergency number to call
- family name, home address, and phone number (Sitters may ''blank out'' while trying to give this critical information over the telephone to the 911 operator.)
- telephone numbers of the family doctor or pediatrician
- telephone numbers, including cellular phones, for reaching parents
- children's full names and dates of birth
- name, address, and phone numbers of neighbors who have agreed to be on call in case of an emergency and numbers of back-up friends or relative
- time parents are expected to come home
- parents' full names, cell phone or pager numbers
- special activities for children and any food restriction for each child
- child's routine, including approved snacks, toileting habits, bedtime, and comfort objects needed for bedtime
- guidelines for the babysitter's personal behavior, such as personal telephone calls or friends visiting
- safety and security procedures, such as what to say when answering the telephone and how to secure all doors

KEY TERMS

Certification course—A course of instruction that covers the basics of babysitting; a certificate of accomplishment is given to teenagers who successfully complete the course.

Parental concerns

Children should be told in advance that their parents are going out and the babysitter will be staying with them. Even if they are initially accepting, it is not unusual for young children to cry when they realize that their parents are leaving. The tears will dry when the parents leave; in response to children's tears, it is useful to minimize the problem and leave happily, assuring young children about the parents' return. The babysitter can use this time to comfort the children. She can distract them by engaging them in conversation or mentioning the next activity.

Parents should plan to stay in the house for at least 15 to 20 minutes after the sitter arrives to give general information about the kids and specific instructions about the home. If the family lives in an apartment building, the parent should point out emergency exits or fire escapes and leave candles and flashlight handy in case of a possible power failure. They should review simple first-aid, where to find bandages and home remedies for bumps or **bruises**. However, juvenile sitters should not be asked to dispense medications to children.

Sitters should know the household routines, approved television programs, recommended bedtime stories, and bedtime. They should be prepared for behaviors that are problematic, such as temper **tantrums**. In all, the sitter's job is to keep the children safe and as happy as possible within the guidelines set by the parents.

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Aliene Linwood, RN, DPA

Bacillary dysentery *see* **Shigellosis**

Bariatric surgery *see* **Obesity**

Bath therapy *see* **Therapeutic baths**

Battered child syndrome

Definition

Battered child syndrome (BCS) refers to non-accidental injuries sustained by a child as a result of physical abuse, usually inflicted by an adult caregiver.

Description

Internal injuries, cuts, **burns**, **bruises**, and broken or fractured bones are all possible results of battered child syndrome. Because adults are so much larger and stronger than children are, an abused can suffer severe injury or death without the abuser intentionally causing such an injury. Shaking an infant can cause bleeding in the brain (**subdural hematoma**), resulting in permanent brain damage or death. Emotional damage to a child is also often the byproduct of **child abuse**, which can result in the child exhibiting serious behavioral problems such as substance abuse or the physical abuse of others.

BCS is alternatively referred to as child physical abuse or non-accidental trauma (NAT).

Demographics

The total abuse rate of children is 25.2 per 1,000 children, with physical abuse accounting for 5.7 per 1,000, sexual abuse 2.5 per 1,000, emotional abuse 3.4

per 1,000, and neglect accounting for 15.9 per 1,000 children. These categories overlap, with sexual and physical abuse often occurring together; physical abuse or neglect seldom occur without emotional abuse. These numbers may be underestimates due to underreporting of the problem or failure of diagnosis by medical personnel.

In 1996, more than 3 million victims of alleged abuse were reported to child protective services in the United States; reports were substantiated in more than one million cases. Parents were abusers in 77 percent of the confirmed cases; other relatives in 11 percent. More than 1,000 children died from abuse in 1996.

Causes and symptoms

Causes

Battered child syndrome (BCS) is found at every level of society, although the incidence may be higher in lower-income households, where adult caregivers may suffer greater stress and social difficulties and have a greater lack of control over stressful situations. Other risk factors include lack of education, single parenthood, and **alcoholism** or other drug addictions. The child abuser most often injures a child in the heat of anger or during moments of stress. Common trigger events that may occur before assaults include incessant crying or whining of infants or children; perceived excessive "fussiness" of an infant or child; a toddler's failed **toilet training**; and exaggerated perceptions of acts of "disobedience" by a child. Sometimes cultural traditions may lead to abuse, including beliefs that a child is property, that parents (especially males) have the right to control their children any way they wish, and that children need to be toughened up to face the hardships of life. Child abusers were often abused as children themselves and do not realize that abuse is not an appropriate disciplinary technique. Abusers also often have poor impulse control and do not understand the consequences of their actions.

Symptoms

Symptoms may include a delayed visit to the emergency room with an injured child; an implausible explanation of the cause of a child's injury; bruises that match the shape of a hand, fist or belt; cigarette burns; scald marks; bite marks; black eyes; unconsciousness; lash marks; bruises or choke marks around the neck; circle marks around wrists or ankles (indicating twisting); separated sutures; unexplained unconsciousness; and a bulging fontanel in small infants.

Emotional trauma may remain after physical injuries have healed. Early recognition and treatment of these emotional "bruises" is important to minimize the long-term effects of physical abuse. Abused children may exhibit:

- a poor self-image
- sexual acting out
- an inability to love or trust others
- aggressive, disruptive, or illegal behavior
- anger, rage, **anxiety**, or **fear**
- self-destructive or self-abusive behavior
- suicidal thoughts
- passive or withdrawn behavior
- fear of entering into new relationships or activities
- school problems or failure
- sadness or other symptoms of depression
- flashbacks or nightmares
- drug or alcohol abuse

Sometimes emotional damage of abused children does not appear until **adolescence** or even later, when abused children become abusing parents who may have trouble with physical closeness, intimacy, and trust. They are also at risk for anxiety, depression, substance abuse, medical illnesses, and problems at school or work. Without proper treatment, abused children can be adversely affected throughout their life.

When to call the doctor

Anyone should call a health care provider or child protective services if they suspect or know that a child is being abused. Reporting child abuse to authorities is mandatory for doctors, teachers, and childcare workers in most states as a means to prevent continued abuse.

Diagnosis

Battered child syndrome is most often diagnosed by an emergency room physician or pediatrician, or by teachers or social workers. Physical examination will detect injuries such as bruises, burns, swelling, retinal hemorrhages (bleeding in the back of the eye), internal damage such as bleeding or rupture of an organ, **fractures** of long bones ore spiral-type fractures that result from twisting, and fractured ribs or skull. **X rays**, and other imaging techniques, such as MRI or scans, may confirm or reveal other internal injuries. The presence of injuries at different stages of healing (i.e., having occurred at different times) is nearly always indicative of BCS. Establishing the diagnosis is often hindered

by the excessive cautiousness of caregivers or by actual concealment of the true origin of the child's injuries, as a result of fear, shame and avoidance or denial mechanisms.

Treatment

Medical treatment for battered child syndrome will vary according to the type of injury incurred. Counseling and the implementation of an intervention plan for the child's parents or guardians are necessary. The child abuser may be incarcerated, and/or the abused child removed from the home to prevent further harm. Decisions regarding placement of the child with an outside caregiver or returning the child to the home will be determined by an appropriate government agency working within the court system, based on the severity of the abuse and the likelihood of recurrence. Both physical and psychological therapy are often recommended as treatment for the abused child. If the child has siblings, the authorities should determine where they have also been abused, for about 20 percent of siblings of abused children are also shown to exhibit signs of physical abuse.

Prognosis

The prognosis for battered child syndrome will depend on the severity of injury, actions taken by the authorities to ensure the future **safety** of the injured child, and the willingness of parents or guardians to seek counseling for themselves as well as for the child.

Prevention

Recognizing the potential for child abuse and the seeking or offering of intervention, counseling, and training in good parenting skills before battered child syndrome occurs is the best way to prevent abuse. The use of educational programs to teach caregivers good parenting skills and to be aware of abusive behaviors so that they seek help for abusive tendencies is critical to stopping abuse. Support from the extended **family**, friends, clergy, or other supportive persons or groups may also be effective in preventing abuse. Signs that physical abuse may occur include parental alcohol or substance abuse; high stress factors in the family life; previous abuse of the child or the child's siblings; history of mental or emotional problems in parents; parents abused as children; absence of visible parental love or concern for the child; and neglect of the child's hygiene.

Parental concerns

Parents who are in danger of abusing their children (for example, when they find themselves becoming inappropriately or excessively angry in response to a child's behavior) should seek professional counseling. Parents may also call the National Child Abuse Hotline (800-4-A-Child; 800-422-4453, a nationwide 24-hour telephone hotline), where they will be counseled through a parenting or caretaking crisis and offered guidance about how to better handle the situation.

Parents should also exercise caution in arranging for or hiring **babysitters** and other caretakers. If they suspect abuse, they should immediately report those suspicions to the police or to their local child protective services agency. They should also teach their children to report abuse to a trusted adult.

KEY TERMS

Child protective services (CPS)—The designated social services agency (in most states) to receive reports, investigate, and provide intervention and treatment services to children and families in which child maltreatment has occurred. Frequently this agency is located within larger public social service agencies, such as Departments of Social Services.

Fontanelle—One of several "soft spots" on the skull where the developing bones of the skull have yet to fuse.

Multiple retinal hemorrhages—Bleeding in the back of the eye.

Subdural hematoma—A localized accumulation of blood, sometimes mixed with spinal fluid, in the space between the middle (arachnoid) and outer (dura mater) membranes covering the brain. It is caused by an injury to the head that tears blood vessels.

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ORGANIZATIONS

National Child Abuse Hotline. 800-4-A-Child (800-422-4453).

National Clearinghouse on Child Abuse and Neglect Information. P.O. Box 1182, Washington, DC 20013-1182. 800-394-3366. Web site: <http://nccanch.acf.hhs.gov>.

Prevent Child Abuse America. 200 South Michigan Avenue, 17th Floor, Chicago, IL 60604. (312) 663-3520. Web site: <http://preventchildabuse.org>.

National Parents Anonymous. 675 West Foothill Blvd., Suite 220m Claremont, CA 91711. (909) 621-6184. Web site: <http://www.parentsanonymous.org/pahtml/paNPLTabout.html>.

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Child Abuse: Types, Symptoms, Causes, and Help. Available online at: <http://www.helpguide.org/mental/child_abuse_physical_emotional_sexual_neglect.htm>.

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Bayley Scales of Infant Development

Definition

The Bayley Scales of Infant Development (BSID) measure the mental and motor development and test the behavior of infants from one to 42 months of age.

Purpose

The BSID are used to describe the current developmental functioning of infants and to assist in diagnosis and treatment planning for infants with developmental delays or disabilities. The test is intended to measure a child's level of development in three domains: cognitive, motor, and behavioral.

Cognitive development

Cognition can be defined as a process by which knowledge is gained from perceptions or ideas. **Cognitive development** refers to how an infant perceives, thinks, and gains an understanding of the world. Within the history of developmental psychology, the work of Jean Piaget (1896–1980), the Swiss psychologist, has had the greatest impact on the study of cognitive development. Piaget's theory is focused on the processes of cognitive development and states that the child is born with an innate curiosity to interact with and understand his/her environment. It is through interaction with others that the child actively constructs his/her development.

Motor development

During the first two years of life, infants grow and develop in many ways. Two types of motor development occur at this stage. Cephalocaudal development occurs in the following sequence: head before arms and trunk and arms and trunk before legs. Proximodistal development occurs as follows: head, trunk, arms before hands and fingers. Motor development has a powerful impact on the social relationships, thinking, and language of infants. Large motor development allows infants to have more control over actions that help them move around their environment, while small motor development gives them more control over movements that allow them to reach, grasp, and handle objects. The sequence of these developments is similar in most children; however, the rate of growth and development varies by individual.

Behavioral development

Temperament is the set of genetically determined traits that organize the child's approach to the world. They are instrumental in the development of the child's distinct personality and behavior. This behavioral style appears very early in life—within the first two months after birth—and undergoes development, centered on features such as intensity, activity, persistence, or emotionality.

Besides measuring normal cognitive, motor, and behavioral developmental levels, the BSID are also used in cases in which there are significant delays in acquiring certain skills or performing key activities in order to qualify a child for special interventions. Specifically, they are also used to do the following:

- identify children who are developmentally delayed
- chart a child's progress after the initiation of an intervention program

- teach parents about their infant's development
- conduct research in developmental psychology

Description

The BSID were first published by Nancy Bayley in *The Bayley Scales of Infant Development* (1969) and in a second edition (1993). The scales have been used extensively worldwide to assess the development of infants. The test is given on an individual basis and takes 45–60 minutes to complete. It is administered by examiners who are experienced clinicians specifically trained in BSID test procedures. The examiner presents a series of test materials to the child and observes the child's responses and behaviors. The test contains items designed to identify young children at risk for **developmental delay**. BSID evaluates individuals along three scales:

- Mental scale: This part of the evaluation, which yields a score called the mental development index, evaluates several types of abilities: sensory/perceptual acuities, discriminations, and response; acquisition of object constancy; memory learning and problem solving; vocalization and beginning of verbal communication; basis of abstract thinking; habituation; mental mapping; complex language; and mathematical concept formation.
- Motor scale: This part of the BSID assesses the degree of body control, large muscle coordination, finer manipulatory skills of the hands and fingers, dynamic movement, postural imitation, and the ability to recognize objects by sense of touch (stereognosis).
- Behavior rating scale: This scale provides information that can be used to supplement information gained from the mental and motor scales. This 30-item scale rates the child's relevant behaviors and measures attention/arousal, orientation/engagement, emotional regulation, and motor quality.

The BSID are known to have high reliability and validity. The mental and motor scales have high correlation coefficients (.83 and .77 respectively) for test-retest reliability.

Precautions

BSID data reflect the U.S. population in terms of race, ethnicity, infant gender, education level of parents, and demographic location of the infant. The BSID was standardized on 1,700 infants, toddlers, and preschoolers between one and 42 months of age. Norms were established using samples that did not include disabled, premature, and other at-risk children. Corrected scores are sometimes used to evaluate these groups, but their use remains controversial.

The BSID has poor predictive value, unless the scores are very low. It is considered a good screening device for identifying children in need of early intervention.

Preparation

Before giving the BSID test to a child, the examiner explains to the parents what will happen during the test procedure. This is to allow the examiner to establish a focused rapport with the child once the procedure has started and avoid diverting attention from the child to the parents during the test. The parents are also asked not to talk to the child during the BSID test to avoid skewing results.

Risks

There are no risks associated with the BSID test.

Parental concerns

As of 2004 it was recognized that parental involvement in the developmental **assessment** of their children is very important. First, because parents are more familiar with their child's behavior, their assessment may indeed be more indicative of the child's developmental status than an assessment that is based on limited observation in an unfamiliar clinical setting. The involvement of parents in their child's development testing also improves their knowledge of child development issues and their subsequent participation in required intervention programs, if any. In cases of developmental problems, parents should bear in mind that the scoring and interpretation of the test results is a highly technical matter that requires years of training and experience. Besides the BSID, parents should be aware that three other infant development scales are commonly used:

- Brazelton Neonatal Behavioral Assessment Scale: This scale tests an infant's neurological development, interactive behavior, and responsiveness to the examiner, and need for stimulation. This test is administered during the newborn period only.
- Gesell Developmental Schedules: These schedules test for fine and **gross motor skills**, language behavior, adaptive behavior including eye-hand coordination, imitation, object recovery, personal-social behavior such as reaction to persons, initiative, independence, and **play** response.
- Denver Developmental Screening Test: This test is used to identify problems or delays that should be more carefully evaluated. It measures four types of development: personal/social, fine-motor/adaptive, language, and gross motor skills.

KEY TERMS

Behavior—A stereotyped motor response to an internal or external stimulus.

Cephalocaudal development—Motor development which occurs in the first two years of life: head before arms and trunk, arms and trunk before legs.

Cognition—The act or process of knowing or perceiving.

Cognitive—The ability (or lack of) to think, learn, and memorize.

Motor skills—Controlled movements of muscle groups. Fine motor skills involve tasks that require dexterity of small muscles, such as buttoning a shirt. Tasks such as walking or throwing a ball involve the use of gross motor skills.

Proximodistal development—Motor development which occurs in the first two years of life: head, trunk, arms before hands and fingers.

Stereognosis—The ability to recognize objects by sense of touch.

Temperament—A person's natural disposition or inborn combination of mental and emotional traits.

See also Cognitive development; Personality development; Personality disorders.

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American Academy of Child & Adolescent Psychiatry (AACAP). 3615 Wisconsin Ave., N.W., Washington, DC. 20016–3007. Web site: <www.aacap.org>.

American Academy of Pediatrics (AAP). 141 Northwest Point Boulevard, Elk Grove Village, IL 60007–1098. Web site: <www.aap.org>.

American Psychological Association (APA). 750 First Street, NE, Washington, DC 20002–4242. Web site: <www.apa.org>.

Child Development Institute (CDI). 3528 E. Ridgeway Road, Orange, CA 92867. Web site: <www.childdevelopmentinfo.com>.

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Bed-wetting

Definition

Bed-wetting, also called enuresis, is the unintentional discharge of urine during **sleep**. Although most children between the ages of three and five begin to stay dry at night, the age at which children are physically and emotionally ready to maintain complete bladder control varies.

Description

Most children wet the bed occasionally, and definitions of the age and frequency at which bed-wetting becomes a medical problem vary somewhat. The word enuresis is derived from a Greek word meaning "to make water." Enuresis is defined as the repeated voiding of urine into the bed or clothes at least twice a week for at least three consecutive months in a child who is at least five years of age. It can be nocturnal

(occurring at night) or diurnal (occurring during the day). Enuresis is a fairly common condition in children. It can be a stressful condition as well for both parents and children. Some children find bed-wetting extremely embarrassing. Parents sometimes become both frustrated and angry.

Enuresis is divided into two classes. A child with primary enuresis has never been consistently dry through the night. A child with secondary enuresis begins to wet after a prolonged dry period. Some children have both nocturnal and diurnal enuresis.

Demographics

The prevalence of bedwetting gradually declines throughout childhood. Of children aged five years, 23 percent have nocturnal enuresis. During elementary school years, the problem remains common, with 20 percent of seven-year-old children and 4 percent of ten-year-old children still experiencing nighttime bedwetting. Nocturnal enuresis is more common in males. It occurs in boys aged seven and ten years at 9 percent and 7 percent, respectively, compared to 6 percent and 3 percent, respectively, in girls.

Causes and symptoms

The causes of bed-wetting are not entirely known. It tends to run in families. Most children with primary enuresis have a close relative—a parent, aunt, or uncle—who also had the disorder. Over 70 percent of children with two parents who wet the bed will also wet the bed. Twin studies have shown that both of a pair of identical **twins** experience enuresis more often than both of a pair of fraternal twins.

Sometimes bed-wetting can be caused by a serious medical problem like diabetes, sickle-cell anemia, or epilepsy. Snoring and episodes of interrupted breathing during sleep (sleep apnea) occasionally contribute to bed-wetting problems. Enlarged adenoids can cause these conditions. Other physiological problems, such as urinary tract infection, severe **constipation**, or **spinal cord injury**, can cause bed-wetting.

Children who wet the bed frequently may have a smaller than normal functional bladder capacity. Functional bladder capacity is the amount of urine a person can hold in the bladder before feeling a strong urge to urinate. When functional capacity is small, the bladder will not hold all the urine produced during the night. Tests have shown that bladder size in these children is normal. Nevertheless, they experience frequent strong urges to urinate. Such children urinate often during the daytime and may wet several times at night. Although a small functional bladder capacity may be caused by a **developmental delay**, it may also be that the child's habit of voiding frequently slows bladder development.

Parents often report that their bed-wetting child is an extremely sound sleeper and difficult to wake. However, several research studies found that bed-wetting children have normal sleep patterns and that bed-wetting can occur in any stage of sleep.

In the early 2000s medical research has found that many children who wet the bed may have a deficiency of an important hormone known as antidiuretic hormone (ADH). ADH helps to concentrate urine during sleep hours, meaning that the urine contains less water and, therefore, takes up less space. This decreased volume of water usually prevents the child's bladder from overfilling during the night, unless the child drinks a lot just before going to bed. Testing of many bed-wetting children has shown that these children do not have the usual increase in ADH during sleep. Children who wet the bed, therefore, often produce more urine during the hours of sleep than their bladders can hold. If they do not wake up, the bladder releases the excess urine and the child wets the bed.

Research demonstrates that in most cases bed-wetting does not indicate that the child has a physical or psychological problem. Children who wet the bed usually have normal-sized bladders and have sleep patterns that are no different from those of non-bed-wetting children. Sometimes emotional stress, such as the birth of a sibling, a death in the **family**, or separation from the family, may be associated with the onset of bed-wetting in a previously toilet-trained child. Daytime wetting, however, may indicate that the problem has a physical cause.

While most children have no long-term problems as a result of bed-wetting, some children may develop psychological problems. Low **self-esteem** may occur when these children, who already feel embarrassed, are further humiliated by angry or frustrated parents who punish them or who are overly aggressive about **toilet training**. The problem can by aggravated when playmates tease or when social activities such as sleep-away camp are avoided for **fear** of teasing.

When to call the doctor

Parents should contact their child's doctor if the child has started wetting the bed after a sustained period of time staying dry. Parents should also notify the physician if their child over the age of five begins to have urinary incontinence during the day, as this may be caused by a physical disorder.

Diagnosis

If a child continues to wet the bed after the age of six, parents may feel the need to seek evaluation and diagnosis by the family doctor or a children's specialist (pediatrician). Typically, before the doctor can make a diagnosis, a thorough medical history is obtained. Then the child receives a physical examination, appropriate laboratory tests, including a urine test, and if necessary, radiologic studies (such as **x rays**).

If the child is healthy and no physical problem is found, which is the case 90 percent of the time, the doctor may not advise treatment but rather may provide the parents and the child with reassurance, information, and advice.

Treatment

Occasionally a doctor will determine that the problem is serious enough to require treatment. Standard treatments for bed-wetting include bladder training exercises, motivational therapy, drug therapy, psychotherapy, and diet therapy.

Bladder training exercises are based on the theory that those who wet the bed have small functional bladder capacity. Children are told to drink a large quantity of water and to try to prolong the periods between voiding. These exercises are designed to increase bladder capacity but are only successful in resolving bed-wetting in a small number of patients.

In motivational therapy, parents attempt to encourage the child to combat bed-wetting, but the child must want to achieve success. Positive reinforcement, such as praise or rewards for staying dry, can help improve self-image and resolve the condition. Punishment for wet nights hamper the child's self-esteem and compound the problem.

The following motivational techniques are commonly used:

- Behavior modification: This method of therapy is aimed at helping children take responsibility for their nighttime bladder control by teaching new behaviors. For example, children are taught to use the bathroom before bedtime and to avoid drinking fluids after dinner. While behavior modification generally produces good results, it is long-term treatment.
- Alarms: This form of therapy uses a sensor placed in the child's pajamas or in a bed pad. This sensor triggers an alarm that wakes the child at the first sign of wetness. If the child is awakened, he or she can then go to the bathroom and finish urinating. The intention is to condition a response to awaken when the bladder is full. Bed-wetting alarms require the motivation of both parents and children. They were considered the most effective form of treatment available as of 2004.

A number of drugs are also used to treat bed-wetting. These medications are usually fast acting; children often respond to them within the first week of treatment. Among the drugs commonly used are a nasal spray of desmopressin acetate (DDAVP), a substance similar to the hormone that helps regulate urine production; and imipramine hydrochloride, a drug that helps to increase bladder capacity. Studies show that imipramine is effective for as many as 50 percent of patients. However, children often wet the bed again after the drug is discontinued, and it has some side effects. Some bed-wetting with an underlying physical cause can be treated by surgical procedures. These causes include enlarged adenoids that cause sleep apnea, physical defects in the urinary system, or a spinal tumor.

Psychotherapy is indicated when the child exhibits signs of severe emotional distress in response to events such as a death in the family, the birth of a new child, a change in schools, or **divorce**. Psychotherapy is also indicated if a child shows signs of persistently low self-esteem or depression.

In rare cases, **allergies** or intolerances to certain foods—such as dairy products, citrus products, or chocolate—can cause bed-wetting. When children have **food sensitivities**, bed-wetting may be helped by discovering the substances that trigger the allergic response and eliminating these substances from the child's diet.

Prognosis

Occasional bed-wetting is not a disease, and it does not have a cure. If the child has no underlying physical or psychological problem that is causing the bed-wetting, in most cases he or she will outgrow the condition without treatment. About 15 percent of bed-wetters become dry each year after age six. If bed-wetting is frequent, accompanied by daytime wetting, or falls into the American Psychiatric Association's diagnostic definition of enuresis, a doctor should be consulted. If treatment is indicated, it usually successfully resolves the problem. Marked improvement is seen in about 75 percent of cases treated with wetness alarms.

Prevention

Although preventing a child from wetting the bed is not always possible, parents can take steps to help the child keep the bed dry at night. These steps include:

KEY TERMS

Antidiuretic hormone (ADH)—Also called vasopressin, a hormone that acts on the kidneys to regulate water balance.

Nocturnal enuresis—Involuntary discharge of urine during the night.

Void—To empty the bladder.

- encouraging and praising the child for staying dry instead of punishing when the child wets
- reminding the child to urinate before going to bed, if he or she feels the need
- limiting liquid intake at least two hours before bedtime

Parental concerns

Bed-wetting often leads to behavioral problems because of the embarrassment and guilt the child may feel. Parents should not attempt to make their child feel guilty about wetting the bed. They should let the child know that bedwetting is not their fault. Punishment is an inappropriate response to enuresis and will not resolve the problem.

Resources

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Bee sting *see* **Bites and stings**

Bee sting allergy *see* **Insect sting allergy**

Beery-Buktenica test

Definition

The Beery-Buktenica visual-motor integration test is a neuropsychological test that analyzes visual construction skills. It identifies problems with visual perception, motor coordination, and visual-motor integration such as **hand-eye coordination**.

Purpose

The Beery-Buktenica Test, also known as Developmental Test of Visual-Motor Integration or VMI, is designed to identify deficits in visual perception, **fine motor skills**, and hand-eye coordination. It may be used to diagnose **cognitive development** disorders in young children through an analysis of visual construction skills. It can be administered to individuals from age two through young adulthood and can also be used to test adults of all ages, particularly those who have been disabled by **stroke**, injury, or Alzheimer's disease.

The Beery-Buktenica VMI test is used by physicians, psychologists, neuropsychologists, learning disability specialists, counselors, educators, and other professionals. It can be effectively used for the following purposes:

- to identify individuals who are having visual-motor difficulties
- to help diagnose visual-motor deficits
- to make referrals to specific professionals or services
- to test individual learning levels and educational programs
- to monitor the progress of individuals with known visual-motor or developmental difficulties

Description

One of the basic aspects of an individual's ability to think and know (cognition) is how one is able to perceive certain stimuli. Assessing perception skills—observing how individuals may respond to things they see, hear, and touch—is, therefore, a basic part of assessing cognitive function. Children with possible **developmental delay** may be tested for their perception of visual, auditory, and tactile stimuli, not just to understand their ability to see, hear, and touch, but to understand how they perceive stimuli and what conclusions they make as a result. This information can help pediatricians and child psychologists evaluate the child's nervous system

(neurological) functioning and psychological development. Visual testing may include color perception, object recognition, visual organizational abilities, and the ability to differentiate figures from the background against which they appear. It also includes visual construction tests. Some visual construction tests are designed to test memory by asking the child to draw a familiar object. Others, such as the Beery-Buktenica test, are designed to test visual motor skills as a factor of visual perception and integration.

Visual-motor integration or VMI can be evaluated as a factor in child development by providing the child with geometric designs ranging from simple line **drawings** to more complex figures and asking that the designs be copied. The construction skills used in the test have been shown to indicate visual motor impairment, such as problems with fine motors skills of the hand and hand-eye coordination. The developers of the test, Keith E. Beery and Norman A. Buktenica, have established adequate norms for visual motor performance by children in various age groups. The test is considered especially useful to help evaluate children with other disabilities or disabling conditions. It can also be used for the evaluation of motor skills such as handwriting.

The Beery-Buktenica test is usually administered individually but can also be given in groups. The child is given a booklet containing increasingly complex geometric figures and asked to copy them without any erasures and without rotating the booklet in any direction. The test is given in two versions: the Short Test Form containing 15 figures is used for ages three through eight; the Long Test Form, with 24 figures, is used for older children, adolescents, and adults with developmental delay. A raw score based on the number of correct copies is converted based on norms for each age group, and results are reported as converted scores and percentiles. The test is untimed but usually takes 10–15 minutes to administer.

Precautions

There are no precautions involved in visual motor testing.

Preparation

More successful testing is achieved when no preparatory steps are taken. The test can be explained briefly to the child beforehand.

Aftercare

No particular care is recommended after administration of the test. Further testing may be recommended as well as specific intervention to help correct any deficits noted. Depending upon the specific deficits found, intervention may include occupational therapy, physical therapy, counseling, behavior modification, **play** therapy, and medication for certain neuropsychological disorders.

KEY TERMS

Cognition—The act or process of knowing or perceiving.

Deficit—A shortfall or slowdown in development, possibly related to a disorder that slows or interrupts normal childhood development.

Developmental—Referring to the growth process, particularly the growth patterns and associated skills acquired in childhood.

Developmental delay—The failure of a child to meet certain developmental milestones, such as sitting, walking, and talking, at the average age. Developmental delay may indicate a problem in development of the central nervous system.

Motor coordination (MC)—Related to movement of parts of the body, particularly the use of the hands and coordination of eye-hand motion.

Neurological—Relating to the brain and central nervous system.

Neuropsychological—Referring to the interaction between the nervous system and cognitive function, the influence of one function on the other.

Spatial skills—The ability to locate objects in a three-dimensional world using sight or touch.

Visual perception (VP)—The ability to perceive or understand what is being seen; the integration of an image with an idea of what it represents.

Risks

There are no risks associated with taking the Beery-Buktenica VMI test.

Normal results

Children who perform well on VMI testing may still have visual perception or motor coordination deficits. Visual conceptualization and motor coordination should be evaluated separately to confirm the results.

Children who do not perform well on VMI testing may have impairment of visual-motor skills including the following types:

- visual analysis and visual spatial ability
- motor coordination (MC)
- visual conceptualization (VC)
- visual motor integration

Parental concerns

Parents may be apprehensive about the performance of their child in the Beery-Buktenica testing process. Results are carefully analyzed, and parents are advised not to judge the child's skills until they have discussed the test with the pediatrician, neurologist, or psychologist who will use the results in conjunction with other developmental tests in order to make a diagnosis or recommendations for therapy.

See also Cognitive development; Fine motor skills.

Resources

ORGANIZATIONS

National Institute of Neurological Disorders and Stroke. National Institutes of Health, Bethesda, MD 20892. Web site: <http//www.nindsnih.gov>.

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Bejel

Definition

Bejel, also known as endemic syphilis, is a chronic but curable disease that is seen mostly in children in dry regions, such as parts of Africa (Sudan, southern Rhodesia, and South Africa), parts of the Middle East (among nomadic/Bedouin tribes of Saudi Arabia, Iraq, and Syria), and parts of Asia (Turkey, southeast Asia, and the western Pacific). Unlike venereal syphilis, endemic syphilis is not sexually transmitted. Similar to venereal syphilis, however, it begins with skin sores and has a latent period followed by a more severe stage, which includes bone infections and additional skin lesions.

Demographics

Bejel occurs predominately in children aged two to 15 years. Twenty-five percent of the cases occur in those younger than six years of age, and 55 percent of the cases occur before the age of 16 years. The remaining 20 percent of the cases occur in adults who are in close contact with infected children. Bejel is only rarely reported in the United States and then usually among immigrants and people arriving from areas where the disease is common. Both sexes are equally susceptible to bejel.

Description

Bejel has many other names depending on the locality, including siti (Gambia), njovera (southern Rhodesia), therlijevo (Croatia), and frenjak (Balkans). Bejel is related to yaws and **pinta**, and together the three diseases are referred to as treponematoses. Yaws, which also affects the skin and bones, occurs in the humid equatorial countries, while pinta, which only affects the skin, is common among the native peoples of Mexico, Central America, and South America.

Transmission

Treponema pallidum subspecies *endemicum*, the bacteria that causes bejel, is very closely related to the one that causes the sexually transmitted form of syphilis, but the method of transmission is different. In bejel, transmission is by direct contact, with broken skin or contaminated hands, or indirectly by sharing drinking vessels and eating utensils. *T. pallidum* subspecies *endemicum* is passed on mostly among children living in poverty in unsanitary environments and with poor hygiene.

Causes and symptoms

The skin, bones, and mucous membranes are all affected by bejel. The disease begins with slimy patches on the inside of the mouth, followed by blisters on the trunk, arms, and legs. Bone infections, mainly in the legs, develop later and cause **pain** deep within the bones. Eventually, the bones may become deformed because of bone and cartilage destruction. In later stages, soft gummy lesions may form in the nasal passages, destroying nasal cartilage and in the roof of the mouth, even breaking through the mouth palate. The lesions associated with bejel are destructive and may leave disfiguring scars.

Diagnosis

T. pallidum subspecies *endemicum* can be detected by microscopic study of samples taken from the sores or lymph fluid. However, since antibody tests do not distinguish between the types of syphilis, specific diagnosis of

KEY TERMS

Endemic disease—An infectious disease that occurs frequently in a specific geographical locale. The disease often occurs in cycles.

Lymph fluid—Clear, colorless fluid found in lymph vessels and nodes. The lymph nodes contain organisms that destroy bacteria and other disease causing organisms (also called pathogens).

Syphilis—This disease occurs in two forms. One is a sexually transmitted disease caused by A systemic infection caused by the spirochete *Treponema pallidum*. It is most commonly transmitted by sexual contact.

the type of syphilis depends on the child's history, symptoms, and environment.

When to call the doctor

The doctor should be called if symptoms of bejel develop in a child. Travel information is invaluable in diagnosis of the disease.

Treatment

Large doses of benzathine penicillin G given by injection into the muscle can cure this disease in any stage, although it may take longer and require additional doses in later stages. If penicillin cannot be given, alternative **antibiotics** are chloramphenicol and tetracycline. Since tetracycline can permanently discolor new teeth that are still forming, it is usually not prescribed for children unless no viable alternative is available.

Prognosis

Bejel is usually completely curable with antibiotic treatment. Death from bejel is uncommon. Follow-up care is recommended to detect treatment failures and reinfection.

Prevention

The World Health Organization (WHO) has worked with many countries to prevent this and other diseases, and the number of cases of bejel has been reduced somewhat. Widespread use of penicillin has been responsible for reducing the number of existing cases, but the only way to eliminate bejel is by improving living and sanitation conditions and through continuing health education. Since the disease is very contagious, public health personnel must seek out and treat infected children and their contacts in order to prevent additional cases.

Parental concerns

When traveling in areas where bejel is endemic, parents should ensure that their children avoid contact with children with lesions and avoid shared drinking and eating utensils.

See also Pinta.

Resources

ORGANIZATIONS

National Organization for Rare Disorders Inc. 55 Kenosia Ave, PO Box 1968, Danbury, CT 06813–1968. Web site: <www.rarediseases.org>.

WEB SITES

"Endemic syphilis." *eMedicine*, October 28, 2004. Available online at <www.emedicine.com/derm/topic117.htm> (accessed December 6, 2004).

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Bell's palsy

Definition

The National Institute of Neurological Disorders and Stroke (NINDS), a part of the National Institute of Health (NIH), defines Bell's palsy as "a form of facial paralysis resulting from damage to the seventh (facial) cranial nerve." This condition is considered to be normally a transient phenomenon and not permanently disabling. It is named for Sir Charles Bell, a Scottish surgeon who, over two hundred years ago, did much of the earliest research regarding the anatomy and pathology of the cranial nerves.

Description

There are 12 sets of bilateral cranial nerves originating in the posterior portion of the brain stem, called the pons. These nerves control various functions in the upper portion of the body, especially within the face and head. The seventh cranial nerve enters the facial region through a small opening in the bony area behind the ear called the stylomastoid foramen. From the stylomastoid

foramen, the nerve enters the parotid gland and divides into an estimated 7,000 nerve fibers that control a wide range of facial and neck activity. Seventh cranial nerve endings control neck, eyelid, and forehead muscles; are responsible for facial expression, the secretion of saliva, the volume at which sound is perceived; and a myriad of other functions.

The taste sensations for the front two-thirds of the tongue are sent to the brain via the seventh cranial nerve. In Bell's palsy, this nerve becomes compressed due to swelling and inflammation that is a part of the body's reaction to an infectious disease process. This compression results in weakness or paralysis that normally occurs on one side of the face only. However, though highly unusual (occurring in only 1 percent of all incidences), it is also possible to have bilateral Bell's palsy, that is, paralysis on both sides of the face at the same time, caused by compression of both seventh cranial nerves.

Transmission

Bell's palsy, in itself, is not contagious. Many of the agents that cause it, however, are conditions that have already caused an infection in the body.

Demographics

In the past, Bell's palsy was thought to be a highly uncommon occurrence. It is now known that this nerve disorder is the most common cause of one-sided facial weakness for children. It affects on average approximately one in every five thousand people worldwide, and nearly 40,000 Americans each year. Because diseases that compromise the immune system such as **HIV infection** or sarcoidosis can also result in Bell's palsy, there are geographical variations in the incidence of the disease. Bell's palsy is seen more commonly in areas where **AIDS** or sarcoidosis are more prevalent, but its incidence overall throughout the world remains constant.

The majority of Bell's palsy sufferers are adults. This disorder is much more likely to occur in old age or in the last trimester of pregnancy than in childhood. Diabetics are four times more apt to contract Bell's palsy than non-diabetics. Though children are considered far less likely than adults to contract Bell's palsy, they are not immune from it. There is no difference in the incidence of Bell's palsy between males and females, nor does race seem to be a factor. In addition to incidence, severity of symptoms and recovery rates appear to be equal across both gender and racial lines. The number of children that contract left-sided Bell's palsy is no different from the number that get the right-sided form.

Causes and symptoms

As noted previously, Bell's palsy occurs as a manifestation of the body's reaction to microbial infection of the structures surrounding the seventh cranial nerve. The most commonly responsible germs are viruses that are members of the herpes family. The herpes family of viruses share some common characteristics, including the capacity for long life, going into a dormant phase that in some cases can literally last decades following infection, having an affinity for nerve tissue. Herpes viruses are the cause of infections as diverse as **sexually transmitted diseases**, chickenpox and cold sores. As early as 1970, a study by researcher Shingo Murakami identified HSV-1 as the primary cause of Bell's palsy. Several subsequent studies have consistently verified Murakami's research. HSV-1, also known as herpes simplex and the usual cause of cold sores, has been shown to be the infecting agent in at least 60–70 percent of all Bell's palsy cases.

HSV-1 is a herpes virus that nearly all of the human race has been exposed to, with exposure beginning in early childhood. It is spread through kissing, sharing towels, and/or sharing eating and drinking utensils. It is now known that HSV-1 often infects children but does not always manifest itself by the creation of cold sores. (In fact, only 15 percent of people exposed to HSV-1 develop cold sores.) Because the virus becomes dormant following its initial infection of the body, a large number of HSV-1 carriers are thus produced, most often without the infected person or others even being aware that HSV-1 is present. Other herpes viruses such as Epstein-Barr, responsible for mononucleosis as well as the viruses causing the **common cold**, **influenza** (the flu) are all potential culprits for causing this condition. The bacterial infection involved in **Lyme disease** has also been demonstrated as causing some cases of Bell's palsy. The same causative agents infect both adults and children.

Impairment of the immune system has been unquestionably determined to be the reason why Herpes Viruses are reactivated from a dormant state and re-infect children causing Bell's palsy. Such a weakening of the immune system can be long-term, caused by chronic disease such as leukemia or autoimmune disorders such as lupus, or short-term. The most common causes for short-term or temporary impaired immunity are:

- stress created by difficult situations for the child either at home or at school
- lack of sleep
- non-life-threatening illness such as upper respiratory infection (URI)
- physical trauma

It is also worth noting that in 2004, the World Health Organization (WHO) Global Advisory Committee on Vaccine Safety reported that in October of 2000 an increased incidence of Bell's palsy in Switzerland was observed following the initiation of an internasal **flu vaccine**. Due to this adverse effect, the vaccine manufacturer discontinued research and production. In 2003, another internasal flu vaccine was licensed in the United States, and this vaccine has so far shown no increased occurrence of Bell's palsy. However, the Global Advisory Committee on Vaccine Safety continues to monitor these vaccines, and the use of internasal vaccines should be discussed with the family healthcare provider.

Clearly the overwhelming majority of children that contract mononucleosis, cold sores, Lyme disease, cold or flu do not develop Bell's palsy. But for some, a reaction of their immune system to viral, or in some cases bacterial, infection causes the production of antibodies which in turn produces inflammation and swelling. In Bell's palsy, this process typically occurs after the seventh cranial nerve's passage through the stylomastoid foramen into a tiny bony tube called the fallopian canal. If the inflammation within the fallopian canal is severe enough, it will exert sufficient pressure on the seventh cranial nerve to make it impossible for the nerve to carry messages to and from the brain.

As noted previously, such messages normally carried by healthy seventh cranial nerves control the actions of several facial muscles, each side acting in synchronization to "tell" eyelids to close, tears to form, saliva to be created within the mouth, or the mouth to turn up in a smile. When the nerve is unable to transmit the message to facial muscles to relax or contract, facial muscles quickly become paralyzed or weakened. Such paralysis normally lasts only for the period of time that the nerve is unable to transmit messages. Because this swelling and infection usually affects only the seventh cranial nerve on one side of the head, the resultant paralysis normally occurs solely on one side of the face and affects only the facial areas that the seventh cranial nerve transmits to.

Because there is a wide variance in the severity of symptoms, signs of Bell's palsy may not be immediately noticed by parents. However, classical symptoms of Bell's palsy include:

- Though not always present, the child may complain of **headache** or **pain** behind or in front of the ear a few days prior to the onset of Bell's palsy.
- One side of the face droops, feels stiff or numb. (Though one side of the face is always affected, there are varying degrees of severity of this facial paralysis. Some children have only very mild weakness of facial muscles while others may be totally unable to move that side of their face.)
- An over-all droopy appearance of the child's facial expression.
- Swelling of the child's face.
- The child has a continually runny or stuffy nose.
- The child has either excessive or reduced production of saliva.
- The child is having difficulty speaking.
- The child is unable to blink or completely close one eye.
- Drooping of one side of the child's mouth is noted.
- The child has either excessive tears or marked dryness and inability to make tears in one eye.
- There are problems with the child holding food or fluids in the affected side of the mouth, resulting in drooling or difficulty swallowing.
- The child complains of either diminished, distorted or complete inability to taste food or drink.
- The child is experiencing *Hyperacusis*, or hearing sounds as seeming louder than they really are.
- The child is experiencing photosensitivity, or sensitivity to light.
- The child complains of dizziness.

When to call the doctor

Signs and symptoms of Bell's palsy typically manifest themselves within 14 days after a child has had a viral or bacterial infection. There is usually a very rapid onset once facial paralysis or weakness makes an appearance, and Bell's palsy normally reaches its peak symptoms within 48 hours of onset. In some rare cases, symptoms may take longer than this, but have very seldom been shown to take longer than two weeks to develop. It is of tremendous importance to clarify the diagnosis, and assure that it is truly Bell's palsy that a child is suffering from as soon as possible. This is because there are several other, far more serious and even life-threatening possible causes for facial paralysis in children.

These possible causes include:

- head trauma such as blunt force injuries, including temporal bone **fractures** or damage to the brain stem
- brain pathology, including neuromas, brain tumors, or cysts

- otitis media
- mastoiditis
- abcess of the temporal bone
- accidental surgical injury
- less likely causes such as congenital conditions, lupus, diabetes, or thyroid conditions

These conditions are considerably more dangerous to a child or teen than Bell's palsy and will require immediate, possibly emergency treatment as quickly as feasible. It is important to remember that paralysis in any other part of the body than the face is definitely not Bell's palsy and should be evaluated by a medical professional as soon as possible. As the facial paralysis of Bell's palsy is usually perceived correctly by parents to be a neurological condition, neurologists are often consulted. However, pediatricians and otolaryngologists (ENT—ear, nose and throat specialists) also treat Bell's palsy.

Diagnosis

Reaching a diagnosis of Bell's palsy is a process of ruling out other possible causes for the child's complaints and the observed symptoms. As noted previously, other, more serious possible causes of facial paralysis need to be eliminated before diagnosis can be made. Paralysis located in any other part of the body than the face definitely rules out Bell's palsy, and should be considered a more serious potential problem. A detailed history, including queries about recent injuries or falls; as well as various imaging tests such as **magnetic resonance imaging** (MRI), **computed tomography** (CT) scans, **x rays**, and electromyography (EMG) assure that the correct diagnosis is made.

Ramsey-Hunt syndrome

Another differential cause of facial paralysis similar to Bell's palsy is Ramsey-Hunt Syndrome. Ramsey-Hunt's chief differences from Bell's palsy are both its causative agent and the severity of some symptoms. It has been conclusively proven that another herpes virus—varicella zoster virus (VZV), the cause of both chickenpox and shingles—is the culprit for Ramsey-Hunt syndrome. This syndrome is usually an adult disease whose incidence increases after the age of 50. However, children and young adults found to have Ramsey-Hunt syndrome are considered at risk for, and in need of evaluation for, having autoimmune diseases.

Some of the symptoms that differentiate Ramsey-Hunt syndrome from Bell's palsy include:

- shingles, or painful skin eruptions, that last for two to five weeks
- more severe ear pain, often located inside of the ear
- more severe and longer-lasting dizziness
- loss of hearing (This occurs because Ramsey-Hunt syndrome also affects the eighth cranial nerve that is responsible for hearing.)
- swollen, painful lymph nodes near the area involved

Treatment

General treatment

Though most nerve compression in Bell's palsy is mild and temporary for children, the primary goal is to assure that no further damage to the seventh cranial nerve occurs. Careful monitoring is necessary, and in some cases aggressive treatment may include eliminating the swelling and inflammation that is compressing the nerve as quickly as possible. Typically the ideal time for reducing this inflammation is within the first seven days after diagnosis. A 2001 NINDS study showed steroids such as prednisone and the antiviral medication acyclovir offer some relief of these symptoms, but are considered a more controversial treatment by some health care professionals when prescribed for children. Mild **analgesics** such as **acetaminophen** (Tylenol) may be ordered if there is pain. Because of changes in saliva production and difficulty swallowing, extra care in **oral hygiene** for the child may be necessary. As in any infection or injury, rest and good **nutrition** is of paramount importance in allowing the body to heal itself.

Monitoring the state of, and providing care to the affected eye is very important. Tears may not be produced at all, or if produced, run out without actually lubricating the eye. This can cause a stinging or burning sensation in the child's eye due to dryness. Under normal circumstances, we protect our eyes by blinking every five to seven seconds. This provides moisture by moving tears across the eye and stops the entrance of debris from the external world. When the eye is unable to produce tears or close completely or to blink, as often occurs in Bell's palsy, there is danger of doing permanent damage to the cornea of the eye. Children with Bell's palsy who are old enough to follow instructions and are showing eye symptoms should be taught to manually "blink" the eye by holding the lid shut every few minutes with one finger, especially when the eye feels dry. Artificial tear products may be ordered by the pediatrician or specialist. Tinted **eyeglasses** or sunglasses may be helpful. A patch and eye ointment can be necessary at night if the child is unable to close an eye. If the eye is seriously affected, an ophthalmologist should be consulted to develop the best means of protection for the eye.

When facial paralysis persists

Though most cases of Bell's palsy resolve uneventfully in children, some do not. It is possible that rehabilitation, including retraining the brain through facial **exercises**, or even surgical correction for weakened facial muscles can be necessary in extreme cases. In the early stage of Bell's palsy, when facial muscles are the most flaccid, it is desirable to allow the muscles to simply rest and recover on their own. Gentle massage and moist warmth may provide pain relief and improve circulation, but stronger interventions should wait. Usually facial exercises will not be necessary for children with Bell's palsy unless the paralysis does not resolve itself and there is long-term damage to nerves. However facial exercises such as wrinkling the forehead, flaring and sniffling the nostrils, curling and puckering the lips, and several others may be used to retrain the brain's messages to facial muscles. Even younger children can often be taught to do these exercises, and they can be presented by parents or therapists as playing a game—making faces in the mirror. Sessions of facial exercise should be brief and performed two to three times a day. A surgical procedure involving decompression of the facial nerve through extremely delicate microsurgery has, in severe cases, also been done. But its effectiveness in Bell's palsy remains at issue among child health-care providers. Benefits of this surgery are considered by some child health specialists to be insufficient compared to the risks involved.

Nutritional concerns

Because compromise of the immune system is so often a facet of children contracting Bell's palsy, good nutrition is necessary to rebuild and strengthen that immune system. This involves following the American Dietetic Association (ADA) nutritional guidelines for children, and possibly the addition of a multivitamin if the pediatrician feels it is advisable. Semi-solid foods such as yogurt, jello, pudding, or ice cream may be easier to take in than liquids if the child is experiencing swallowing difficulty.

ADA nutritional guidelines for children include:

- Grain group: Six servings per day. Includes, per serving, one slice of bread, one-half cup cooked rice or pasta, one-half cup cooked cereal or 1 oz (28 g) of ready-to-eat cereal.
- Vegetable group: Three servings per day. Includes, per serving, one-half cup of chopped raw or cooked vegetables, one cup of raw, leafy vegetables.
- Fruit group: Two servings per day. Includes, per serving, one piece of fruit or melon wedge, three-quarters cup of fruit juice, one-half cup of canned fruit, one-quarter cup of dried fruit.
- Milk group: Two servings per day. Includes, per serving, one cup of milk or yogurt, or 2 oz (57 g) of cheese.
- Meat group: Two servings per day. Includes, per serving, 2–3 oz (57–85 g) of cooked lean meat, poultry or fish, one-half cup of cooked dry beans, one egg, or two tablespoons of peanut butter.
- Fats and sweets group: Should be limited as much as possible.

Prognosis

The potential outcome from Bell's palsy is quite hopeful. NINDS notes that the majority of all Bell's palsy sufferers improve dramatically, with or without treatment, within two weeks. The Bell's Palsy Information Site notes that half of all people contracting this condition recover completely within "a short time," and another 35 percent have "good recoveries within a year." The outlook for children is better. Eighty-five percent of children with this disease recover completely. Ten percent of the children who contract Bell's palsy will have mild weakness remaining afterward, and 5 percent will have severe residual facial weakness. Statistically, 7 percent of all children that develop Bell's palsy will have a recurrent episode in the future.

Prevention

Because of the prevalence of HSV-1, the primary cause of Bell's palsy, it is extremely difficult to prevent children from coming in contact with it. Teaching children to routinely wash their hands, and to not share towels, face-cloths, cups, or silverware can be helpful. However none of these will probably stop a visiting relative or friend from kissing a child or teen, passing along the HSV-1 virus that this friend or relative may carry. Assuring that children get sufficient rest and do not become fatigued can help in maintaining and building up an immune system that can fight off these infecting agents. This strengthening or maintenance of the immune system is even more important following any childhood illness.

Parental concerns

Clearly the notion of a child having permanent facial paralysis can be quite frightening for parents as well as the child suffering from Bell's palsy. The realization of looking different—not being able to smile,

KEY TERMS

Autoimmune disorder—One of a group of disorders, like rheumatoid arthritis and systemic lupus erythematosus, in which the immune system is overactive and has lost the ability to distinguish between self and non-self. The body's immune cells turn on the body, attacking various tissues and organs.

Cornea—The clear, dome-shaped outer covering of the eye that lies in front of the iris and pupil. The cornea lets light into the eye.

Dormant—The biological state of being relatively inactive or in a resting state in which certain processes are slowed down or suspended.

Electrooculography (EOG)—A diagnostic test that records the electrical activity of the muscles that control eye movement.

Herpesvirus—A family of viruses including herpes simplex types 1 and 2, and herpes zoster (also called varicella zoster). Herpes viruses cause several infections, all characterized by blisters and ulcers, including chickenpox, shingles, genital herpes, and cold sores or fever blisters.

Human immunodeficiency virus (HIV)—A transmissible retrovirus that causes AIDS in humans. Two forms of HIV are now recognized: HIV-1, which causes most cases of AIDS in Europe, North and South America, and most parts of Africa; and HIV-2, which is chiefly found in West African patients. HIV-2, discovered in 1986, appears to be less virulent than HIV-1 and may also have a longer latency period.

Lyme disease—An acute, recurrent, inflammatory disease involving one or a few joints, and transmitted by the bite of ticks carrying the spiral-shaped bacterium *Borrelia burgdorferi*. The condition was originally described in the community of Lyme, Connecticut, but has also been reported in other parts of the United States and other countries. Knees and other large joints are most commonly involved with local inflammation and swelling.

Mastoiditis—An inflammation of the bone behind the ear (the mastoid bone) caused by an infection spreading from the middle ear to the cavity in the mastoid bone.

Mononucleosis—An infection, caused by the Epstein-Barr virus, that causes swelling of lymph nodes, spleen, and liver, usually accompanied by extremely sore throat, fever, headache, and intense long-lasting fatigue. Also called infectious mononucleosis.

Neurological disorders—Pathological conditions relating to the brain and/or nervous system.

Neuromas— Usually benign tumors affecting nerve tissue.

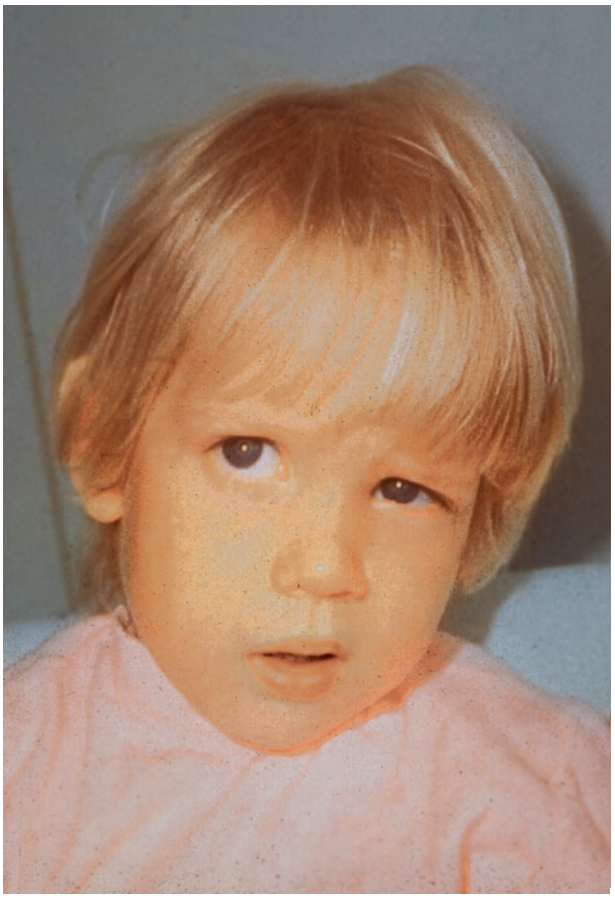
Otitis media—Inflammation or infection of the middle ear space behind the eardrum. It commonly occurs in early childhood and is characterized by ear pain, fever, and hearing problems.

Sarcoidosis—A chronic disease that causes the formation of granulomas, masses resembling small tumors composed of clumps of immune cells, in any organ or tissue. Common sites include the lungs, spleen, liver, mucous membranes, skin, and lymph nodes.

Shingles—An disease caused by an infection with the *Herpes zoster* virus, the same virus that causes chickenpox. Symptoms of shingles include pain and blisters along one nerve, usually on the face, chest, stomach, or back.

Temporal bones—The compound bones that form the left and right sides of the skull and contain various cavities associated with the ear.

close an eye or even hold fluids in the mouth properly is highly upsetting to parents, and embarrassing and frustrating for the child. Once the diagnosis of Bell's palsy is made, parents can feel reasonably optimistic that this is a condition that normally resolves itself within a set period of time, usually a matter of days or weeks. When Bell's palsy is understood, parents can generally feel some personal reassurance and transmit a sense of comfort and hope to the child. As noted previously, the paramount concern is reaching the correct diagnosis as other causes of facial or any other bodily paralysis can be of a much more serious nature. When the diagnosis has been verified by a health-care professional, accurate information about Bell's palsy can greatly alleviate further fears. The Bell's Palsy Information website <http://www.bellspalsy.ws> provides



Boy's facial paralysis caused by a tick-borne meningoradiculitis. *(Photo Researchers, Inc.)*

extensive information regarding all aspects of this disease, including measures that parents can take, and even products that can be helpful in making the child more comfortable.

Resources

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Joan Schonbeck, RN

Benzoyl peroxide *see* **Antiacne drugs**

Biliary atresia

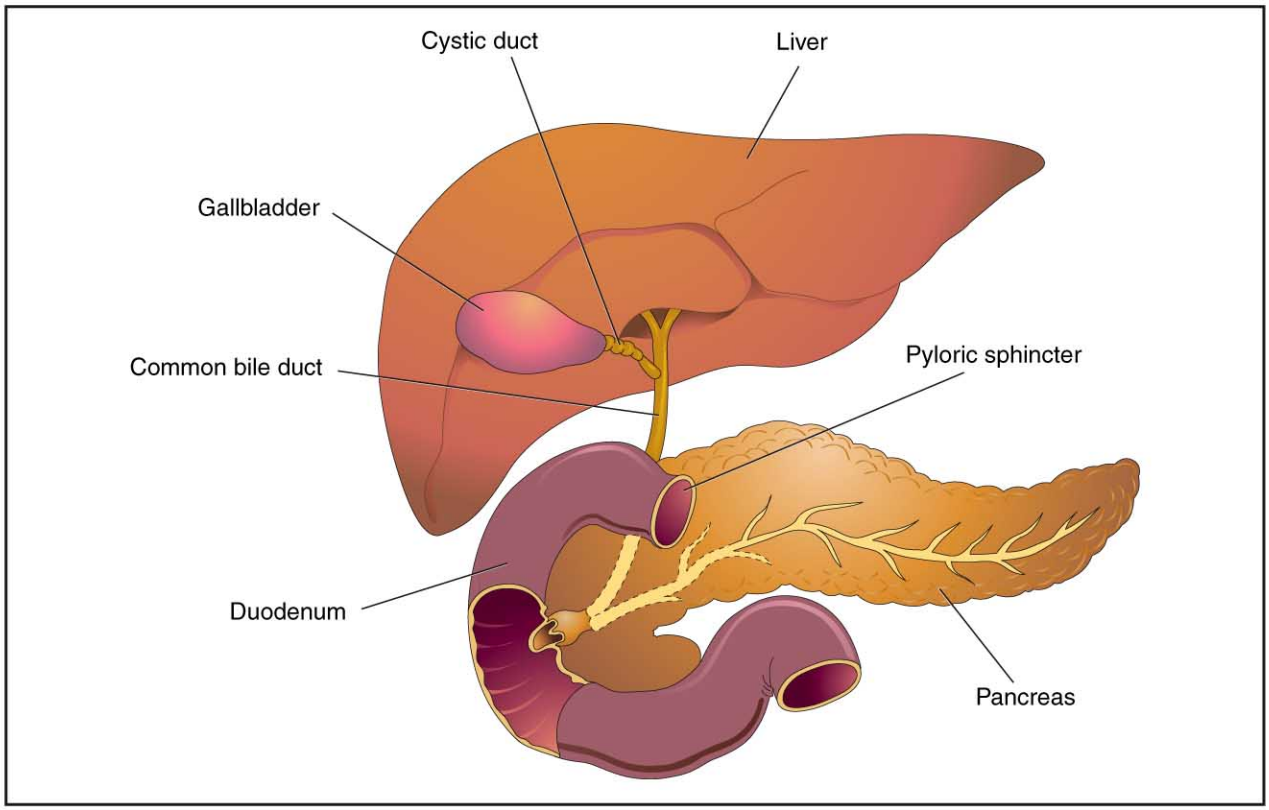
Definition

Biliary atresia is the congenital failure of a fetus to develop an adequate pathway for bile to drain from the liver to the intestine.

Description

Biliary atresia is the congenital absence or closure of the ducts that drain bile from the liver. Bile is a liquid mixture of cholesterol, bile salts, and waste products, including bilirubin, which the liver excretes through thousands of tiny biliary ducts to the intestine, where the bile aids in the digestive process of dietary fats. These ducts merge into larger and larger channels, like streams flowing into rivers, until they all pour into a single duct that empties into the duodenum (first part of the small intestine). Between the liver and the duodenum this duct has a side channel connected to the gall bladder. The gall bladder stores bile and concentrates it, removing much of its water content. Then when food enters the stomach, the gall bladder contracts and empties its contents.

If bile cannot get out because the ducts are absent or blocked, it backs up into the liver (referred to as biliary stasis) and eventually into the rest of the body. The major pigment in bile is a chemical called bilirubin, which is yellow. Bilirubin is a breakdown product of hemoglobin (the red chemical in blood that carries oxygen). If the body accumulates an excess of bilirubin, it turns yellow (jaundiced). Bile also turns the stool brown; without it, stools are pale gray-, white- or fawn-colored. Bile trapped within the liver causes damage and scarring to the liver cells (cirrhosis of the liver). Scarring of the liver can cause portal **hypertension** (high blood pressure in the portal vein, which is the main vein carrying blood from the intestine to the liver). Portal hypertension may result in the development of fragile veins in the intestinal lining, stomach, or esophagus, which can bleed and require emergency medical attention.



Biliary atresia is a congenital condition in which the pathway for bile to drain from the liver to the intestine is undeveloped. It is the most common lethal liver disease in children. *(Illustration by Electronic Illustrators Group.)*

Demographics

Biliary atresia is the most common lethal liver disease in children, occurring once every 10,000 to 15,000 live births. In the United States, approximately 300 cases of biliary atresia are diagnosed each year. Females are affected slightly more often than males. The incidence of biliary atresia is highest in Asian populations. The disorder also occurs in black infants at a rate approximately two times higher than that in white infants.

Causes and symptoms

The cause of biliary atresia is unknown. However, there are indications that viral infections or autoimmune mechanisms may be responsible for the development of biliary atresia. About 10 percent of children with biliary atresia also have other associated congenital defects in blood vessels, heart, spleen, or intestines.

The affected infant appears normal at birth and during the newborn period. After about two to three weeks, the infant develops **jaundice**. The infant has yellow eyes and skin and dark yellow or brown urine due to build-up of bilirubin, and the stools are probably light-colored. The child's abdomen begins to swell because of a firm, enlarged liver, and the infant gets progressively more ill. Weight loss and irritability will increase as the effects of jaundice increase. Some infants may develop intense **itching** (pruritis), which makes them even more uncomfortable. Nearly all untreated children die of liver failure within two years.

When to call the doctor

The doctor should be called if an infant older than two weeks of age exhibits jaundice or has other symptoms typical of biliary atresia.

If, after surgery for biliary atresia, an infant becomes jaundiced, has a high temperature for more than 24 hours, or if there is a change in the color of the stools or urine. Also after surgery, the infant may experience an abnormal collection of fluid in the abdomen, referred to as ascites, so the doctor should be consulted if the infant's stomach is distended.

If a child has black stools, pallor, or **vomiting** of blood due to the development of portal hypertension, emergency medical attention is required to treat the bleeding.

Diagnosis

The persistence or development of jaundice beyond the second week in a newborn who also has light-colored

stools indicates obstruction to the flow of bile. An immediate evaluation that includes blood tests and imaging of the biliary system (through ultrasound, specialized x-ray techniques, or radioactive screens of the liver) are required to confirm the diagnosis. Other liver diseases that cause symptoms similar to biliary atresia must be ruled out through the testing process. In addition, in most cases, a liver biopsy or a surgical exploration of the infant's abdomen is necessary for a definitive diagnosis.

Treatment

Surgery is the only means to treat biliary atresia. The surgeon must create an adequate pathway for bile to escape the liver into the intestine. The altered anatomy of the biliary system is different in every case, calling upon the surgeon's skill and experience to select and execute the most effective among several options. If the obstruction is only between the gall bladder and the intestine, it is possible to attach a piece of intestine directly to the gall bladder.

If the upper biliary system is also inadequate, the surgeon will attach a piece of intestine directly to the liver using the Kasai procedure, named after Morio Kasai, the Japanese surgeon who developed the procedure. The tiny bile ducts in that part of the liver where the surgery is performed discharge their bile directly into the intestine, and the channels will gradually enlarge. A possible complication after the Kasai operation is an infection in the bile ducts (cholangitis). This infection must be treated immediately with intravenous **antibiotics**. If the child develops ascites (abnormal build-up of fluid in the abdomen), treatment consists of medications and alteration of the diet to maintain calorie intake but to reduce salt and fluid intake.

The operation is most successful in infants under the age of eight weeks. However, in many cases, liver damage may continue to occur, and without further intervention, cirrhosis of the liver and associated complications may develop. Continued problems often develop because there are also obstructed ducts within the liver that cannot be surgically treated. In these cases, liver transplantation is required. Improved techniques of liver transplantation, which allow transplantation in children of any age, and development of drugs that help overcome the problems of organ rejection offer significant hope to children with biliary atresia who are not successfully treated with surgical techniques.

Nutritional concerns

A low- or modified-fat diet with supplementary **vitamins** is often required after surgery, since the absorption of fats and vitamins can be impaired. Postoperative breastfeeding is encouraged whenever possible, as breast milk contains lipases and bile salts to aid in digestion. Infants who are formula-fed should use special formulas (Alimentum, Pregestimil) that contain chemicals to enhance digestion of dietary fats. Extra calories may also be required to help the infant gain weight. A dietary expert should be consulted to guide in the development of feeding requirements for an infant who has been treated surgically for biliary atresia.

KEY TERMS

Cirrhosis—A chronic degenerative disease of the liver, in which normal cells are replaced by fibrous tissue and normal liver function is disrupted. The most common symptoms are mild jaundice, fluid collection in the tissues, mental confusion, and vomiting of blood. Cirrhosis is associated with portal hypertension and is a major risk factor for the later development of liver cancer. If left untreated, cirrhosis leads to liver failure.

Duodenum—The first of the three segments of the small intestine. The duodenum is about 10 in (25 cm) long and connects the stomach and the jejunum.

Hemoglobin—An iron-containing pigment of red blood cells composed of four amino acid chains (alpha, beta, gamma, delta) that delivers oxygen from the lungs to the cells of the body and carries carbon dioxide from the cells to the lungs.

Jaundice—A condition in which the skin and whites of the eyes take on a yellowish color due to an increase of bilirubin (a compound produced by the liver) in the blood. Also called icterus.

Prognosis

Early diagnosis of biliary atresia is essential, for if left untreated, few children survive beyond the age of two years. If surgery is performed before the infant is two months old, success is much more likely, while after three months of age, the success rate is much poorer. Unfortunately for many infants, surgery is not a cure, and complications of cirrhosis of the liver may develop gradually, and the child eventually requires liver transplantation to avoid an early death. Transplantation as of 2004 achieves up to 80 to 90 percent one-year survival rates and promises to prevent the chronic disease that used to accompany earlier surgical procedures.

Prevention

Since the specific cause of this birth defect is unknown, there is no way known as of 2004 to prevent biliary atresia. However, it is not a hereditary condition.

Parental concerns

Parents of children with biliary atresia require help in coping with the strain of this chronic illness as well as the stress associated with waiting for a liver transplant. Parents may also feel guilty because they feel that they may have in some way contributed to the development of biliary atresia, although as of 2004, there is no known way to prevent the disease. The American Liver Foundation organizes and coordinates mutual help groups to provide emotional support for families, to make referrals to specialists as needed, and keep parents aware of research developments.

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Bilingualism/Bilingual education

Definition

Bilingualism is the ability to communicate in two different languages. Bilingual education is the use of two different languages in classroom instruction.

Description

Languages are learned most readily during the toddler and **preschool** years and, to a lesser extent, during elementary school. Therefore, children growing up in bilingual homes and/or receiving bilingual education easily acquire both languages. Throughout much of the world, bilingualism is the norm for both children and adults. In the past, immigrants to the United States often began learning and using English in their homes as soon as possible. In the early 2000s, however, many immigrants choose to maintain their native language at home. Bilingual children are at an advantage in this increasingly multilingual nation.

Bilingual language development

Language acquisition is very similar for monolingual and bilingual children, although some experts view bilingualism as a specialized case of **language development**. Children growing up in homes where two different languages are spoken usually acquire both languages simultaneously. Although their acquisition of each language may be somewhat slower than that of children who are acquiring a single language, their development in the two languages combined is equivalent to that of monolingual children. Bilingual language learners proceed through the same patterns of language and speech development as children acquiring a single language. Their first words usually are spoken at about one year of age, and they begin stringing two words together at about age two. Even if the two languages do not share similarities in pronunciation, children eventually master them both.

There are two major patterns of bilingual language development, both occurring before the age of three. Simultaneous bilingualism occurs when a child learns both languages at the same time. In the early stages of simultaneous bilingual language development, a child may mix words, parts of words, and inflections from both languages in a single sentence. Sometimes this occurs because a child knows a word in one language but not in the other. Some bilingual children initially resist learning words for the same thing in two languages. Children also may experiment with their two languages for effect. During the second stage of bilingual language development, at age four or older, children gradually begin to distinguish between the two languages and use them separately, sometimes depending on where they are. One language may be used less formally to talk about home and **family**, whereas the other language may be used more formally, perhaps for relating events that took place outside the home. Often children find it easier to express a specific idea in one language rather than the other. Bilingual children also go through periods when one language is used more than the other. Some children may begin to prefer one language over the other, particularly if that language is spoken more frequently in their home or school. Bilingual children usually are not equally skilled in both languages. Often they understand more in one language but speak more in the other.

Sequential bilingualism occurs when children use their knowledge of and experience with a first language to

rapidly acquire a second language. The first language may influence the way in which they learn and use their second language. Learning the second language is easier for children if the sounds, words, and vocabulary of the languages are similar.

Bilingual language development usually proceeds more smoothly when both languages are introduced early and simultaneously. When the parents each use a different language with their child, the child is less likely to experience language confusion.

Research indicates that there are numerous advantages to bilingualism. Bilingualism has been reported to improve the following skills:

- verbal and linguistic abilities
- general reasoning
- concept formation
- divergent thinking
- metalinguistic skills, the ability to analyze and talk about language and control language processing

These abilities are important for reading development in young children and may be a prerequisite for later learning to read and write in a new language.

Types of bilingual education

Bilingual education is common throughout the world and involves hundreds of languages. In the United States bilingualism is assumed to mean English and another language, often Spanish. More than 300 languages are spoken in the United States. In New York City schools, classroom instruction is given in 115 different languages. Bilingual education includes all teaching methods that are designed to meet the needs of English-language learners (ELLs), also referred to as ''limited English proficient'' (LEP) students.

There are numerous approaches to bilingual education, although all include English as a second language (ESL). ESL is English language instruction that includes little or no use of a child's native language. ESL classes often include students with many different primary languages. Some school districts use a variety of approaches to bilingual education, designing individual programs based on the needs of each child.

A common approach is transitional bilingual education (TBE). TBE programs include ESL; however, some or all academic classes are conducted in children's primary languages until they are well-prepared for English-only classes. Even children who converse well in English may not be ready to learn academic subjects in English. Often these children spend part of the school day in an intensive ESL program and the remainder of the day receiving instruction in their primary language. Bilingual teachers may help students improve their primary language skills. Bilingual/bicultural programs include instruction in the history and culture of a student's ethnic heritage. Studies have shown that children who receive several years of instruction in their native language learn English faster and have higher overall academic achievement levels that those who do not.

Two-way bilingual or dual-language programs use both English and a second language in classrooms made up of both ELLs and native English speakers. The goal is for both groups to become bilingual. Children in two-way bilingual education programs have been found to outperform their peers academically.

Many educators—and a segment of the public—believe in the English immersion approach, even if ELLs do not understand very much in the classroom. In this approach nearly all instruction is in English, and there is little or no use of other languages. If the teacher is bilingual, students may be allowed to ask questions in their native language, but the teacher answers them in English. Some schools employ structured English immersion or sheltered English, in which teachers use pictures, simple reading words, and other techniques to teach ELLs both English and academic subjects.

History of bilingual education

Although bilingual education has been used in the United States for more than 200 years, the 1968 Title VII amendment to the 1965 Elementary and Secondary Education Act (ESEA) instituted federal grants for bilingual education programs. This legislation led to the development of appropriate teaching and learning materials and training for teachers of bilingual students.

In 1974 the U.S. Supreme Court ruled that the San Francisco school system had violated the Civil Rights Act of 1964 by not providing English-language instruction for Chinese-speaking students. All school districts were directed to serve ELLs adequately, and bilingual education quickly spread throughout the United States. In the 1980s a group called Asian Americans United filed a class-action lawsuit charging that Asian Americans were not being provided with an equitable education because they were not offered bilingual classes. The result of this suit was the creation of sheltered ESL, in which ESL students take all of their classes together.

The No Child Left Behind (NCLB) Act of 2001—President George W. Bush's major education initiative—reauthorized the ESEA. It also imposed penalties on schools that did not raise the achievement levels of ELLs for at least two consecutive years. Although most research indicates that it often takes seven years for ELLs to attain full English

fluency, the new federal law allows these children only three years before they must take standardized tests in English. Schools with large numbers of children speaking many different languages are particularly disadvantaged under the law. A 2003 survey by the National Education Association found that 22,000 schools in 44 states failed to make the required yearly progress on standardized tests, primarily because of low test scores by ELLs and disabled students. The National Association for Bilingual Education claims that NCLB sets arbitrary goals for achievement and uses "invalid and unreliable assessments." Furthermore, although the NCLB requires teachers to be qualified, as of 2004 there is a severe shortage of qualified teachers for ELLs. Some communities have developed early-intervention programs for Spanish-speaking parents and preschoolers to help children develop their Spanish language skills in preparation for entering English-only schools.

In May of 2004, the U.S. Department of Education and faith-based community leaders launched an initiative to inform Hispanic, Asian, and other parents of ELLs about the NCLB. It featured the "Declaration of Rights for Parents of English Language Learners under No Child Left Behind."

As of 2004 American public schools include about 11 million children of immigrants. Approximately 5.5 million students—10 percent of the public school enrollment—speak little or no English. Spanish speakers account for 80 percent of these children. About one-third of children enrolled in urban schools speak a primary language other than English in their homes. Between 2001 and 2004, 19 states reported increases of 50 to 200 percent in Spanish-speaking students. ELLs are the fastest-growing public school population in kindergarten through twelfth grade. Between 2000 and 2002, nationwide ELL enrollment increased 27 percent. About 25 percent of California public school children are ELLs. However, there is a profound shortage of bilingual and ESL teachers throughout the United States. Although 41 percent of U.S. teachers have ELLs in their classrooms, only about 2.5 percent of them have degrees in ESL or bilingual education. The majority of these teachers report that they are not well-prepared for teaching ELLs. About 75 percent of ELLs are in poverty schools, where student turnover is high and many teachers have only emergency credentials.

Opposition to bilingual education

In 1980 voters in Dade County, Florida, made English their official language. In 1981 California Senator S. I. Hayakawa introduced a constitutional amendment to make English the country's official language. In 1983 Hayakawa founded U.S. English, Inc., which grew to include 1.8 million members by 2004. U.S. English argues the following premises:

- The unifying effect of the English language must be preserved in the United States.
- Bilingual education fails to adequately teach English.
- Learning English quickly in English-only classrooms is best for ELLs, both academically and socially.
- Any special language instruction should be short-term and transitional.

In 1986 California voters passed Proposition 63 that made English the state's official language. Other states did the same. In 1998 Californians passed Proposition 227, a referendum that attempted to eliminate bilingual education by allowing only one year of structured English immersion, followed by mainstreaming. Similar initiatives have appeared on other state ballots. However, only 9 percent of the California children attained English proficiency in one year, and most remained in the immersion programs for a second year. Prior to the new law only 29 percent of California ELLs were in bilingual programs, in part because of a shortage of qualified teachers. Since the law allowed parents to apply for waivers, 12 percent of the ELLs were allowed to remain in bilingual classes.

In January of 2004, as part of a lawsuit settlement, the California State Board of Education was forced to radically revise the implementation of their "Reading First" program. Previously California had withheld all of the $133 million provided by NCLB from ELLs enrolled in alternative bilingual programs.

Common problems

Language delay

Language and learning difficulties occur with the same frequency in monolingual and bilingual children. However, as the number of bilingual children in the United States increases, it becomes increasingly important for parents and pediatricians to understand the normal patterns of bilingual language development in order to recognize abnormal language development in a bilingual child.

If a bilingual child has a speech or language problem, it should be apparent in both languages. However detecting language delays or abnormalities in bilingual children can be difficult. Signs of possible **language delay** in bilingual children include the following:

- not making sounds between two and six months of age
- fewer than one new word per week in children aged six to 15 months
- fewer than 20 words in the two languages combined by 20 months of age

- limited vocabulary without word combinations in children aged two to three years of age
- prolonged periods without using speech
- difficulty remembering words
- missing normal milestones of language development in the first language of a sequentially bilingual child

Language development in bilingual children can be assessed by a bilingual speech/language pathologist or by a professional who has knowledge of the rules and structure of both languages, perhaps with the assistance of a translator or interpreter.

English-only education

ELLs in English-only programs often fall behind academically. Many ELLs who are assessed using traditional methods are referred for **special education**. Such children often become school drop-outs.

Parental concerns

Parents in bilingual households can help their children by taking the following steps:

- speaking the language in which they are most comfortable
- being consistent regarding how and with whom they use each language
- using each language's grammar in a manner that is appropriate for the child's developmental stage
- keeping children interested and motivated in language acquisition

See also Language development.

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Elementary and Secondary Education Act (ESEA)—The 1965 federal law that is reauthorized and amended every five years.

English as a second language (ESL)—English language instruction for English language learners (ELLs) that includes little or no use of a child's native language; a component of all bilingual education programs.

English language learner (ELL)—A student who is learning English as a second language; also called limited English proficient (LEP).

Immersion—A language education approach in which English is the only language used.

Limited English proficient (LEP)—Used to identify children who have insufficient English to succeed in English-only classrooms; also called English language learner (ELL).

Metalinguistic skills—The ability to analyze language and control internal language processing; important for reading development in children.

No Child Left Behind (NCLB) Act—The 2001 reauthorization of the ESEA, President George W. Bush's major education initiative.

Sequential bilingualism—Acquiring first one language and then a second language before the age of three.

Sheltered English—Structured English immersion; English instruction for ELLs that focuses on content and skills rather than the language itself; uses simplified language, visual aids, physical activity, and the physical environment to teach academic subjects.

Sheltered ESL—Bilingual education in which ESL students attend all of their classes together.

Simultaneous bilingualism—Acquiring two languages simultaneously before the age of three.

Structured English immersion—Sheltered English; English-only instruction for ELLs that uses simplified language, visual aids, physical activity, and the physical environment to teach academic subjects.

Transitional bilingual education (TBE)—Bilingual education that includes ESL and academic classes conducted in a child's primary language.

Two-way bilingual education—Dual language programs in which English and a second language are both used in classes consisting of ELLs and native-English speakers.

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National Association for Multicultural Education. 733 15th St., NW, Suite 430, Washington, DC 20005. Web site: <http://nameorg.org>.

National Clearinghouse for English Language Acquisition. Office of English Language Acquisition, Language Enhancement & Academic Achievement for Limited English Proficient Students, U.S. Department of Education, George Washington University Graduate School of Education and Human Development, 2121 K St., NW, Suite 260, Washington, DC 20037. Web site: <www.ncela.gwu.edu>.

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Bilirubin test

Definition

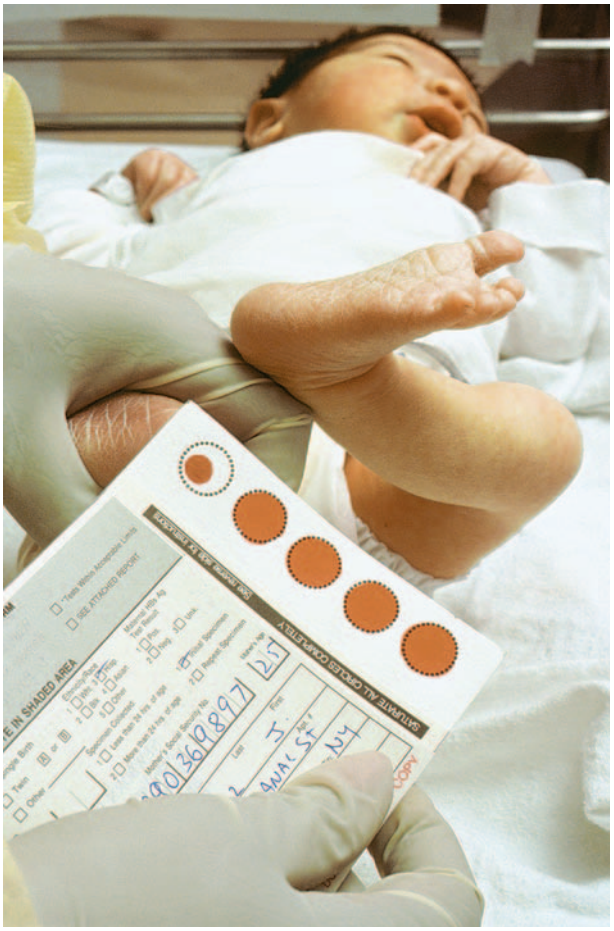
A bilirubin test is a diagnostic blood test performed to measure levels of bile pigment in an individual's blood serum and to help evaluate liver function.

Purpose

The bilirubin test is an important part of routine newborn (neonatal) diagnostic screening tests. The level of bilirubin in a newborn's blood serum is measured to determine if the circulating level of bilirubin is normal or abnormal. Bilirubin is a yellow-orange bile pigment produced during the breakdown of hemoglobin, the iron-bearing and oxygen-carrying protein in red blood cells. All individuals produce bilirubin daily as part of the normal turnover of red cells. A higher than normal (elevated) bilirubin test can reflect accelerated red blood cell destruction or may indicate that bilirubin is not being excreted as it should be, suggesting that liver function problems or other abnormalities may be present. Neonatal bilirubin screening often reveals an elevated bilirubin (hyperbilirubinemia). The bilirubin test will determine if hyperbilirubinemia is present and, along with other diagnostic tests, help determine if the condition is relatively normal (benign) or possibly related to liver function problems or other conditions.

Description

Usually all newborns (neonates) delivered in the hospital will have total serum bilirubin (TSB) measured in the



Blood taken from the heel of a newborn to test the level of bilirubin. *(© Ted Horowitz/Corbis.)*

clinical laboratory on one or more blood samples as requested by attending pediatricians. To obtain a blood sample for TSB, a phlebotomist takes blood from the infant's tissue (usually the heel) rather than from a vein, as the veins of newborns are extremely small and easily damaged. After sterilizing the surface of the site with alcohol and/or an antibacterial solution such as betadine, a heel puncture is made and blood from the puncture is drawn into a tiny capillary tube about 2 inches (5 cm) long that is stoppered at each end when full. This tube is spun down in a special centrifuge in the laboratory to separate serum, the liquid part of blood, from red cells. In the TSB test, spectrophotometry is used to identify and quantify the amount of bilirubin in a specific amount of serum by measuring the amount of ultraviolet light absorbed by bilirubin pigment in the sample. The test method requires only minutes and a very small amount of blood serum to produce accurate results, measuring the results in milligrams per desiliter (mg/dL). The amount of total bilirubin in circulating blood can be calculated from the results of a single bilirubin test. Results are compared to known normal values to determine if the individual has normal or abnormal levels.

All newborn infants begin to destroy fetal red blood cells (RBCs) in their first few days of life, replacing them with new red blood cells. The rapid destruction of red blood cells and subsequent release of fetal hemoglobin into the bloodstream results in the production of bilirubin. As a waste product, bilirubin is filtered out of blood (cleared) by the liver and excreted in bile, eliminated normally in stool produced by the large intestine. However, immediately after birth, more bilirubin is produced than the infant's immature liver can handle, and the excess remains circulating in the blood. This situation results in **jaundice** in over 60 percent of newborns, usually due to the presence of fetal hemoglobin released into the blood during the normal destruction of fetal red blood cells. Even healthy infants may appear to have a yellow stain in their skin (physiological jaundice or icterus) and the whites of the eyes (sclerae) in the first week after birth. This may first be noticed by pediatric nurses as they care for the infant. Visual evaluation of jaundice is not considered a reliable way, however, to determine its cause or the risk of continued rising of bilirubin and possible complications. Performing bilirubin tests is the first step in making sure that normal degrees of jaundice do not become more severe and that liver dysfunction or other causative conditions, if present, are identified and treated early.

Besides normal red cell destruction after birth, neonatal hyperbilirubinemia may also be caused by the following:

- low birth weight
- feeding or **nutrition** problems
- glucose 6-phospho-dehydrogenase (G6PD) deficiency
- insufficient intestinal bacteria
- incompatibility of major blood groups (ABO) between mother and baby
- blood type (Rh) incompatibility (rare due to treatment of Rh negative mothers)
- genetic abnormalities linked to a history of jaundice among siblings
- liver dysfunction

From 8 to 9 percent of newborns develop severe hyperbilirubinemia. Severe hyperbilirubinemia is of great concern to pediatricians because it may lead to bilirubin-related brain damage (kernicterus). Persistent elevated levels of bilirubin in the body can place infants at risk of neurotoxicity or bilirubin-induced neurologic dysfunction (BIND). The risk of liver dysfunction has been shown to be higher in infants who were born before term (less than

37 weeks' gestation) or who have other abnormalities in addition to an elevated total serum bilirubin.

Some pediatricians order bilirubin tests at defined times within 24 to 48 hours after birth to monitor the rate of increase of bilirubin and to help determine associated risks on an individual basis. Infants with a low rate of rise in bilirubin (less than 17mg/dL per hour) are considered lower risk and are likely to be discharged without further testing or treatment. Those who show visual jaundice at birth or within several hours after birth and whose rate of bilirubin rises more rapidly are considered at higher risk for severe hyperbilirubinemia and associated kernicterus, especially if the bilirubin level is still rising at time of discharge.

Some newborns are placed under special lamps (phototherapy) to help correct the jaundice caused by elevated bilirubin levels and to bring down the bilirubin level. Supervision of breastfeeding and supplemental nutritional support may be needed to help infants who are not getting their nutritional needs met. Exchange transfusions may be given for high-risk infants, especially those with blood group (ABO) or type (Rh positive infants born to Rh negative mothers) incompatibilities. Additional tests may be required to evaluate G6PD deficiency, genetic abnormalities, or liver function.

After discharge from the hospital, about 25 percent of otherwise healthy infants who are still showing signs of jaundice may continue to be tested for bilirubin levels. An elevated bilirubin usually goes down on its own if the hyperbilirubinemia is benign; if liver dysfunction or other abnormalities exist, bilirubin levels may remain elevated or continue to rise, indicating that further diagnostic testing, clinical evaluation, and treatment are needed.

Precautions

Performance of the bilirubin test itself is a precaution against the serious consequences that can occur when bilirubin levels continue to rise in jaundiced infants. Visual jaundice present at birth may predict rapid rises in bilirubin and risk of liver dysfunction or other abnormalities.

Preparation

No preparation is needed before performing bilirubin tests on infants' blood samples. Proper identification and careful handling of the infant are important when a blood sample is being obtained for testing. A site, usually on the infant's heel, is chosen by the phlebotomist who draws the infant's blood sample. The area is prepared by wrapping the baby's foot in a warm cloth for a few minutes to bring blood to the surface and allow it to flow more easily. The heel is then wiped with alcohol and/or an antibacterial solution such as betadine to sterilize the surface. The heel is then punctured with a lancet, avoiding the center of the heel, in order to prevent inflammation of the bone. The blood sample is drawn in tiny capillary tubes, properly labeled, and taken to the laboratory for testing. In rare instances, a phlebotomist is not able to draw sufficient blood from a heel puncture, and a physician may draw venous blood from a femoral vein in the groin area, which is larger than veins in an infant's arms.

Aftercare

The site from which blood is withdrawn must be kept clean after the procedure and must be checked regularly for bleeding. A small adhesive patch may be used to protect the site.

KEY TERMS

Bilirubin—A reddish yellow pigment formed from the breakdown of red blood cells, and metabolized by the liver. When levels are abnormally high, it causes the yellowish tint to eyes and skin known as jaundice. Levels of bilirubin in the blood increase in patients with liver disease, blockage of the bile ducts, and other conditions.

Glucose-6-phosphate dehydrogenase (G6PD) deficiency—A sex-linked hereditary disorder in which the body lacks an enzyme that normally protects red blood cells from toxic chemicals. When people with this condition take certain drugs, their red blood cells break down, causing anemia.

Hyperbilirubinemia—A condition characterized by a high level of bilirubin in the blood. Bilirubin is a natural byproduct of the breakdown of red blood cells, however, a high level of bilirubin may indicate a problem with the liver.

Kernicterus—A potentially lethal disease of newborns caused by excessive accumulation of the bile pigment bilirubin in tissues of the central nervous system.

Neurotoxic—Refers to a substance that is harmful to the nervous system.

Phlebotomist—A person who draws blood from a vein.

Spectrophotometry—A testing method that measures the amount of ultraviolet light absorbed by specific substances such as bilirubin pigment. A spectrophotometer can accurately measure how much bilirubin is in a blood sample and the result can be compared to known normal values.

Risks

The performance of bilirubin tests carries no significant risk. Drawing blood for the test may involve light bleeding or bruising at the site of puncture, or blood may accumulate under the puncture site (hematoma), requiring that a new location be found for subsequent tests. Not performing bilirubin tests, however, may have significant risks for some infants. Infants with rising bilirubin levels are at risk of neurotoxicity and developing kernicterus, making the monitoring of bilirubin in the first week of life critical for these infants.

Normal results

At birth, a newborn's TBS is normally 1 or 2 mg/dL, peaking at 6 mg/dL in three or four days. In 10 days to two weeks, a healthy infant's TBS is expected to be less than 0.3 mg/dL.

During the first seven days of the infant's life, TBS results are rated for risk of bilirubin toxicity or bilirubin-related brain damage within percentile ranges representing degrees of hyperbilirubinemia. TBS values less than 20 mg/dL are lower-risk percentile ranges below the 95th percentile, with an incidence of one in nine infants. TBS values greater than 20 mg/dL are in the 98th percentile, with an incidence of one in 50 infants; greater than 25 mg/dL are in the 99.9 percentile, with an incidence of one in 700 infants; and TBS values greater than 30 ng/dL are at the highest level of risk at 99.99 percentile, indicating almost certain neurotoxicity. One in 10,000 infants are in the 99.99 percentile.

Parental concerns

Parents will usually be informed by the pediatrician about any risks associated with an elevated bilirubin, such as liver dysfunction or possible kernicterus. Parents concerned about these risks can be made aware that bilirubin levels usually return to normal in most infants (more than 60%) and the related jaundice goes away gradually. Testing after the baby is discharged is sometimes necessary (in 25% of infants) and is a preventive measure rather than a cause for concern. Repeat testing is necessary to monitor bilirubin levels. Parents should be aware that, although the baby's heel may be bruised, elevated bilirubin levels can cause serious complications, and testing is critical to help prevent them.

See also Neonatal jaundice.

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L. Lee Culvert

Binge eating disorder

Definition

Binge eating disorder (BED) is characterized by loss of control over eating behaviors. The binge eater consumes unnaturally large amounts of food in a short time period, but unlike a bulimic, does not regularly engage in any inappropriate weight-reducing behaviors such as excessive **exercise**, induced **vomiting**, or taking **laxatives** following the binge episodes.

Description

BED typically strikes individuals sometime between **adolescence** and the early twenties. Because of the nature of the disorder, most BED patients are overweight or obese. Studies of weight loss programs have shown that an average of 30 percent of individuals enrolling in these programs report binge eating behavior.

Demographics

Binge eating affects an equal numbers of females and males. Although there are no good statistics on how many children suffer from the condition, an estimated 1 to 2 million Americans of all ages are binge eaters. Many of them report that their condition started in childhood.

Causes and symptoms

Binge eating episodes may act as a psychological release for excessive emotional stress. Other circumstances that may make a child or adolescent more likely to engage in binge eating include heredity and certain psychological affective disorders such as major depression. BED patients are also more likely to have a comorbid (co-existing) diagnosis of impulsive behaviors such as compulsive buying, post-traumatic stress disorder (PTSD), panic disorder, or **personality disorders**.

Individuals who develop BED often come from families who put an extreme emphasis on the importance

of food as a source of comfort in times of emotional distress. Children with BED may have been taught to clean their plates regardless of their satiety or that their finishing a meal makes them a "good" girl or boy. Cultural attitudes towards beauty and thinness may also be a factor in whether a person binges.

During binge episodes, BED patients experience a definite loss of control over their eating. They eat quickly and to the point of discomfort even if they are not hungry. They typically binge alone two or more times a week and often feel depressed and guilty when the episode concludes.

Diagnosis

Binge eating disorder is usually diagnosed and treated by a psychiatrist and/or a psychologist. In addition to an interview with the child, personality and behavioral inventories, such as the **Minnesota Multiphasic Personality Inventory** (MMPI), may be administered as part of the **assessment** process. One of several clinical inventories, or scales, may also be used to assess depressive symptoms, including the Hamilton Depression Scale (HAM-D) or Beck Depression Inventory (BDI). These tests may be administered in an outpatient or hospital setting.

Treatment

Many BED individuals binge after long intervals of excessive dietary restraint; therapy helps normalize this pattern. The initial goal of BED treatment is to teach the patient to gain control over the eating behavior by focusing on eating regular meals and avoiding snacking. Cognitive-behavioral therapy (learning new behavior), group therapy, or interpersonal psychotherapy may be employed to uncover the emotional motives, distorted thinking, and behavioral patterns behind the binge eating.

Because the prevalence of depression in BED patients is high, treatment with **antidepressants** may also be prescribed. Once the binge eating behavior is curbed and depressive symptoms are controlled, the physical symptoms of BED can be addressed. The overweight BED patient may be placed on a moderate exercise program and a nutritionist may be consulted to educate the patient on healthy food choices and strategies for weight loss.

Prognosis

If left unchecked, the poor dietary habits and **obesity** that are symptomatic of BED can lead to serious health problems, such as high blood pressure, heart attacks, and type 2 diabetes. BED is a chronic condition that requires ongoing medical and psychological management. Some of these conditions such as diabetes can occur in young people. To bring long-term relief to the BED patient, it is critical to address the underlying psychological causes for binge eating behaviors. It appears that up to 50 percent of BED patients stop bingeing with cognitive behavioral therapy.

KEY TERMS

Bulimia nervosa—An eating disorder characterized by binge eating and inappropriate compensatory behavior, such as vomiting, misusing laxatives, or excessive exercise.

Cognitive-behavioral therapy—A type of psychotherapy in which people learn to recognize and change negative and self-defeating patterns of thinking and behavior.

Parental concerns

Binge eating can lead to excessive weight, a risk for serious current and future diseases including heart disease, type 2 diabetes, and **cancer**. Overweight children also suffer from psychological distress, particularly when teased or shunned by peers. Parents should be aware that antidepressant drugs used to treat BED as of 2004 contain a warning that recommends close observation of pediatric patients treated with the drugs. In some cases, worsening depression or emergence of suicidal tendencies may occur.

See also Bulimia nervosa.

Resources

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American Psychological Association (APA). 750 First St. NE, Washington, DC 20002–4242. Web site: <www.apa.org>.

Eating Disorders Awareness and Prevention. 603 Stewart St., Suite 803, Seattle, WA 98101. Web site: <www.edap.org>.

National Eating Disorders Association (NEDA). 603 Stewart St., Suite 803, Seattle, WA 98101. Web site: <www.nationaleatingdisorders.org>.

Overeaters Anonymous World Service Office. 6075 Zenith Ct. NE, Rio Rancho, NM 87124. Web site: <www.overeatersanonymous.org>.

Christine Kuehn Kelly

Bipolar disorder

Definition

Bipolar, or manic-depressive disorder, is a mood disorder that causes radical emotional changes and mood swings, from manic highs to depressive lows. The majority of bipolar individuals experience alternating episodes of mania and depression.

Description

The Diagnostic and Statistical Manual of Mental Disorders, fourth edition (*DSM-IV*), the diagnostic standard for mental health professionals in the United States, defines four separate categories of bipolar disorder: bipolar I, bipolar II, cyclothymia, and bipolar not-otherwise-specified (NOS).

Bipolar I disorder is characterized by manic episodes, the "high" of the manic-depressive cycle. A person with bipolar disorder experiencing mania often has feelings of self-importance, elation, talkativeness, increased sociability, and a desire to embark on goal-oriented activities, coupled with the characteristics of irritability, impatience, impulsiveness, hyperactivity, and a decreased need for **sleep**. Usually this manic period is followed by a period of depression, although a few bipolar I individuals may not experience a major depressive episode. Mixed states, where both manic or hypomanic symptoms and depressive symptoms occur at the same time, also occur frequently with bipolar I patients (for example, depression with the racing thoughts of mania). Also, dysphoric mania is common (mania characterized by anger and irritability).

Bipolar II disorder is characterized by major depressive episodes alternating with episodes of hypomania, a milder form of mania. Bipolar depression may be difficult to distinguish from a unipolar major depressive episode. Patients with bipolar depression tend to have extremely low energy, retarded mental and physical processes, and more profound fatigue (for example, hypersomnia, a sleep disorder marked by a need for excessive sleep or sleepiness when awake) than unipolar depressives.

Cyclothymia refers to the cycling of hypomanic episodes with depression that does not reach major depressive proportions. One third of patients with cyclothymia develop bipolar I or II disorder later in life.

A phenomenon known as rapid cycling occurs in up to 20 percent of bipolar I and II patients. In rapid cycling, manic and depressive episodes must alternate frequently, at least four times in 12 months, to meet the diagnostic definition. In some cases of "ultra-rapid cycling" the patient may bounce between manic and depressive states several times within a 24-hour period. This condition is very hard to distinguish from mixed states.

Bipolar NOS is a category for bipolar states that do not clearly fit into the bipolar I, II, or cyclothymia diagnoses.

Demographics

According to the American Academy of Child and Adolescent Psychiatry, up to one third of American children and adolescents diagnosed with depression develop early onset bipolar disorder. The average age of onset of bipolar disorder is from **adolescence** through the early twenties. However, because of the complexity of the disorder, a correct diagnosis can be delayed for several years or more. In a survey of bipolar patients conducted by the National Depressive and Manic Depressive Association (MDMDA), one half of respondents reported visiting three or more professionals before receiving a correct diagnosis, and over one third reported waiting ten years or more before they were correctly diagnosed.

Causes and symptoms

The cause of bipolar disorder had not as of 2004 been clearly defined. Because two thirds of bipolar patients have a **family** history of affective or emotional disorders, researchers have searched for a genetic link to the disorder. Several studies have uncovered a number of possible genetic connections to the predisposition for bipolar disorder. A 2003 study found that **schizophrenia** and bipolar disorder could have similar genetic causes that arise from certain problems with genes associated with myelin development in the central nervous system. (Myelin is a white, fat-like substance that forms a sheath around nerve fibers.) Another possible biological cause under investigation is the presence of an excessive calcium build-up in the cells of bipolar patients. Dopamine and other neurochemical transmitters appear to be implicated in bipolar disorder, and these are under investigation as well.

Over one-half of patients diagnosed with bipolar disorder have a history of substance abuse, which may be an issue in adolescent patients. There is a high rate of association between cocaine abuse and bipolar disorder. Some studies have shown up to 30 percent of abusers meet the criteria for bipolar disorder. The emotional and

physical highs and lows of cocaine use correspond to the manic depression of the bipolar patient, making the disorder difficult to diagnose.

For some bipolar patients, manic and depressive episodes coincide with seasonal changes. Depressive episodes are typical during winter and fall, and manic episodes are more probable in the spring and summer months.

Symptoms of bipolar depressive episodes include low energy levels, feelings of despair, difficulty concentrating, extreme fatigue, and psychomotor retardation (slowed mental and physical capabilities). Manic episodes are characterized by feelings of euphoria, lack of inhibitions, racing thoughts, diminished need for sleep, talkativeness, risk taking, and irritability. In extreme cases, mania can induce hallucinations and other psychotic symptoms such as grandiose delusions.

When to call the doctor

When symptoms of bipolar disorder are present, a child should be taken to a qualified medical healthcare professional as soon as possible for evaluation. If a child or teen diagnosed with bipolar disorder reveals at any time that they have had recent thoughts of self-injury or **suicide**, or if they demonstrate behavior that compromises their **safety** or the safety of others, professional assistance from a mental healthcare provider or care facility should be sought immediately.

Diagnosis

Bipolar disorder usually is diagnosed and treated by a psychiatrist and/or a psychologist. In addition to an interview with the child and her parents, several clinical inventories or scales may be used to assess the patient's mental status and determine the presence of bipolar symptoms. These include the Children's Global **Assessment** Scale (C-GAS), General Behavior Inventory (GBI), Beck Depression Inventory (BDI), **Minnesota Multiphasic Personality Inventory** Adolescent (MMPI-A), the Youth Inventory (YI-4), and the Young Mania Rating Scale (YMRS). The tests are verbal and/or written and are administered in both hospital and outpatient settings.

Bipolar symptoms often present differently in children and adolescents. Manic episodes in these age groups are typically characterized by more psychotic features than in adults, which may lead to a misdiagnosis of schizophrenia. Children and adolescents also tend to demonstrate irritability and aggressiveness instead of the elation of mania in adults. Further, symptoms tend to be chronic, or ongoing, rather than acute, or episodic. Bipolar children are easily distracted, impulsive, and hyperactive, which can lead to a misdiagnosis of attention deficit hyperactivity disorder (ADHD). Their aggression can lead to violence, which may be misdiagnosed as a **conduct disorder**.

Psychologists and psychiatrists typically use the criteria listed in the *Diagnostic and Statistical Manual of Mental Disorders,* fourth edition (*DSM-IV*) as a guideline for diagnosis of bipolar disorder and other mental illnesses. *DSM- IV* describes a manic episode as an abnormally elevated or irritable mood lasting a period of at least one week that is distinguished by at least three of the mania symptoms: inflated **self-esteem**, decreased need for sleep, talkativeness, racing thoughts, distractibility, increase in goal-directed activity, or excessive involvement in pleasurable activities that have a high potential for painful consequences. If the mood of the patient is irritable and not elevated, four of the symptoms are required.

Although many clinicians find the criteria too rigid, a hypomanic diagnosis requires a duration of at least four days with at least three of the symptoms indicated for manic episodes (four if mood is irritable and not elevated). *DSM-IV* notes that unlike manic episodes, hypomanic episodes do not cause a marked impairment in social or occupational functioning, do not require **hospitalization**, and do not have psychotic features. In addition, because hypomanic episodes are characterized by high energy and goal-directed activities and often result in a positive outcome or are perceived in a positive manner by the patient, bipolar II disorder can go undiagnosed.

Substance abuse can mask or mimic the presence of bipolar disorder and can make diagnosis more difficult in adolescents. When substance abuse or **addiction** is present, a patient must ordinarily undergo a period of detoxification and abstinence before a mood disorder can be accurately diagnosed.

Treatment

The manic and depressive symptoms of bipolar disorder are usually controlled by a combination of prescription medications, including lithium, antipsychotics, anticonvulsants, and **antidepressants**.

Lithium

Lithium (Cibalith-S, Eskalith, Lithane, Lithobid, Lithonate, Lithotabs) is one of the oldest and most frequently prescribed drugs available for the treatment of adult bipolar mania and depression. Because the drug takes four to ten days to reach a therapeutic level in the bloodstream, it sometimes is prescribed in conjunction with neuroleptics and/or benzodiazepines to provide more immediate relief of a manic episode. Lithium also has been shown to be effective in regulating bipolar depression, but is not recommended for mixed mania. Lithium may not be an effective long-term treatment

option for rapid cyclers, who typically develop a tolerance for it, or may not respond to it. Possible side effects of the drug include weight gain, thirst, **nausea**, and hand tremors. Prolonged lithium use also may cause **hyperthyroidism**.

Antipsychotics

Clozapine (Clozaril) is an atypical antipsychotic medication used to control manic episodes in adult patients who have not responded to typical mood stabilizing agents. The drug has also been a useful prophylactic, or preventative treatment, in some bipolar patients. Common side effects of clozapine include tachycardia (rapid heart rate), hypotension, **constipation**, and weight gain. Agranulocytosis, a potentially serious but reversible condition in which the white blood cells that typically fight infection in the body are destroyed, is a possible side effect of clozapine. Patients treated with the drug should undergo weekly blood tests to monitor white blood cell counts.

Risperidone (Risperdal) is another atypical antipsychotic that has been successful in controlling mania in several clinical trials when low doses were administered. The side effects of risperidone are mild compared to many other antipsychotics (constipation, coughing, **diarrhea**, dry mouth, **headache**, heartburn, increased length of sleep and dream activity, nausea, runny nose, **sore throat**, fatigue, and weight gain).

Olanzapine (Zyprexa) was approved in 2003 for use in combination with lithium or valproate for treatment of acute manic episodes associated with bipolar disorder. In 2004 it received additional approval for long-term maintenance of bipolar disorder. Possible side effects include drowsiness, **dizziness**, weight gain, dry mouth, rapid heartbeat, nausea, and muscle weakness.

Quetiapine (Seroquel) was approved by the FDA in 2004 for the treatment of acute mania associated with bipolar disorder. Potential side effects of the drug include dizziness, sleepiness, dry mouth, weight gain, and constipation.

Ziprasidone (Geodon) is a schizophrenia drug that is often prescribed to treat bipolar mania. Common side effects associated with ziprasidone include dizziness, fatigue, constipation, and rash. Unlike the other antipsychotic drugs, however, it does not promote weight gain.

Atypical antipsychotics have been associated with **hyperglycemia** (high blood sugar) and diabetes in some patients. Their use may be contraindicated (i.e., not recommended) in children and teens with type 1 or type 2 diabetes.

Anticonvulsants

Valproate (divalproex sodium, or Depakote; valproic acid, or Depakene) is one of the few drugs available that has been proven effective in treating rapid cycling bipolar and mixed states patients. It is also approved for the treatment of mania. Valproate is prescribed alone or in combination with carbamazepine and/or lithium. Stomach cramps, indigestion, diarrhea, hair loss, appetite loss, nausea, and unusual weight loss or gain are some of the common side effects of valproate. A 2003 study found that the risk of suicide from death is about two and one half times higher in people with bipolar disorder taking divalproex than those taking lithium.

Gabapentin (Neurontin) has been prescribed by some physicians for the treatment of bipolar disorder, although there is no conclusive clinical evidence as to its effectiveness.

Carbamazepine (Tegretol, Atretol) is an anticonvulsant drug usually prescribed in conjunction with other mood stabilizing agents. The drug often is used to treat bipolar patients who have not responded well to lithium therapy. Blurred vision and abnormal eye movement are two possible side effects of carbamazepine therapy. Clinical trials continue as of 2004 in an attempt to obtain FDA approval of carbamazepine for use in bipolar treatment.

Lamotrigine (Lamictal, or LTG), an anticonvulsant medication, is often used in patients with a history of rapid cycling and antidepressant-induced mania. A University of Cincinnati one-year study of the drug in patients with bipolar I disorder found that it provided sustained relief of depressive symptoms. Lamotrigine may be used in conjunction with divalproex (divalproate) and/or lithium. Possible side effects of lamotrigine include skin rash, dizziness, drowsiness, headache, nausea, and **vomiting**.

Antidepressants

Because antidepressants may stimulate manic episodes in some bipolar children and teens, their use is typically short-term. Some researchers have hypothesized that the use of antidepressants for depression may even trigger bipolar disorder in children who are genetically predisposed.

When antidepressants are prescribed for episodes of bipolar depression, they are usually selective serotonin reuptake inhibitors (SSRIs) or, less often, monoamine oxidase inhibitors (MAO inhibitors). Tricyclic antidepressants used to treat unipolar depression may trigger rapid cycling in bipolar patients and are, therefore, not a preferred treatment option for bipolar depression.

SSRIs, such as fluoxetine (Prozac), sertraline (Zoloft), and paroxetine (Paxil) regulate depression by regulating levels of serotonin, a neurotransmitter. **Anxiety**, diarrhea, drowsiness, headache, sweating, nausea, sexual problems, and insomnia are all possible side effects of SSRIs.

MAOIs such as tranylcypromine (Parnate) and phenelzine (Nardil) block the action of monoamine oxidase (MAO), an enzyme in the central nervous system. Patients taking MAOIs must cut foods high in tyramine (found in aged cheeses and meats) out of their diet to avoid hypotensive side effects.

Bupropion (Wellbutrin) is a heterocyclic antidepressant. The exact neurochemical mechanism of the drug is not known, but it has been effective in regulating bipolar depression in some patients. Side effects of bupropion include agitation, anxiety, confusion, tremor, dry mouth, fast or irregular heartbeat, headache, and insomnia.

In 2004, 10 antidepressant drugs (including fluoxetine, sertraline, paroxetine, and bupropion) came under scrutiny when the FDA issued a public health advisory and announced it was requesting the addition of a warning statement in drug labeling that outlined the possibility of worsening depression and increased suicide risk. These developments were the result of several clinical studies that found that some children taking these antidepressants had an increased risk of suicidal thoughts and actions. The FDA announced at the time that the agency would embark on a more extensive analysis of the data from these clinical trials and decide if further regulatory action was necessary.

Electroconvulsive therapy

Electroconvulsive therapy (ECT) has a high success rate for treating both unipolar and bipolar depression and mania. However, because of the convenience of drug treatment and the stigma sometimes attached to ECT therapy, ECT usually is employed after all pharmaceutical treatment options have been explored. ECT is given under anesthesia, and patients are given a muscle relaxant medication to prevent convulsions. The treatment consists of a series of electrical pulses that move into the brain through electrodes on the patient's head. Although the exact mechanisms behind the success of ECT therapy are not known, it is believed that this electrical current alters the electrochemical processes of the brain, consequently relieving depression. Headaches, muscle soreness, nausea, and confusion are possible side effects immediately following an ECT procedure. Temporary memory loss has also been reported in ECT patients. In bipolar patients, ECT is often used in conjunction with drug therapy.

Adjunct therapies

Other drugs that may be use as adjunct therapies (i.e., in addition to regular treatment) to treat manic episodes include the following:

- Calcium channel blockers: Nimodipine (Nimotop, Admon) and verapamil (Calan, Covera, Isoptin), typically used to treat angina and hypotension, have been found effective in a few small studies, for treating rapid cyclers. Calcium channel blockers stop the excess calcium build up in cells that is thought to be a cause of bipolar disorder. They usually are used in conjunction with other drug therapies such as carbamazepine or lithium.
- Long-acting benzodiazepines: Lorazepam (Ativan), clonazepam (Klonapin), and alprazolam (Xanax) are used for rapid treatment of manic symptoms to calm and sedate patients until mania or hypomania have waned and mood stabilizing agents can take effect. Sedation is a common effect, and clumsiness, lightheadedness, and slurred speech are other possible side effects of benzodiazepines.
- Neuroleptics: Chlorpromazine (Thorazine) and haloperidol (Haldol) are also used to control mania while a mood stabilizer such as lithium or valproate takes effect. Because the side effects of these drugs can be severe (difficulty in speaking or swallowing, paralysis of the eyes, loss of balance control, **muscle spasms**, severe restlessness, stiffness of arms and legs, tremors in fingers and hands, twisting movements of body, and weakness of arms and legs), benzodiazepines are generally preferred over neuroleptics.

Because bipolar disorder is thought to be biological in nature, therapy and/or counseling is recommended as a companion to, but not a substitute for, pharmaceutical treatment of the disease. Psychotherapy, such as cognitive-behavioral therapy, can be a useful tool in helping patients and their families adjust to the disorder, in encouraging compliance to a medication regimen, and in reducing the risk of suicide. A 2003 report revealed that people on medication for bipolar disorder had better results if they also participated in family-focused therapy.

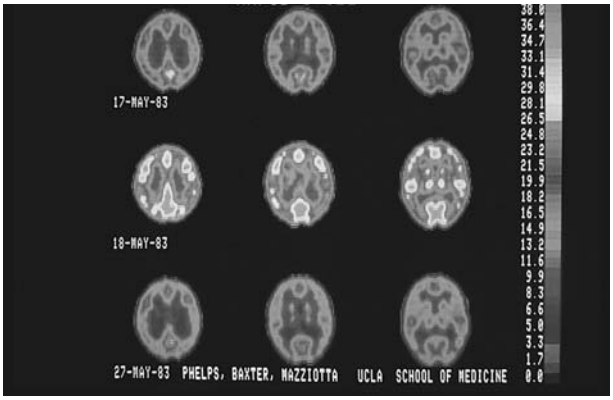
Alternative treatment

General recommendations include maintaining a calm environment, avoiding overstimulation, getting plenty of rest, regular **exercise**, and proper diet. Biofeedback may be effective in helping some children and adolescents control symptoms such as irritability, poor self control, racing thoughts, and sleep problems. A diet low in vanadium (a mineral found in meats and other foods) and high in vitamin C may be helpful in reducing depression.

Repeated transcranial magnetic stimulation (rTMS) is a new and still experimental treatment for the depressive phase of bipolar disorder. In rTMS, a large magnet is placed on the patient's head and magnetic fields of different frequency are generated to stimulate the left front cortex of the brain. Unlike ECT, rTMS requires no anesthesia and does not induce seizures.

Prognosis

While most children show some positive response to treatment, response varies widely, from full recovery to



Bipolar disorder shown in a series of positron emission tomography (PET) scans. *(Dr. Michael E. Phelps.)*

a complete lack of response to all drug and/or ECT therapy. Drug therapies frequently need adjustment to achieve the maximum benefit for the patient. Bipolar disorder is a chronic recurrent illness in over 90 percent of those afflicted, and one that requires lifelong observation and treatment after diagnosis. Patients with untreated or inadequately treated bipolar disorder have a suicide rate of 15 to 25 percent and a nine-year decrease in life expectancy. With proper treatment, the life expectancy of the bipolar patient will increase by nearly seven years and work productivity increases by 10 years.

According to the American Psychiatric Association, bipolar children and adolescents experiencing a manic episode have a one-year recovery rate of 37.1 percent and a relapse rate of 38.3 percent. Discontinuing lithium treatment too early may increase the risk of relapse in adolescents with bipolar disorder. In one 1990 study, 92 percent of adolescents hospitalized for mania who stopped taking the drug experienced a relapse of symptoms within 18 months of discharge, compared to 37 percent of those who stayed on lithium therapy.

Children and teens with bipolar disorder are at a greater risk for substance abuse than their non-bipolar peers, and substance abuse can worsen or complicate bipolar treatment. In a 1999 two-year follow-up study of adolescents hospitalized for manic episodes, patients who had ongoing drug or alcohol abuse problems had more manic episodes and poorer functioning than those patients who were not substance abusers. In addition, some studies have indicated that children who develop bipolar disorder in adolescence are more likely to develop a substance abuse problem than those who have early-onset of bipolar disorder in childhood.

Prevention

The ongoing medical management of bipolar disorder is critical for preventing relapse, or recurrence, of manic episodes. Even in carefully controlled treatment programs, bipolar patients may experience recurring episodes of the disorder. Education in the form of psychotherapy or self-help groups is crucial for training bipolar patients and their caregivers to recognize signs of mania and depression and to take an active part in their treatment program.

Parental concerns

Children with bipolar disorder may require special accommodations in the classroom. Section 504 of the Rehabilitation Act of 1973 enables parents to develop both a Section 504 plan (which describes a child's medical needs) and an individualized education plan (IEP), which describes what special accommodations a child requires to address those needs. The IEP may cover issues such as allowing extra time on tests, modifying assignments, and providing home tutoring or a classroom aide when necessary.

Children who are diagnosed with bipolar disorder should be reassured that the condition is due to factors beyond their control (i.e., genetics, neurochemical imbalance) rather than any fault of their own. For those children and teens who feel stigmatized or self-conscious about their diagnosis, arranging psychotherapy sessions outside school hours may lessen their burden. Any child on prescription medication for bipolar disorder should be carefully monitored for any sign of side effects, and these should be reported to their physician when they do occur. A dosage adjustment or medication change may be warranted if side effects are disruptive or potentially dangerous.

See also Depressive disorders; Minnesota Multiphasic Personality Inventory.

Resources

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KEY TERMS

Affective disorder—An emotional disorder involving abnormal highs and/or lows in mood. Now termed mood disorder.

Anticonvulsant—Drugs used to prevent convulsions or seizures. They often are prescribed in the treatment of epilepsy.

Antipsychotic drug—A class of drugs used to control psychotic symptoms in patients with psychotic disorders such as schizophrenia and delusional disorder. Antipsychotics include risperidone (Risperdal), haloperidol (Haldol), and chlorpromazine (Thorazine).

Benzodiazepine—One of a class of drugs that have a hypnotic and sedative action, used mainly as tranquilizers to control symptoms of anxiety. Diazepam (Valium), alprazolam (Xanax), and chlordiazepoxide (Librium) are all benzodiazepines.

***Diagnostic and Statistical Manual of Mental Disorders, Fourth Edition* (DSM-IV)**—This reference book, published by the American Psychiatric Association, is the diagnostic standard for most mental health professionals in the United States.

Electroconvulsive therapy (ECT)—A psychological treatment in which a series of controlled electrical impulses are delivered to the brain in order to induce a seizure within the brain. This type of therapy is used to treat major depression and severe mental illness that does not respond to medications.

Hyperthyroidism—A condition characterized by abnormal over-functioning of the thyroid glands. Patients are hypermetabolic, lose weight, are nervous, have muscular weakness and fatigue, sweat more, and have increased urination and bowel movements. Also called thyrotoxicosis.

Hypomania—A milder form of mania that is characteristic of bipolar II disorder.

Mania—An elevated or euphoric mood or irritable state that is characteristic of bipolar I disorder. This state is characterized by mental and physical hyperactivity, disorganization of behavior, and inappropriate elevation of mood.

Mixed mania—A mental state in which symptoms of both depression and mania occur simultaneously. Also called mixed state.

Neurotransmitter—A chemical messenger that transmits an impulse from one nerve cell to the next.

Psychomotor retardation—Slowed mental and physical processes characteristic of a bipolar depressive episode.

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Paula Ford-Martin
Teresa Odle

Birth control *see* **Contraception**

Birth control pills *see* **Oral contraceptives**

Birth order

Definition

Birth order is the chronological order of sibling births in a **family**.

Description

Alfred Adler (1870–1937) was a pioneer in the study of birth order. His research suggested that the position a child had by the order of birth significantly affected the child's growth and personality. Research in the late twentieth century and early twenty-first century

shows even greater influence, contributing to **intelligence**, career choice, and, to a certain degree, success in adulthood.

Being born first, last, or somewhere in the middle of itself is not of significance. What matters is how that birth order affects how a child is treated by parents and other siblings and how that child feels about it. Other factors also influence the child's socialization and the parents' expectations.

Birth spacing, gender, physical attributes, and being a twin also affect personality formation and the interpretation of birth order and behavior. These factors influence how parents treat children and how each child is viewed by the other siblings.

Birth spacing changes the dynamics of strict birth order, too. If there is a gap of five or more years between children, each child may be treated as an only child or as a firstborn. If there is a large gap between groups of children in a large family, each group may be treated as a separate birth order family. For example, if child 1, 2, and 3 are three years apart and there is a gap of six years before child 4 is born and child 5 and 6 follow in two year intervals, then child 1, 2, and 3 form a birth order grouping of firstborn, middle, and last, and child 4, 5, and 6 form another grouping of first, middle, and lastborn.

Gender also has a major impact on how a child is treated within the birth order arrangement. The firstborn of either gender, no matter where in the sibling order the child falls, will often be treated as a firstborn. For example if a family has two daughters then has two sons, the first daughter and the first son will be treated as firstborns. The daughter is the true firstborn, but the first son is the first child in the household to be treated with what the family perceives as maleness. Historically, this held true and usually contributed to older sisters not having a claim to inheritance because of their gender.

In addition, if there is only one daughter in a family of three boys, the daughter will often be treated as a first born no matter where in the birth order she is born. The simple fact that she is the only one of her sex allows her to take on the characteristics of a firstborn and be treated as such. This obviously also applies to one son in a household of daughters.

That sense of specialness also applies to children's physical attributes and conditions. If a child of any birth order has a serious medical problem or a physical or mental disability, that child rises either to firstborn status or lastborn status because parental attention is placed on this special child. Robust health and beauty can also skew birth order expectations. For example, if there are two sons and the younger is bigger and more athletic, the younger may be treated as a firstborn because parental favor and expectations are higher for this child. Likewise, if the younger of two daughters is extremely pretty and her older sister is plain, the younger may either be treated as a favored lastborn or as a high-achieving firstborn.

Twins and other birth multiples also skew birth order predictions. Each twin or multiple grouping has its own birth rank. The firstborn twin usually takes on leadership roles for the twin pair. The secondborn usually is more compliant and willing to follow. For the single birth children born after twins or other multiples, birth order is skewed because the twins or multiples have become special children and, in the case of multiples, are their own birth order unit.

Birth order research focuses on five ordinal birth positions: firstborn, secondborn, middle, last, and only children.

Firstborns

In general, firstborn children have been found to be responsible, assertive, task-oriented, perfectionistic, and supporters of authority. Because they often look after their younger siblings, they get experience leading and mentoring others, often rising to leadership positions as adults. Nearly half of all U.S. presidents were firstborns; only four were lastborns. Studies have also linked firstborn children with higher academic achievement and possibly higher intelligence scores when compared to later-born children. This may be due to more exposure to adult language and greater interactions with parents. Firstborns often choose professions that require precision, such as careers in science, medicine, law, engineering, computer science, or accounting.

Firstborns can harbor some resentment toward siblings because parental attention has to be shared. They strive to hang onto parental affection by conforming, either to their parents' wishes, their teachers', or society's. If this does not bring the attention they want, some firstborns defy authority and misbehave or rebel.

Secondborns and middle children

Many secondborns are also middle children. They often report feeling inferior to older children because they do not possess their sibling's advanced abilities. Sometimes, they are very competitive with their firstborn sibling. Others choose to focus their energies in areas different from those in which their older sibling is already established. This competition with firstborns drives secondborns and middleborns to innovation, doing or being different from their older siblings in order to make

themselves stand out in the family dynamic. In truth, they often are more competent at an earlier age than their older siblings because they have had their example to follow.

Middle children can feel forgotten or overlooked because of the attention or demands of either the firstborns or the lastborns. Some of these children never seem to find their place in the social order, and they try to rebel or misbehave in order to draw attention to themselves. Some of these troubled middle children bully younger siblings or children at school.

Other middle children capitalize on the injustice they feel as children and become trial lawyers or social activists because such roles allow them to fight against other social injustices. Some middleborns become very socially skilled because they have learned to negotiate and compromise daily with their siblings and their parents. Some of these children are often called the peacemakers of the household.

Middle children have also been found to succeed in team **sports**, and both they and lastborns have been found to be more socially adjusted if they come from large families.

Lastborns

Lastborns are generally considered to be the family "baby" throughout their lives. Because of nurturing from many older family members and the example of their siblings, lastborns from large families tend to develop strong social and coping skills and may even be able to reach some milestones earlier. As a group, they have been found to be the most successful socially and to have the highest **self-esteem** of all the birth positions.

Youngest children may feel weak and helpless because they compare themselves with older siblings who are able to do more things physically and socially. They may feel that they always have more growing up to do in order to have the privileges they see their older siblings have. Some lastborns develop self-esteem problems if older siblings or parents take power away from these lastborns so that they cannot make decisions or take responsibility. Because of this powerlessness, some lastborns may be grandiose, with big plans that never work out.

Some lastborns transfer this powerlessness into a personal asset by becoming the boss of the family, coyly eliciting or openly demanding their own way. Some families jump to and cater to these lastborns.

Other lastborns engage in **sibling rivalry** because of the injustices they think they experience because they are the youngest. Some ally with firstborns against middleborns.

Only children

Only children may demonstrate characteristics of firstborns and lastborns. Firstborns, after all, are only children until the first sibling is born. Only children grow up relating to adults in the family but have trouble relating to peers. However, this changes as they reach adulthood and get along well with adults.

Only children are achievement-oriented and most likely to attain academic success and attend college. They may also be creative. But only children can be pampered and spoiled as lastborns and can be self-centered. They may rely on service from others rather than their exert their own efforts. They sometimes please others if it suits them but may also be uncooperative. They can also be over-protected.

Some only children become hypercritical, not tolerating mistakes or failure in themselves or others. They can also transform this perfectionist tendency into rescuing behavior, agonizing over the problems of others and rushing to take over and solve everything without letting others help themselves.

Common problems

Sibling rivalry is a normal part of family life. All children become jealous of the love and attention that siblings receive from parents and other adults. When a new baby comes into the family, older children feel betrayed by their parents and may become angry, directing their anger first toward the parents and later toward the intruder who is usurping their position. Jealousy, resentment, and competition are most intense between siblings spaced less than three years apart. Although a certain amount of sibling rivalry is unavoidable, there are measures that parents can take to reduce its severity and its potential effects on their children.

An older child should be prepared for a new addition to the family by having the situation explained and being told in advance about who will take care of her while her mother is in the hospital having the baby. The child's regular routine should be disturbed as little as possible; it is preferable for the child to stay at home and under the care of the father or another close family member. If there is to be a new babysitter or other caretaker unknown to the child, it is helpful for them to meet at least once in advance. If sibling visits are allowed, the child should be taken to visit the mother and new baby in the hospital.

Once the new baby is home, it is normal for an older child to feel hurt and resentful at seeing the attention lavished on the newcomer by parents, other relatives, and family friends. It is not uncommon for the emotional

turmoil of the experience to cause disturbances in eating or sleeping. Some children regress, temporarily losing such attainments as weaning, bowel and bladder control, or clear speech, in an attempt to regain lost parental attention by becoming babies again themselves.

There are a number of ways to ease the unavoidable jealousy of children whose lives have been disrupted by the arrival of a younger sibling. When friends or relatives visit to see the new baby, parents can make the older child feel better by cuddling him or giving him special attention, including a small present to offset the gifts received by the baby. The older child's self-esteem can be bolstered by involving him in the care of the newborn in modest ways, such as helping out when the baby is being diapered or dressed or helping push the stroller. The older child should be made to feel proud of the achievements and responsibilities that go along with his more advanced age—things the new baby cannot do yet because he or she is too young. Another way to make older children feel loved and appreciated is to set aside some quality time to spend alone with each of them on a regular basis. It is also important for parents to avoid overtly comparing their children to each other, and every effort should be made to avoid favoritism.

In general, the most stressful aspect of sibling rivalry is fighting. Physical, as opposed to verbal, fights usually peak before the age of five. It is important for parents not to take sides but rather to help children work out disagreements, calling for a "time out" for feelings to cool down, if necessary. Over-insistence that siblings share can also be harmful. Children need to retain a sense of individuality by developing boundaries with their siblings in terms of possessions, territory, and activities. Furthermore, it is especially difficult for very young children to share their possessions.

Parents should take time to praise cooperation and sharing between siblings as a means of positive reinforcement. The fact that siblings quarrel with each other does not necessarily mean that they will be inconsiderate, hostile, or aggressive in their dealings with others outside the family. The security of family often makes children feel free to express feelings and impulses they are unable to express in other settings.

Parental concerns

Firstborns

Firstborns often feel pressure to succeed or perform well, either by parents or through their own inner drives. They often are called on to take care of younger siblings or do chores because they are responsible. Firstborns also feel pressure to be good examples for their siblings.

Some parents are quick to punish firstborns for not measuring up. Others constantly correct firstborns because they think it will help these children succeed. If firstborns cannot meet these expectations or **fear** that they cannot, they often become depressed and sometimes resort to **suicide** to escape the **pain** they feel.

Parents need to realize that firstborns need not be perfect in order to succeed. They are already eager to please and criticism should be limited to broad strokes rather than focus on minor imperfections. Responsibilities should be meted out in small batches according to their age appropriate abilities. In addition, parents should acknowledge firstborns as people, not the products of their efforts.

When placed in leadership or mentoring roles with their younger siblings, some firstborns may demonstrate aggressive or domineering behavior. They may boss their brothers or sisters around or lord it over them. These behaviors can also transfer to the school setting, making these children uncooperative with their peers. Parents should monitor leadership behavior to make sure these children learn to lead with kindness while respecting other people's feelings.

Secondborns and middle children

Secondborns and middle children often feel invisible. Parents need to make a special effort to seek out their opinions in family discussions. Finding out what special talents or interests these children have and encouraging them through classes or events makes them feel like they matter and are as important as firstborns or lastborns. All of the children in family then feel special and loved as the unique individuals they are.

Lastborns

Youngest children are not usually very responsible because they have not been given the opportunity. Parents can foster responsibility and self- reliance by giving even the youngest child some responsibility, such as setting the table or putting clean clothing in their dresser drawers.

If lastborns are being bullied by older siblings, parents need to step in. Children need help developing strategies for working out difficulties. They can also benefit from hearing parents tell older siblings that it took time for them to do the things that lastborns are struggling to do.

Only children

Parents need to help their only children socialize with other children. They also need to help them accept imperfection in themselves and others by being tolerant of it themselves. In order to keep only children from being

KEY TERMS

Birth multiples—Children born in multiple births; e.g. twins, triplets, quads, etc.

Sibling rivalry—Competition among brothers and sisters in a nuclear family. It is considered to be an important influence in shaping the personalities of children who grow up in middle-class Western societies but less relevant in traditional African and Asian cultures.

rescuers, parents need to help these children develop patience and understanding of differences in others.

See also Sibling rivalry.

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Janie Franz

Birthmarks

Definition

Birthmarks are areas of discolored and raised spots found on the skin. Birthmarks are groups of malformed pigment cells or blood vessels.

Description

Vascular birthmarks are benign (noncancerous) skin growths comprised of rapidly growing or poorly formed blood vessels or lymph vessels. Found at birth (congenital) or developing later in life (acquired) anywhere on the body, they range from faint spots to dark swellings covering wide areas.

Birthmarks are most often found on the head or neck but can be anywhere on the body. The common appearing birthmark is a tiny red or purple mark. A specific group of birthmarks, called "strawberry spot," "port-wine stain," and "stork bite," are medically called hemangiomas. These birthmarks are essentially an overgrowth of blood vessel tissue in a specific area on the body.

Many birthmarks disappear without any special treatment, but some remain the same size or enlarge. In rare cases, the strawberry mark may cover large area of the face and body.

Demographics

About one in every three infants has a birthmark. Twice as many girls as boys have birthmarks. For appearance or cosmetic reasons, medical treatment may be necessary if the birthmark does not disappear on its own. Treatment for most birthmarks is delayed until the child is older.

About 10 in every 100 babies have vascular birthmarks. Skin angiomas, also called vascular nevi (marks), are overgrown blood vessel tissue (hemangiomas) or lymph vessel tissue (lymphangiomas) beneath the skin's surface. Hemangiomas are on the face and neck (60%), trunk (25%), or the arms and legs (15%). Congenital hemangiomas, 90 percent of which appear at birth or within the first month of life, grow quickly and disappear over time. They occur in 1–10 percent of full-term infants, and 25 percent of premature infants. About 65 percent are capillary hemangiomas (strawberry marks), 15 percent are cavernous (deep) hemangiomas, and the rest are mixtures.

Vascular malformations are poorly formed blood or lymph vessels that appear at birth. One type, the salmon patch (nevus simplex), is a pink mark comprised of dilated capillaries (also called a stork bite). It appears on the back of the neck in 40 percent of newborns and on the forehead and eyelids (also called an angel's kiss) in 20 percent. Stork bites appear in 70 percent of white and 60 percent of black newborns.

Fewer than 1 percent of newborns have port-wine stains (nevus flammeus), birthmarks. These vascular malformations of dilated capillaries appear in the upper and lower layers of the skin on the face, neck, arms, and legs.

Nevus flammeus are often permanent; these flat pink to red marks develop into dark purple bumpy areas in later life; 85 percent appear on only one side of the body.

Causes and symptoms

As of 2004 there were no known causes for congenital skin angiomas or birthmarks. Most birthmarks do not hurt; most do not cause any health problems and do not need treatment. Birthmarks may be an inherited weakness of vessel walls.

The birthmark is discoloration of the skin that starts before or just after birth. These marks can appear to be a red rash or lesion. Birthmarks tend to be different color from the skin. They are mostly flat, but some are raised, bumpy, and hairy. Many birthmarks fade or disappear altogether during the **preschool** years, but some never disappear completely.

Diagnosis

Patients are treated by pediatricians, dermatologists (skin disease specialists), plastic surgeons (doctors who specialize in correcting abnormalities of the appearance), and ophthalmologists (eye disease specialists), depending on the type and severity of the birthmark.

Angiomas and vascular malformations are not difficult to diagnose. The doctor takes a medical history and performs a physical examination, including visual inspection and palpation (feeling with the hands) of the marks. The skin is examined for discoloration, scarring, bleeding, infection, or ulceration. The type, location, size, number, and severity of the marks are recorded. The doctor may empty the mark of blood by gentle pressure. Biopsies or specialized **x rays** or scans of the abnormal vessels and their surrounding areas may confirm the diagnosis. Patients with port-wine stains near the eye may need skull x rays, **computed tomography** scans, and vision and central nervous system tests. Most insurance plans pay for diagnosis and treatment of these conditions.

Types of birthmarks

There are many types of birthmarks. Certain types of raised or flat red, pink, or bluish birthmarks need close watching by a qualified medical expert as the child grows. Description of common variations in skin color and birthmarks is as follows:

- Port-wine stains: These flat, pink, red, or purple colored birthmarks are caused by a concentration of dilated tiny blood vessels call capillaries. The stains usually occur on the head, face, and neck. They may be small, or they may cover large areas of the child's body. Port-wine stains do not change color when gently pressed and do not disappear over time. They may become darker and may bleed when the child is older or as an adult. Skin-colored cosmetics will cover small port-wine stains. The most effective way of treating port-wine stains is with a special laser when the child is older.
- Stork bites or salmon patches (called angel kisses when occurring on forehead or eyelids): These small pink or red patches are often found on the baby's eyelids or forehead, between the eyes, on the upper lip, and back of the neck. The name comes from the marks on the back of the neck where, as the myth goes, a stork may have picked up the baby. This concentration of immature blood vessels is most visible when the baby is crying. Most of these fade and disappear.
- Strawberry hemangiomas: These bright or dark red, raised or swollen, bumpy areas look like a strawberry. Hemangiomas are a concentration of tiny, immature blood vessels. Most of these occur on the head. They may not appear at birth but often develop in the first two months. Strawberry hemangiomas are more common in premature babies and in girls. These birthmarks often grow in size for several months (they stop growing around the first birthday), then the birthmarks gradually begin to fade. By age five, the birthmarks fade in half the children affected, and they disappear by age nine.
- Mongolian spots: These blue or purple-colored splotches on the baby's lower back or buttocks occur on over 80 percent of African-American, Asian, and Indian babies. They also occur in dark-skinned babies of all races. The spots, a concentration of pigmented cells, usually disappear in the first four years of life.

Treatment

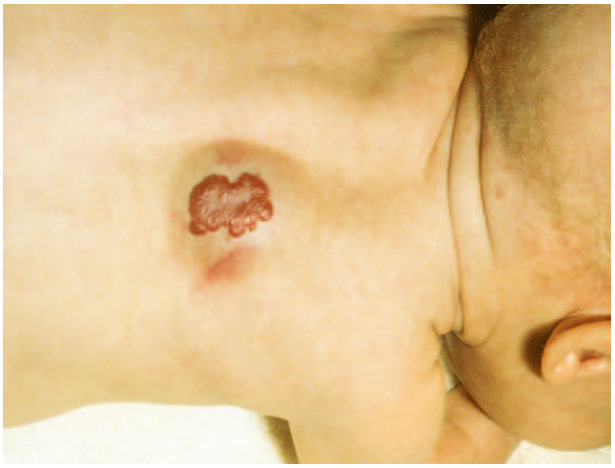
Treatment choices for skin angiomas and vascular malformations depend on their type, location, severity, and degree of disfigurement.

Watchful waiting

Birthmarks are regularly examined until they disappear or require treatment. This approach is appropriate for most hemangiomas, since many eventually shrink by themselves.

Complications

When birthmarks (hemangiomas) form in an area that can interfere with the baby's normal development (for example, blocking vision or causing difficulty breathing or hearing), treatment may be necessary. If the mark begins bleeding, parents should apply pressure firmly to control the bleeding. About 5 percent of



Strawberry nevus, commonly known as a strawberry mark, on the back of an infant. *(© Mike Devlin/Photo Researchers, Inc.)*

hemangiomas become ulcerated, especially if they are in an area that is under pressure or touched often.

Port-wine stains are on the forehead, sides of the face, or eyelids. They are occasionally linked with an increase of blood vessels in the brain or glaucoma. An increase in pressure in the eye can lead to blindness if not treated. Port-wine stains on the legs or arm may be linked to an overgrowth of that extremity.

Other complications, including congestive heart failure from large lesions, Kippel-Trenaunay-Weber syndrome, and Kasabach-Merrit syndrome, are rare.

Drugs

CORTICOSTEROIDS Parents may consider treatment for hemangiomas that do not shrink or fade by the time the child is four years old. The treatments include the use of high doses of steroids (either orally or injected into the lesion) to stop the growth. Steroids prevent the marks from growing; they do not make the birthmarks smaller. Other treatments include interferon alpha, laser therapy, and surgical removal.

INTERFERON ALPHA-2A This drug reduces cell growth in vascular marks that affect vision and that are unresponsive to corticosteroids. Given in daily injections under the skin, the response rate is 50 percent after seven months. Side effects include **fever**, chills, muscle and joint **pain**, vision disorders, low white and red blood cell counts, fatigue, elevated liver enzymes, **nausea**, blood clotting problems, and nerve damage.

ANTIBIOTICS Oral or topical (applied to the skin) **antibiotics** are prescribed for infected marks.

Surgery

Birthmarks may be removed by laser surgery. Lasers create intensive heat that destroys abnormal blood vessels beneath the skin, without damaging normal skin. Laser surgery is not usually painful but can be uncomfortable. Children are usually sedated or anesthetized. Healing occurs within two weeks. Side effects include bruising, skin discoloration, swelling, crusting, and minor bleeding.

In some cases, the birthmark can be surgically excised, or removed. Under local or general anesthesia, the skin is cut and vascular marks or their scars are removed. The cut is repaired with stitches or skin clips.

Cryosurgery is another technique used to remove small birthmarks. Vascular marks can be frozen with a substance that is sprayed onto the skin. **Wounds** heal with minimal scarring.

Birthmarks can also be treated by electrodesiccation. In this procedure, affected vessels are destroyed with the current from an electric needle.

Other treatments

Other treatments include the following:

- Sclerotherapy: Injection of a special solution causes blood clotting and shrinkage with little scarring. Side effects include stinging, swelling, bruising, scarring, muscle cramping, and allergic reactions. This treatment is used most commonly for spider angiomas.
- Embolization: A special material is injected into the vessel blocks blood flow, which helps control blood loss from a bleeding birthmark or reduces the size of inoperable growths. A serious side effect, **stroke**, can occur if a major blood vessel becomes blocked.
- Make-up: Special brands designed to cover birthmarks are sold. Two of these are Covermark and Derma blend.
- Cleaning and compression: Bleeding marks are cleaned with soap and water or hydrogen peroxide, and compressed with a sterile bandage for five to 10 minutes.

Prognosis

Many birthmarks fade or disappear before the child is school age. Some may never go away. Most of them are benign and do not need treatment. Babies with birthmarks are examined and diagnosed by the doctor. Those birthmarks that cause complications in normal childhood growth and development may require medical and surgical treatment.

KEY TERMS

Angioma—A tumor (such as a hemangioma or lymphangioma) that mainly consists of blood vessels or lymphatic vessels.

Benign—In medical usage, benign is the opposite of malignant. It describes an abnormal growth that is stable, treatable, and generally not life-threatening.

Capillaries—The tiniest blood vessels with the smallest diameter. These vessels receive blood from the arterioles and deliver blood to the venules. In the lungs, capillaries are located next to the alveoli so that they can pick up oxygen from inhaled air.

Corticosteroids—A group of hormones produced naturally by the adrenal gland or manufactured synthetically. They are often used to treat inflammation. Examples include cortisone and prednisone.

Cutaneous—Pertaining to the skin

Cutaneous angiolipomas—Benign growths consisting of fat cells and blood vessels just underneath the skin.

Hemangioma—A benign skin tumor composed of abnormal blood vessels.

Hereditary—Something which is inherited, that is passed down from parents to offspring. In biology and medicine, the word pertains to inherited genetic characteristics.

Incidence—The rate of development of a disease in a given population over time.

Kasabach-Merrit syndrome—A combination of rapidly enlarging hemangioma and thrombocytopenia; it is usually clinically evident during early infancy, but occasionally the onset is later. The hemangiomas are large and may increase in size rapidly and may cause severe anemia in infants.

Lymphangioma—A benign skin tumor composed of abnormal lymph vessels.

Lymphatic vessels—Part of the lymphatic system, these vessels connect lymph capillaries with the lymph nodes. They carry lymph, a thin, watery fluid resembling blood plasma and containing white blood cells. Also called lymphatic channels.

Nevus—Any pigmented blemish of the skin present at birth, including moles and various types of birthmarks.

Seizure—A sudden attack, spasm, or convulsion.

Subcutaneous—Referring to the area beneath the skin.

Syndrome—A group of signs and symptoms that collectively characterize a disease or disorder.

Ulcer—A site of damage to the skin or mucous membrane that is characterized by the formation of pus, death of tissue, and is frequently accompanied by an inflammatory reaction.

Vascular malformation—Abnormally formed blood or lymph vessels.

The various types of birthmarks have different prognoses:

- Capillary hemangiomas: Fewer than 10 percent need treatment. Without treatment, 50 percent disappear by age five; 70 percent by age seven; and 90 percent by age nine. No skin changes are found in half, while others have some discoloration, scarring, or wrinkling. From 30 to 90 percent respond to oral corticosteroids, and 45 percent respond to injected corticosteroids; 50 percent respond to interferon Alpha-2a. About 60 percent improve after laser surgery.
- Cavernous hemangiomas: Many do not disappear and are complicated by ulceration or infection. About 75 percent respond to laser surgery but have scarring.
- Spider angiomas: These fade in children but may recur. About 90 percent respond to sclerotherapy, electrodesiccation, or laser therapy.
- Cherry angiomas: These are easily removed by electrodesiccation.
- Lymphangiomas: These marks require surgery.
- Salmon patches on eyelid marks: These marks disappear by six to 12 months of age, and forehead marks fade by age six; however, 50 percent of stork bites on the neck persist into adulthood.
- Port-wine stains (flat birthmarks): These marks are easily covered with make-up. Treatment during infancy or childhood improves results. About 95 percent of the stains respond to FPDL surgery with minimal scarring; 25 percent will disappear, and

70 percent will partially disappear. For unknown reasons, 5 percent show no improvement.

Prevention

Birthmarks are congenital hemangiomas or vascular malformations and cannot be prevented.

Parental concerns

Though no treatment is needed in many of these cases, a child with a hemangiomas should be watched carefully by a doctor skilled in pediatric skin disorders. The hardest part for parents is to wait until the birthmarks begin to fade on their own or, in the case of a birthmark that does not fade, waiting until the child is old enough for surgical management.

When to call the doctor

Parents should report any birthmarks they notice to the child's pediatrician. They should call the pediatrician if they notice bleeding from the birthmark, if a sore develops on the birthmark, if the mark is growing larger.

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Bisexuality *see* **Homosexuality and Bisexuality**

Bites and stings

Definition

Humans can be injured by the bites or stings of many kinds of insects and animals. These range from the bites from a neighbor's dog or cat to bites from fellow humans and spiders to the stings from bees, wasps, snakes, and marine animals such as jellyfish and stingrays.

Description

Mammals

DOGS With more than 60 million pet dogs in the United States, plus thousands of strays, it is not surprising that an estimated 4.7 million Americans a year are bitten by a dog. Although most dog-bite injuries are minor, about 400,000 children seek medical attention for a dog bite every year, according to the Centers for Disease Control and Prevention. Of those injured, about 386,000 require emergency treatment and about a dozen die from their injuries. However, most of the injuries suffered by children seeking treatment in emergency rooms are of low severity and result in treatment and quick release from the hospital.

Children aged five to nine are most likely to be injured by dog bites. Males are more likely than females to require emergency treatment. About two-thirds of injuries to children aged four years or younger are to the head and neck.

Studies also show that most dog bites are from pets or other dogs known to the child who is bitten. In fact, more than half of the bites seen by emergency departments occur at home. Many of the bites result from attempting to break up fights between animals.

CATS Although cats are found in nearly one third of U.S. households, cat bites are far less common than dog bites. According to one study, cats inflict perhaps 400,000 harmful bites in the United States each year. The tissue damage caused by cat bites is usually limited but carries a high risk of infection. Whereas the infection rate for dog bite injuries is 15 to 20 percent, the infection rate for cat bites is 30 to 40 percent. Cat bites are also more likely to be provoked. A typical person who has been bitten is a young girl playing with a pet.

HUMANS Bites from mammals other than dogs and cats are uncommon, with one exception—human bites. There are approximately 70,000 human bites each year in the United States. Because the human mouth contains a multitude of potentially harmful microorganisms, human bites are more infectious than those of most other mammals.

Arthropods

Arthropods are invertebrates belonging to the phylum *Arthropoda*, the jointed-leg, spineless creatures of the world. *Arthropoda* include insects, arachnids

(spiders), crustaceans, and other subgroups. There are more than 700,000 species in all. The list of arthropods that bite or sting humans is extensive and includes lice, bedbugs, fleas, mosquitoes, black flies, ants, chiggers, ticks, centipedes, scorpions, and other species. Spiders, ants, bees, and wasps are the four kinds of arthropod that most often bite people.

SPIDERS In the United States, only two kinds of venomous spider are truly dangerous: black widow spiders and brown recluse (violin or fiddle) spiders. The black widow, which is found in every state but Alaska, prefers dark, dry places, such as barns, garages, and outhouses, and also lives under rocks and logs. Disturbing a female black widow or its web may provoke a bite. Brown recluse spiders also prefer sheltered places, including clothing, and may bite if disturbed.

ANTS, BEES, AND WASPS Ants, bees, and wasps will sting to defend their nests or if they are disturbed. Ants sting more than 9.3 million people each year. Other hymenoptera account for more than 1 million stings annually. Species common to the United States include fire ants, honeybees, bumblebees, yellow jackets, bald-faced hornets, brown hornets, and paper wasps. The Africanized bee species, also called "killer bees," is as of 2004 found in the United States.

More than 50 Americans die each year after being stung by a bee or wasp. Almost all of those deaths are the result of allergic reactions to the sting and not of exposure to the venom itself.

Snakes

There are 20 species of venomous snakes in the United States. These snakes are found in every state except Maine, Alaska, and Hawaii. Each year about 8,000 Americans receive a venomous snakebite, but no more than about 15 die, mostly from rattlesnake bites.

The venomous snakes of the United States are divided into two families: the Crotalidae (pit vipers) and the Elapidae. Pit vipers, named after the small heat-sensing pit that lies between each eye and nostril, are responsible for about 99 percent of the venomous snakebites suffered by Americans. Rattlesnakes, copperheads, and cottonmouths (also called water moccasins) are pit vipers. This family of snakes delivers its venom through two long, hinged fangs in the upper jaw. Some pit vipers carry potent venom that can threaten the brain and spinal cord. The venom of others, such as the copperhead, is less harmful.

The Elapidae family includes two kinds of venomous coral snakes indigenous to the southern and western states. Because coral snakes are creatures that come out only at night, they almost never bite humans; they are held responsible for approximately 25 bites a year in the United States. Coral snakes also have short fangs and a small mouth, which lowers the risk of a bite actually forcing venom into the human body. However, their venom is highly poisonous.

Marine animals

Several varieties of marine animal may bite or sting. Jellyfish and stingrays are two kinds that pose a threat to people who live or vacation in coastal communities.

Causes and symptoms

Mammals

DOGS A typical dog bite results in a laceration, tear, puncture, or crush injury. Bites from large, powerful dogs may even cause **fractures** and dangerous internal injuries. Also, dogs trained to attack may bite repeatedly during a single episode. Infected bites usually cause **pain**, inflammation of the connective tissues, and a pus-filled discharge at the wound site within eight to 24 hours. Most infections are confined to the wound site, but many of the microorganisms in the mouths of dogs can cause systemic and possibly life-threatening infections. Examples are bacteremia and **meningitis**, especially severe in children with health conditions that increase their susceptibility to infection. **Rabies** is rare among pet dogs in the United States, most of which have been vaccinated against the disease. **Tetanus** is also rare but can be transmitted by a dog bite if the victim is not immunized.

CATS The mouths of cats and dogs contain many of the same microorganisms. Cat scratches and bites are also capable of transmitting the *Bartonella henselae* bacterium, which can lead to **cat-scratch disease**, an unpleasant but usually not life-threatening illness.

Cat bites are mostly found on the arms and hands. Sharp cat teeth typically leave behind a deep puncture wound that can reach muscles, tendons, and bones, which are vulnerable to infection because of their comparatively poor blood supply. This is why cat bites are much more likely to become infected than dog bites. Also, people are less inclined to view cat bites as dangerous enough to require immediate attention. The risk that infection has set in by the time a medical professional is consulted is, therefore, greater.

HUMANS Humans bites result from fights, sexual activity, medical and dental treatment, and seizures. Bites also may be sign of **child abuse**. Children often bite other children, but those bites are hardly ever severe. Human bites are capable of transmitting a wide range of dangerous diseases, including **hepatitis B**, syphilis, and **tuberculosis**.

Human bites fall into two categories: occlusional bites and clenched-fist injuries. Occlusional bites result from an actual bite and present a lower risk of infection. The clenched-fist injury happens when a fist hits a mouth during a fight and may lead to an infected hand.

Arthropods

SPIDERS As a rule, children rarely see a black widow bite them, nor do they feel the bite when it happens. The first (and possibly only) evidence that a child has been bitten may be a mild swelling of the injured area and two red puncture marks. Within a short time, however, some victims begin to experience severe **muscle cramps** and rigidity of the abdominal muscles. Other possible symptoms include excessive sweating, **nausea**, **vomiting**, headaches, and vertigo, as well as breathing, vision, and speech problems.

A brown recluse spider's bite can lead to necrotic arachnidism, in which the tissue in an area of up to several inches around the bite becomes necrotic (dies), producing an open sore that can take months or years to disappear. In most cases, however, the bite simply produces a hard, painful, itchy, and discolored area that heals without treatment in two to three days. The bite may also be accompanied by **fever**, chills, edema (an accumulation of excess tissue fluid), **nausea and vomiting**, **dizziness**, muscle and joint pain, and a rash.

BEES, WASPS AND ANTS The familiar symptoms of bee, wasp, and ant stings include pain, redness, swelling, and itchiness in the area of the sting. Multiple stings can have much more severe consequences, such as **anaphylaxis**, a life-threatening allergic reaction that occurs in children who are hypersensitive to the venom.

The fire ant sting usually produces immediate pain followed by a red, swollen area that disappears within 45 minutes. A blister or a red, swollen, itchy patch then develops. The blister may rupture and become infected.

Snakes

Venomous pit viper bites usually begin to swell within ten minutes and sometimes are painful. Other symptoms include skin blisters and discoloration, weakness, sweating, nausea, faintness, dizziness, bruising, and tender lymph nodes. Severe **poisoning** can also lead to **tingling** in the scalp, fingers, and toes, muscle contractions, an elevated heart rate, rapid breathing, large drops in body temperature and blood pressure, vomiting of blood, and coma.

Many pit viper and coral snake bites (20%) fail to poison their victim or introduce only a small amount of venom into the victim's body. The **wounds**, however, can still become infected by the harmful microorganisms that snakes carry in their mouths.

Coral snake bites are painful but may be hard to see. One to seven hours after the bite, the victim begins to experience the effects of the venom, which include tingling at the wound site, weakness, nausea, vomiting, excessive salivation, and irrational behavior. Major nerves of the body can become paralyzed for six to 14 days, causing double vision, difficulty swallowing and speaking, respiratory failure, and other problems. Six to eight weeks may be needed before normal muscular strength is regained.

Marine animals

JELLYFISH Jellyfish venom is delivered by barbs called nematocysts, which are located on the creature's tentacles and penetrate the skin of people who brush up against them. Instantly painful and itchy red lesions usually result. The pain can continue up to 48 hours. Severe cases may lead to skin necrosis, **muscle spasms and cramps**, vomiting, nausea, **diarrhea**, headaches, excessive sweating, and other symptoms. In rare instances, cardiorespiratory (heart/lung) failure may also occur.

STINGRAYS Tail spines are the delivery mechanism for stingray venom. Deep puncture wounds result that can cause an infection if pieces of spine become embedded in the wound. A typical stingray injury scenario involves a person who inadvertently steps on a resting stingray and is lashed in the ankle by its tail. Stingray venom produces immediate, excruciating pain that lasts several hours. Sometimes the victim suffers a severe reaction, including vomiting, diarrhea, hemorrhage (bleeding), a drop in blood pressure, and cardiac arrhythmia (disordered heart beat).

Diagnosis

Mammals

DOGS Gathering information on the circumstances of a dog attack is a crucial part of treatment. Medical professionals need to know:

- when the attack occurred (The chances of infection increase dramatically if the wound has been left untreated for more than eight hours.)

- what led to the attack (Unprovoked attacks are more likely to be associated with rabies.)
- the child's general health, including tetanus immunization history and information about **allergies** to medication and pre-existing health problems that may increase the risk of infection

A physical examination requires careful scrutiny of the wound, with special attention to possible bone, joint, ligament, muscle, tendon, nerve, or blood-vessel damage caused by deep punctures or severe crush injuries. Serious hand injuries should be evaluated by a specialized surgeon. Most of the time, laboratory tests for identifying the microorganisms in bite wounds are performed if infection is present. **X rays** and other diagnostic procedures may also be necessary.

CATS The diagnostic procedures used for dog bites also apply to cat bites.

HUMAN Testing the blood of a person who has been bitten for immunity to hepatitis B and other diseases is always necessary after a human bite. Ideally, the biter should be tested as well for the presence of transmissible disease. Physicians can advise if this is necessary if the biter is another small child. Medical professionals will also look for indications of child abuse when evaluating human bites.

Arthropods

SPIDERS Because bites from widow spiders and brown spiders require different treatment, capturing and identifying the spider helps to establish diagnosis.

Snakes

Diagnosis relies on a physical examination of the victim, information about the circumstances of the bite, and a look at the snake itself (if it can safely be killed and brought in for identification). Blood tests and urinalysis supply important data on the victim's condition. Chest x rays and electrocardiography (a procedure for measuring heart activity) may also be necessary.

Treatment

Mammals

DOGS Minor dog bites can be treated at home. The American Academy of Family Physicians recommends gently washing the wound with soap and water and then applying pressure to the injured area with a clean towel to stop the bleeding. The next step is to apply antibiotic ointment and a sterile bandage to the wound. To reduce swelling and fend off infection, ice should be applied and the injured area kept elevated above the level of the heart. The wound should be cleaned and covered with ointment twice a day until it heals.

Any dog bite that does not stop bleeding after 15 minutes of pressure must be seen by a medical professional. The same is true for bites that are deep or gaping; for bites to the head, hands, or feet; and for bites that may have broken a bone, damaged nerves, or caused a major injury of another kind. Bite victims must also watch for infection. A fever is a sign of infection, as are redness, swelling, warmth, increased tenderness, and pus at the wound site. Children with diabetes or **cancer** who have not had a tetanus shot in five years or who have a medical problem that can increase susceptibility to infection should seek medical treatment no matter how minor the bite appears.

Medical treatment of dog bites involves washing the wound with an anti-infective solution. Removal of dead and damaged tissue (under local, regional, or general anesthetic) may be required after the wound has been washed, and any child whose tetanus shots are not up-to-date should receive a booster injection. Some wounds are left open and allowed to heal on their own, while others require stitches (stitching may be delayed a few days if infection is a concern). Many emergency departments prescribe **antibiotics** for all people with dog bites, but some researchers suggest that antibiotics are usually unnecessary and should be limited to those whose injuries or other health problems make them likely candidates for infection. A follow-up visit after one or two days is generally required for anyone who has received bite treatment.

CATS Because of the high risk of infection, parents of children who are bitten by a cat should always call the child's doctor. Most cat scratches do not require professional medical treatment unless the wound appears infected or the scratched person has a weakened immune system.

Medical treatment for cat bites generally follows the procedures used for dog bites. Experts advise, however, that cat-bite wounds should always be left open to prevent infection. Persons who have been bitten by cats generally receive antibiotics as a preventive measure.

HUMANS Human bites should always be examined by a doctor. Such bites are usually treated with antibiotics and left open because of the high risk of infection. A person who has been bitten may also require

immunization against hepatitis B and other diseases. This is usually not necessary if the biter is a child.

Arthropods

SPIDERS No spider bite should be ignored. The antidote for severe widow spider bites is a substance called antivenin, which contains antibodies taken from the blood serum of horses injected with spider venom. Doctors exercise caution in using antivenin, however, because it can trigger anaphylactic shock, a potentially deadly (though treatable) allergic reaction, and serum sickness, an inflammatory response that can give rise to joint pain, a fever, **rashes**, and other unpleasant, though rarely serious, consequences.

An antivenin for brown spider bites exists as well, but it is not readily available in the United States. The drug dapsone, used to treat leprosy, can sometimes stop the tissue death associated with a brown spider bite. Necrotic areas (areas of dead tissue) may need debridement (removal of dead and damaged tissue) and skin grafts. Pain medications, **antihistamines**, antibiotics, and tetanus shots are a few of the other treatments that are sometimes necessary after a bite from a brown spider or widow spider.

BEES, WASPS, AND ANTS Most stings can be treated at home. A stinger that is stuck in the skin can be scraped off with a blade, fingernail, credit card, or piece of paper (using tweezers may push more venom out of the venom sac and into the wound). The area should be cleaned and covered with an ice pack. Aspirin and other pain medications, oral antihistamines, and calamine lotion are good for treating minor symptoms. Putting meat tenderizer on the wound has no effect.

Persons who have been stung and experience an allergic reaction or who are at risk due to their medical history require immediate medical attention. The danger signs, which usually begin ten minutes after an individual is stung (though possibly not for several hours) include nausea, faintness, chest pain, abdominal cramps, diarrhea, and difficulty swallowing or breathing.

Snakes

Although most snakes are not venomous, any snakebite should immediately be examined at a hospital. While waiting for emergency help to arrive, the victim should wash the wound site with soap and water and then keep the injured area still and at a level lower than the heart. Ice should never be used on the wound site nor should attempts be made to suck out the venom. Making a cut at the wound site is also dangerous. It is important to stay calm and wait for emergency medical aid if it can arrive quickly. Otherwise, the child should proceed directly to a hospital.

When the child arrives at a hospital, the medical staff must determine whether the bite was inflicted by a venomous snake and, if so, whether envenomation (venom is injected into the victim) occurred and how much venom the person has received. Patients may develop low blood pressure, abnormal blood clotting, or severe pain, all of which require aggressive treatment. Fortunately, the effects of some snake bites can be counteracted with antivenin. Minor rattlesnake envenomations can be successfully treated without antivenin, as can copperhead and watermoccasin bites. However, coral snake envenomations and the more dangerous rattlesnake envenomations require antivenin, sometimes in large amounts. Other treatment measures include antibiotics to prevent infection and a tetanus booster injection.

Marine animals

JELLYFISH Vinegar and other acidic substances are used to neutralize jellyfish nematocysts still clinging to the skin, which are then scraped off. Anesthetic ointments, antihistamine creams, and steroid lotions applied to the skin are sometimes beneficial. Other measures may be necessary to counter the many harmful effects of jellyfish stings, which, if severe, require emergency medical care.

STINGRAYS Stingray wounds should be washed with saltwater and then soaked in very hot water for 30 to 90 minutes to neutralize the venom. Afterwards, the wound should be examined by a doctor to ensure that no pieces of spine remain.

Prognosis

Mammals

Prompt treatment and recognizing that even apparently minor bites can have serious consequences are the keys to a good outcome after a mammal bite. Infected bites can be fatal if neglected. Surgery and **hospitalization** may be needed for severe bites.

Arthropods

SPIDERS Even without treatment, most children recover from black widow bites after two to three days. In the case of brown spider bites, the risk of death is greatest for children, though rare.

BEES, WASPS, AND ANTS The pain and other symptoms of a bee or wasp sting normally fade away after a

few hours. Children who are allergic to such stings, however, can experience severe and occasionally fatal anaphylaxis (life-threatening allergic reaction to bites and stings).

Snakes

A snakebite victim's chances of survival are excellent if medical aid is obtained in time. Some bites, however, can result in amputation, permanent deformity, or loss of function in the injured area, although this is rare.

Marine animals

STINGRAYS Stingray venom kills its human victims on rare occasions.

Prevention

Mammals

DOGS The risk of a dog-bite injury can be reduced by avoiding sick or stray dogs, staying away from dogfights (children can get bitten when they try to separate the animals), and not behaving in ways that might provoke or upset dogs, such as wrestling with them or bothering them while they are sleeping, eating, or looking after their puppies. Infants and young children must never be left alone with a dog. Pit bulls, rottweilers, and German shepherds (responsible for nearly half of all fatal dog attacks in the United States in 2000) are potentially dangerous pets in households where children live or visit. For all breeds of dog, obedience training as well as spaying or neutering lessen the chances of **aggressive behavior**.

CATS Prevention involves warning children to stay away from strange cats and to avoid rough **play** and other behavior that can anger cats and cause them to bite.

Arthropods

SPIDERS Common-sense precautions include clearing webs out of garages, out buildings, and other places favored by venomous spiders; teaching children to keep their hands away from places where spiders may be lurking; and, when camping or vacationing, checking clothing, shoes, and sleeping areas.

BEES, WASPS, AND ANTS Children should avoid the nests of bees, wasps, and ants. When playing outside in an area where these insects are found, children also should avoid eating sweet food or wearing bright clothing, perfumes, or cosmetics that attract bees, wasps, and ants.

Emergency medical kits containing self-administrable epinephrine to counter anaphylactic shock are available for allergic children and should be carried by them at all times. Children who are suspected of being allergic should consult an allergist about shots that can reduce reactions to bee and wasp venom (venom immunotherapy). Venom immunotherapy in children leads to a significantly lower risk of reaction to stings up to ten to 20 years after treatment is stopped.

Mosquito bite behind the ear of a girl. *(© Deb Yeske/Visuals Unlimited.)*

Snakes

Snakes should not be kept as pets. Measures such as mowing the lawn, keeping hedges trimmed, and removing brush from the yard also discourages snakes from living close to human dwellings. Tongs should be used to move brush, lumber, and firewood, to avoid exposing one's hands to snakes that might be lying underneath. Children should be prevented from playing in weedy, vacant lots and other places where snakes may live. Leather boots and long pants offer hikers and campers some protection from bites. Approaching a snake, even a dead one, can be dangerous since the venom of recently killed snakes may still be active.

Marine animals

JELLYFISH Prevention of jellyfish stings includes obeying posted warning signs at the beach. Also, jellyfish tentacles may be transparent and up to 120 feet (36.5 m) long; therefore, great caution must be exercised whenever a jellyfish is sighted nearby.

STINGRAYS Kicking the sand while walking through shallow areas that may be inhabited by stingrays will disturb the water, causing the animal to move before it can be stepped on.

KEY TERMS

Anaphylaxis—Also called anaphylactic shock; a severe allergic reaction characterized by airway constriction, tissue swelling, and lowered blood pressure.

Antibiotics—Drugs that are designed to kill or inhibit the growth of the bacteria that cause infections.

Antibody—A special protein made by the body's immune system as a defense against foreign material (bacteria, viruses, etc.) that enters the body. It is uniquely designed to attack and neutralize the specific antigen that triggered the immune response.

Antihistamine—A drug used to treat allergic conditions that blocks the effects of histamine, a substance in the body that causes itching, vascular changes, and mucus secretion when released by cells.

Arachnid—A large class of arthropods that includes spiders, scorpions, mites, and ticks.

Arachnidism—Poisoning resulting from the bite or sting of an arachnid.

Bacteremia—Bacterial infection of the blood.

Blood serum—A component of blood.

Immune system—The system of specialized organs, lymph nodes, and blood cells throughout the body that work together to defend the body against foreign invaders (bacteria, viruses, fungi, etc.).

Killer bees—Hybrids of African bees accidentally introduced into the wild in South and North America in 1956 and first reported in Texas in 1990. They were first imported by Brazilian scientists attempting to create a new hybrid bee to improve honey production.

Lymph nodes—Small, bean-shaped collections of tissue located throughout the lymphatic system. They produce cells and proteins that fight infection and filter lymph. Nodes are sometimes called lymph glands.

Pus—A thick, yellowish or greenish fluid composed of the remains of dead white blood cells, pathogens, and decomposed cellular debris. It is most often associated with bacterial infection.

Parental concerns

Children frequently play in areas where they can be exposed to stings and bites. Children who are sensitive to certain stings and bites are at risk for serious anaphylactic reactions.

See also Insect sting allergy.

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American Academy of Emergency Medicine. 611 East Wells Street, Milwaukee, WI 53202. Web site: <www.aaem.org>.

American Academy of Family Physicians. 11400 Tomahawk Creek Parkway, Leawood, KS 66211–2672. Web site: <www.aafp.org>.

American Medical Association. 515 N. State Street, Chicago, IL 60610. Web site: <www.ama-assn.org>.

Christine Kuehn Kelly

Biting *see* **Aggressive behavior**

Bladder anomalies *see* **Congenital bladder anomalies**

Bleeding disorders *see* **Coagulation disorders**

Blended family *see* **Stepfamilies**

Blood sugar tests

Definition

Blood sugar or plasma glucose tests are used to determine the concentration of glucose in blood. These tests are used to detect an increased blood glucose (**hyperglycemia**) or a decreased blood glucose (**hypoglycemia**).

Purpose

Blood glucose tests are used in a variety of situations, including the following:

- Screening persons for **diabetes mellitus**: The American Diabetes Association (ADA) recommends that a fasting plasma glucose (fasting blood sugar) be used to diagnose diabetes. If the person already has symptoms of diabetes, a blood glucose test without fasting, called a casual plasma glucose test, may be performed. In difficult diagnostic cases, a glucose challenge test called a two-hour oral glucose tolerance test is recommended. If the result of any of these three tests is abnormal, it must be confirmed with a second test performed on another day. The same test or a different test can be used, but the result of the second test must be abnormal as well in order to establish a diagnosis of diabetes.
- Blood glucose monitoring: Daily measurement of whole blood glucose identifies diabetics who require intervention to maintain their blood glucose within an acceptable range as determined by their physician. The Diabetes Control and Complications Trial (DCCT) demonstrated that persons with diabetes who maintained blood glucose and glycated hemoglobin (hemoglobin with glucose bound to it) at or near normal decreased their risk of complications by 50 to 75 percent. Based on results of this study, the American Diabetes Association (ADA) recommends routine glycated hemoglobin testing to measure long-term control of blood sugar.
- Diagnosis and differentiation of hypoglycemia: Low blood glucose may be associated with symptoms such as confusion, memory loss, and seizure. Demonstration that such symptoms are the result of hypoglycemia requires evidence of low blood glucose at the time of symptoms and reversal of the symptoms by glucose. In documented hypoglycemia, blood glucose tests are used along with measurements of insulin and C-peptide (a fragment of proinsulin) to differentiate between fasting and postprandial (after a meal) causes.

Description

The body uses glucose to produce the majority of the energy it needs to function. Glucose is absorbed from the gastrointestinal tract directly and is also derived from digestion of other dietary carbohydrates. It is also produced inside cells by the processes of glycogen breakdown (glycogenolysis) and reverse glycolysis (gluconeogenesis). Insulin is made by the pancreas and facilitates the movement of glucose from the blood and extracellular fluids into the cells. Insulin also promotes cellular production of lipids and glycogen and opposes the action of glucagons, which increases the formation of glucose by cells.

Diabetes may result from a lack of insulin or a subnormal response to insulin. There are three forms of diabetes: Type I or insulin dependent (IDDM), type II or noninsulin dependent (NIDDM), and gestational diabetes (GDM). Type I diabetes usually occurs in childhood and is associated with low or absent blood insulin and production of ketones even in the absence of stressed metabolic conditions. It is caused by autoantibodies to the islet cells in the pancreas that produce insulin, and persons must be given insulin to control blood glucose and prevent ketosis. Type II accounts for 85 percent or more of persons with diabetes. It usually occurs after age 40 and is usually associated with **obesity**. Persons who have a deficiency of insulin may require insulin to maintain glucose, but those who have a poor response to insulin may not. Ketosis does not develop under normal metabolic conditions but may occur with stress. Gestational diabetes is a form of glucose intolerance that first appears during pregnancy. It usually ends after delivery, but over a 10-year span approximately 30 to 40 percent of females with gestational diabetes go on to develop noninsulin dependent diabetes.

There are a variety of ways to measure a person's blood glucose.

Whole blood glucose tests

Whole blood glucose testing can be performed by a person in his or her home, or by a member of the healthcare team outside the laboratory. The test is usually performed using a drop of whole blood obtained by finger puncture. Care must be taken to wipe away the first drop of blood because this is diluted with tissue fluid. The second drop is applied to the dry reagent test strip or device.

Fasting plasma glucose test

The fasting plasma glucose test requires an eight-hour fast. The person must have nothing to eat or drink except water. The person's blood is usually collected by a nurse or phlebotomist by sticking a needle into a vein. Either serum, the liquid portion of the blood after it clots, or plasma may be used. Plasma is the liquid portion of unclotted blood that is collected. The ADA recommends a

normal range for fasting plasma glucose of 55–109 mg/dL. A glucose level equal to greater than 126 mg/dL is indicative of diabetes. A fasting plasma glucose level of 110–125 gm/dL is referred to as "impaired fasting glucose."

Oral glucose tolerance test (OGTT)

The oral glucose tolerance test is done to see how well the body handles a standard amount of glucose. There are many variations of this test. A two-hour OGTT as recommended by the ADA is described below. The person must have at least 150 grams of carbohydrate each day, for at least three days before this test. The person must take nothing but water and abstain from **exercise** for 12 hours before the glucose is given. At 12 hours after the start of the fast, the person is given 75 grams of glucose to ingest in the form of a drink or standardized jelly beans. A healthcare provider draws a sample of venous blood two hours following the dose of glucose. The serum or plasma glucose is measured. A glucose concentration equal to or greater than 200 mg/dL is indicative of diabetes. A level below 140 mg/dL is considered normal. A level of 140–199 mg/dL is termed "impaired glucose tolerance."

The glycated (glycosylated) hemoglobin test is used to monitor the effectiveness of diabetes treatment. Glycated hemoglobin is a test that indicates how much glucose was in a person's blood during a two- to three-month window beginning about four weeks prior to sampling. The test is a measure of the time-averaged blood glucose over the 120-day life span of the red blood cells. The normal range for glycated hemoglobin measured as HbA_{1c} is 3 to 6 percent. Values above 8 percent indicate that a hyperglycemic episode occurred sometime during the window monitored by the test (two to three months beginning four weeks prior to the time of blood collection).

The ADA recommends that glycated hemoglobin testing be performed during a person's first diabetes evaluation, again after treatment is begun and glucose levels are stabilized, then repeated semiannually. If the person does not meet treatment goals, the test should be repeated quarterly.

A related blood test, fructosamine assay, measures the amount of albumin in the plasma that is bound to glucose. Albumin has a shorter half-life than red blood cells, and this test reflects the time-averaged blood glucose over a period of two to three weeks prior to sample collection.

Precautions

Diabetes must be diagnosed as early as possible. If left untreated, it results in progressive vascular disease that may damage the blood vessels, nerves, kidneys, heart, and other organs. Brain damage can occur from glucose levels below 40 mg/dL and coma from levels above 450 mg/dL. For this reason, plasma glucose levels below 40 mg/dL or above 450 mg/dL are commonly used as alert values. Point-of-care and home glucose monitors measure glucose in whole blood rather than plasma and are accurate generally within a range of glucose concentration between 40 and 450 mg/dL. In addition, whole blood glucose measurements are generally 10 percent lower than serum or plasma glucose.

Other endocrine disorders and several medications can cause both hyperglycemia and hypoglycemia. For this reason, abnormal glucose test results must be interpreted by a physician.

Glucose is a labile (affected by heat) substance; therefore, plasma or serum must be separated from the blood cells and refrigerated as soon as possible. Splenectomy can result in an increase and hemolytic anemia can result in a decrease in glycated hemoglobin.

Exercise, diet, anorexia, and **smoking** affect the results of the oral glucose tolerance test. Drugs that decrease tolerance to glucose and affect the test include steroids, **oral contraceptives**, estrogens, and thiazide diuretics.

Preparation

Blood glucose tests require either whole blood, serum, or plasma collected by vein puncture or finger puncture. No special preparation is required for a casual blood glucose test. An eight-hour fast is required for the fasting plasma or whole-blood glucose test. A 12-hour fast is required for the two-hour OGTT and three-hour OGTT tests. In addition, the person must abstain from exercise in the 12-hour fasting period. Medications known to affect carbohydrate metabolism should be discontinued three days prior to an OGTT test if possible, and the person must maintain a diet of at least 150 grams of carbohydrate per day for at least three days prior to the fast.

Aftercare

After the test or series of tests is completed (and with the approval of his or her doctor), the person should eat, drink, and take any medications that were stopped for the test.

The patient may feel discomfort when blood is drawn from a vein. Bruising may occur at the puncture site, or the person may feel dizzy or faint. Pressure should be applied to the puncture site until the bleeding stops to reduce bruising. Warm packs can also be placed over the puncture site to relieve discomfort.

Diabetic children learning how to monitor glucose levels with a blood test. *(© Roger Ressmeyer/Corbis.)*

Risks

The patient may experience weakness, fainting, sweating, or other reactions while fasting or during the test. If this occurs, he or she should immediately inform the physician or nurse.

Normal results

Normal values listed below are for children. Results may vary slightly from one laboratory to another depending upon the method of analysis used.

- fasting plasma glucose test: 55–109 mg/dL
- oral glucose tolerance test at two hours: less than 140 mg/dL
- glycated hemoglobin: 3–6 percent
- fructosamine: 1.6–2.7 mmol/L for adults (5% lower for children)
- gestational diabetes screening test: less than 140 mg/dL
- cerebrospinal glucose: 40–80 mg/dL
- serous fluid glucose: equal to plasma glucose
- synovial fluid glucose: within 10 mg/dL of the plasma glucose
- urine glucose (random semiquantitative): negative

For the diabetic person, the ADA recommends an ongoing blood glucose goal of less than or equal to 120 mg/dL.

The following results are suggestive of diabetes mellitus and must be confirmed with repeat testing:

- fasting plasma glucose test: greater than or equal to 126 mg/dL
- oral glucose tolerance test at two hours: equal to or greater than 200 mg/dL
- casual plasma glucose test (nonfasting, with symptoms): equal to or greater than 200 mg/dL

Parental concerns

The needle used to withdraw the blood only causes **pain** for a moment. If a child needs to take glucose tests

KEY TERMS

Diabetes mellitus—The clinical name for common diabetes. It is a chronic disease characterized by the inability of the body to produce or respond properly to insulin, a hormone required by the body to convert glucose to energy.

Glucose—A simple sugar that serves as the body's main source of energy.

Glycated hemoglobin—A test that measures the amount of hemoglobin bound to glucose. It is a measure of how much glucose has been in the blood during a two to three month period beginning approximately one month prior to sample collection.

regularly at home, the parent will need to keep track of the testing schedule and the results.

When to call a doctor

If the needle puncture site continues to bleed, or if hours or days later the site looks infected (red and swollen), then a doctor should be contacted.

See also Diabetes.

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Mark A. Best

Body piercing *see* **Piercing and tattoos**

Bonding

Definition

Bonding is the formation of a mutual emotional and psychological closeness between parents (or primary caregivers) and their newborn child. Babies usually bond with their parents in the minutes, hours, or days following birth.

Description

Bonding is essential for survival. The biological capacity to bond and form attachments is genetically determined. The drive to survive is basic in all species. Infants are defenseless and must depend on a caring adult for survival. The baby's primary dependence and the maternal response to this dependence causes bonding to develop.

Bonding and attachment are terms that describe the affectional relationships between parents and the infants. An increased awareness of the importance of bonding has led to significant improvements in routine birthing procedures and postpartum parent-infant contact. Bonding begins rapidly, shortly after birth, and reflects the feelings of parents toward the newborn; attachment involves reciprocal feelings between parent and infant and develops gradually over the first year. The focus of this entry is bonding in the newborn period. Attachment develops over the larger period of infancy and is treated in a separate entry.

Many parents, mothers in particular, begin bonding with their child before birth. The physical dependency the fetus has with the mother creates a basis for emotional and psychological bonding after birth. This attachment provides the foundation that allows babies to thrive in the world. When the umbilical cord is cut at birth,

physical attachment to the mother ceases, and emotional and psychological bonding begins. A firm bond between mother and child affects all later development, and it influences how well children will react to new experiences, situations, and stresses.

Bonding research

American pediatricians John Kennell and Marshall Klaus pioneered scientific research on bonding in the 1970s. Working with infants in a neonatal intensive care unit, they noted that infants were taken away from their mothers immediately after birth for emergency medical procedures. These babies remained in the nursery for several weeks before being allowed to go home with their families. Although the babies did well in the hospital, a troubling percentage of them seemed not to prosper at home and were even victims of battering and abuse. Kennell and Klaus also noted the mothers of these babies were often uncomfortable with them, sometimes not believing that their babies had survived birth. Even mothers who had successfully raised previous infants have special difficulties when their children had been in the intensive care nursery. Kennell and Klaus surmised the separation immediately after birth interrupted a fundamental relationship between the mother and the new baby. They experimented with giving mothers of both premature and healthy full-term babies extra contact with their infants immediately after birth and in the few days following birth. Mothers with more access to their babies in the hospital developed better rapport with their infants, held them more comfortably, and smiled at and talked to them more often.

Gradually bonding research brought about widespread changes in hospital obstetrical practice in the United States. Fathers and **family** members often remain with the mother during labor and delivery. Mothers hold their infants immediately after birth, and babies often remain with their mothers throughout their hospital stay. Bonding research has also led to increased awareness of the natural capabilities of the infant at birth, and so it has encouraged many others to deliver their babies without anesthesia (which depresses mother and infant responsiveness).

Infancy

Emotionally and physically healthy mothers and fathers are attracted to their infant. They naturally feel a physical longing to smell, cuddle, and rock their infant. They look at their baby and communicate to the baby. In turn the infant responds with snuggling, babbling, smiling, sucking, and clinging. Usually, the parents' behaviors bring pleasure and nourishment to the infant, and the infant's behaviors bring pleasures and satisfaction to the parents. This reciprocal positive maternal and paternal-infant interaction initiates attachment.

One important part in the parents' ability to bond with the infant after birth is the healthy, drug-free newborn is in a "quiet alert" state for 45 to 60 minutes after birth. Immediately after birth the newborn can see, can hear, will turn his head toward a spoken voice, and will move in rhythm to his mother's voice. Mothers and fathers who have the opportunity to interact with their newborns within an hour after birth bond with their baby quickly. The act of holding, rocking, laughing, singing, feeding, gazing, kissing, and other nurturing behaviors involved in caring for infants (and young children) are bonding experiences. The most important ways to create attachment is positive physical contact such as hugging, holding, and rocking. It should be no surprise that nurturing behaviors cause specific neurochemical actions in the brain. These actions lead to organization of brain systems responsible for attachment.

Physical changes occur in the mother after birth, such as hormonal increases triggered by the infant licking or sucking her nipples and increased blood flow to her breasts when she hears the infant cry. Instinctive behaviors triggered in the mother in response to the infant immediately after birth promote her bonding with the infant and thus support the infant's survival.

Toddlerhood

Bonding experiences lead to healthy relations for children in the earliest years of life. During the first three years of life, the human brain develops to 90 percent of adult size. The brain puts in place most of the systems and structures that are responsible for future emotional, behavioral, social, and physiological functioning. Bonding experiences must be present at certain critical times for the brain parts responsible for attachment to develop normally. These critical periods appear in the first year of life and are related to the capacity of the infant and parent or caregiver to develop a positive interactive relation. Problems with bonding and attachment can lead to a fragile biological and emotional foundation for later experiences.

Common problems

Any problem with bonding experiences can interfere with attachment capacities. When the interactive, reciprocal "dance" between the parent and infant is disrupted or becomes difficult, bonding experiences are difficult to maintain. Disruptions can occur because of medical problems with the infant or the parent, the environment, or the fit between the infant and the parent.

The infant's personality or **temperament** influences bonding. If an infant is difficult to comfort, is irritable, or unresponsive, the baby may have more difficulty developing a secure bond. Moreover, the infant's ability to take part in the maternal-infant interaction may be compromised because of a medical condition, such as **prematurity**, birth defect, or illness.

The parent's or caregiver's behavior can also hinder bonding. Critical, rejecting, and interfering parents have children who may avoid emotional intimacy. Abusive parents have children who become uncomfortable with intimacy and withdraw. The child's mother may be unresponsive to the child because of maternal depression, substance abuse, or overwhelming personal problems that interfere with her ability to be consistent and nurturing for the child.

The environment is also a factor. A major impediment to healthy bonding is **fear**. If an infant is distressed because of **pain**, pervasive threat, or a chaotic environment, the baby may have a difficult time engaging in a sympathetic care-giving relationship. Infants or children living amid domestic violence, in refugee shelters, in areas besieged by community violence, or in war zones are at risk for developing attachment problems.

The fit between the infant's temperament and capabilities and those of the mother and father is important. Some parents can bond with a calm infant but are overwhelmed by an irritable infant. Understanding each other's nonverbal cues and responding appropriately is essential to preserving the bonding experiences that build healthy attachments. Sometimes a style of communication and response familiar to a mother from one of her other children may not fit her new infant. The mutual frustration of being "out of sync" can undermine bonding.

Since the first phase of bonding takes place in the womb, researchers believe difficult and unwanted pregnancies and planned adoptions interfere with mother and infant bonding. Teenagers and immature mothers often conceal and reject their pregnancies. This behavior and feeling may result in **abandonment**, neglect, and the absence of bonding at birth. Often there is also an emotional detachment from a fetus that causes emotional or physical pain to the mother during pregnancy. Mothers may have difficulty bonding with an infant if prenatal testing suggests the child will have a birth defect or is likely to be mentally retarded and malformed. And babies planned for **adoption** at birth may be "given up" emotionally by the birth mother during pregnancy. Any or all of these circumstances can interfere with the infant-parent bonding process.

Mothers who develop secure bonds with their infants tend to spend time holding them and making eye contact. *(Photograph by Goujon/Jerrican. Photo Researchers, Inc.)*

Parental concerns

The birth of a premature infant is documented to be a time of stress and crisis for parents and infants. Among these stressors are perceived losses and grief from the early abrupt termination of pregnancy, feelings of guilt and failure in inability to carry the infant to term, uncertainty regarding the infant's future health and developmental potential, and immediate and long-term separation of the infant from the family.

Parental involvement in the care of sick or premature newborns is a major concern of many pediatricians and nursery staff. Touching, stroking, and talking, and, later, massaging are encouraged during frequent parental visits to the nursery. It is hoped that the emotional bonding of parents with low birth-weight infants will increase the baby's chance of doing well despite prematurity.

KEY TERMS

Attachment—A bond between an infant and a caregiver, usually its mother. Attachment is generally formed within the context of a family, providing the child with the necessary feelings of safety and nurturing at a time when the infant is growing and developing. This relationship between the infant and his caregiver serves as a model for all future relationships.

Because premature infants are sometimes seem fragile, parents may handle them less. Skin to skin contact is important to the growing infant, premature or full term. Lack of this contact may predispose the child to psychological problems as well as diminish opportunities for learning.

The practice of "kangaroo care," first introduced by two South American neonatologists, is a method of skin-to-skin contact to promote parent and infant bonding especially for premature infants. This method involves holding infants dressed only in a diaper and a hat between the mother's bare breasts or against the father's chest, similar to a kangaroo carrying their young. Through contact with their parents' skin, the babies are kept warm and allowed close interaction with their parents. This decreases some of the stressors associated with premature births and helps infants needing neonatal intensive care.

Parents who have experienced kangaroo care have expressed excitement and joy with the practice and many have felt like parents for the first time since their infant's birth. Infants have been observed in a restful **sleep** state while in the kangaroo position. As well, kangaroo care has been found to promote parent and infant bonding, breastfeeding, and early discharge for premature infants.

Kangaroo care is offered to stable babies who are less than 1,500 grams and are breathing on their own. Babies needing oxygen or nasal continuous positive airway pressure (CPAP) may also be eligible. Cardiopulmonary monitoring and oximetry may be continued during kangaroo care. The nurse remains nearby to monitor the infant as necessary.

See also Attachment between infant and caregiver.

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Bone cancer *see* **Sarcomas**

Bordetella pertussis infection *see* **Whooping cough**

Botulism

Definition

Botulism is an acute, progressive condition caused by botulinum toxin, a natural poison produced by the spore-forming bacteria *Clostridium botulinum*. Exposure to the botulinum toxin usually occurs from eating contaminated food although, in infants, it may be caused by specific types of clostridia obtained from soil or inhaled spores, causing growth of the bacteria in the infant's intestine. Botulinum toxin is a neurotoxin that blocks the ability of motor nerves to release acetylcholine, the neurotransmitter that relays nerve signals to muscles, a process that may result in unresponsive muscles, a condition known as flaccid paralysis. Breathing may be severely compromised in progressive botulism because of failure of the muscles that control the airway and breathing.

Description

Botulism occurs only rarely, but its high fatality rate makes it a great concern for those in the general public and in the medical community. Clinical descriptions of botulism reach as far back in history as ancient Rome and Greece. However, the relationship between contaminated food and botulism was not defined until the late 1700s. In 1793 the German physician, Justinius Kerner (1786–1862), deduced that a substance in spoiled sausages, which he called *wurstgift* (German for sausage poison), caused botulism. The toxin's origin and identity remained vague until Emile van Ermengem (1851–1932), a Belgian professor, isolated *Clostridium botulinum* in 1895 and identified it as the source of **food poisoning**.

Three types of botulism have been identified: food-borne, wound, and infant botulism. The main difference between types hinges on the route of exposure to the toxin. Food-borne botulism accounts for 25 percent of all botulism cases and can usually be traced to eating contaminated home-preserved food. Infant botulism accounts for 72 percent of all cases. About 98 percent of infants recover with proper treatment. Although domestic food poisoning is a problem worldwide, concern is growing regarding the use of botulism toxin in biological warfare. At the end of the twentieth century 17 countries were known to be developing biological weapons, including the culture of botulism toxins.

Transmission

Botulism is not spread from one individual to another, but through exposure to the deadly botulinum toxin, a natural poison produced by certain *Clostridium* bacteria that may be found in preserved, especially canned, foods and sometimes in the intestines of infants. Botulism spores can cause widespread illness if introduced into the environment.

Demographics

Botulism occurs worldwide, with 90 percent of the comparatively rare cases occurring in the United States. Approximately 110 cases of botulism are reported annually in the United States, with 50 percent of cases in California alone. Infant botulism accounts for 72 percent of all cases, far exceeding both food-borne and wound botulism. Food-borne botulism accounts for 25 percent of all cases, primarily due to eating contaminated home-preserved food.

Causes and symptoms

Toxins produced by the bacterium *Clostridium botulinum* are the main culprit in botulism. Other members of the *Clostridium* genus can produce botulinum toxin, namely *C. argentinense*, *C. butyricum*, and *C. baratii*, but these are minor sources. To grow, these bacteria require a low-acid, oxygen-free environment that is warm (40–120°F or 4.4–48.8°C) and moist. Lacking these conditions, the bacteria transform themselves into spores that, like plant seeds, can remain dormant for years. Clostridia and their spores exist all over the world, especially in soil and aquatic sediments. They do not threaten human or animal health until the spores encounter an environment that favors growth. The spores then germinate, and the growing bacteria produce the deadly botulism toxin.

Scientists have discovered that clostridia can produce at least seven types of botulism toxin, identified as A, B, C, D, E, F, and G. Humans are usually affected by A, B, E, and very rarely F; infants are affected by types A and B. Domesticated animals such as dogs, cattle, and mink are affected by botulism C toxin, which also affects birds and has caused massive die-offs in domestic bird flocks and wild waterfowl. Botulism D toxin can cause illness in cattle, and horses succumb to botulism A, B, and C toxin. There have been no confirmed cases of human or animal botulism linked to the G toxin.

In humans, botulinum toxin latches onto specific proteins in nerve endings and irreversibly destroys them. These proteins control the release of acetylcholine, a neurotransmitter that stimulates muscle cells. With acetylcholine release blocked, nerves are not able to stimulate muscles. Ironically, this action of the botulinum toxin has given it a beneficial niche in the world of medicine. Certain medical disorders are characterized by involuntary and uncontrollable muscle contractions. Medical researchers have discovered that injecting a strictly controlled dose of botulinum toxin into affected muscles inhibits excessive muscle contractions. The muscle is partially paralyzed and normal movement is retained.

Human botulism (caused by botulism toxins A, B, and E) may stem from contaminated food, wound contamination, or the intestinal botulism toxin found in infants. Each produces multiple symptoms as follows:

- Food-borne botulism. Food that has been improperly preserved or stored can harbor botulinum toxin-producing clostridia. Canned or jarred baby food has also been known to cause botulism. Symptoms of food-borne botulism typically appear within 18 to 36 hours of eating contaminated food, with extremes of four hours to eight days. Initial symptoms include blurred or double vision and difficulty swallowing and speaking. Possible gastrointestinal problems include **constipation**, **nausea**, and **vomiting**. As botulism progresses, the victim experiences weakness or paralysis, starting with the head muscles and progressing down the body. Breathing becomes increasingly difficult. Without medical care, respiratory failure and death are very likely.
- Infant botulism. Infant botulism was first described in 1976. Unlike adults, infants younger than 12 months are vulnerable to *C. botulinum* colonizing the intestine. Infants ingest spores in honey or simply by swallowing spore-containing dust or dirt. The spores germinate in the large intestine and, once colonized, toxin is produced and absorbed into the infant's body from the entire intestinal tract. The first symptoms include constipation, lethargy, and poor feeding. As infant botulism progresses, sucking and swallowing (thus eating)

become difficult. A nursing mother will often notice her own breast engorgement as the first sign of her infant's illness. The baby suffers overall weakness and cannot control head movements. Because of the flaccid paralysis of the muscles, the baby appears floppy. Breathing is impaired, and death from respiratory failure is a very real danger.

- Wound botulism. Confirmed cases of wound botulism have been linked to trauma such as severe crush injuries to the extremities, surgery, and illegal drug use. Wound botulism occurs when *Clostridia* colonize an infected wound and produce botulinum toxin. The symptoms usually appear four to 18 days after an injury occurs and are similar to food-borne botulism, although gastrointestinal symptoms may be absent.

When to call the doctor

Infant botulism may be hard for parents to identify because the symptoms occur slowly. Parents should call the doctor or take the infant or child to emergency services as soon as the child shows symptoms such as weakness or listlessness, lethargy, irritability, and poor eating (or nursing) along with decreased bowel movements or constipation. An affected child may be so weak as to appear floppy and not in control of muscle movements, especially movement of the neck and head. Whether parents are aware of a possible source of the botulism toxin, the suggestive symptoms should not be ignored.

Diagnosis

Differential diagnosis of botulism can be complex because the symptoms mimic those of other diseases, especially diseases characterized by muscle weakness. Botulism must be differentiated from diseases such as the following:

- Guillain-Barré syndrome
- meningoencephalitis
- myasthenia gravis
- systemic poisoning or sepsis
- reactions to therapeutic drugs
- nervous system infection
- carbon monoxide or atropine intoxication
- severe allergic reactions to bee sting, shell fish, and other allergens
- failure to thrive

Sepsis is the most common initial diagnosis for actual infant botulism, and meningoencephalitis may also be the diagnosis if irritability and lethargy are present. Infant botulism was at one time linked to 5 to 15 percent of cases of **sudden infant death syndrome** (SIDS, crib death) because of spores found in 4 to 15 percent of cases; however, a subsequent 10-year study did not find a significant influence of botulism on SIDS.

Laboratory tests are used to make a definitive diagnosis, but if botulism seems likely, treatment starts immediately without waiting for test results, which may take up to two days. Diagnostic tests focus on identifying the organism causing the illness. This may involve performing a culture on contaminated material from the suspect food or the nose or throat of the affected individual. In infant botulism, the infant's stool may be cultured to isolate the organism; this test may be performed by the state health department or the Centers for Disease Control (CDC). Culture results are available from the microbiology laboratory as soon as bacteria grow in a special plate incubated at temperatures at or above body temperature. The growth of *Clostridium* confirms the diagnosis. Sometimes the organism cultured is not *Clostridium* as suspected. The microbiology laboratory may use samples of the bacteria grown to perform other special techniques in order to help identify the causative organism.

While waiting for diagnostic test results, doctors ask about recently consumed food, possible open sores, recent activities and behavior, and other factors that may help to rule out other disease possibilities. A physical examination is done with an emphasis on the nervous system and muscle function. As part of this examination, imaging studies such as CT and MRI may be done and electrodiagnostic muscle function tests (electromyogram) or lumbar punctures may be ordered. Laboratory tests look for the presence of botulinum toxin or *Clostridia* in suspected foods and/or the child's blood serum, feces, or other specimens for traces of botulinum toxin or *Clostridia*. Magnesium levels may be measured, since magnesium increases the activity of *Clostridium*. Additional diagnostic tests may be done to rule out other diseases or conditions with similar symptoms.

Treatment

Drugs

Older children and adults with botulism are sometimes treated with an antitoxin derived from horse serum that is distributed by the Centers for Disease Control and Prevention. The antitoxin (effective against toxin types A, B, and E) inactivates only the botulinum toxin that is unattached to nerve endings. Early injection of the antitoxin, ideally within 24 hours of onset of symptoms, can preserve nerve endings, prevent progression of the disease, and reduce mortality.

Unfortunately, infants cannot receive the antitoxin used for adults. For them, human botulism immune globulin (BIG) is the preferred treatment. It is available in the United States through the Infant Botulism Treatment and Prevention Program in Berkeley, California. BIG neutralizes toxin types A, B, C, D, and E before they can bind to nerves. This antitoxin can provide protection against A and B toxins for approximately four months. Though many infants recover with supportive care, BIG cuts hospital stay in half and, therefore, reduces hospital costs by 50 percent as well.

Aside from the specific antitoxin, no therapeutic drugs are used to treat botulism. **Antibiotics** are not effective for preventing or treating botulism because the *Clostridium* group of toxins are not sensitive to them. In fact, antibiotic use is discouraged for infants because bacteria could potentially release more toxin into a baby's system as they are killed. Antibiotics can be used, however, to treat secondary respiratory tract and other infections.

Respiratory support

Treatment for infants usually requires them to be in an intensive care unit, involving intensive respiratory support and nasogastric tube feeding for weeks or even months. Once an infant can breathe unaided, physical therapy is initiated to help the child relearn how to suck and swallow. In older children and adults, a respirator is often required to assist breathing; a tracheostomy may be necessary in some cases.

Surgery

Surgery may be necessary to clean an infected wound (debridement) and remove the source of the bacteria producing the toxin. Antimicrobial therapy may be necessary.

Gastric lavage

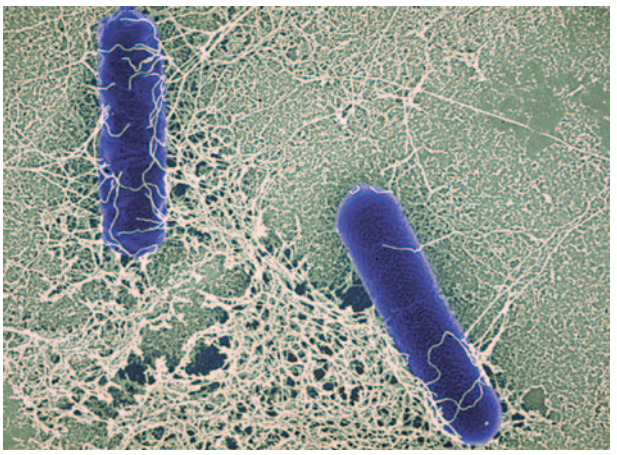
When botulism in older children or adults is caused by food, it often is necessary to flush the gastrointestinal tract (gastric lavage). Often cathartic agents or enemas are used. It is important to avoid products that contain magnesium, since magnesium enhances the effect of the toxin.

Nutritional concerns

Parents should avoid feeding honey to infants younger than 12 months because it is one known source of botulism spores.

Prognosis

With medical intervention, botulism victims can recover completely, though it may be a very slow recovery. It takes weeks to months to recover from botulism, and severe cases can take years before a total recovery is attained. Recovery depends on the nerve endings building new proteins to replace those destroyed by botulinum toxin.



Electron micrograph of the *Clostridium botulinum* bacteria which cause botulism. *(Photograph by Gary Gaugler. Visuals Unlimited.)*

Prevention

Vaccines have not been developed directed against botulism, which makes prevention of infant botulism or other forms of the disease difficult, since exposure to the botulinum toxic is typically unrecognized. Food safety is the surest prevention for botulism. Botulinum toxin cannot be seen, smelled, or tasted, so the wisest course is to discard any food that seems spoiled; avoid eating food from dented, rusty, or bulging cans; avoid refreezing meats once they have been thawed; and avoid buying broken containers of food or eating food that has been stored at room temperature or above for more than a few hours. People who like to can food at home must be diligent about using sterile equipment and following U.S. Department of Agriculture canning guidelines.

Infant botulism is difficult to prevent, because controlling what goes into an infant's mouth is often beyond control, especially in regard to airborne spores. One concrete preventative is to never feed honey to infants younger than 12 months as it is one known source of botulism spores. As infants begin eating solid foods, the same food precautions should be followed as for older children and adults.

Parental concerns

Because symptoms of infant botulism appear slowly, parents may be concerned that they will be missed or not found early. Normal watchfulness of the parents is sufficient, paying attention to any change in feeding, a decrease in bowel movements, or a lack of normal responses such

KEY TERMS

Acetylcholine—A chemical called a neurotransmitter that functions primarily to mediate activity of the nervous system and skeletal muscles.

Antitoxin—An antibody against an exotoxin, usually derived from horse serum.

Computed tomography (CT)—An imaging technique in which cross-sectional x rays of the body are compiled to create a three-dimensional image of the body's internal structures; also called computed axial tomography.

Culture—A test in which a sample of body fluid is placed on materials specially formulated to grow microorganisms. A culture is used to learn what type of bacterium is causing infection.

Electrooculography (EOG)—A diagnostic test that records the electrical activity of the muscles that control eye movement.

Flaccid paralysis—Paralysis characterized by limp, unresponsive muscles.

Lumbar puncture—A procedure in which the doctor inserts a small needle into the spinal cavity in the lower back to withdraw spinal fluid for testing. Also known as a spinal tap.

Magnetic resonance imaging (MRI)—An imaging technique that uses a large circular magnet and radio waves to generate signals from atoms in the body. These signals are used to construct detailed images of internal body structures and organs, including the brain.

Neurotoxin—A poison that acts directly on the central nervous system.

Neurotransmitter—A chemical messenger that transmits an impulse from one nerve cell to the next.

Sepsis—A severe systemic infection in which bacteria have entered the bloodstream or body tissues.

Spore—A dormant form assumed by some bacteria, such as anthrax, that enable the bacterium to survive high temperatures, dryness, and lack of nourishment for long periods of time. Under proper conditions, the spore may revert to the actively multiplying form of the bacteria. Also refers to the small, thick-walled reproductive structure of a fungus.

Toxin—A poisonous substance usually produced by a microorganism or plant.

Tracheostomy—A procedure in which a small opening is made in the neck and into the trachea or windpipe. A breathing tube is then placed through this opening.

as turning of the head and body movements. It may be helpful to remember how rare botulism is, how easy it is to assure food safety, and also that morbidity and mortality can be avoided with early recognition of the symptoms.

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ORGANIZATIONS

Centers for Disease Control and Prevention. 1600 Clifton Rd., NE, Atlanta, GA 30333. Web site: <www.cdc.gov>.

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Braces, body *see* **Immobilization**
Braces, dental *see* **Orthodontics**

Brachial plexopathy, obstetric

Definition

Brachial plexopathy is any injury to the brachial plexus—the nerve bundles located on each side of the neck that give rise to the individual nerves controlling the muscles of the shoulders, arms, and hands.

Description

Brachial plexopathy occurs most often during birth, when an infant's neck and shoulders are stretched apart

during passage through the birth canal. Injury to the brachial plexus is referred to by various names. Some names, such as obstetric Erb's palsy, refer to the specific region of the brachial plexus where the injury has occurred. Other more general names for the condition include:

- obstetric brachial plexus
- brachial plexus injury (BPI)
- brachial plexus palsy (BPP)
- brachial palsy in the neonate

Brachial plexus injuries

The nerves of the brachial plexus are the fifth through the eighth cervical nerves (C5, C6, C7, and C8) and the first thoracic nerve (T1). They run from nerve roots on each side of the upper spinal cord to regions beneath the collarbone where they branch out as the major nerves of the shoulders, arms, wrists, and hands.

Every brachial plexus injury is different, depending on the affected nerve or nerves and the extent and severity of the injury. There are four general types of injury, and an individual brachial plexopathy may include any or all of these injury types, on one or both (bilateral) sides of the body. The types are as follows:

- A stretch, praxis (damage), or traction (pulling or tension-creating) injury, in which the nerve has been overstretched and damaged but is not torn. Most brachial plexus injuries in newborns are of this type.
- A rupture, in which there has been a separation within a nerve. A single nerve may have more than one rupture.
- A neuroma, in which scar tissue has formed around a nerve injury. The scar tissue puts pressure on the nerve and interferes with nerve signal conduction to the muscles.
- An avulsion is the most severe form of brachial plexus injury. It is the detachment of a nerve from the spinal cord.

A minor brachial plexus injury can be a stretched nerve that results in a short-circuit in a few of the nerve fibers, causing temporary paralysis. A more severe injury may involve a complete disruption of the nerve, in which the nerve fibers around the injury become disorganized, all nerve function is lost, and scarring prevents the nerve from healing. Such an injury can result in permanent paralysis.

Obstetrical brachial plexopathy

Most injuries to the brachial plexus during birth involve the C5 and C6 nerve roots, affecting the movement of the shoulder, upper arm, and elbow. Limited shoulder motion can affect the function and strength of the hand. Obstetrical brachial plexopathies are classified according to the extent of the injury.

ERB'S PALSY Obstetric Erb's palsy is an injury in the upper brachial plexus involving C5 and C6 and sometimes C7. It usually affects the upper arm and the rotation of the lower arm. Erb's palsy is known also by the following names:

- Erb's paralysis
- Duchenne's paralysis
- Erb-Duchenne palsy or paralysis
- Duchenne-Erb syndrome or paralysis
- upper brachial plexus palsy or paralysis, Erb-Duchenne type

KLUMPKE'S PALSY Klumpke's palsy or Klumpke's paralysis is an injury to the lower brachial plexus: C7, C8, and sometimes T1. Children with Klumpke's palsy have normal use of the shoulder and elbow but weakness or paralysis in the hand and fingers. An infant also may have a drooping eyelid on the opposite side of the body from the affected hand.

TOTAL PLEXUS PALSY Also called Erb/Klumpke palsy, total plexus palsy involves all of the nerve roots of the brachial plexus to at least some extent. The entire upper extremity is affected.

Demographics

It is estimated that in North America between one and three of every 1,000 newborns are affected to some extent by brachial plexopathy. It appears to be less common than in the past, due to improvements in infant delivery methods and the increased use of cesarean sections (c-sections) to surgically remove the infant through the abdomen. However, some physicians are concerned that the late 1990s and early 2000s trend toward decreasing elective c-section deliveries will result in more cases of brachial plexopathy among newborns.

Erb's palsy is the most common type of obstetric brachial plexopathy. Total plexus palsy accounts for about 10 percent of obstetric brachial plexopathies and Klumpke's palsy accounts for less than 1 percent.

Causes and symptoms

Causes

Brachial plexopathy can result when the following events occur. An individual brachial plexopathy may include any or all of these injury types, on one or both (bilateral) sides of the body. The events are as follows:

- An infant's shoulder becomes stuck on the mother's pelvic bone during birth; the infant's neck may be stretched and the brachial plexus injured as the physician pulls on the baby to free it before circulatory or respiratory stress occurs.
- An infant's head and neck are pulled to one side while the shoulders pass through the birth canal.
- There is excessive stress on the infant's shoulders during a head-first delivery.
- There is pressure on the infant's raised arms during a breech (feet-first) delivery.
- An infant's shoulder is too large to fit easily through the birth canal (shoulder dystocia). An individual brachial plexopathy may include any or all of these injury types, on one or both (bilateral) sides of the body. (However, the association between brachial plexopathy and shoulder dystocia is controversial, with some studies indicating that 4 to 40 percent of shoulder dystocias result in brachial plexopathy and other studies finding no evidence of an association.)

Causes of brachial plexopathy, other than injuries during birth, include:

- any trauma or injury to the brachial plexus, such as might occur with vehicular accidents, **sports injuries**, puncture **wounds**, or surgery
- congenital abnormalities that affect the cervical ribs
- pressure from tumors in the region
- damage from radiation therapy
- exposure to some toxins, drugs, or chemicals

RISK FACTORS Although brachial plexus injuries can occur during any birth, there are particular risk factors. The highest rates of brachial plexus injury (7.8%) occur in newborns weighing over 10 lb (4.5 kg) who are born by assisted vaginal delivery to diabetic mothers. Premature and underdeveloped newborns are at a decreased risk for brachial plexopathy. Other risk factors for this injury are as follows:

- Some 50–70 percent of brachial plexus injuries occur in larger-than-average newborns, usually those over 7.7 lb (3.5 kg).
- About 44 percent of brachial plexopathies occur in newborns who experienced fetal distress.
- Breech deliveries increase the risk of brachial plexopathy by 175-fold, often causing bilateral injuries to the lower nerve roots of the brachial plexus.
- Prolonged labor and difficult or abnormal labor or delivery increase the risk of injury.
- Use of forceps or a vacuum device to deliver a baby increases risk.
- Injuries occur more frequently in births to mothers who have had several prior births.
- A mother who has had previous brachial-plexus-injured infants is at a 14-fold-increased risk of having another infant with brachial plexopathy.

Symptoms

The symptoms of brachial plexopathy vary greatly depending on the extent and severity of the damage. Some children with brachial plexopathy have the following limitations:

- They have no feeling or muscle control in the arm or hand.
- They can move their arm but have little control over the wrist and hand.
- They can use their hands normally but cannot use their shoulder or elbow muscles.

Typical symptoms that may be recognized at birth or shortly thereafter include the following:

- lack of spontaneous movement in the upper or lower arm or hand
- weakness in an arm
- weak or no grip
- no Moro reflex (a startled response when an infant's head drops suddenly, characterized by spreading the arms with the palms up and fingers flexed; as the reflex ends, the arms return to the body and the elbows flex and relax)
- weak or absent normal infant position with the arm flexed at the elbow and held against the body
- a limp or paralyzed arm
- lack of sensation in the arm or hand (a completely flaccid arm or part of an arm indicates an avulsion injury)

Other common symptoms of brachial plexopathy include:

- limited range-of-motion (ROM) in the arm or part of the arm
- sensation changes in the arm
- weakness in specific muscle groups
- inability to perform typical movements
- inability to bear weight in the arm
- neglect of the arm
- atypical positioning of the arm
- developmental delays
- torticollis (a shortened neck muscle, causing the head to tilt to one side)

Additional symptoms of brachial plexopathy include:

- arm **pain**
- facial paralysis on the affected side
- inability to sit up without assistance
- inability to crawl without a therapeutic device

Symptoms of Erb's palsy include:

- decreased abduction (turning outward) and external rotation of the shoulder
- decreased elbow flexion (bending)
- decreased supination (rotation of the forearm so that the palm of the hand is turned forward or upward)
- grasp reflex but no biceps reflex
- normal hand movement but abnormal Moro reflex
- sensory deficits
- paralysis of the diaphragm on the affected side in about 5 percent of Erb's palsy cases

Symptoms of total plexus palsy include:

- paralysis extending from the shoulder to the hand with no grasp reflex
- sensory loss
- Horner's syndrome (pupil contraction, receding eyeball, and sometimes inability to sweat on the affected side of the face) in about one-third of total plexus palsy cases

When to call the doctor

Although obstetric brachial plexopathy usually heals quite rapidly on its own, the infant may begin physical therapy within the first two weeks of life and should be evaluated by a specialist, such as a pediatric neurologist, by six weeks of age.

Diagnosis

A newborn lacking movement in an arm is examined first for **fractures** of the collarbone, ribs, humerus (the long bone of the upper arm, extending from the shoulder to the elbow), and even the femur (thighbone), as well as dislocations of the shoulder or elbow. Symptoms of such fractures and dislocations may be similar to those of brachial plexopathy and can cause infants to not move their arms. Those at risk for obstetric brachial plexopathy are also at risk for fractures and dislocations during birth. An infant who is not moving a fractured arm because of the pain will still exhibit a Moro reflex. However, when the infant is rolled from side to side, a brachial-plexopathy-affected arm may flop.

Brachial plexopathy is diagnosed by the following:

- an unpredictable or patchy pattern of sensory, motor, and reflex dysfunction in the arm
- **x rays** that rule out other causes for the symptoms
- a nerve conduction velocity (NCV) test detecting nerve damage (An electrode on the skin stimulates the nerve with a mild impulse and other electrodes record the resulting electrical impulse; the distance between the electrodes and the time the impulse takes to travel determines the conduction velocity; below-normal nerve conduction may indicate damage to the nerve.)
- electromyography (EMG) measures of the muscle response to nerve impulses (A needle electrode is inserted through the skin into the muscle and records the electrical activity of the muscle; EMG can reveal loss of nerve activity within one week of birth and can help determine which nerves are damaged.)
- magnetic resonance imaging (MRI; use magnets and radio waves to obtain images) of the brachial plexus determining the location and type of injury (MRI can be performed when the infant reaches two to three months of age.)
- computed tomography (CT) or computed axial tomography (CAT) scans performed at two to three months of age to reveal injury (CT scans use a thin, rotating x-ray beam to obtain an image as the x rays pass through body parts.)
- myelograms (x rays or CT scans taken after a dye is injected into the spinal fluid) revealing the fluid space surrounding the spinal cord in the neck (Although they may help to determine the location and type of nerve injury, myelograms generally have been replaced by MRI scans of the brachial plexus due to false positives and the invasiveness of procedure.)

Treatment

Nerve regeneration in newborns occurs at 1.8 mm. per day. Therefore, stretch or praxis injuries to the brachial plexus usually heal on their own within about three months, leading to complete recovery. However, many children with brachial plexopathy require treatment. Specialists who may become involved in a child's treatment include:

- physical therapists
- occupational therapists
- neurologists
- pediatric neurosurgeons
- orthopedic surgeons
- plastic surgeons

Physical therapy

Gentle massage and range-of-motion (ROM) exercises usually are initiated immediately, even in infants with very mild brachial plexopathy. Therapy regimes are individualized for each child, depending on the specific injury and its effects.

Treatment for brachial plexopathy may include a combination of exercises and focused physical, occupational, and aquatic therapies. Typically the therapist teaches the child's **family** to perform the following:

- position the infant properly
- never lift the child under the armpits
- perform passive range-of-motion (PROM) exercises on the infant
- assist with weight-bearing activities, even in a newborn
- help the infant avoid atypical movement patterns
- help thwart the infant's tendency to neglect the affected region
- detect muscle contractions even when no movement is evident
- help avoid tightening of the infant's muscles
- make adaptive equipment for the infant

At home parents may be instructed to do the following:

- perform ten repetitions of all prescribed PROMs two to three times daily
- begin gentle movements with the child's forearm rotated and the palm upward or forward (supination) to increase joint flexibility and muscle tone
- perform joint compression and weight-bearing exercises throughout the affected extremity
- practice aquatic exercises as prescribed
- practice tactile stimulation on the affected extremities, using textured materials, soft balls, vibration, and massage to increase sensory awareness
- actively involve the affected extremity in developmentally appropriate activities to increase strength and coordination, working first without the effects of gravity and later working against gravity
- place pillows or stuffed animals under the child's armpit or along side the affected arm during rest or **sleep** to obtain a sustained stretch

Play therapy

Play therapies are used to extend ROM from six months of age on in children with brachial plexopathy. General body activities include:

- any activity that forces the child to reach
- any activity that puts pressure on the affected arm or hand
- playing while lying on the stomach
- stacking empty boxes
- playing circle games
- playing "Simon says"
- "making angels" while lying on the floor
- throwing a beach ball overhead
- riding a tricycle or bicycle
- playing and walking in water
- climbing on play equipment
- climbing and sliding down a slide

Activities to increase hand coordination include:

- folding paper napkins
- copying and drawing basic shapes
- using scissors, paste, or toy tools
- painting
- writing on a chalkboard
- sewing cards
- playing with **toys** in the sink or tub
- playing with wooden puzzles, dominoes, Legos, or blocks

- stringing beads
- rolling dough
- playing in sand
- throwing a soft ball
- picking up and sorting small objects

Surgery

About ten percent of brachial plexus injuries in infants require surgery. Children with total plexus palsy who have not improved by three months of age and children with Erb's palsy who cannot strongly bend their elbow or raise their shoulder by six months of age are candidates for exploratory surgery to examine the nerve damage and perform possible nerve grafting. Surgeries are most successful if performed when the child is five to seven months of age. Surgeries are less likely to be successful if performed after 12 months of age.

Rupture injuries usually require surgery. Avulsion injuries require surgery to reattach the nerve root to the spinal cord. Surgeries that may be performed by a pediatric neurosurgeon include:

- neuroplastysurgery to repair the nerve, including stretching the nerve to relieve tension or reconnecting torn nerves
- neurolysis to destroy damaged nerve tissue or to loosen or remove scar tissue (neuroma) around the nerve
- nerve grafting by transplanting nerves from another part of the body, such as the ankle, to bridge a torn nerve or to reconnect the nerve root after an avulsion injury

Other types of surgery include:

- muscle surgery on a child aged 18 to 24 months if physical therapy has not restored shoulder rotation
- muscle or tendon transfer surgeries to restore function (For example, a child without elbow function might have a leg muscle transferred to the elbow and attached to the nerves.)
- muscle transfer surgeries in which the muscles are rearranged in an attempt to prevent permanent abnormalities
- plastic surgeries

A variety of other surgical procedures may be considered depending upon the specific situation. At least 90 percent of children improve following surgery. **Exercise** and massage are eventually reinitiated, depending on the type of surgery.

Other treatments

Other treatments for brachial plexopathy can be used. For example, various types of splints are available to position and support the extremity during activities and to increase weight-bearing ability. Also, casts are sometimes used to allow the nerves to heal. Finally, electrical stimulation, in conjunction with EMG, can deliver a small amount of electrical current to the muscle to prevent atrophy; it may be performed either by a therapist or at home. The child is examined regularly, both during and after treatment, for muscle recovery and proper joint development.

Prognosis

About 85 percent of infants with brachial plexopathy make a complete neurological recovery within three to six years. One study of 59 children found that 88 percent recovered by four months of age, 92 percent recovered by 12 months, and 93 percent recovered by 48 months. In another study of 28 infants with damage to the upper brachial plexus and 38 infants with total plexus palsy, 92 percent recovered spontaneously.

However, the prognosis for an individual brachial plexopathy depends on the location, severity, and extent of the damage and may be difficult to predict. In general, damage to the nerve sheath (outer covering) alone has a good prognosis. Praxis-type injuries, in which the nerve is damaged but not torn, usually improve within three months and eventually heal completely. Stretch injuries heal on their own, with 90 to 100 percent of function returning within one to two years. Severed nerves, particularly avulsion injuries in which the nerve is severed at the root, have poorer prognoses. In severe cases there may be permanent partial or total loss of nerve function in the affected nerves and weakness or paralysis of the arm may be permanent.

Erb's palsy has the best prognosis since, although shoulder, elbow, and forearm function may be affected, the hands and fingers are not affected. However, infants with Erb's palsy that involves C7 as well as C5 and C6 have a poorer prognosis. In addition Erb's palsy may lead to secondary deformities as the child grows. The most common problem is internal rotation contracture (permanent muscle contraction) of the shoulder.

Complete recovery from brachial plexopathy may be difficult to define. A Swedish study found that about 30 percent of children who had recovered the use of their shoulder, biceps, and hand by the age of three months still had disabilities at age five, including a weakened hand grip or difficulty dressing or running. The delay in normal functioning caused by brachial plexopathy and

any muscle imbalances across a joint can have a major impact on the child's growing skeleton and can result in permanent muscular-skeletal abnormalities.

Long-term effects of brachial plexography may include:

- a weak shoulder girdle
- muscle atrophy
- joint contractures
- a bent elbow (called Erb Engram) with shoulder adduction (pulled in toward the body)
- impaired limb growth
- progressive bone deformities

Prevention

The primary prevention for obstetrical brachial plexopathy is the avoidance of a potentially difficult delivery by choosing **cesarean section**. Failure to anticipate a particularly large baby before delivery is an important risk factor. Some physicians suggest that women whose previous children had shoulder dystocia should be offered an elective cesarean delivery. However, cesarean deliveries also have risks associated with them, and it appears that increasing the frequency of c-sections would prevent few cases of brachial plexopathy since large-scale studies have shown that 3 percent of brachial plexus injuries occur during cesarean deliveries.

The use of an epidural (local) anesthetic during labor may contribute to the risk of brachial plexopathy since the anesthetic decreases the mother's ability to push during labor and may force the physician to use forceps or a vacuum device to pull the baby out.

Parental concerns

Promoting recovery

Although exercises required to treat brachial plexopathy in infants may be painful, they are essential for preventing much more serious pain and suffering as the child grows. In addition to performing prescribed massage and ROM exercises, parents should:

- always first offer objects or food to the child's affected side
- not allow the child to use compensatory movements, particularly those involving the trunk of the body
- have the child use the unaffected arm as a guide for the affected one so that the affected arm experiences what the unaffected arm is doing

Parents should help their child to become self-sufficient in the movements involved in the following daily tasks:

- toileting
- personal hygiene
- dressing
- performing simple household chores such as picking up toys, cleaning a room, or setting a table

Daily activities that increase ROM are essential even if the child is seeing a physical therapist. Parents should do the following:

- encourage a consistent daily routine and participation in activities
- try to incorporate therapy into daily play and other activities
- have the child participate in play activities that increase ROM for 15 to 30 minutes twice a day
- provide enjoyable and challenging activities
- encourage movement and use of affected joints
- encourage the child to focus on using the affected arm
- focus on the child's abilities rather than lack of abilities
- avoid doing something for the child simply because the child finds it difficult
- reward the child with verbal praise or a treat for attempting or initiating an activity
- allow the child enough time for each activity

Activities

Numerous activities have been found useful for promoting a child's recovery. Activities to increase shoulder flexibility include:

- placing and removing objects from a board or mirror
- popping bubbles
- rolling out dough with a rolling pin
- raising a dowel over the head
- playing basketball

Activities to increase shoulder abduction (movement of the arm away from the body) include playing bird or airplane and turning a jump rope.

Activities to increase elbow flexion (bending) include:

- bringing food items from hand to mouth

KEY TERMS

Abduction—Turning away from the body.

Adduction—Movement toward the body.

Avulsion—The forcible separation of a piece from the entire structure.

Brachial plexus—A group of lower neck and upper back spinal nerves supplying the arm, forearm and hand.

Cervical nerves—The eight pairs of nerves (C1C8) originating in the cervical (neck) region of the spinal cord.

Cesarean section—Delivery of a baby through an incision in the mother's abdomen instead of through the vagina; also called a C-section, Cesarean birth, or Cesarean delivery.

Computed tomography (CT)—An imaging technique in which cross-sectional x rays of the body are compiled to create a three-dimensional image of the body's internal structures; also called computed axial tomography.

Contracture—A tightening or shortening of muscles that prevents normal movement of the associated limb or other body part.

Dystocia—Failure to progress in labor, either because the cervix will not dilate (expand) further or because the head does not descend through the mother's pelvis after full dilation of the cervix.

Electromyography (EMG)—A diagnostic test that records the electrical activity of muscles. In the test, small electrodes are placed on or in the skin; the patterns of electrical activity are projected on a screen or over a loudspeaker. This procedure is used to test for muscle disorders, including muscular dystrophy.

Erb's palsy or paralysis—A condition caused by an injury to the upper brachial plexus, involving the cervical nerves C5, C6, and sometimes C7, affecting the upper arm and the rotation of the lower arm.

Flexion—The act of bending or condition of being bent.

Klumpke's palsy or paralysis—A condition caused by an injury to the lower brachial plexus, involving the cervical nerves C7 and C8, and sometimes the thoracic nerve T1, causing weakness or paralysis in the hands and fingers.

Magnetic resonance imaging (MRI)—An imaging technique that uses a large circular magnet and radio waves to generate signals from atoms in the body. These signals are used to construct detailed images of internal body structures and organs, including the brain.

Moro reflex—A startle response in a newborn, characterized by spreading the arms with the palms up and fingers flexed; the reflex usually disappears by two months of age.

Nerve condition velocity (NCV)—Technique for studying nerve or muscle disorders, measuring the speed at which nerves transmit signals.

Neurolysis—The destruction of nerve tissue or removal of scar tissue surrounding a nerve.

Neuroma—Scar tissue that forms around a nerve; a tumor derived from nerve tissue.

Neuroplasty—Surgery to repair nerves.

Palsy—Uncontrolable tremors.

Total plexus palsy—Erb/Klumpke palsy; a condition resulting from injury involving all of the brachial plexus nerves and affecting the entire upper extremity of the body.

- using big jacks or chew toys
- talking into a play microphone
- playing a musical instrument

Activities to increase elbow extension include **crawling** and reaching out for objects.

Activities to increase wrist extension include:

- shaking rattles
- pulling pegs from a pegboard
- knocking on a door
- playing drums or a xylophone
- banging with a hammer
- splashing water
- painting

If a child has no hand function, double sided Velcro can be placed around the hand and used to hold rattles and toys with Velcro attached to them. An older child can use a Velcro mitt and balls.

Support groups

There are numerous support groups across the United States for the families of children with Erb's palsy and other brachial plexopathies. Support groups offer encouragement and advice on the following topics:

- coping with pain and crying during therapy
- coping with daily routines
- play therapies and other activities
- sibling issues
- daycare and school
- social issues facing the child

Resources

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Erb's Palsy Lawyers Network. Web site: <www.erbspalsy.com/index.htm>.

National Brachial Plexus/Erb's Palsy Association Inc. PO Box 23, Larsen, WI 54947. Web site: <www.nbpepa.org>.

National Institute of Arthritis and Musculoskeletal and Skin Diseases. National Institutes of Health, 1 AMS Circle, Bethesda, MD 20892–3675. Web site: <www.nih.gov/niams>.

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Margaret Alic, Ph.D.

Brain defects, congenital *see* **Congenital brain defects**

Breast development

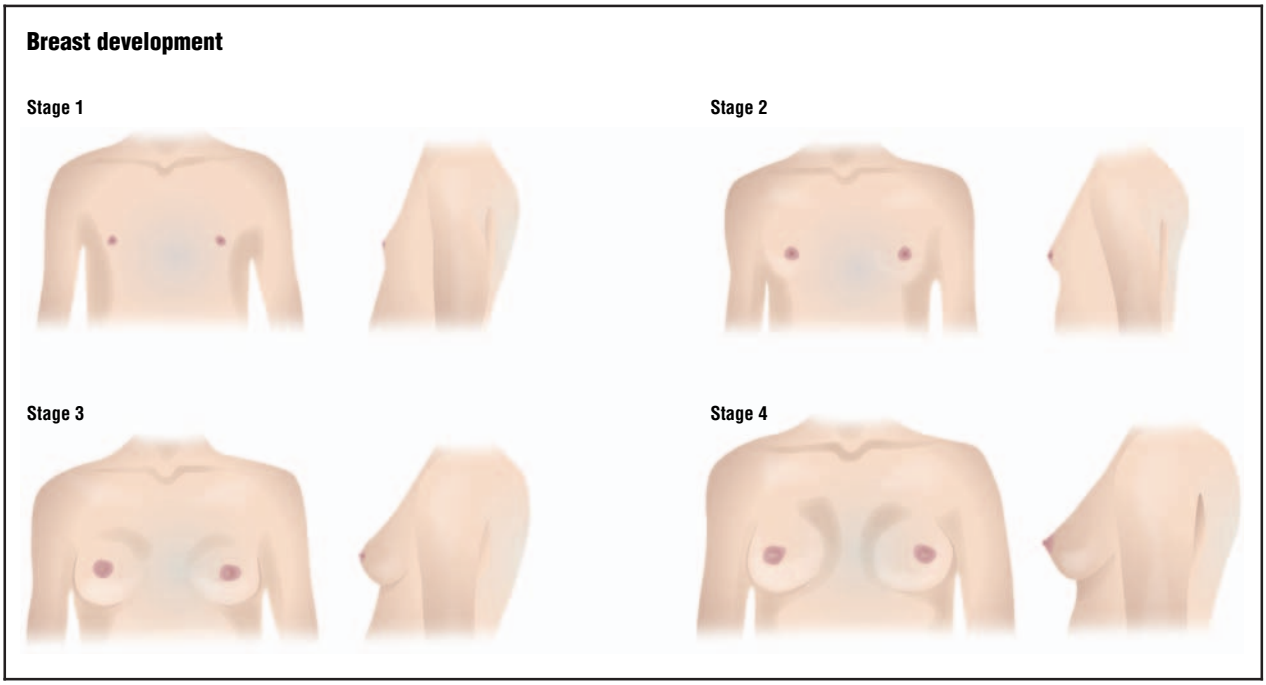
Definition

A newborn baby has nipples, areolas, and the beginnings of breast tissue, but most of breast development occurs in two different periods of time in a woman's life: first in **puberty**, then during pregnancy. Breast development is a vital part of puberty in the human female. Interestingly, humans are the only mammals whose breasts develop before they are needed to serve their biological purpose—breastfeeding.

Description

The first stage of breast development begins at about six weeks of fetal development with a thickening called the mammary ridge or the milk line. By six months of development, this ridge extends all the way down to the groin, but then regresses. Solid columns of cells form from each breast bud, with each column becoming a separate sweat gland. Each of these has its own separate duct leading to the nipple. By the final months of fetal development, these columns have become hollow, and by the time a female infant is born, a nipple and the beginnings of the milk-duct system have formed.

As a girl approaches puberty, the first outward signs of breast development begin to appear. When the ovaries start to secrete estrogen, fat in the connective tissue begins to accumulate causing the breasts to enlarge and the duct system begins to grow. Breast development normally begins about one to two years before the men-



The four stages of breast development. In Stage 1 shows the flat breasts of childhood. By Stage 2, breast buds are formed as milk ducts and fat tissue develop. In Stage 3, the breast become round and full, and the areola darkens. Stage 4 shows fully mature breasts. *(Illustration by GGS Information Services.)*

strual period begins. Usually these signs are accompanied by the appearance of pubic hair and hair under the arms.

Once ovulation and **menstruation** begin, the maturing of the breasts begins with the formation of secretory glands at the end of the milk ducts. The breasts and duct system continue to grow and mature with the development of many glands and lobules. The rate at which breasts grow varies significantly and is different for each young woman. Breast development occurs in five stages:

- Stage One: In preadolescence, the breasts are flat and only the tip of the nipple is raised.
- Stage Two: Buds appear, breast and nipple are raised, fat tissue begins to form and the areola (dark area of skin that surrounds the nipple) enlarges.
- Stage Three: Breasts are slightly larger with glandular breast tissue present. Initially this happens in a conical shape and later in a rounder shape. The areola begins to darken.
- Stage Four: The nipple and areola become raised and form a second mound above the rest of the breast. Menstruation typically starts within two years of reaching this stage, and some girls skip this stage completely.
- Stage Five: Mature adult breast is rounded and only the nipple is raised.

The entire process from the breast bud stage through stage five usually takes about three to five years, but for some girls it takes close to ten years. After these five stages, the breast is still not considered mature or fully developed. Only pregnancy brings about the fullness of breast growth and development.

Each month, women experience fluctuations in hormones that make up the normal menstrual cycle. Estrogen, which is produced by the ovaries in the first half of the menstrual cycle, stimulates the growth of milk ducts in the breasts. The increasing level of estrogen leads to ovulation halfway through the cycle, and then the hormone progesterone takes over in the second half of the cycle, stimulating the formation of the milk glands. These hormones are believed to be responsible for the cyclical changes such as the swelling, **pain**, and tenderness that many women experience in their breasts just before menstruation. Many women also experience changes in breast texture, with breasts feeling particularly lumpy. This, too, is related to the glands in the breast enlarging in preparation for a possible pregnancy. If pregnancy does not occur, the breasts return to normal size.

KEY TERMS

Anorexia nervosa—An eating disorder marked by an unrealistic fear of weight gain, self-starvation, and distortion of body image. It most commonly occurs in adolescent females.

Bulimia nervosa—An eating disorder characterized by binge eating and inappropriate compensatory behavior, such as vomiting, misusing laxatives, or excessive exercise.

Congenital—Present at birth.

Puberty—The point in development when the ability to reproduce begins. The gonads begin to function and secondary sexual characteristics begin to appear.

Common problems

Breast development can start in a girl as early as eight years or as late as 13 years. If a girl does not have breast buds or pubic hair, which is the first signs of puberty, by age 14, there may be other medical problems. Most girls begin menstruating between ages nine and 18, with an average around 12 years of age. Primary **amenorrhea** is the absence of any menstrual flow in a girl who has never menstruated by the age of 16. Primary amenorrhea is not considered to have occurred until a girl is beyond age 16, if she has undergone other normal changes that occur during puberty. Primary amenorrhea may occur with or without other signs of puberty, but this condition is rare in the United States occurring in only about 0.1 percent of all girls.

There are many possible causes of primary amenorrhea, including **malnutrition**, extreme **obesity**, genetic disorders, endocrine (hormonal) disorders, pituitary tumors, congenital abnormalities, **anorexia nervosa**, bulimia, and, of course, pregnancy. Emotional distress or crisis about being different from friends or **family** can occur.

Parental concerns

Parents should become concerned if their daughter shows no signs of breast development by age 14 and if by the age of 16, there has not been a menstrual period.

When to call the doctor

Parents should call their health provider if their daughter is older than 16 and has not yet begun menstruating.

See also Puberty.

Resources

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Association of Women's Health, Obstetric and Neonatal Nursing. 2000 L Street, NW, Suite 740, Washington, DC 20036. Web site: <www.awhonn.org>.

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Breastfeeding *see* **Lactation**

Breath holding spells

Definition

Breath holding spells are episodes of brief, involuntary cessations of breathing that occur in children in response to stimuli such as anger, frustration, **fear**, or injury.

Description

A breath holding spell (BHS) is a reflexive response that occurs in some healthy young children, usually between the ages of about eight months and two years. A typical breath holding spell lasts between two and 20 seconds. The child cries or gasps, forcibly exhales, stops

breathing, and turns either blue (cyanotic form) or pale (pallid form). In a simple BHS the child may faint or briefly lose consciousness. If the child recovers before fainting, some physicians do not consider it to be a true breath holding spell. In a severe or complicated BHS, the child may have a small seizure while unconscious. The entire episode usually lasts less than one minute. The child regains consciousness and normal breathing, and color resume immediately.

The frequency of breath holding spells varies from several times a day to perhaps only once a year. About one-third of affected children have two to five spells daily whereas another one-third of affected children average one spell per month. It is not uncommon for a child with only sporadic or rare breath holding spells to have several in a single day.

Cyanotic form

About 60 percent of affected children have the cyanotic form of BHS, in which the skin and lips turn bluish. This type of breath holding spell also is called type 1, red-blue form, or cyanotic infantile syncope. It usually occurs when the child is angry or frustrated and may be a component of a temper tantrum.

Pallid form

About 20 percent of affected children have the pallid form of BHS in which the child turns pale. It also is called type 2, white breath-holding, reflex anoxic seizure, or pallid infantile syncope. The pallid form of BHS typically follows a frightening or painful experience.

An additional 20 percent of affected children have both types of breath holding spells at different times.

Demographics

Breath holding spells occur in about 5 percent of healthy children between the ages of six months and six years. They are most common between six and 18 months of age. Most affected children have had their first episode before the age of 18 months and almost all affected children have had an episode by the age of two. Breath holding spells are rare before six months of age. Although they are less common after the age of five, some children continue to have episodes until age six or seven.

Breath holding spells appear to occur equally among boys and girls. However, some studies have suggested that boys are more susceptible and that the average onset of BHS in boys is earlier (13–18 months) compared with girls (19–24 months). Studies have found no significant behavioral differences between children who have breath holding spells and those who do not. Nor is there a correlation between the frequency of spells and scores on behavioral profiles. Cyanotic breath holding spells typically begin when children are in a negative or oppositional phase and are starting to assert their independence. They typically end as the child learns to express emotions in words.

Causes and symptoms

Causes

Although the exact physiological mechanism of breath holding spells is not understood, they are an involuntary reflex caused by the interplay between the respiratory control center of the central nervous system, the autonomic nervous system, and the cardiopulmonary system.

CYANOTIC BHS Cyanotic BHS may be a component of a temper tantrum or a child's attempt to gain control over a situation. A cyanotic breath holding spell may also occur in response to the following:

- anger or frustration
- failure of the child to get its way
- a scolding or some other upsetting event
- **pain**
- being startled
- fear
- a confrontational situation
- a traumatic event

A breath holding spell is an involuntary reflex because it follows exhalation rather than inhalation. In a cyanotic BHS the long exhalation following crying causes breathing to stop. However, in some situations a child may learn how to trigger a cyanotic BHS. A child over the age of two with daily spells may have learned that intense crying or a temper tantrum can trigger a spell. If past breath holding spells have earned children lavish attention or enabled them to get their own way, the children may intentionally cause the spells to trigger an episode.

PALLID BHS Pallid breath holding spells are unpredictable. They usually occur in response to being startled, frightened, in pain, immunized, or injured, particularly after hitting the head.

In a pallid BHS the brain sends a signal via the vagus nerve that severely slows the heart rate, leading to a temporary cessation of breathing and loss of consciousness.

Risk factors

There appears to be a genetic component to at least some breath holding spells. About 25 percent of affected children—particularly those who experience pallid BHS—have a **family** history of BHS or fainting.

In some cases breath holding spells may be associated with anemia (a reduced number of red blood cells) caused by an iron deficiency, although this is controversial. Treatment may decrease the number of spells in some anemic children; however, treatment with iron increases the frequency of spells in children who are not anemic.

Symptoms

CYANOTIC BHS Once a parent or caregiver has witnessed a breath holding spell the symptoms are obvious. Children may do the following:

- cry vigorously for less than 30 seconds
- hyperventilate (over-breathe)
- have a pause in breathing followed by a long forced exhalation
- turn red in the face from anger
- stop breathing (apnea)
- have a strained face as if they are crying, although there is no sound
- turn bluish-purple, particularly in the face and around the lips, due to lack of oxygen in the brain.

The spell may end at this point and the child resumes breathing. Alternatively, the following may occur:

- The child may faint, become limp, or lose consciousness, usually for just a few seconds, due to a lack of oxygen reaching the brain.
- If breath holding lasts ten seconds or more, the unconscious child may experience muscle twitching, one or two jerky movements, back arching, body stiffening, or a true seizure.
- The child takes a deep breath and resumes normal breathing within 30 to 60 seconds.
- Consciousness and normal skin color return.

Most children recover completely within less than one minute after the start of the episode and resume normal activities. Some children may cry or scream for a period, and other children may fall asleep for an hour.

PALLID BHS In a pallid breath holding spell a child do the following:

- gasp and the lower jaw may quiver, but there is little or no crying
- experience a slowing heart rate or the heart may even stop briefly
- turn pale
- sweat
- stop breathing
- lose muscle strength and go limp
- faint or lose consciousness
- experience muscle twitching or body stiffness while unconscious
- have a seizure

Following these responses, the child's heart speeds up, breathing resumes on its own, and consciousness returns. The child usually recovers completely within one minute but may feel sleepy.

Seizures are much more likely with the pallid form than with the cyanotic form of BHS. Seizures during breath holding spells are more likely if breath holding lasts longer than usual. A child may vomit or urinate during a seizure.

When to call the doctor

Breath holding spells may have symptoms in common with various seizure disorders or other medical conditions. Therefore, a physician should be consulted if any of the following occurs:

- It is the first time a child has had a breath holding spell.
- The child is under six months of age, particularly if the spells occur during feeding or diaper changing.
- The child has a first breath holding spell at four-and-a-half years of age or older.
- The spells become more frequent.
- The spells become more severe.
- The pattern of the spells changes.
- The pallid form of BHS occurs frequently.
- The spells last more than one minute, with continuous body stiffening and relaxing.

Diagnosis

Diagnosis of breath holding spells usually is based on the medical history of children and their families and on complete physical and neurological examinations to

rule out other causes. Breath holding spells usually are diagnosed in the following way:

- a child's history of breath holding spells
- the exact sequence of events, which can be written down or videotaped
- lack of incontinence
- lack of post-convulsion symptoms
- blood tests to determine if a child has iron-deficient anemia

In addition the physician inquire about the following:

- if the child has ever been diagnosed with a medical condition
- if there have been recent changes in the child's behavior that are cause for concern
- if there have been recent changes in the child's life such as moving, a new sibling, or divorce
- if the parents have concerns about how other people may be treating the child

Ruling out other causes

Medical conditions that may cause breath holding spells include the following:

- Rett syndrome, a rare genetic disorder affecting girls
- Batten disease, the juvenile form of a group of progressive neurological disorders known as neuronal ceroid lipofuscinoses
- Riley-Day syndrome, a rare genetic disorder
- familial dysautonomia, a rare genetic disorder that can cause involuntary breath holding spells in a child who is already seriously ill

Breath holding spells can be distinguished from epileptic seizures using the following criteria:

- BHS are provoked by an event or situation.
- BHS seizures are brief.
- Recovery from BHS is rapid.
- The change in skin color and loss of consciousness with BHS occur before any seizure-type jerking.
- With epilepsy, convulsions and muscle weakness precede the loss of skin color.
- An **electroencephalogram** (EEG) that records electrical activity in the brain is normal in all forms of BHS, whereas it may be abnormal with epilepsy.

An electrocardiogram (ECG, EKG) that records the electrical activity in the heart may be used to check for heart rhythm abnormalities, such as long QT syndrome, in children who have had a pallid breath holding spell. Children with long QT syndrome may have breath holding spells in response to exertion or excitement. However, because long QT syndrome is so serious, some physicians recommend that all children with breath holding spells have a baseline EKG.

Treatment

The primary treatment for BHS is to reassure the parent or caregiver that the spells are completely harmless and that they usually disappear by the age of two or two-and-a-half. The child may be put in bed to rest after recovering from the spell. The only treatment for cyanotic BHS is to not encourage or reward the behavior. It is possible that behavior therapy may help a child who suffers from frequent cyanotic spells.

If a child is anemic, iron (at 6 mg per kg [2.2 lb]) of body weight per day for at least three months) may reduce the frequency of breath-holding spells. If pallid breath holding spells are frequent and severe, a preventative anti-cholinergic medicine such as atropine sulfate may be prescribed, in consultation with a neurologist or cardiologist. The dosage is usually 0.1 mg of oral atropine three times daily. Anti-convulsive medications have no effect on breath holding spells.

Prognosis

There are no long-term effects of breath holding spells. Both types of BHS cease without treatment as the child's brain and body develops and matures. The cyanotic form usually peaks at about two years of age and is rare past the age of five. Both types of BHS disappear by the age of four or five in about 50 percent of affected children and in 90 percent of children by the age of six.

Up to 17 percent of children with pallid BHS will experience syncope (fainting spells) as adults, usually in response to fear, injury, or emotional stress. Children with cyanotic episodes are not at a greater risk for syncope as adults.

Prevention

As of 2004 there is no known prevention for pallid breath holding spells since the trigger for such spells is unpredictable. It sometimes may be possible to prevent or interrupt a cyanotic spell by doing the following:

- avoiding situations or events that may lead to **tantrums** or have caused previous breath holding spells

- distracting the child
- intervening in temper tantrums with soothing words and gestures
- encouraging the child to express emotion with words
- placing a cold cloth on the child's face, particularly within the first 15 seconds

Parenting strategies that may help avoid cyanotic BHS include the following:

- ensuring that the child gets plenty of rest, including daytime rest periods and adequate **sleep** at night
- not allowing the child to become too hungry, because hunger can contribute to frustration
- minimizing unnecessary frustration
- avoiding unnecessary discipline
- helping the child to learn other means of expressing anger and frustration
- maintaining a regular daily routine
- maintaining a calm home atmosphere
- allowing the child to make simple choices
- praising accomplishments and good behavior
- helping the child to feel secure
- helping the child to become more independent and self-confident

Parental concerns

Breath holding spells can be extremely frightening for parents, siblings, and caregivers. Families need to be reassured that BHS is not a harmful or dangerous event and that no treatment is needed. It is important that caregivers understand the cause of breath holding spells and the proper response.

During a breath holding spell parents should:

- Protect children from injury and prevent their arms, legs, and head from hitting something hard or sharp.
- Lay children down on their back or side, preferably on a padded surface such as a carpeted floor; this increases blood flow to the brain and helps prevent muscle jerking.
- Check for food in the mouth if the child ate just before a spell. Parents should not try to remove the food; rather the child's head should be turned to one side so that the food can come out on its own.
- Touch and talk to the child.

KEY TERMS

Apnea—The temporary absence of breathing. Sleep apnea consists of repeated episodes of temporary suspension of breathing during sleep.

Cyanosis—A bluish tinge to the skin that can occur when the blood oxygen level drops too low.

Electrocardiagram (ECG, EKG)—A record of the electrical activity of the heart, with each wave being labeled as P, Q, R, S, and T waves. It is often used in the diagnosis of cases of abnormal cardiac rhythm and myocardial damage.

Electroencephalogram (EEG)—A record of the tiny electrical impulses produced by the brain's activity picked up by electrodes placed on the scalp. By measuring characteristic wave patterns, the EEG can help diagnose certain conditions of the brain.

Pallor—Extreme paleness in the color of the skin.

Syncope—A loss of consciousness over a short period of time, caused by a temporary lack of oxygen in the brain.

- Allow children to wake from the spell on their own.
- Time the spell with a watch.

Following a breath holding spell, parents should do the following:

- Acknowledge the child's behavior and emotions.
- Reassure any other children present that everything is okay and it is not their fault.
- Hug the child and walk away.

Parents should NOT do the following:

- overreact
- call 911 or use mouth-to-mouth resuscitation or **cardiopulmonary resuscitation** (CPR)
- place anything in the child's mouth which could cause **choking** or vomiting
- give the child any medications during the episode
- do anything that could reinforce the behavior, including paying undue attention to the child, making a fuss about the episode, or giving in to the child's demands
- try to keep children from all frustration by overprotecting or sheltering them

A parent who cannot watch a child having a breath holding spell without intervening should leave the room.

Parents who have difficulty dealing with a child's frequent breath holding spells may choose to seek counseling.

If a child does not begin breathing on his or her own within one minute, it is not a normal breath holding spell. The parent should call 911 or other emergency services and begin rescue breathing to maintain the child's air passage until help arrives.

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Breech birth

Definition

Breech birth is the delivery of a fetus (unborn baby) in a bottom- or foot-first position. Between 3 to 4 percent of fetuses start labor in the breech position, which is a potentially dangerous situation.

Description

Throughout most of pregnancy the developing fetus is completely free to move around within the uterus. Between 32 and 36 weeks, however, the fetus becomes so large that movement is restricted. It is much harder for the fetus to turn over, so whatever position it has assumed by this point is likely to be the same position that he or she will be in when labor begins.

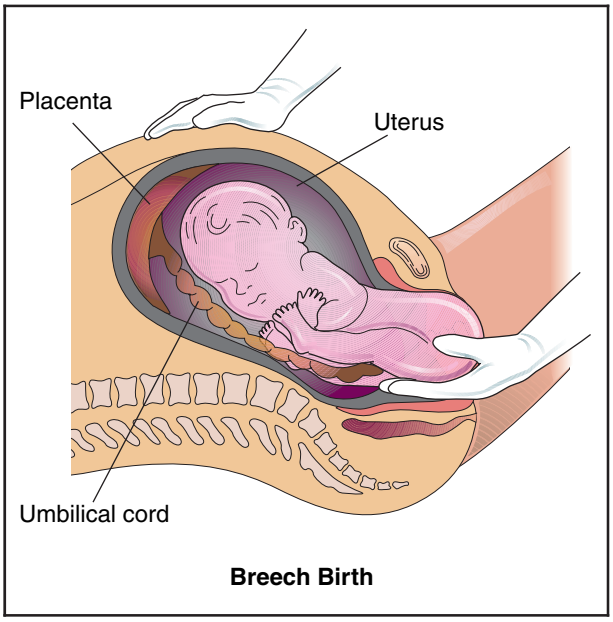
For reasons that are not fully understood, almost all unborn babies settle into a head down, or vertex, position. The fetus is upside down in the uterus, and the head will dilate the cervix (or vaginal opening) and lead the way during the birth process.

Some fetuses, however, present in a breech position. There are three breech positions: frank, complete, and incomplete. In a complete breech, the buttocks lead the way out of the uterus, and the legs are folded in front of the body. A frank breech baby also has his buttocks down, but his legs will stretch straight up with his feet by his head. An incomplete breech, also known as a footling breech, presents with one or both legs down so that the feet drop into the birth canal at delivery.

Of course, many babies are safely delivered from the breech position. There are certain factors that make a breech delivery more likely to be Successful; if ultrasound (a technique that uses sound waves to visualize the fetus) shows that the fetus is in the frank breech position, the fetus's chin is tucked on its chest, and the fetus is not big, it is more likely that an uncomplicated breech delivery is possible.

The biggest part of the fetus's body is usually its head. If the head fits through the mother's pelvis, then the rest of the fetus's body should slip out fairly easily. In addition, when the baby's head comes first, the soft bones of the skull "mold" to the shape of the birth canal during labor (which is what gives newborns that cone-headed appearance). If the fetus is born bottom first, it is possible that the body will fit through the mother's pelvis, but the baby's head will get stuck at the level of the chin. This condition, known as a entrapment, has the potential to cause serious injury to the fetus, and surgical intervention may be required to complete the birth.

There is also a possibility of umbilical cord prolapse with a breech birth. The baby continues to get its oxygen supply from its mother exclusively from the blood in the umbilical cord until the head is delivered and baby breathes on her own. In some cases of breech birth, part of the umbilical cord enters the birth canal before or with the baby's feet or buttocks and pressure on the cord cuts



Approximately 3–4% of babies will start labor in the breech (buttocks first) position. While this is a potentially dangerous situation, many full-term babies can be safely delivered from the breech position. *(Illustration by Electronic Illustrators Group.)*

off the blood and oxygen supply. This situation is known as cord prolapse.

Demographics

Breech presentation occurs in 3 to 4 percent of all births, and up to 95 percent of women with a breech fetus choose **cesarean section** for birth. The earlier a birth occurs in pregnancy, the higher the chances are that the fetus will be in a breech position. Twenty-five percent of premature infants born before 28 weeks are breech.

Causes and symptoms

The cause of breech birth is not known. Women with multiple gestations (i.e., **twins** or more) are more likely to have at least one fetus in a breech position simply due to space constraints in the womb. There are generally no identifiable symptoms of a breech fetus. However, some women may be able to detect the position of the fetus by where they feel the fetus kicking.

Diagnosis

A healthcare provider can often tell the position of the fetus by feeling it through the wall of the mother's abdomen. Another clue to the position is the location where the heartbeat is heard best. If the fetal heartbeat is best heard below the level of the mother's navel, it is likely to be positioned head first. On the other hand, if the heartbeat is best heard above the level of the navel, it is likely to be breech. The most accurate way to determine breech position is using ultrasound.

Treatment

If a fetus is in the breech position in the last weeks of pregnancy, there are three possible courses of action: cesarean section (or c-section), attempted external cephalic version, or vaginal breech delivery.

Some women choose vaginal breech delivery. This should only be attempted if ultrasound shows that the fetus is in a favorable breech position. The frank breech position is the preferred position for successful vaginal breech birth, and the majority of breech fetuses are in this position. Most babies will do very well during a breech delivery, but there is a risk of fetal injury. Some providers may use forceps or a vacuum extraction device to help a breech baby out of the birth canal, a procedure known as assisted breech birth.

During an external cephalic version (also known as version), the obstetrician attempts to turn the fetus to a head first position before labor begins by manipulating the outside of the abdomen. The obstetrician places his or her hands on the mother's abdomen to feel the location of the unborn baby's buttocks and head. The buttocks are lifted up slightly and the doctor pushes on the baby's head to encourage him to perform a sideways somersault. It may take several tries before the fetus cooperates, but about half will eventually turn.

A version should only be done in a hospital, with an ultrasound machine used to guide the obstetrician in turning the fetus. The fetus should be monitored with a fetal monitor before and after the version. The mother is given medication to relax the uterus, minimize discomfort, and prevent premature contractions.

A version is not appropriate for every fetus who is in the breech position at the end of pregnancy. It can only be tried if there is one fetus in the uterus, if the placenta is not lying in front of the fetus, and if the umbilical cord does not appear to be wrapped around the fetus at any point.

Cesarean section is the most common way to deliver a breech baby and is the method recommended by the American College of Gynecology and Obstetrics if a version has failed. A c-section is performed by an obstetrician, who makes an incision in the lower abdomen through which the baby is delivered. Like any surgical procedure, c-section carries a risk of infection and hemorrhage. Postpartum recovery is also longer with

KEY TERMS

Complete breech—A breech position in which the baby is "sitting" bottom first on the cervix with legs crossed.

External cephalic version—Manual manipulation of the abdomen in order to turn a breech baby; also known as version.

Frank breech—A breech position where the baby is bottom first and his legs are extended upward so that his feet are near his head.

Incomplete breech—Also called a footling breech, in this position the baby has one or both feet down towards the pelvis so that his leg(s) are poised to deliver first.

Umbilical cord prolapse—A birth situation in which the umbilical cord, the structure that connects the placenta to the umbilicus of the fetus to deliver oxygen and nutrients, falls out of the uterus and becomes compressed, thus preventing the delivery of oxygen.

Vertex—The top of the head or highest point of the skull.

c-section than with vaginal delivery. However, in difficult breech presentations, or in cases where there are multiple fetuses and one or more are breech, it may be considered the best option for delivery.

Prognosis

Version is successful in turning a breech baby approximately 50 percent of the time. However, some babies who are successfully turned will turn back to the breech position after the procedure is done, particularly if version is attempted too early before the onset of labor.

Manipulations to deliver an entrapped head or stuck shoulder or arm can cause injury to the baby. Both entrapment and cord prolapse can be potentially fatal to an infant if delivery is delayed.

Among breech babies born after the full nine-month term, smaller babies usually do better. The exception to this is premature babies. C-section is generally the delivery mode of choice for premature babies due to the other risks these infants face (such as lung immaturity).

Prevention

There is no way to prevent a fetus from settling into the breech position at the end of pregnancy. A woman who has had one breech fetus is at an increased risk for having another breech fetus in subsequent pregnancies.

See also Cesarean section; Childbirth.

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Paula Ford-Martin

Brittle bone disease *see* **Osteogenesis imperfecta**

Bronchiolitis

Definition

Bronchiolitis is a lung infection that affects children of any age; however, it is much more severe when it occurs in young infants.

Description

The bronchioles are small branches off of the more major bronchi or airway tubes that run through the lungs. When these bronchioles are infected, they become inflamed, and breathing may become difficult.

Bronchiolitis is a particularly important problem in babies who are born prematurely or who have other chronic medical illness. These children are at greatly increased risk of contracting bronchiolitis and of having a more severe course of the illness. Bronchiolitis is the most common reason that babies are hospitalized in the winter. Most cases of bronchiolitis occur between the months of December and May.

Demographics

Every year, 1–2 percent of all babies under 12 months of age require **hospitalization** due to bronchiolitis. At highest risk are boys, premature infants, infants living in urban locations, babies who have not been breastfed, and babies with chronic pulmonary, cardiac, or immune conditions.

Causes and symptoms

Most cases of bronchiolitis are caused by viruses, the most common of which is respiratory syncytial virus. Other common viral causes include parainfluenza, **influenza**, and adenovirus. Like most types of respiratory viruses, the viruses that cause bronchiolitis are usually contracted through breathing in infected droplets that are sprayed out by another ill individual during coughing or sneezing.

Most cases of bronchiolitis start with symptoms of a cold: sneezing, runny nose, fatigue, decreased appetite, **fever**. After two or three days of these symptoms, the bronchiole inflammation becomes severe enough to cause **cough**, wheezing, and rapid breathing.

Severely ill babies or children show signs of difficulty breathing. Their neck muscles and the muscles between their ribs will contract with each effort to breathe, and their chest may cave in as well. Smaller babies may make grunting sounds as they struggle to take in air. Babies will have difficulty nursing or taking bottles and may not be able to feed at all.

When to call the doctor

A doctor should always be called when a child appears to be in any respiratory distress. Fast breathing rates, wheezing, abnormal muscle contractions, or a blue cast to the lips or fingernails should all alert the parent that the child is having difficulty breathing and should be seen immediately by a healthcare provider.

Diagnosis

Initial diagnosis of respiratory distress is made based on clinical signs of difficulty breathing. A pulse oximeter or arterial blood gas measurement reveals the presence of decreased oxygen in the blood. Chest **x rays** may show characteristic patterns of lung involvement. Nasal swabs can be taken in order to identify the causative viral agent, although viral culture takes long enough that the patient is usually on the way to recovery by the time the viral agent has been identified.

KEY TERMS

Bronchiole—Tubes in the lungs that carry air from the bronchi to lung tissues.

Treatment

Treatment at home should consist of **acetaminophen** for fever and comfort (not aspirin, which has been implicated in **Reye's syndrome** in children), increased intake of liquids, and a cool water vaporizer. The utility of **asthma** medications, like bronchodilators, is as of 2004 still undecided.

Children who require hospitalization receive fluids intravenously and supplemental oxygen through a mask or nasal cannulae (small tubes into the openings of the nostrils). Ten percent of all hospitalized infants require mechanical ventilation. Children who are severely ill may be given antiviral medications, such as ribavirin, which is thought to shorten the length of illness and decrease its severity.

Prognosis

Most children recover uneventfully from bronchiolitis, although some studies have suggested that children who have had bronchiolitis may be at higher risk for reactive airway disease throughout the remainder of their lives.

Prevention

Bronchiolitis is spread the same way that most other respiratory viruses are communicated, through droplets and contact with infected nasal secretions. Good hand washing is paramount to prevention, as is keeping children out of public places while they are acutely ill and coughing and sneezing.

Parental concerns

A doctor should always be called when a child appears to be in any respiratory distress. Severe breathing difficulties need immediate medical treatment. Parent should educate their children about good personal hygiene to avoid spreading the germs that cause colds and bronchiolitis.

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Bronchitis

Definition

Bronchitis is an inflammation of the air passages between the nose and the lungs, including the windpipe or trachea and the larger air tubes of the lung that bring air in from the trachea (bronchi). Bronchitis can either be of brief duration (acute) or have a long course (chronic). Acute bronchitis is usually caused by a viral infection but can also be caused by a bacterial infection and can heal without complications. Chronic bronchitis is a sign of serious lung disease that may be slowed but cannot be cured. This form is found almost exclusively in adult smokers. Bronchitis in children is often misdiagnosed as **asthma**.

Description

Acute bronchitis is most prevalent in winter. It is most often caused by a viral infection and may be accompanied by a secondary bacterial infection. Acute bronchitis resolves within two weeks, although the **cough** may persist longer. Acute bronchitis, like any upper airway inflammatory process, can increase a child's likelihood of developing **pneumonia**.

Demographics

Acute bronchitis is one of the more common illnesses affecting **preschool** and school-age children. It is more commonly diagnosed among children under age five than any other age group. It occurs more often in young males. It can occur anytime but is more frequent during the winter months. In otherwise healthy children complications are few.

Causes and symptoms

Acute bronchitis usually begins with the symptoms of a cold, such as a runny nose, sneezing, and dry cough. However, the cough soon becomes deep and painful. Coughing brings up a greenish yellow phlegm or sputum. These symptoms may be accompanied by a **fever** of up to 102°F (38.8°C). Wheezing after coughing is common.

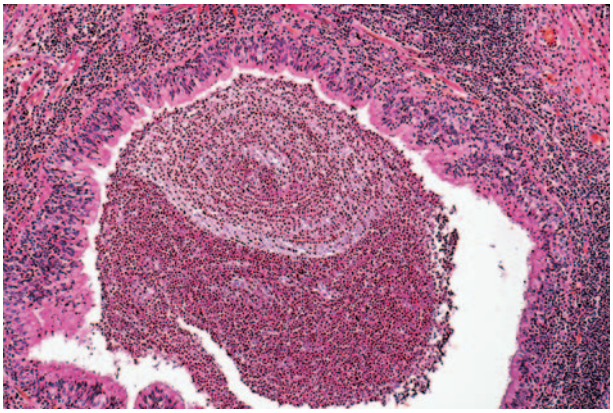
In uncomplicated acute bronchitis, the fever and most other symptoms, except the cough, disappear after three to five days. Coughing may continue for several weeks. Acute bronchitis is often complicated by a bacterial infection, in which case the fever and a general feeling of illness persist. To be cured, the bacterial infection should be treated with **antibiotics**. A cough that does not go away may be a sign of another problem such as asthma or pneumonia.

Physical findings of acute bronchitis vary with the age of the child, and the stage of the disease, but may include the following:

- runny nose
- dry, hacking unproductive cough that may change to a loose cough with increased mucus
- sore throat
- back and other muscle pains
- chills and low grade fever
- headache and general malaise (feeling unwell)

Diagnosis

Initial diagnosis of bronchitis is based on observing the child's symptoms and health history. The physician will listen to the child's chest with a stethoscope for specific sounds that indicate lung inflammation, such as moist rales and crackling, and wheezing, that indicate airway narrowing. Moist rales is a bubbling sound heard



Light micrograph of a five-year-old revealing acute bronchitis, shown by a bronchial tube filled with pus. *(© Gladden Willis, M.D./Visuals Unlimited.)*

with a stethoscope that is caused by fluid secretion in the bronchial tubes.

A sputum culture may be performed, particularly if the sputum is green or has blood in it, to determine whether a bacterial infection is present and to identify the disease-causing organism so that an appropriate antibiotic can be selected. Normally, the patient will be asked to cough deeply then spit the material that comes up from the lungs (sputum) into a cup. This sample is then grown in the laboratory to determine which organisms are present. The results are available in two to three days.

Occasionally, in diagnosing a chronic lung disorder, the sample of sputum is collected using a procedure called a bronchoscopy. In this procedure, the patient is given a local anesthetic, and a tube is passed into the airways to collect a sputum sample.

To better determine what type of obstructive lung disease a patient has, the doctor may do a chest x ray and order blood tests. Other tests may be used to measure how effectively oxygen and carbon dioxide are exchanged in the lungs.

Treatment

When no secondary infection is present, acute bronchitis is treated in the same way as the **common cold**. Home care includes drinking plenty of fluids, resting, not **smoking**, increasing moisture in the air with a cool mist humidifier, and taking **acetaminophen** (Datril, Tylenol, Panadol) for fever and **pain**. Aspirin should not be given to children because of its association with the serious illness **Reye's syndrome**.

Cough suppressants are used only when the cough is dry and produces no sputum. If the patient is coughing up phlegm, the cough should be allowed to continue. The purpose of the cough is to bring up extra mucus and irritants from the lungs. When coughing is suppressed, the mucus accumulates in the plugged airways and can become a breeding ground for pneumonia bacteria.

Expectorant cough medicines, unlike cough suppressants, do not stop the cough. Instead they are used to thin the mucus in the lungs, making it easier to cough up. This type of cough medicine may be helpful to individuals suffering from bronchitis. People who are unsure about what type of medications are in over-the-counter cough syrups should ask their pharmacist for an explanation.

If a secondary bacterial infection is present, the infection is treated with an antibiotic. Patients need to take the entire amount of antibiotic prescribed. Stopping the antibiotic early can lead to a return of the infection. Tetracycline or ampicillin is often used to treat adults. Other possibilities include trimethoprim/sulfamethoxazole (Bactrim or Septra) and the newer erythromycin-like drugs, such as azithromycin (Zithromax) and clarithromycin (Biaxin). Children under age eight are usually given amoxicillin (Amoxil, Pentamox, Sumox, Trimox) because tetracycline discolors permanent teeth that have not yet come in.

For some children with acute bronchitis, doctors may prescribe medicines often used to treat asthma. These medicines can help open the bronchial tubes and clear out mucus. Bronchial dilators are usually given with an inhaler. An inhaler sprays the medicine right into the bronchial tree.

Prognosis

When treated, acute bronchitis normally resolves in one to two weeks without complications, although a cough may continue for several more weeks. The progression of chronic bronchitis, on the other hand, may be slowed, but an initial improvement in symptoms may be achieved.

Prevention

Parents should make sure their children are getting adequate **nutrition** and rest to boost their immunity during cold and flu season. Children should be taught to wash their hands regularly to avoid spreading bacteria and viruses. Other preventative steps include avoiding chemical and environmental irritants, such as air pollution. Immunizations against certain types of pneumonia (as well as **influenza**) are an important preventative measure for the very young or those children with chronic diseases.

KEY TERMS

Acute—Refers to a disease or symptom that has a sudden onset and lasts a relatively short period of time.

Bronchi—Singular, bronchus; the large tubular passages that carry air to the lung and allow air to be expelled from the lungs.

Chronic—Refers to a disease or condition that progresses slowly but persists or recurs over time.

Parental concerns

Parents should encourage fluids by frequent offers of small amounts of the child's favorite liquids. Humidifiers should produce moist air to keep mucus from drying and to make it easier for the child to breathe. The child should be checked for signs of **dehydration**, including daily weights. Acetaminophen is given for temperatures over 101°F (38.3°C). Quiet activity provides a diversion for the sick child.

In caring for a child with acute bronchitis, parents should make the following observations:

- Is there a decrease in coughing and mucus production?
- Does the child have periods of rest and sleep?
- Is the child's intake enough for his or her age?
- Has the child kept a normal body temperature for 24 hours?

Parents should be aware that there is a significant association between high levels of air pollution, smoking, and increased incidence of chronic bronchitis. Air pollutants aggravate chronic pulmonary disease in children and cause decreased pulmonary performance in exercising children and teenagers. Teenagers should be questioned and taught about the ill effects of smoking either tobacco or marijuana. Teenagers should also be questioned about industrial fumes or automobile exhaust exposure at school or work.

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National Heart, Lung, and Blood Institute. PO Box 30105, Bethesda, MD 20824–0105. Web site: <www.nhlbi.nih.gov>.

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Bruises

Definition

Bruises, or ecchymoses, are a discoloration and tenderness of the skin or mucous membranes due to the leakage of blood from an injured blood vessel into the tissues. Purpura refers to bruising as the result of a disease condition. A very small bruise is called a petechia. These often appear as many tiny red dots clustered together and could indicate a serious problem.

Description

Bruises change colors over time in a predictable pattern, so that it is possible to estimate when an injury occurred by the color of the bruise. Initially, a bruise will be reddish, the color of the blood under the skin. After one to two days, the red blood cells begin to break down, and the bruise will darken to a blue or purplish color. This color fades to green at about day six. Around the eighth or ninth day, the skin over the bruised area will have a brown or yellowish appearance, and it will gradually fade back to its normal color.

Long periods of standing cause blood that collects in a bruise to seep through the tissues. Bruises are actually made of little pools of blood, so the blood in one place

may flow toward the ground, and the bruise may appear in another location. For instance, bruising in the back of the abdomen may eventually appear in the groin; bruising in the thigh or the knee will work its way down to the ankle.

Demographics

All persons develop bruises at many times during their lives. The condition is entirely natural and normal.

Causes and symptoms

Healthy people may develop bruises from any injury that does not break through the skin. Vigorous **exercise** may also cause bruises due to bringing about small tears in blood vessels walls. In a condition known as purpura simplex, there is a tendency to bruise easily due to an increased fragility of the blood vessels. Bruises also develop easily in the elderly, because the skin and blood vessels have a tendency to become thinner and more fragile with aging, and there tends to be an increased use of medications that interfere with the blood clotting system. In the condition known as purpura senilis, the elderly develop bruises from minimal contact that may take up to several months to completely heal.

The use of nonsteroidal anti-inflammatories such as ibuprofen and naproxen sodium may lead to increased bruising. Aspirin, **antidepressants**, **asthma** medications, and cortisone medications also have this effect. The anticlotting medications also known as blood thinners, especially the drug warfarin (Coumadin), may be the cause of particularly severe bruising.

Sometimes bruises are linked with more serious illnesses. There are a number of diseases that cause excessive bleeding or bleeding from injuries too slight to have consequences in healthy people. An abnormal tendency to bleed may be due to hereditary bleeding disorders, certain prescription medications, diseases of the blood such as leukemia, and diseases that increase the fragility of blood vessels. If there are large areas of bruising or bruises develop very easily, this may herald a problem. Other causes that should be ruled out include liver disease, **alcoholism**, drug **addiction**, and acquired immune deficiency syndrome (**AIDS**). Bruising that occurs around the navel may indicate dangerous internal bleeding; bruising behind the ear, called Battle's sign, may be due to a skull fracture; and raised bruises may point to autoimmune disease.

When to call the doctor

A physician or healthcare professional should be consulted when accidents involve extensive bruising or when bruises do not heal in a timely manner (seven to 10 days). A physician should be called if bruises appear in unusual locations on the body such as on the back or around the eyes or wrists. Such injuries are often the result of abuse.

Diagnosis

Bruising is usually a minor problem that does not require a formal medical diagnosis. However, faced with extensive bruising, bruising with no apparent cause, or bruising in certain locations, a physician will pursue an evaluation that includes a number of blood tests. If the area of the bruise becomes hard, an x ray may be required.

Treatment

A bruise by itself usually requires no medical treatment. It is often recommended that ice packs be applied on and off during the first 24 hours after injury to reduce the bruising. After that, heat, especially moist heat, is recommended to increase the circulation and the healing of the injured tissues. Rest, elevation of the affected part, and compression with a bandage will also retard the accumulation of blood. Rarely, if a bruise is so large that the body cannot completely absorb it or if the site becomes infected, it may have to be surgically removed.

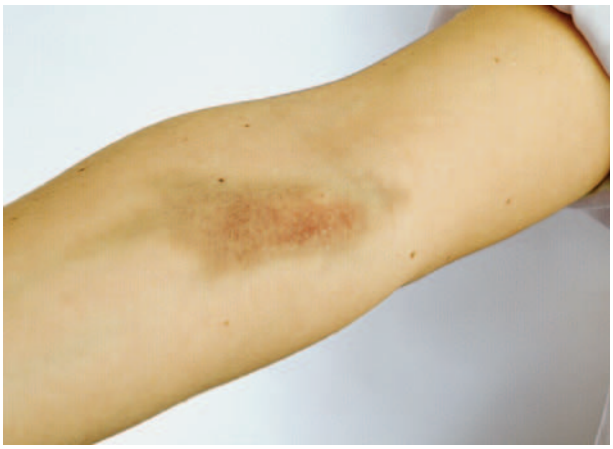
Several types of alternative treatments are often recommended to speed healing and to reduce the **pain** associated with bruises. Most of these treatments are topical in nature and frequently include vitamin K cream can be applied directly to the site of injury. Astringent herbs such as witch hazel, *Hamamelis virginiana*, can be used. This treatment will tighten the tissues and therefore diminish the bruising. The homeopathic remedy, *Arnica montana*, can be applied as a cream or gel to unbroken skin.

Oral homeopathic remedies may reduce bruising, pain, and swelling as well. *Arnica montana*, at 30 ml (1 oz), taken one to two times per day is highly recommended.

Prognosis

The blood under the skin which causes the discoloration of bruising should be totally reabsorbed by the body in three weeks or less. At that time, the skin color should have completely returned to normal.

Sometimes a bruise may become solid and increase in size instead of dissolving. This may indicate blood trapped in the tissues, which may need to be drained. This condition is referred to as a hematoma. Less



Bruised arm of a child. *(© Garo/Photo Researchers, Inc.)*

commonly, the body may develop calcium deposits at the injury site in a process called heterotopic ossification.

Prevention

Vitamin K promotes normal clotting in the blood and, therefore, may help reduce the tendency to bruise easily. Green leafy vegetables, alfalfa, broccoli, seaweed, and fish liver oils are good dietary sources of vitamin K. Other good foods to eat are those containing bioflavonoids, such as reddish-blue berries. These can assist in strengthening the connective tissue, which decreases the spread of blood and bruising. Zinc and vitamin C supplements are also recommended for this purpose.

Nutritional concerns

A balanced diet that includes green leafy vegetables and broccoli should provide a sufficient source of vitamin K. Vitamin C and zinc supplements are also helpful.

Parental concerns

Parents should provide a balanced diet for their children. They should also provide appropriate care for bruises that inevitably occur.

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Ecchymosis—The medical term for a bruise, or skin discoloration caused by blood seeping from broken capillaries under the skin.

Petechia—Plural, petechiae. A tiny purple or red spot on the skin resulting from a hemorrhage under the skin's surface.

Purpura—A group of disorders characterized by purplish or reddish brown areas of discoloration visible through the skin. These areas of discoloration are caused by bleeding from broken capillaries.

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American Academy of Family Physicians. 11400 Tomahawk Creek Parkway, Leawood, KS 66211–2672. Web site: <www.aafp.org/>.

American Academy of Pediatrics. 141 Northwest Point Boulevard, Elk Grove Village, IL 60007–1098. Web site: <www.aap.org/>.

American Academy of Physical Medicine and Rehabilitation. One IBM Plaza, Suite 2500, Chicago, IL 60611–3604. Web site: <www.aapmr.org/>.

American College of Emergency Physicians. PO Box 619911, Dallas, TX 75261–9911. Web site: <www.acep.org/>.

American College of Osteopathic Emergency Physicians. 142 E. Ontario Street, Suite 550, Chicago, IL 60611. Web site: <www.acoep.org/>.

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Bruton's agammaglobulinemia

Definition

Bruton's agammaglobulinemia is a disorder that is present at birth (congenital) and is characterized by low or completely absent levels of immunoglobulins in the bloodstream. Bruton's agammaglobulinemia is also known as X-linked agammaglobulinemia (XLA).

Description

Children with XLA have very low, or completely absent, levels of immumoglobulins in their blood. Immunoglobulins are protein molecules in blood serum that function like antibodies. Without them, the body lacks a fully functioning immune system. Individuals with XLA are vulnerable to repeated, potentially fatal, bacterial infections.

Although persons with XLA carry the genes to produce immunoglobulins, a genetic defect on the X chromosome prevents their formation. This defect is not associated with the immunoglobulins themselves, but rather with the B cells in the bloodstream that ordinarily secrete the immunoglobulins.

B cells are a type of white blood cell. They are the sole producers of immunoglobulins in the body. B cells are produced in the bone marrow and carried to the spleen, lymph nodes, and other organs as they mature. The maturation process depends on an enzyme called Bruton's agammaglobulinemia tyrosine kinase (Btk). If Btk is missing or defective, the B cells cannot mature and cannot produce immunoglobulins.

The gene that controls the production of Btk is on the X chromosome. Certain changes (mutations) in this gene result in defective Btk. Males have one X and one Y chromosome (XY). Females have two X chromosomes (XX). The mother passes one of her two X chromosomes down to her child, and the father passes either an X or a Y chromosome to the child. The mutated gene that produces XLA is a recessive gene. This means that as long as one good copy is present, the disease will not occur. Boys only have one copy of the gene, because they only have one X chromosome. Girls have two copies of the gene. This means that for boys to have XLA they must only inherit one copy of the defective gene, but for girls to have the disease they have to inherit two copies, one from each parent. This is why diseases associated with X linked genes are usually much more common in boys than in girls. To date, no cases of XLA in girls have actually been reported.

Demographics

XLA occurs in one in every 50,000 to one in every 100,000 newborns. Males are overwhelmingly more likely to have it than girls. Children who have an affected relative are more likely to be at risk, because the defect causing the disorder is inherited.

Causes and symptoms

XLA is caused by a defect in the gene that controls the production of the enzyme Btk. This defect blocks B cells from maturing. Only mature B cells produce immunoglobulins. Because other portions of the immune system are functional, people with XLA can fight off some types of infection, such as fungal and most viral infections. Immunoglobulins, however, are vital for combating bacterial infections.

Infants with XLA usually do not show symptoms of the disorder during the first six months of life, because immunoglobulins from their mothers are circulating in their bloodstreams. Over time, their immunoglobulin levels begin to decrease because they cannot successfully produce their own. As the immunoglobulin levels decrease, the baby becomes increasingly vulnerable to bacterial infections.

Common symptoms of immunoglobulin deficiency usually appear after the infant is six months old. They include frequent ear and sinus infections, **pneumonia**, and **gastroenteritis**. Certain viruses, such as hepatitis and **polio** viruses, can also pose a threat. Children with XLA often have small tonsils and lymph nodes and may develop chronic skin infections. Approximately 20 percent of these children develop arthritis, possibly as a result of joint infections.

KEY TERMS

Antibody—A special protein made by the body's immune system as a defense against foreign material (bacteria, viruses, etc.) that enters the body. It is uniquely designed to attack and neutralize the specific antigen that triggered the immune response.

B cell—A type of white blood cell derived from bone marrow. B cells are sometimes called B lymphocytes. They secrete antibodies and have a number of other complex functions within the human immune system.

Bruton's agammaglobulinemia tyrosine kinase (Btk)—An enzyme vital for the maturation of B cells.

Carrier—A person who possesses a gene for an abnormal trait without showing signs of the disorder. The person may pass the abnormal gene on to offspring. Also refers to a person who has a particular disease agent present within his/her body, and can pass this agent on to others, but who displays no symptoms of infection.

Enzyme—A protein that catalyzes a biochemical reaction without changing its own structure or function.

Immunoglobulin G (IgG)— Immunoglobulin type gamma, the most common type found in the blood and tissue fluids.

Mutation—A permanent change in the genetic material that may alter a trait or characteristic of an individual, or manifest as disease. This change can be transmitted to offspring.

X chromosome—One of the two sex chromosomes (the other is Y) that determine a person's gender. Normal males have both an X and a Y chromosome, and normal females have two X chromosomes.

When to call the doctor

If a child has had many more infections, especially serious infections, than is normal for a child of his or her age there may be an immune system problem such as XLA and a doctor should be consulted.

Diagnosis

Frequent bacterial infections, a lack of mature B cells, and low-to-nonexistent levels of immunoglobulins point to a diagnosis of XLA. A sample of the child's blood serum can be analyzed for the presence of immunoglobulins by a technique called immunoelectrophoresis. To make a definitive diagnosis, the child's X chromosome is analyzed for defects in the Btk gene. Similar analysis can be used for prenatal diagnosis or to detect carriers of the defective gene.

Treatment

Treatment of XLA consists of regular intravenous doses of commercially prepared gamma globulin (sold under the trade names Gamimune or Gammagard) to ward off infections. **Antibiotics** are used to treat infections as they occur. Children with XLA must be treated promptly for even minor cuts and scrapes and taught to avoid crowds and people with active infections.

Prognosis

Prior to the era of gamma globulin and antibiotic treatment, approximately 90 percent of XLA individuals died before the age of eight. Early diagnosis and therapy in the early 2000s allows most individuals with XLA to reach adulthood and lead relatively normal lives. Infants who develop polio or persistent viral infections, however, have a poorer prognosis.

Prevention

There is no known way to prevent XLA. However, if an individual believes a **family** member may have XLA, it is possible to get genetic counseling prior to pregnancy to determine if the individual is a carrier of the gene. If the person is a carrier various options can be discussed.

Parental concerns

Children with XLA can have normal lives. Most medical care can be managed on an outpatient basis or even by home care. Special attention needs to be paid to the beginning of infections so that they can be treated promptly, but in general, children should be encouraged to participate in normal activities such as school and **play**. Usually, children with XLA are not given any vaccines containing live viruses (such as **measles**, **mumps**, or polio) because there is a small but dangerous risk that the child will actually get the disease that the vaccine was intended to prevent.

Resources

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Immune Deficiency Foundation. 40 W. Chesapeake Ave Suite 308 Towson, MD 21204. Web site: <www.primaryimmune.org>.

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Bulimia nervosa

Definition

Bulimia nervosa is a serious and sometimes life-threatening eating disorder affecting mainly young women. People with bulimia, known as bulimics, consume large amounts of food (binge) and then try to rid themselves of the food and calories (purge) by fasting, excessive **exercise**, **vomiting**, or using **laxatives**. The behavior often serves to reduce stress and relieve **anxiety**. Because bulimia results from an excessive concern with weight control and self-image and is often accompanied by depression, it is also considered to be a psychiatric illness.

Description

Bulimia nervosa is a serious health problem for over 2 million adolescent girls and young women in the United States. The bingeing and purging activity associated with this disorder can cause severe damage, even death, although the risk of death is not as high as for **anorexia nervosa**, an eating disorder that leads to excessive weight loss.

In rare instances, binge eating may cause the stomach to rupture. In the case of purging, heart failure can result due to loss of vital **minerals** such as potassium. Vomiting causes other serious problems, including acid-related scarring of the fingers (if used to induce vomiting) and damage to tooth enamel. In addition, the tube that delivers food from the mouth to the stomach (the esophagus) often becomes inflamed, and salivary glands can become swollen. Irregular menstrual periods can also result, and interest in sex may diminish.

Most bulimics find it difficult to stop their behavior without professional help. Many typically recognize that the behavior is not normal, but they feel helpless to control it. Some bulimics struggle with other compulsive, risky behaviors such as drug and alcohol abuse. Many also suffer from other psychiatric illnesses, including clinical depression, anxiety, and **obsessive-compulsive disorder** (OCD).

Bulimic behavior is often carried out in secrecy, accompanied by feelings of guilt or shame. Outwardly, many people with bulimia appear healthy and successful, while inside they have feelings of helplessness and low **self-esteem**.

Demographics

Most bulimics are females in their teens to early 20s. Males account for only 5 to 10 percent of all cases. People of all races develop the disorder, but most of those diagnosed are white.

Causes and symptoms

The cause of bulimia is as of 2004 unknown. Researchers believe that it may be caused by a combination of genetic and environmental factors. Bulimia tends to run in families. Research shows that certain brain chemicals, known as neurotransmitters, may function abnormally in acutely ill people with bulimia nervosa. Scientists also believe there may be a link between bulimia and other psychiatric problems, such as depression and OCD. Environmental influences include participation in work or **sports** that emphasize thinness, such as modeling, dancing, or gymnastics. **Family** pressures also may play a role. One study found that mothers who are extremely concerned about their daughters' physical attractiveness and weight may in part cause bulimia in them. In addition, girls with eating disorders tend to have fathers and brothers who criticize their weight. Bulimia tends to run in families.

According to the American Anorexia/Bulimia Association Inc., warning signs of bulimia include the following:

- eating large amounts of food uncontrollably (bingeing)
- vomiting, abusing laxatives or diuretics, or engaging in fasting, dieting, or vigorous exercise (purging)
- preoccupation with body weight
- using the bathroom frequently after meals
- depression or mood swings
- irregular menstrual periods

- onset of dental problems, swollen cheeks or glands, heartburn, or bloating

When to call the doctor

A healthcare professional should be consulted at the first sign of behaviors associated with bulimia.

Diagnosis

Bulimia is treated most successfully when diagnosed early. However, because the bulimic may deny there is a problem, getting medical help is often delayed. A complete physical examination in order to rule out other illnesses is the first step to diagnosis.

According to the American Psychiatric Association, a diagnosis of bulimia requires that a person have all of the following symptoms:

- recurrent episodes of binge eating (minimum average of two binge-eating episodes a week for at least three months)
- a feeling of lack of control over eating during the binges
- regular use of one or more of the following to prevent weight gain: self-induced vomiting, use of laxatives or diuretics, strict dieting or fasting, or vigorous exercise
- persistent over-concern with body shape and weight

Bulimia nervosa

Criteria

1. Recurrent episodes of binge eating, which is characterized by 1) consumption of an amount of food that is definitely larger than most people would eat during a similar period of time and under similar circumstances AND 2) a sense of lack of control over eating during the episode.
2. Recurrent inappropriate compensatory behavior in order to prevent weight gain, such as self-induced vomiting; misuse of laxatives, diuretics, or enemas, or other medications; or fasting or excessive exercise.
3. Binge eating and compensatory behaviors both occur an average of twice a week for three months.
4. Self-evaluation is unduly influenced by body shape and weight.
5. The disturbance does not occur exclusively during episodes of anorexia nervosa.

Purging type: Regular episodes of self-induced vomiting or misuse of laxatives, diuretics, or enemas.

Nonpurging type: No regular episodes of self-induced vomiting or misuse of laxatives, diuretics, or enemas. Patient uses fasting or excessive exercise to avoid weight gain.

SOURCE: *Diagnostic and Statistical Manual of Mental Disorders IV.*

(Table by GGS Information Services.)

Treatment

Early treatment is important; otherwise, bulimia may become chronic, with serious health consequences. A comprehensive treatment plan is called for in order to address the complex interaction of physical and psychological problems in bulimia. A combination of drug and behavioral therapies is commonly used.

Behavioral approaches include individual psychotherapy, group therapy, and **family therapy**. Cognitive-behavioral therapy, which teaches people how to change abnormal thoughts and behavior, is also used. **Nutrition** counseling and self-help groups are often helpful.

Antidepressants commonly used to treat bulimia include desipramine (Norpramin), imipramine (Tofranil), and fluoxetine (Prozac). These medications also may treat any co-existing depression.

In addition to professional treatment, family support plays an important role in helping the bulimic person. Encouragement and emotional support may convince the sick person to get help, stay with treatment, or try again after a failure. Family members can help locate resources, such as eating disorder clinics in local hospitals or treatment programs in colleges designed for students.

Light therapy—exposure to bright, artificial light—may be useful in reducing bulimic episodes, especially during the dark winter months. Some feel that massage may prove helpful, putting people in touch with the reality of their own bodies and correcting misconceptions of body image. Hypnotherapy may help resolve unconscious issues that contribute to bulimic behavior.

Prognosis

Bulimia may become chronic and lead to serious health problems, including seizures, irregular heartbeat, and thin bones. In rare cases, it may be fatal. Timely therapy and medication can effectively manage the disorder and help the bulimic live a normal, productive, and fulfilling life.

Prevention

There is as of 2004 no known method for preventing bulimia. However, parents can promote healthy eating habits in their children and encourage them to embrace realistic, rather than overly thin, body images.

KEY TERMS

Binge—A pattern of eating marked by episodes of rapid consumption of large amounts of food; usually food that is high in calories.

Diuretics—A group of drugs that helps remove excess water from the body by increasing the amount lost by urination.

Neurotransmitters—Chemicals in the brain that transmit nerve impulses.

Obsessive-compulsive disorder—An anxiety disorder marked by the recurrence of intrusive or disturbing thoughts, impulses, images, or ideas (obsessions) accompanied by repeated attempts to supress these thoughts through the performance of certain irrational and ritualistic behaviors or mental acts (compulsions).

Purge—To rid the body of food and calories, commonly by vomiting or using laxatives.

Nutritional concerns

Abnormal food intake and purging may result in abnormal nutrition. Purging may lead to a loss of potassium and other essential metabolic ions. These can become life threatening.

Parental concerns

Parental remarks about body size and shape often trigger bulimia. Parents of bulimics must be supportive and participate in treatment if the condition is to be successfully treated.

See also Binge eating disorder.

Resources

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American Academy of Pediatrics. 141 Northwest Point Boulevard, Elk Grove Village, IL 60007–1098. Web site: <www.aap.org/default.htm>.

American Psychiatric Association. 1400 K Street NW, Washington, DC 20005. Web site: <www.psych.org/>.

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Bullies

Definition

Bullies are aggressive children who repeatedly physically or emotionally abuse, torment, or victimize smaller, weaker, or younger children.

Description

Bullying usually involves an older or larger child or children victimizing a single child who is unable to defend himself or herself. Bullying is generally viewed as a form of harassment committed by a child or children who are older, stronger, or otherwise more powerful socially, upon weaker adolescents. Often, the power of the bully is dependent on the perception of the victim, with the bullied child often too intimidated to effectively resist the bully.

Although the stereotypical bully is male, girls engage in bullying behavior almost as often as boys. Their tactics differ, however, in that they are less visible. Boys who are bullies tend to resort to one-on-one physical aggression, while girls tend to bully as a group through social exclusion and the spreading of rumors. Girls who would never bully individually will often take part in group bullying activities.

Bullying begins at a very early age; it is not uncommon to find bullies in **preschool**. Until about age seven, bullies appear to choose their victims at random. After that, they single out specific children to torment on a regular basis. Nearly twice as much bullying goes on in grades two to four as in grades six to eight, and, as bullies grow older, they tend to use less physical abuse and more verbal abuse.

Bullies are often popular among their peers until about sixth grade. They average two or three friends, and other children seem to admire them for their physical toughness. By high school, however, their social acceptance diminishes to the point that their only "friends" are other bullies. Despite their unpopularity, bullies have relatively high **self-esteem**, perhaps because they process social information inaccurately.

For example, bullies attribute hostile intentions to people around them and therefore perceive provocation where it does not exist. "What are you staring at?" is a common opening line of bullies. For the bully, these perceived slights serve as justification for **aggressive behavior**.

Children who become the targets of bullies generally have a negative view of violence and go out of their way to avoid conflict. They tend to be "loners" who exhibit signs of vulnerability before being singled out by a bully. Being victimized leads these children, who already may lack self-esteem, to feel more anxious, thereby increasing their vulnerability to further bullying. Being the target of a bully leads to social isolation and rejection by peers, and victims tend to internalize others' negative views, further eroding their self-esteem. Although bullying actually lessens during **adolescence**, this is the period when peer rejection is most painful for victims.

Sometimes the victims of bullies are larger, stronger, or older than the bully but allow the bullying to continue because they are intimidated, do not believe in violence, or are taught non-violence by their parents.

Studies show that students who are gay or bisexual or are perceived as gay or bisexual experience an extremely high rate of bullying, not only by other students, but often by teachers and other school personnel. Also, bullying against gay and bisexual students is often ignored or sometimes encouraged by homophobic school staff members.

According to the American School Health Association, students who discover they are gay or bisexual often experience rejection, discrimination, isolation, and violence, and this fact makes it all the more important for teachers and administrators to be supportive and sensitive to them. Schools are obligated under federal law to protect students from discrimination and harassment, from other students as well as teachers and all other school employees. In 1996, a federal appeals court ruled that school officials can be held liable under the Equal Protection Clause of the U.S. Constitution for not protecting gay and bisexual students from harassment and discrimination. The ruling still stood as of 2004.

Extensive long-term research indicates that bullying is not a phase a child outgrows. In a study of more than 500 children, University of Michigan researchers discovered that children who were viewed as the most aggressive by their peers at age eight grew up to commit increasingly more serious crimes as adults. Other studies indicate that, as adults, bullies are far more likely to abuse their spouses and children.

Modern schools tend to discourage bullying with programs designed to teach students cooperation and train peers in bullying intervention techniques. However, some schools have a zero tolerance for violence so if two students are discovered in a fight, both are disciplined, often by suspension, even though one may be the bully and the other the victim.

Experts say that school violence often is rooted in bullying. While bullying is often verbal threats and

harassment, it can get out of control and turn into violence, including the use of weapons.

Researchers who have studied bullying have concluded that up to 15 percent of children say they are regularly bullied, and it occurs most frequently at school in areas where there is inadequate or no adult supervision, such as the playground, hallways, cafeteria, and in classrooms before lessons start. Bullying usually starts in elementary school, peaks in middle school, and drops in high school. It does not disappear, however. Although boys are more often the perpetrators and victims of bullying, girls tend to bully in indirect ways, such as manipulating friendships, ostracizing classmates, and spreading malicious rumors. Boys who are regularly bullied tend to be more passive and physically weaker than the bullies. In middle school, girls who mature early are commonly victims of bullying, according to some findings.

Preschool

Bullying behavior can be seen as early as preschool. However, little data exists regarding the prevalence of bullying in preschool. Preschool-age children may bully others to get attention, show off, or to get another child's possessions, such as **toys**, clothing, or use of playground equipment. They may also be jealous of the children they are bullying or may be getting bullied themselves. Preschool bullying usually begins with name-calling and can escalate into physical violence if left unchecked. Preschool teachers are urged to intervene immediately to stop bullying and to teach acceptable behavior. If teachers or staff at a preschool do not do enough to stop bullying, parents should find another preschool.

School age

A 2001 report by the National Institute of Child Health and Human Development (NICHD) found that 17 percent of the respondents had been bullied sometimes or weekly; 19 percent had bullied others sometimes or weekly, and 6 percent had both bullied others and been bullied. The researchers estimated that 1.6 million children in grades six through 10 in the United States are bullied at least once a week and 1.7 million children bully others as frequently.

The survey, the first nationwide research on the problem in this country, questioned 15,686 public and private school students, grades six through 10, on their experiences with bullying. In a study of 6,500 middle school students in rural South Carolina, 23 percent said they had been bullied regularly during the previous three months, and 20 percent admitted bullying another child regularly during the same time.

Bullying appears to be rapidly increasing, according to statistics from the U.S. Department of Justice. Among sixth-grade students, rates of bullying rose from 10.5 percent in 1999 to 14.3 percent in 2001; among eighth-grade students victimization by bullies went from 5.5 percent in 1999 to 9.2 percent in 2001. In the tenth grade, bullying rose from 3.2 percent in 1999 to 4.6 percent in 2001, and among twelfth graders, it doubled from 1.2 percent in 1999 to 2.4 percent just two years later.

A bully's behavior does not exist in isolation. Rather, it may indicate the beginning of a generally antisocial and rule-breaking behavior pattern that can extend into adulthood. Programs to address the problem, therefore, must reduce opportunities and rewards for bullying behavior. The Olweus Bullying Prevention Program, developed, refined, and systematically evaluated in Norway in the mid-1980s, is the best-known initiative designed to reduce bullying among elementary, middle, and junior high school children. The strategy behind the program is to involve school staff, students, and parents in efforts to designed to develop awareness about bullying, improve peer relations, intervene to stop intimidation, develop clear rules against bullying behavior, and support and protect victims.

The program intervenes on three levels:

- School: Faculty and staff survey students anonymously to determine the nature and prevalence of the school's bullying problem, increase supervision of students during breaks, and conduct school-wide assemblies to discuss the issue. Teachers receive in-service training on how to implement the program.
- Classroom: Teachers and other school personnel introduce and enforce classroom rules against bullying, hold regular classroom meetings with students to discuss bullying, and meet with parents to encourage their participation.
- Individual: Staff intervention with bullies, victims, and their parents to ensures that the bullying stops.

The Bergen research showed that the program was highly effective among students in elementary, middle, and junior high schools: Bullying dropped by 50 percent or more during the program's two years. Behavior changes were more pronounced the longer the program was in effect. The school climate improved, and the rate of **antisocial behavior**, such as theft, vandalism, and **truancy**, declined during the two-year period.

Common problems

The NICHD study found that bullying has long-term and short-term psychological effects on both those who

bully and those who are bullied. Victims experience loneliness and report having trouble making social and emotional adjustments, difficulty making friends, and poor relationships with classmates. Victims of bullying often suffer humiliation, insecurity, and a loss of self-esteem, and often develop a **fear** of going to school. The impact of frequent bullying often accompanies these victims into adulthood; they are at greater risk of suffering from depression and other mental health problems, including **schizophrenia**. In rare cases, they commit **suicide**.

Bullying behavior has been linked to other forms of antisocial behavior, such as vandalism, shoplifting, skipping and dropping out of school, fighting, and using alcohol and other drugs. Research suggests that bullying can lead to criminal behavior later in life: 60 percent of males who were bullies in grades six through nine were convicted of at least one crime as adults, compared with 23 percent of males who did not bully; 35 to 40 percent of these former bullies had three or more convictions by age 24, compared with 10 percent of those who did not bully.

The NICHD study found that those who bully and are bullied appear to be at greatest risk of experiencing the following: loneliness, trouble making friends, lack of success in school, and involvement in problem behaviors such as **smoking**, illegal drug use, and drinking.

Parental concerns

Parents should be aware of common signs that a child is being bullied. These include trouble sleeping, bedwetting, stomachaches, headaches, lack of appetite, fear of going to school, crying before or after school, lack of interest in social events, low self-esteem, unexplained loss of personal items and money, unexplained **bruises** and injuries, and **acting out** aggressively at home.

Parents should teach their children proper **communication skills** that they may need to seek assistance if they are being bullied, according to the Web site <www.bullying.org>. Other advice for parents from the Web site include:

- Be involved with the child's school and talk to other parents about the problem.
- Meet with school officials and make sure the school has an anti-bullying policy and that it is strictly enforced. If a child is being bullied, meet with school officials to find out what they are doing about it. If no action is being taken, demand that it be done.

KEY TERMS

Antisocial behavior—Behavior characterized by high levels of anger, aggression, manipulation, or violence.

Harassment—The persistent annoying, attacking, or bothering of another person.

Schizophrenia—A severe mental illness in which a person has difficulty distinguishing what is real from what is not real. It is often characterized by hallucinations, delusions, and withdrawal from people and social activities.

- Talk to the child's teacher or teachers to determine if they have seen any bullying problems in the classroom or playground.
- Talk to a school counselor and ask that person to discuss bullying with children.
- Report all verbal or physical threats against a child to school authorities and insist they take action. If they do not take action, report the problem to local police.

When to call the doctor

Bullying is violence. If both bullies and their victims are not offered help, there can be serious long-term consequences for both. Bullies and their victims may need professional counseling or psychological help. Parents should seek immediate help for children who are depressed or suicidal. Parents of bullies also need to seek psychological help for their child if the bullying continues for even a short period of time.

See also Antisocial behavior.

Resources

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Bullying *see* **Bullies**

Burns

Definition

Burns are injuries to tissues that are caused by heat, friction, electricity, radiation, or chemicals.

Description

Burns are characterized by degree, based on the severity of the tissue damage. A first-degree burn causes redness and swelling in the outermost layers of skin (epidermis). A second-degree burn involves redness, swelling and blistering, and the damage may extend beneath the epidermis to deeper layers of skin (dermis). A third-degree burn, also called a full-thickness burn, destroys the entire depth of skin, causing significant scarring. Damage also may extend to the underlying fat, muscle, or bone.

Demographics

The severity of the burn is also judged by the amount of body surface area (BSA) involved. Healthcare workers use the "rule of nines" to determine the percentage of BSA affected in people more than 9 years of age: each arm with its hand is 9 percent of BSA; each leg with its foot is 18 percent; the front of the torso is 18 percent; the back of the torso, including the buttocks, is 18 percent; the head and neck are 9 percent; and the genital area (perineum) is 1 percent. This rule cannot be applied to a young child's body proportions, so BSA is estimated using the palm of a person's hand as a measure of 1 percent area.

The severity of the burn determines the type of treatment and also where the burned person should receive treatment. Minor burns may be treated at home or in a doctor's office. These are defined as first- or second-degree burns covering less than 15 percent of an adult's body or less than 10 percent of a child's body, or a third-degree burn on less than 2 percent BSA. Moderate burns should be treated at a hospital. These are defined as first- or second-degree burns covering 15 percent to 25 percent of an adult's body or 10 percent to 20 percent of a child's body, or a third-degree burn on 2 percent to 10 percent BSA. Critical, or major, burns are the most serious and should be treated in a specialized burn unit of a hospital. These are defined as first- or second-degree burns covering more than 25 percent of an adult's body or more than 20 percent of a child's body, or a third-degree burn on more than 10 percent BSA. In addition, burns involving the hands, feet, face, eyes, ears, or genitals are considered critical. Other factors influence the level of treatment needed, including associated injuries such as bone **fractures** and **smoke inhalation**, presence of a chronic disease, or a history of abuse. Also, children and the elderly are more vulnerable to complications from burn injuries and require more intensive care.

Causes and symptoms

Burns may be caused by even a brief encounter with heat greater than 120°F (49°C). The source of this heat may be the sun (causing a **sunburn**), hot liquids, steam, fire, electricity, friction (causing rug burns and rope burns), and chemicals (causing caustic burn upon contact).

Signs of a burn are localized redness, swelling, and **pain**. A severe burn will also blister. The skin may also

peel, appear white or charred, and feel numb. A burn may trigger a **headache** and **fever**. Extensive burns may induce shock, the symptoms of which are faintness, weakness, rapid pulse and breathing, pale and clammy skin, and bluish lips and fingernails.

When to call the doctor

A physician or healthcare professional should be consulted whenever first or second degree burns cover more than 15 percent of a person's body surface area (BSA) or third degree burns involve more than 2 percent of a victim's BSA.

Diagnosis

A physician will diagnose a burn based on visual examination and will also ask the burned person or **family** members questions to determine the best treatment. He or she may also check for smoke inhalation, **carbon monoxide poisoning**, cyanide **poisoning**, other event-related trauma, or, if suspected, evidence of **child abuse**.

Treatment

Burn treatment consists of relieving pain, preventing infection, and maintaining body fluids, electrolytes, and calorie intake while the body heals. Treatment of chemical or electrical burns is slightly different from the treatment of thermal burns but the objectives are the same.

Thermal burn treatment

The first act of thermal burn treatment is to stop the burning process. This may be accomplished by letting cool water run over the burned area or by soaking it in cool (not cold) water. Ice should never be applied to a burn. Cool (not cold) wet compresses may provide some pain relief when applied to small areas of first- and second-degree burns. Butter, shortening, or similar salve should never be applied to the burn because these prevent heat from escaping and drive the burning process deeper into the skin.

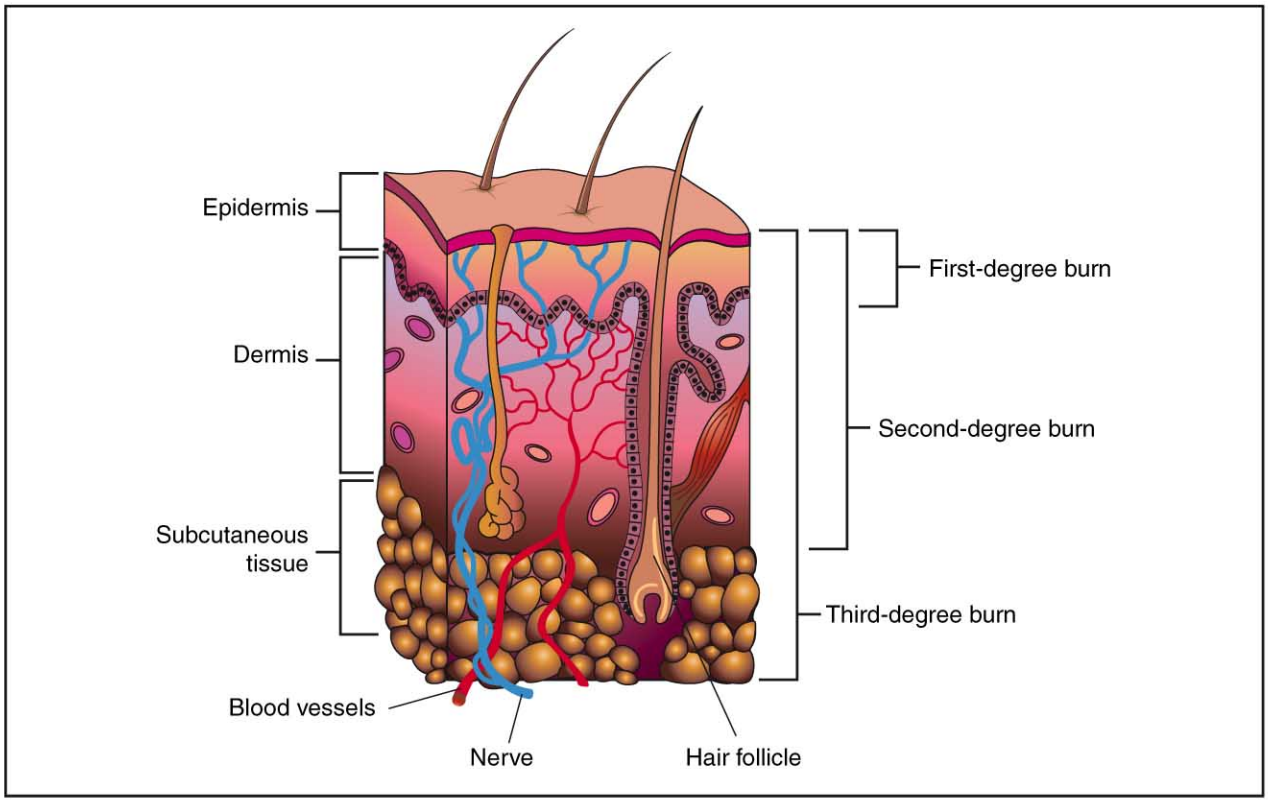
If the burn is minor, it may be cleaned gently with soap and water. Blisters should not be broken. If the skin of the burned area is unbroken and it is not likely to be further irritated by pressure or friction, the burn should be left exposed to the air to promote healing. If the skin is broken or apt to be disturbed, the burned area should be coated lightly with an antibacterial ointment and covered with a sterile bandage. Aspirin, **acetaminophen**, or ibuprofen may be taken to ease pain and relieve inflammation. A doctor should be consulted if these signs of infection appear: increased warmth, redness, pain, or swelling; pus or similar drainage from the wound; swollen lymph nodes; or red streaks spreading away from the burn.

In situations in which a person has received moderate or critical burns, lifesaving measures take precedence over burn treatment, and emergency medical assistance must be called. A person with serious burns may stop breathing, and artificial respiration (also called mouth-to-mouth resuscitation or rescue breathing) should be administered immediately. Also, a person with burns covering more than 12 percent BSA is likely to go into shock; this condition may be prevented by laying the person flat and elevating the feet about 12 inches (30 cm). Burned arms and hands should also be raised higher than the person's heart.

In rescues, a blanket may be used to smother any flames as the person is removed from danger. The person whose clothing is on fire should "stop, drop, and roll" or be assisted in lying flat on the ground and rolling to put out the fire. Afterwards, only burned clothing that comes off easily should be removed; any clothing embedded in the burn should not be disturbed. Removing any smoldering apparel and covering the person with a light, cool, wet cloth, such as a sheet but not a blanket or towel, will stop the burning process.

At the hospital, the staff provide further medical treatment. A tube to aid breathing may be inserted if the person's airways or lungs have been damaged, as can happen during an explosion or a fire in an enclosed space. Also, because burns dramatically deplete the body of fluids, replacement fluids are administered intravenously. The person is also given **antibiotics** intravenously to prevent infection, and he or she may also receive a **tetanus** shot, depending on his or her immunization history. Once the burned area is cleaned and treated with antibiotic cream or ointment, it is covered in sterile bandages, which are changed two to three times a day. Surgical removal of dead tissue (debridement) also takes place. As the burns heal, thick, taut scabs (eschar) form, which the doctor may have to cut to improve blood flow to the more elastic healthy tissue beneath. The person will also undergo physical and occupational therapy to keep the burned areas from becoming inflexible and to minimize scarring.

In cases where the skin has been so damaged that it cannot properly heal, a skin graft is usually performed. A skin graft involves taking a piece of skin from an unburned portion of the person's body (autograft) and transplanting it to the burned area. When doctors cannot immediately use the individual's own skin, a temporary graft is performed using the skin of a human donor (allograft), either alive or dead, or the skin of an animal (xenograft), usually that of a pig.



There are three classifications of burns: first-degree, second-degree, and third-degree burns. *(Illustration by Electronic Illustrators Group.)*

The burn victim also may be placed in a hyperbaric chamber, if one is available. In a hyperbaric chamber (which can be a specialized room or enclosed space), the person is exposed to pure oxygen under high pressure, which can aid in healing. However, for this therapy to be effective, the burned individual must be placed in a chamber within 24 hours of being burned.

Chemical burn treatment

Burns from liquid chemicals must be rinsed with cool water for at least 15 minutes to stop the burning process. Any burn to the eye must be similarly flushed with water. In cases of burns from dry chemicals such as lime, the powder should be completely brushed away before the area is washed. Any clothing which may have absorbed the chemical should be removed. The burn should then be loosely covered with a sterile gauze pad and the person taken to the hospital for further treatment. A physician may be able to neutralize the offending chemical with another before treating the burn like a thermal burn of similar severity.

Electrical burn treatment

Before electrical burns are treated at the site of the accident, the power source must be disconnected if possible and the victim moved away from it to keep the person giving aid from being electrocuted. Lifesaving measures again take priority over burn treatment, so breathing must be checked and assisted if necessary. Electrical burns should be loosely covered with sterile gauze pads and the person taken to the hospital for further treatment.

Alternative treatment

In addition to the excellent treatment of burns provided by traditional medicine, some alternative approaches may be helpful as well. (Major burns should always be treated by a medical practitioner.) The homeopathic remedies *Cantharis* and *Causticum* can assist in burn healing. A number of botanical remedies, applied topically, can also help burns heal. These include aloe (*Aloe barbadensis*), oil of St. John's wort (*Hypericum perforatum*), calendula (*Calendula officinalis*), comfrey (*Symphytum officinale*), and tea tree oil (*Melaleuca* spp.). Supplementing the diet with vitamin C, vitamin E, and zinc also is beneficial for wound healing.

Prognosis

The prognosis is dependent upon the degree of the burn, the amount of body surface covered, whether

critical body parts were affected, any additional injuries or complications like infection, and the promptness of medical treatment. Minor burns may heal in five to ten days with no scarring. Moderate burns may heal in ten to 14 days and may leave scarring. Critical or major burns take more than 14 days to heal and leave significant scarring. Scar tissue may limit mobility and functionality, but physical therapy may overcome these limitations. In some cases, additional surgery may be advisable to remove scar tissue and restore appearance.

Prevention

Burns are commonly received in residential fires. Properly placed and working smoke detectors in combination with rapid evacuation plans minimize a person's exposure to smoke and flames in the event of a fire. Children must be taught never to **play** with matches, lighters, fireworks, gasoline, and cleaning fluids.

Burns by scalding with hot water or other liquids may be prevented by setting the water heater thermostat no higher than 120°F (49°C), checking the temperature of bath water before getting into the tub, and turning pot handles on the stove out of the reach of children. Care should be used when removing covers from pans of steaming foods and when uncovering or opening foods heated in a microwave oven.

Thermal burns are often received from electrical appliances. Care should be exercised around stoves, space heaters, irons, and curling irons.

Sunburns may be avoided by the liberal use of a sunscreen containing either an opaque active ingredient such as zinc oxide or titanium dioxide or a nonopaque active ingredient such as PABA (para-aminobenzoic acid) or benzophenone. Hats, loose clothing, and umbrellas also provide protection, especially between 10 a.m. and 3 p.m. when the most damaging ultraviolet rays are present in direct sunlight.

Electrical burns may be prevented by covering unused electrical outlets with **safety** plugs and keeping electrical cords away from infants and toddlers who might chew on them. Persons should also seek shelter indoors during a thunderstorm to avoid being struck by lightning.

Chemical burns may be prevented by wearing protective clothing, including gloves and eyeshields. Chemical agents should always be used according to the manufacturer's instructions and properly stored when not in use.

Nutritional concerns

Adequate **nutrition**, including liquids and electrolytes, is essential when recovering from burns.

KEY TERMS

Debridement—The surgical removal of dead tissue and/or foreign bodies from a wound or cut.

Dermis—The basal layer of skin; it contains blood and lymphatic vessels, nerves, glands, and hair follicles.

Epidermis—The outermost layer of the human skin.

Shock—A medical emergency in which the organs and tissues of the body are not receiving an adequate flow of blood. This deprives the organs and tissues of oxygen and allows the build-up of waste products. Shock can be caused by certain diseases, serious injury, or blood loss.

Parental concerns

Parents should fire-proof their homes to protect small children. They should teach fire safety to their children from a very young age. Smoke detectors should be installed and tested at least twice each year. Parents are advised to discuss fire and escape routes (including alternates) from their home with their children. Holding a fire drill at night may be momentarily unpopular but may save lives and prevent serious injuries. Proper **childproofing** tools can prevent young children from being burned in the kitchen and bathroom.

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American Academy of Emergency Medicine. 611 East Wells Street, Milwaukee, WI 53202. Web site: <www.aaem.org/>.

American Academy of Family Physicians. 11400 Tomahawk Creek Parkway, Leawood, KS 66211–2672. Web site: <www.aafp.org/>.

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C

Caffeine

Definition

Caffeine is a drug that stimulates the central nervous system.

Description

Caffeine is found naturally in coffee, tea, and chocolate. Colas and some other soft drinks contain it. Caffeine also comes in tablet and capsule forms and can be bought without a prescription. Over-the-counter caffeine brands include No Doz, Overtime, Pep-Back, Quick-Pep, Caffedrine, and Vivarin. Some **pain** relievers, medicines for migraine headaches, and **antihistamines** also contain caffeine.

General use

Caffeine makes people more alert, less drowsy, and improves coordination. Combined with certain pain relievers or medicines for treating migraine **headache**, caffeine makes those drugs work more quickly and effectively. Caffeine alone can also help relieve headaches. Antihistamines are sometimes combined with caffeine to counteract the drowsiness that those drugs cause. Caffeine is also sometimes used to treat other conditions, including breathing problems in newborns and in young babies after surgery.

Precautions

Caffeine cannot replace **sleep** and should not be used regularly for staying awake as the drug can lead to serious **sleep disorders**, like insomnia.

People who use large amounts of caffeine over long periods build up a tolerance to it. When that happens, they have to use more and more caffeine to get the same effects. Heavy caffeine use can also lead to dependence. If the person then stops using caffeine abruptly, withdrawal symptoms may occur. These can include throbbing headaches, fatigue, drowsiness, yawning, irritability, restlessness, **vomiting**, or runny nose. These symptoms can go on for as long as a week if caffeine is avoided. Then the symptoms usually disappear.

If taken too close to bedtime, caffeine can interfere with sleep. Even if it does not prevent a person from falling asleep, it may disturb sleep during the night.

The notion that caffeine helps people sober up after drinking too much alcohol is a myth. In fact, using caffeine and alcohol together is not a good idea. The combination can lead to an upset stomach, **nausea**, and vomiting.

Older people may be more sensitive to caffeine and thus more likely to have certain side effects, such as irritability, nervousness, **anxiety**, and sleep problems.

Children under the age of 12 should normally avoid caffeine.

Side effects

Although caffeine is used to treat headaches, regular consumption of large quantities of caffeine containing beverages can cause severe headaches.

Excess use of caffeine by children leads to decreased nighttime sleep, but increased daytime sleep.

Interactions

Certain drugs interfere with the breakdown of caffeine in the body. These include **oral contraceptives** that contain estrogen, the antiarrhythmia drug mexiletine (Mexitil), and the ulcer drug cimetidine (Tagamet).

Caffeine interferes with drugs that regulate heart rhythm, such as quinidine and propranolol (Inderal). Caffeine may also interfere with the body's absorption of iron. Anyone who takes iron supplements should take them at least an hour before or two hours after using caffeine.

Serious side effects are possible when caffeine is combined with certain drugs. For example, taking

KEY TERMS

Arrhythmia—Any deviation from a normal heart beat.

Central nervous system—Part of the nervous system consisting of the brain, cranial nerves, and spinal cord. The brain is the center of higher processes, such as thought and emotion and is responsible for the coordination and control of bodily activities and the interpretation of information from the senses. The cranial nerves and spinal cord link the brain to the peripheral nervous system, that is the nerves present in the rest of body.

Withdrawal symptoms—A group of physical and/or mental symptoms that may occur when a person suddenly stops using a drug or other substance upon which he or she has become dependent.

caffeine with the decongestant phenylpropanolamine can raise blood pressure. Very serious heart problems may occur if caffeine and monoamine oxidase (MAO) inhibitors are taken together. These drugs are used to treat Parkinson's disease, depression, and other psychiatric conditions. People who use these drugs should consult a pharmacist or physician about which drugs can interact with caffeine.

Because caffeine stimulates the nervous system, anyone taking other central nervous system (CNS) stimulants should be careful about using caffeine.

Parental concerns

Moderate amounts of caffeine are not normally associated with adverse effects. As a rule, a daily intake of 300 milligrams should not present a problem. The following list gives the estimated amount of caffeine in common foods, but actual concentrations may be higher or lower.

- coffee, 115 mg
- black tea, 40 mg
- cola and other soft drinks, 18 mg
- chocolate milk, 5 mg
- milk chocolate (1 ounce) 6 mg

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University of Minnesota Extension Service. Office of the Director, 240 Coffey Hall, 1420 Eckles Ave. St. Paul, MN 55108–6068.

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C-section *see* **Cesarean section**

California Achievement Tests

Definition

The California Achievement Tests (CAT) are among the most widely used tests of basic academic skills for children from kindergarten through grade 12. The most recent edition of the CAT (the sixth) is also called *TerraNova, Second Edition* (or alternately, *TerraNova CAT*).

Purpose

The CAT is often administered to determine a child's readiness for promotion to a more advanced grade level and may also be used by schools to satisfy state or local testing requirements.

Description

First introduced in 1950, the CAT is a paper-and-pencil test for children from kindergarten through grade 12 that is designed to measure academic competency in a variety of areas. The test is available in six different forms: CAT Complete Battery, CAT Basic Battery, CAT Survey (grades two through 12 only), and CAT Plus.

- CAT Multiple Assessments. Uses multiple choice and open-ended test questions to assess reading/language arts, mathematics, science, and social studies skills.
- CAT Basic Multiple Assessments. Uses multiple choice and open-ended test questions to assess reading/language arts and mathematics skills.
- CAT Complete Battery. Uses multiple choice questions to assess reading/language arts, mathematics, science, and social studies skills.
- CAT Basic Battery. Uses multiple choice questions to assess reading/language arts and mathematics skills.
- CAT Survey. A shortened version of the complete battery, this form uses multiple choice questions to assess reading/language arts, mathematics, science, and social studies skills.
- CAT Plus. Add-on assessments that address the academic components of word analysis, vocabulary, language mechanics, spelling, and mathematics computation.

The CAT is a standardized test, meaning that norms were established during the design phase of the test by administering the test to a large, representative sample of the test population (in the case of the CAT, over 300,000 students). The test is given in a group, classroom setting, and can take anywhere from one-and-a-half to over five hours to complete depending on the test form and grade level. A teacher typically administers the CAT. When testing is complete, the test is sent back to the company that publishes the CAT (CTB/McGraw Hill) for scoring, and then scoring information is returned to the school in the form of individual test reports.

The test report includes a scale score, which is the basic measurement of how a child performs on the **assessment**, and a national percentile (NP), which reflects the percentage of students in the national norm group who have scores below the student's score (e.g., an NP of 80 means that 80 percent of students scored lower than the student). The scale score may be derived one of two ways—a straight score determined by the total number of test items correct or through item-pattern scoring (also called item response theory, or IRT). Item-pattern scoring examines not only the number of correct responses, but also the difficulty level of the questions answered right and the interrelationship of the pattern of answers. Other scoring information may also be included in the test report depending on the scoring report format.

KEY TERMS

Norms—A fixed or ideal standard; a normative or mean score for a particular age group.

Representative sample—A random sample of people that adequately represents the test-taking population in age, gender, race, and socioeconomic standing.

Standardization—The process of determining established norms and procedures for a test to act as a standard reference point for future test results.

Preparation

For students who are unfamiliar with the mechanics of taking a standardized test, a practice test session given by a teacher shortly before the CAT testing session begins may be appropriate. Because the CAT is designed to be a measurement of a child's current educational achievement level, the test publisher recommends that no pre-test coaching or test study programs be used.

Parental concerns

Test anxiety can have a negative impact on a child's performance, so parents should attempt to take the stress off their child by making sure the child understands that it is the effort and attention they give the test, not the final score, that matters. Parents can also ensure that their children are well rested on the testing day and have a nutritious meal beforehand.

When test results are available, parents should schedule a meeting with their child's teacher to discuss the test's implications. Results from CAT testing can help parents and teachers identify academic strengths and weaknesses and develop strategies for capitalizing on the former and building skills in the latter.

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Paula Ford-Martin

Cancer

Definition

Cancer is a group of diseases characterized by uncontrolled growth of tissue cells in the body and the invasion by these cells into nearby tissue and migration to distant sites.

Description

Cancer results from alterations (mutations) in genes that make up DNA, the master molecule of the cell. Genes make proteins, which are the ultimate workhorses of the cells, responsible for the many processes that permit humans to breathe, think, and move, among other functions. Some of these proteins control the orderly growth, division, and reproduction of normal tissue cells. Gene mutations can produce faulty proteins, which in turn produce abnormal cells that no longer divide and reproduce in an orderly manner. These abnormal cells divide uncontrollably and eventually form a new growth known as a tumor or neoplasm. A healthy immune system can usually recognize neoplastic cells and destroy them before they divide. However, mutant cells may escape immune detection and become tumors or cancers.

Studies of the origins of cancer have shown that a combination of genetic influences and environmental causes over time triggers gene mutations, which may explain why most cancers are seen in adults of middle age or older (60%) and cancer is rare children. Many cancers have been shown to result from exposure to environmental toxins (carcinogens) and related alterations in DNA. Faulty DNA can also be inherited, predisposing an individual to develop cancer, although fewer than 10 percent of cancers are purely hereditary. Hereditary links have been shown in cancers of the breast, colon, ovaries, and uterus. Inherited physiological traits can also contribute to cancer, such as inheriting fair skin increasing the risk of skin cancer, but only if accompanied by prolonged exposure to intensive sunlight.

Tumors can be benign or malignant. A benign tumor is not cancer. It is slow growing, does not invade surrounding tissue, and once removed, does not usually recur. A malignant tumor is cancerous. It invades surrounding tissue and spreads to nearby or distant organs (metastasis). If the cancer cells have spread to surrounding tissue, even after the malignant tumor is removed, it will typically recur.

Cancer falls into several general categories:

- Carcinoma (90% of all cancer) are solid tumors arising in the layer of cells (epithelium) covering the body's surface and lining internal organs and glands. Adenocarcinomas develop in an organ or gland and squamous cell carcinomas originate in the skin.
- Melanoma originates in the skin, usually in pigment cells (melanocytes).
- Sarcoma is cancer of supporting tissue such as bone, muscle, and blood vessels.
- Leukemias and lymphomas are cancers of the blood and lymph glands.
- Gliomas are cancers of the nerve tissue.

The most common cancers affecting adults are cancer of the skin, lung, colon, breast, and prostate. Cancer of the kidneys, ovaries, uterus, pancreas, bladder, rectum, and the leukemias and lymphomas are among the 12 major cancers affecting Americans of all ages. Although children and adolescents do develop solid tumors, the most common high-risk cancers among children are:

- acute myeloid leukemia
- acute lymphoblastic leukemia
- neuroblastoma
- glioma
- sarcoma of bone (osteosarcoma) and soft tissue

Demographics

Childhood cancer is rare, occurring in about 14 in 100,000 children in the United States each year. However, in the entire U.S. population, one of every four deaths is from cancer, second only to deaths from heart disease. About 1.2 million cancer cases are diagnosed annually and more than 500,000 die, of whom 2,700 are children or adolescents.

Causes and symptoms

Genetic predisposition, environmental causes, and individual developmental problems are responsible for most childhood cancer. The presence of other disorders, such as **Down syndrome**, has also been shown to be

associated with cancer in children. The major risk factors that apply to adult cancer are tobacco, alcohol, sexual and reproductive behavior, and occupation, none of which increases risk in children. Other well-known risk factors, such as **family** history, infectious agents, diet, environmental toxins, and pollution, can apply equally to children.

Tobacco

Approximately 80 to 90 percent of lung cancer cases occur in smokers. **Smoking** is also the leading cause of bladder cancer and has been shown to contribute to cancers of the upper respiratory tract, esophagus, larynx, kidney, pancreas, stomach, and possibly breast as well. Second-hand smoke (passive smoking) has been shown to increase cancer risk in children and adults who live with smokers.

Infectious agents

Cancer deaths worldwide can be traced to viruses, bacteria, or parasites. Epstein-Barr virus (EBV), for example, is associated with lymphoma, the hepatitis viruses are associated with liver cancer, HIV is associated with Kaposi's sarcoma, and the bacteria *Helicobacter pylori* is associated with stomach cancer.

Genetic predisposition

Certain cancers such as breast, colon, ovarian, and uterine cancer recur generation after generation in some families. Eye cancer (**retinoblastoma**), a type of colon cancer, and early-onset breast cancer have been shown to be linked to the inheritance of specific genes.

Environmental sources

Radiation is believed to cause 1 to 2 percent of all cancer deaths. Ultraviolet radiation from the sun accounts for a majority of melanoma deaths. Other sources of radiation are x-rays, radon gas, and ionizing radiation from nuclear material.

Pollution

Studies have established links between environmental toxins, such as asbestos, and cancer. Chlorination of water may account for a small rise in cancer risk. However, the main danger from pollutants occurs when toxic industrial chemicals are released into the surrounding environment. As of 2004 an estimated 1 percent of cancer deaths are believed to be due to air, land, and water pollution.

Cancer is a progressive disease that goes through several stages, each producing a number of symptoms. Early symptoms can be produced by the growth of a solid tumor in an organ or gland. A growing tumor may press on nearby nerves, organs, and blood vessels, causing **pain** and pressure that may be the first warning signs of cancer. Other symptoms can include sores that do not heal, growths on the skin or below the skin, unusual bleeding, difficulty digesting food or swallowing, and changes in bowel or bladder function. **Fever** can be present as well as fatigue and weakness.

When to call the doctor

Despite the fact that there are hundreds of different types of cancer, each producing different symptoms, the American Cancer Society has established the following seven symptoms as possible warning signals of cancer:

- changes in the size, color, or shape of a wart or a mole
- a sore that does not heal
- persistent **cough**, hoarseness, or sore throat
- a lump or thickening in the breast or elsewhere
- unusual bleeding or discharge
- chronic indigestion or difficulty swallowing
- any change in bowel or bladder habits

Parents should report any such symptoms to the pediatrician along with unexplained fever or frequent infections. Vision problems, weight loss, lack of appetite, depression, swollen glands, paleness, or general weakness are other reasons for parents to consult the pediatrician. Generally, the earlier cancer is diagnosed and treated, the better the chance of a cure, although not all cancers have early symptoms.

Diagnosis

Diagnosis begins with a complete medical history, including family history of cancer, and a thorough physical examination. The doctor observes and palpates (applies pressure by touch) different parts of the body in order to identify any variations from normal size, feel, and texture of an organ or tissue. The doctor looks inside the mouth for abnormalities in color, moisture, surface texture, or the presence of any thickening or sores in the lips, tongue, gums, the roof of the mouth, or the throat. The doctor observes the front of the neck for swelling and may gently manipulate the neck and palpate the front and side surfaces of the thyroid gland at the base of the neck, looking for nodules or tenderness. The doctor also palpates the lymph nodes in the neck, under the arms, and in the groin, looking for enlargement. The skin is examined for sores that are slow to heal, especially

those that bleed, ooze, or crust; irritated patches that may itch or hurt; and any change in the size of a wart or a mole.

In adolescent females, a pelvic exam may be conducted to detect cancers of the ovaries, uterus, cervix, and vagina. The doctor first looks for abnormal discharges or the presence of sores. Then the internal pelvic organs such as the uterus and ovaries are palpated (touched while applying gentle pressure) to detect abnormal masses. Breast examination evaluates unevenness, discoloration, or scaling; both breasts are palpated to feel for masses or lumps.

In adolescent males, inspection of the rectum and prostate may be included in the physical examination. The doctor inserts a gloved finger into the rectum and rotates it slowly to feel for growths, tumors, or other abnormalities. The testes are examined visually, looking for unevenness, swelling, or other abnormalities. The testicles are palpated to identify lumps, thickening or differences in size, weight, or firmness.

If an abnormality is detected on physical examination, or symptoms suggestive of cancer are noted, diagnostic tests will be performed. Laboratory studies of sputum, blood, urine, and stool can detect abnormalities that may confirm cancer. Sputum cytology involves the microscopic examination of phlegm that is coughed up from the lungs. Tumor markers, specific proteins released by certain types of cancer cells, can be detected by performing a test on venous blood. If leukemia or lymphoma is suspected, a complete blood count (CBC) with peripheral smear (differential) is done to evaluate the number, appearance, and maturity of red blood cells (RBCs) and white blood cells (WBCs) and to measure hemoglobin, hematocrit, and **platelet count**. A bone marrow biopsy may be done to determine what type of cells is present in the bone marrow. Blood chemistries will be done to help determine if liver or kidney problems are present. Blood chemistries are also useful in monitoring the effectiveness of treatment for all types of cancer and in following the course of the disease and detecting recurrences.

Diagnostic imaging techniques such as **computed tomography** (CT scans), **magnetic resonance imaging** (MRI), ultrasound, and fiberoptic scope examinations (such as colonoscopy or sigmoidoscopy) can help determine the location, size, and characteristics of a tumor even if it is deep within the body. Conventional **x rays** are often used for initial evaluation, because they are relatively cheap, painless, and easily accessible. In order to increase the information obtained from a conventional x ray, air or contrast media (such as barium or iodine) may be used to enhance the images.

The most definitive diagnostic test for cancer is a biopsy, which is the surgical removal of a piece of suspect tissue for staining and microscope examination (cytochemistry). By examining certain cell characteristics, abnormalities can be identified and the presence of specific types of cells can be diagnostic for certain cancers. The biopsy provides information about the type of cancer, its stage, the aggressiveness of the cancer in invading nearby tissue or organs, and the extent of metastases at diagnosis. The pathologist who evaluates cancer cells in biopsied tissue designates the cancer as being stage I, II, III, or IV, in terms of the degree of metastasis.

Newer molecular and cellular diagnostic testing, such as polymerase chain reaction (PCR), allows the molecular genetic analysis of tumors. Cytogenetic analysis of tumor chromosomes, for example, can identify structural abnormalities that may explain the unique origins of cancer in an individual child. Spectral karyotyping (SKY), an advanced method of screening chromosomes for numeric and structural abnormalities, is used to evaluate pediatric tumors. Gene sequences can also be evaluated in a method (comparative genomic hybridization) that compares samples from a tumor and normal tissue after both have been exposed to the same radioactive material. This method can determine gains and losses in DNA in the region of the tumor, detecting alterations that have caused the cancer. The developing science of proteomics studies specific proteins in cells and may someday be able to provide detailed assessment of cancer cells.

Treatment

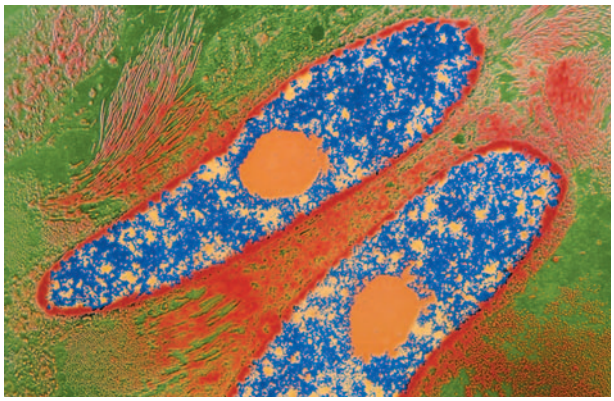
The aim of cancer treatment is to remove or destroy all or as much of the primary tumor as possible and to prevent its recurrence or metastases. While devising a treatment plan for cancer, the likelihood of curing the cancer has to be weighed against the side effects of the treatment. If the cancer is highly aggressive and cure is not likely, treatment will be aimed at relieving symptoms and controlling the cancer for as long as possible.

Cancer treatment is always tailored to the individual. The treatment choice depends on the type and location of cancer, the extent to which it has already spread, and the age, sex, and general health status of the individual. The major types of treatment are: surgery, radiation, **chemotherapy**, immunotherapy, hormone therapy, and bone-marrow transplantation.

Advances in molecular biology and cancer genetics have contributed greatly to the development of therapies that provide cell-targeted treatment. Genetic testing uses molecular probes to identify gene mutations that have

been linked to specific cancers. In the early 2000s ongoing research is focused on new treatment and prevention methods, including molecular-targeted therapies, virus therapy, immunotherapy, and drug therapy that stimulates the self-destruction of cancer cells (apoptosis).

Targeted molecular therapy, although as of 2004 still the subject of concentrated research, was being used effectively in pediatric study subjects where it has been shown to reduce the toxicity seen with conventional chemotherapy. Unlike chemotherapy, which treats all cells uniformly, targeted molecular therapy can focus on selected cells without affecting normal cells and tissues. This refinement frees children from some of the long-term toxic effects and complications that can negatively affect quality of life and survival even if the cancer is cured.



A transmission electron micrograph (TEM) of two spindle cell nuclei from a human sarcoma. Sarcomas are cancers of the connective tissue (bone, nerves, smooth muscle). *(Photograph by Dr. Brian Eyden. National Audubon Society Collection/ Photo Researchers, Inc.)*

Surgery

Surgical removal of a solid tumor is most effective with small tumors confined to one area of the body. Surgery removes the tumor (tumor resection) and usually part of the surrounding tissue to ensure that no cancer cells remain in the area. Since cancer usually spreads via the lymphatic system, adjoining lymph nodes are sometimes removed as well. Surgery may also be preventive or prophylactic, removing an abnormal looking area of tissue that is likely to become malignant over time. During surgery biopsies may also be performed on tissue that may be affected by metastases. Surgery is not a typical treatment for leukemia or lymphoma, which arise in the circulatory system and lymphatic systems that extend throughout the body. Children with osteosarcoma (bone cancer) and other solid tumors are candidates for surgery, however.

Surgery may be performed in conjunction with radiation (cytoreductive surgery) or chemotherapy. The surgeon removes as much of the cancer as possible and the remaining area is treated with radiotherapy or chemotherapy or both. In advanced metastatic cancer when cure is unlikely, palliative surgery aims at reducing symptoms. Debulking surgery, for example, removes part of a tumor that is pressing on other organs and causing pain. In tumors that are dependent on hormones, one option is to remove organs that secrete the hormones.

Radiation therapy

Radiation kills tumor cells and is used alone when a tumor is in a poor location for surgery. More often, it is used in conjunction with surgery and chemotherapy. Radiation can be either external or internal. External radiation is aimed at the tumor from outside the body. In internal radiation (brachytherapy), radioactive liquid or pellets are delivered to the cancerous site via a pill, injection, or insertion in a sealed container.

Chemotherapy

Chemotherapy is the administration of drugs that kill cancer cells (cytotoxic drugs). It destroys hard-to-detect cancer cells that have spread (metastasized) through the circulation or lymph system. Chemotherapeutic drugs are given orally or intravenously, either alone or in conjunction with surgery, radiation, or both. When chemotherapy is used before surgery or radiation, it is known as primary chemotherapy or neoadjuvant chemotherapy. Because the cancer cells have not yet been exposed to anti-cancer drugs, they are especially vulnerable, allowing neoadjuvant therapy to effectively reduce tumor size. However, the toxic effects of neoadjuvant chemotherapy may be severe, because normal cells are also destroyed. Chemotherapy may also make the body less tolerant of the side effects of other treatments such as radiation therapy. Adjuvant therapy is the more common type of chemotherapy, used to enhance the effectiveness of other treatments.

Immunotherapy

Immunotherapy uses the body's own immune system, specifically a type of disease-fighting white cell called T-cells, to destroy cancer cells. Tumor-specific proteins that are part of unique genetic mutations in pediatric cancer, for example, are believed to be ideal targets for anti-tumor immune processes. Various immunological agents are as of 2004 still in clinical trials and are not as of that year widely available, though initial results are promising. Monoclonal antibodies are used to

Common childhood cancers

Percentage of total childhood cancers	Type of cancer
39%	Leukemia (white blood cell cancer) and lymphoma (lymph system cancer)
20.7%	Brain cancers (brain and spinal cord tumors)
7.3%	Neuroblastoma (nerve cell cancer, most commonly in the adrenal gland)
6.1%	Wilms' tumor (kidney cancer that can metastasize to lung)
4.7%	Osteosarcoma (bone cancer) and Ewing's sarcoma (cancer in the bone shaft)
3.4%	Rhabdomyosarcoma (muscle tissue cancer, most often in head and neck)
2.9%	Retinoblastoma (malignant eye tumor)
16.4%	Germ cell cancer (ovarian or testicular cancers) and others

SOURCE: Margo Hoover-Regan. http://www.csupomona.edu/~cancerbio/pediatric%20cancer%20-%20Dr.%20Hoover-Regan.htm. Updated May 15, 2000.

(Table by GGS Information Services.)

fight cancer cells in much the same way as antibodies that are produced by the body's own immune system work to fight infection. Other substances are also being used experimentally. They include substances such as interferons, interleukins, growth factors, monoclonal antibodies, and vaccines. Unlike traditional vaccines, cancer vaccines do not prevent cancer but are designed to treat existing disease. They work by boosting the immune system and training immunized cells to destroy cancer cells.

Hormone therapy

Hormone therapy is standard treatment for cancers that are hormone-dependent and grow faster in the presence of specific hormones, such as cancer of the prostate, breast, and uterus. Hormone therapy blocks the production or action of these hormones, slowing growth of the tumor and extending survival for months or years.

Bone marrow transplantation

Bone marrow is the tissue within bone cavities that produces blood cells. Healthy bone marrow tissue constantly replenishes the blood supply and is essential to life. Sometimes drugs or radiation needed to destroy cancer cells also destroys bone marrow and only replacement with healthy cells counteracts this adverse effect. A bone marrow transplant involves removing marrow from a donor and transplanting blood-forming cells to a recipient. While not a therapy in itself, bone marrow transplantation may allow a cancer patient to undergo aggressive therapy.

Many specialists work together to treat cancer patients. The oncologist is a physician who specializes in cancer care and usually coordinates the treatment plan, directing chemotherapy, hormone therapy, and any treatment that does not involve radiation or surgery. The radiation oncologist uses radiation to treat cancer, while the surgical oncologist performs surgery to diagnose or treat cancer. Gynecologist-oncologists and pediatric-oncologists, as their titles suggest, are physicians who treat women's and children's cancers. Radiologists read the x rays, ultrasound images, CT scans, and MRI images to help diagnose cancer. Hematologists specialize in disorders of the blood and bone marrow and are consulted in the evaluation of leukemia, lymphoma, and bone cancer.

Alternative treatment

A range of alternative treatments are available to help treat cancer that can be used in conjunction with, or separate from, surgery, chemotherapy, and radiation. Alternative treatment of cancer is a complicated arena and a trained complementary health practitioner should be consulted.

Although the effectiveness of complementary therapies such as acupuncture in alleviating cancer pain have not as of 2004 been clinically proven, many cancer patients find it safe and beneficial. Bodywork therapies such as massage and reflexology ease muscle tension and may alleviate side effects such as **nausea and vomiting**. Homeopathy and herbal remedies used in Chinese traditional herbal medicine also have been shown to alleviate some of the side effects of radiation and chemotherapy and are being recommended by many doctors.

Prognosis

Most cancers show good cure rates if detected and treated at early stages. The prognosis involves the type of cancer, its degree of invasiveness, and the extent of metastases at diagnosis. In addition, age, general health status, and response to treatment are important factors. Cancer deaths in children have shown consistent declines, decreasing between 1975 and 2000 from 50 in 1 million diagnosed to 25 in 1 million. However, cancer is the leading cause of death among children and adolescents, responsible for 2,700 deaths each year in the United States.

Prevention

Prevention of cancer means being aware of causes and risks, which involve a combination of genetic and environmental factors. Except for family history, specific genetic causes or an inherited predisposition are generally

unknown in individuals until revealed in the diagnostic process. Known environmental causes can be avoided, however. A list of guidelines offered by nutritionists and epidemiologists from leading U.S. universities to reduce the risk of cancer includes some that may apply to children and adolescents:

- Eat plenty of vegetables and fruits, especially cruciferous vegetables such as broccoli, cauliflower, and cabbage.
- Decrease or avoid eating animal fats and red meats.
- Exercise vigorously for at least 20 minutes every day.
- Avoid excessive weight gain.
- Avoid tobacco (including second hand smoke).
- Avoid excessive amounts of alcohol.
- Avoid midday sun (between 11 a.m. and 3 p.m.) when rays are the strongest.
- Avoid risky sexual practices and multiple partners.
- Avoid known carcinogens in the environment or work place.

Certain drugs being used as of 2004 for treatment could also be suitable for prevention, at least prevention of recurrences. For example, the drug tamoxifen has been very effective against breast cancer and is in 2004 being used to prevent recurrence in breast cancer survivors. Similarly, retinoids derived from vitamin A are being tested for their ability to slow the progression of or to prevent head and neck cancers. Certain studies suggest that cancer incidence is lower in areas where soil and foods are rich in the mineral selenium.

Nutritional concerns

Certain foods, including many vegetables, fruits, and grains, are believed to offer protection against various cancers. In laboratory studies, **vitamins** such as A, C, and E, as well as beta-carotene found in carrots and isothiocyanate and dithiolthione compounds found in cruciferous vegetables, such as broccoli, cauliflower, and cabbage, have been shown to provide protection against certain types of cancer. Studies have shown that eating a diet rich in fiber as found in fruits, vegetables, and whole grains can reduce the risk of colon cancer.

Parental concerns

A diagnosis of childhood cancer raises many uncertainties and concerns for parents, including how to acquire the most effective therapy. Advances in molecular and cellular technologies have improved both the diagnosis and treatment of pediatric cancer and also carry with them the possibility of someday curing and preventing cancer in children. While cancer was at one time nearly always fatal in children, as of 2004 more than 75 percent of children diagnosed with cancer

KEY TERMS

Benign—In medical usage, benign is the opposite of malignant. It describes an abnormal growth that is stable, treatable, and generally not life-threatening.

Biopsy—The surgical removal and microscopic examination of living tissue for diagnostic purposes or to follow the course of a disease. Most commonly the term refers to the collection and analysis of tissue from a suspected tumor to establish malignancy.

Bone marrow—The spongy tissue inside the large bones in the body that is responsible for making the red blood cells, most white blood cells, and platelets.

Carcinogenic—A substance that can cause cancer to develop.

Chemotherapy—Any treatment of an illness with chemical agents. The term is usually used to describe the treatment of cancer with drugs that inhibit cancer growth or destroy cancer cells.

Epithelium—The layer of cells that covers body surfaces, lines body cavities, and forms glands.

Hormone therapy—Treating cancers by changing the hormone balance of the body, instead of by using cell-killing drugs.

Immunotherapy—A mode of cancer treatment in which the immune system is stimulated to fight the cancer.

Malignant—Cells that have been altered such that they have lost normal control mechanisms and are capable of local invasion and spread to other areas of the body. Often used to describe a cancer.

Metastasis—A secondary tumor resulting from the spread of cancerous cells from the primary tumor to other parts of the body.

Radiation therapy—A cancer treatment that uses high-energy rays or particles to kill or weaken cancer cells. Radiation may be delivered externally or internally via surgically implanted pellets. Also called radiotherapy.

Sore—A wound, lesion, or ulcer on the skin.

Tumor—A growth of tissue resulting from the uncontrolled proliferation of cells.

enjoyed disease-free survival. Targeted molecular therapy and immunotherapies are the ongoing focus of concentrated research, and studies using these cell-selective technologies in treating children have shown encouraging results, both in earlier responses and reduced toxicity and complications longer term. Parents can be assured of access to the current knowledge base in molecular biology and advanced treatment technologies that promise better outcomes.

See also Leukemias, acute; Leukemias, chronic.

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Woznick, Leigh A., and Carol D. Goodheart. *Living with Childhood Cancer: A Practical Guide to Help Families Cope*. Washington, DC: American Psychological Association (APA), 2002.

ORGANIZATIONS

American Cancer Society. 1599 Clifton Road, NE, Atlanta, GA 30329. Web site: <www.cancer.org>.

Cancer Research Institute (National Headquarters). 681 Fifth Avenue, New York, NY 10022. Web site: <www.cancerresearch.org>.

National Cancer Institute. 9000 Rockville Pike, Building 31, room 10A16, Bethesda, MD 20892. Web site: <wwwicic.nci.nih.gov>.

National Children's Cancer Society. 1015 Locust Suite 600, St. Louis, MO 63101. Web site: <www.nationalchildrenscancer society.com>.

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Candida infection *see* **Candidiasis**

Candidiasis

Definition

Candidiasis is an infection caused by a species of the yeast *Candida*, usually the *Candida albicans* fungus. *Candida* is found on various parts of the bodies of almost all normal people but causes problems in only a few. Candidiasis can affect the skin, nails, and mucous membranes throughout the body including the mouth (thrush), esophagus, vagina (yeast infection), intestines, and lungs.

Description

Candida may cause yeast mouth infections (also known as thrush) in children with reduced immune function or in children taking certain **antibiotics**. Antibiotics may upset the balance of microorganisms in the body and allow an overgrowth of *Candida*. The use of inhaled steroids for the treatment of **asthma** has also been shown to cause oral candidiasis. Many infants acquire candidiasis from their mothers during the process of birth, when the baby comes in contact with naturally existing *Candida* found in the mother's vagina. Candidiasis is not considered harmful to infants unless it lasts more than several weeks after birth. These yeast mouth infections cause creamy white, curd-like patches on the tongue, inside of the mouth, and on the back of the throat. Under the whitish material, there are red lesions that may bleed.

Candida also may infect an infant's **diaper rash**, as it grows rapidly on irritated and moist skin. Children who suck their thumbs or other fingers may also develop candidiasis around their fingernails, causing redness on the nail edges.

Candida is a common cause of vaginal infections in adolescent girls, especially when the normal populations of the bacteria *Lactobacilli* have been reduced due to antibiotic use, allowing the overgrowth of *Candida*. A candidiasis infection in the vagina results in **itching**, burning, soreness, and a thick, white vaginal discharge.

Other risk factors for candidiasis include **obesity**, heat, and excessive sweating that result in the formation of moist skin areas where the yeast organism can grow.

In the early 2000s, several serious categories of candidiasis have become more common, due to overuse of antibiotics, the rise of **AIDS**, the increase in incidence of organ transplantations, the use of **chemotherapy** in **cancer** treatment, and the implantation of invasive devices (e.g., nasogastric tubes, catheters, and artificial joints and valves) into the body—all of which increase a patient's susceptibility to infection. Diabetics are especially susceptible to candidiasis, as they have high levels of sugar in their blood and urine and a low resistance to infection, both of which are conditions that favor the growth of yeast. Also known as invasive candidiasis, deep organ candidiasis is a serious systemic infection

that can affect the esophagus, heart, blood, liver, spleen, kidneys, eyes, and skin. Like vaginal and oral candidiasis, it is an opportunistic disease that strikes when a child's resistance is lowered, often due to another illness. Children with granulocytopenia (deficiency of white blood cells) are particularly at risk for deep organ candidiasis. There are many diagnostic categories of deep organ candidiasis, depending on the tissues involved.

In the past candidiasis was referred to as moniliasis.

Demographics

Candidiasis is an extremely common infection. Thrush occurs in approximately 2–5 percent of healthy newborns and occurs in a slightly higher percentage of infants during their first year of life.

Over 1 million adult women and adolescent girls in the United States develop vaginal yeast infections each year. It is not life-threatening, but the condition can be uncomfortable and frustrating.

Causes and symptoms

Candidiasis is caused by a species of the yeast *Candida*, usually the *Candida albicans* fungus.

In oral candidiasis, the disease is characterized by whitish patches that appear on the tongue, inside of the cheeks, or on the palate. **Pain** or difficulty in swallowing may indicate a fungal infection in the throat, which is a potential complication of AIDS. Most adolescent girls with vaginal candidiasis experience severe vaginal itching and have a discharge that often looks like cottage cheese and has a sweet or bread-like odor. The vulva and vagina can be red, swollen, and painful. The infected skin in diaper rash that includes infection with *Candida* appears fiery red with areas that may have a raised red border.

Effects of deep organ or systematic candidiasis include **meningitis**, arthritis, fungemia (fungi in the blood, causing **fever** and possibly leading to sepsis), endocarditis (heart infection), endophthalmitis (infection and scarring in the eye that can affect vision), and renal or bladder bezoars (colonization and blockage of the urinary tract by *Candida*, which can cause urinary tract infections and kidney failure.

Diagnosis

Often clinical appearance and visual examination give a strong suggestion about the diagnosis. Generally, a doctor takes a sample of the vaginal discharge or swabs an area of oral or skin lesions, and then inspects this material under a microscope, where it is possible to see characteristic forms of yeasts at various stages in the lifecycle.

Fungal blood and stool cultures for detection of the *Candida* organism should be taken for patients suspected of having deep organ candidiasis. Tissue biopsy may be needed for a definitive diagnosis.

When to call the doctor

The doctor should be called if a child exhibits any symptoms of the various types of candidiasis.

Treatment

Treatment of candidiasis is primarily accomplished through the use of antifungal drugs. Oral candidiasis is usually treated with prescription lozenges or mouthwashes. Some of the most-used prescriptions are nystatin mouthwashes (Nilstat or Nitrostat) and clotrimazole lozenges. Skin infections can be treated with topical antifungal creams. Highly inflamed skin lesions can also be treated with corticosteroid creams.

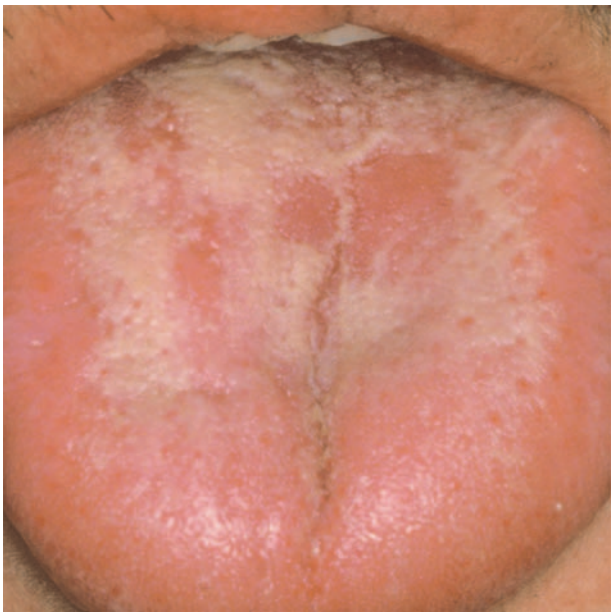
For infants with oral candidiasis, pacifiers should be sterilized or discarded. Bottle nipples should be discarded and new ones used as the infant's mouth begins to heal.

The risk of diaper rash complicated with candidiasis can be reduced by preventing irritating **dermatitis** through the use of absorbent diapers and prevention of excessive exposure to urine or feces through frequent changing of diapers. The use of plastic pants that do not allow air circulation over the diaper area is not recommended. Children may still attend child care; however, childcare providers should follow good hygienic practices, including thorough hand washing and disposal of materials that may contain nasal and oral secretions of infected children, in order to prevent transmitting the infection to other children.

In most cases, vaginal candidiasis can be treated successfully with a variety of over-the-counter antifungal creams or suppositories, including Monistat, Gyne-Lotrimin, and Mycelex. However, infections often recur. If an adolescent girl has frequent recurrences, she should consult her doctor about prescription drugs such as Vagistat-1, Diflucan, and others.

The early 2000s increase in deep organ candidiasis has led to the creation of treatment guidelines, including, but not limited to, the following:

- Catheters should be removed from children with candidiasis.



This patient's tongue is infected with candidiasis, or thrush. *(Photograph by Edward H. Gill, Custom Medical Stock Photo Inc.)*

KEY TERMS

Biopsy—The surgical removal and microscopic examination of living tissue for diagnostic purposes or to follow the course of a disease. Most commonly the term refers to the collection and analysis of tissue from a suspected tumor to establish malignancy.

Granulocytopenia—A condition characterized by a deficiency of white blood cells.

Nasogastric tube—A long, flexible tube inserted through the nasal passages, down the throat, and into the stomach.

Opportunistic—Infection caused by microorganisms that are usually harmless, but which can cause disease when a host's resistance is lowered.

Systemic—Relating to an entire body system or the body in general.

- Antifungal therapy may be used during chemotherapy to prevent candidiasis.
- Drugs should be prescribed based on a child's specific history and immune defense status (this is especially critical for children with AIDS). Stronger antifungal drugs, such as ketoconazole or fluconazole, may be necessary.
- Diabetes mellitus should be controlled with appropriate medication and dietary changes.

Alternative treatment

Home remedies for vaginal candidiasis include vinegar douches or insertion of a paste made from *Lactobacillus acidophilus* powder into the vagina. In theory, these remedies make the vagina more acidic and, therefore, less hospitable to the growth of *Candida*. Fresh garlic (*Allium sativum*) is believed to have antifungal action, so incorporating it into the diet or inserting a gauze-wrapped, peeled garlic clove into the vagina may be helpful. The insert should be changed twice daily. Some women report success with these remedies; however, they should try a conventional treatment if an alternative remedy is not effective.

Prognosis

Oral and skin candidiasis, though painful, are usually cured with the use of antifungal medications. However, in premature infants, in children with poor or compromised immune systems, or in children with deep organ or systematic infections, eradication of the infections may be more difficult to achieve. Mortality in low birth-weight premature infants with systemic candidiasis may reach 50 percent.

Prevention

Often candidiasis can be prevented through good sanitation procedures, such as keeping the body cool and dry, wearing natural fabric underclothes, changing underclothes frequently, wiping from front to back after bowel movements, and washing hands often. For children who are susceptible to candidiasis because of immune deficiencies, the regular use of antifungal drugs to prevent infections may be required.

Parental concerns

Parents need to practice good hygienic procedures as they care for their children, in order to prevent the development of candidiasis.

Resources

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Canker sores

Definition

Canker sores (aphthous ulcers) are small shallow sores or ulcers that appear inside the lips, inside the cheeks, or on the gums. They begin as small, reddish swellings. Then they burst, and the rupture sores are covered with a white or yellow membrane. The edges of the sores are still red and look like a painful red halo. Without treatment, canker sores heal in two weeks or less.

Description

Canker sores occur inside the mouth, usually inside the lips, cheeks, or soft palate. They can also occur on or under the tongue and in the throat. Often, several canker sores appear at the same time and maybe in clusters. Canker sores appear as a whitish, round area with a red border. The sores are painful and sensitive to touch. The average canker sore is about 0.25 inch (0.6 cm) in size, although they can occasionally be larger. Canker sores are not infectious.

Demographics

Anyone can get canker sores, but they are most common among teenagers and people in their twenties. Females are slightly more likely to get canker sores than males. Some people have one or two episodes a year; others have almost continual problems. Females are likely to have canker sores during their premenstrual period.

Approximately 20 percent of the U.S. population is affected by recurring canker sores. That means as of 2004 some 56 million Americans suffer annually from these small but painful ulcers. Fortunately, certain safe, natural remedies are effective in treating canker sores.

Canker sores are sometimes mistaken as cold sores (also known as fever blisters). Cold sores are caused by the **herpes simplex** virus. The sores caused by this disease, also known as oral herpes, can occur anywhere on the body. Most commonly, herpes infection occurs on the outside of lips and much less often inside the mouth. Cold sores are infectious.

Causes and symptoms

There is some evidence that canker sores are due in part to nutritional deficiencies and a lack of vitamin B_{12}, **folic acid**, and iron. Gastrointestinal problems correlate with canker sores as well. Frequent recurrent canker sores may suggest a metabolic imbalance. The sores appear during times of stress or as a reaction to hormonal imbalances in women. Pregnancy causes remission. A tendency to get canker sores may be inherited.

As of 2004 data suggest that aphthous ulcers are a form of autoimmune disease. Other proposed causes for canker sores are trauma from toothbrush and toothpick scrapes (trauma), hormones, and **food allergies**.

Symptoms

The first symptom is a tingling or mildly painful **itching** sensation in the area where the sore will appear. After one to several days, a small red swelling appears. The sore is round and is a whitish color with a grayish colored center. Usually, there is a red ring of inflammation surrounding the sore. The main symptom is **pain**. Canker sores can be very painful, especially if they are touched repeatedly by the tongue or silverware.

When to call the doctor

Infants and children may have difficulty sleeping because of the pain. The doctor should be called for pain relief remedies or medication to help the child through this period. The doctor should be called if the child runs a fever, refuses to eat or drink, and if the child shows signs of **dehydration**.

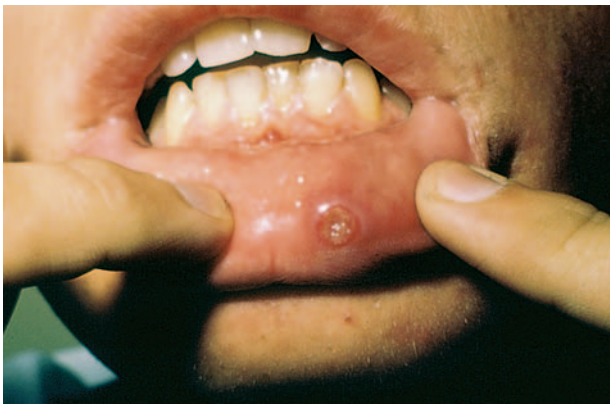
Diagnosis

Canker sores are diagnosed by observation of the blister, which generally appears in the mouth or throat. Canker sores are bacterial infections and not contagious.

Recurrent canker sores may indicate a metabolic imbalance, dietary deficiency, stress, and a lack of rest. Children who have frequent canker sores may benefit from dietary supplements of B-complex vitamin or may undergo blood and **allergy tests** to see if some other underlying cause can be identified.

Treatment

Since canker sores heal by themselves, professional treatment is not usually necessary. Topical anesthetics



Apthous ulcer, or canker sore, on the inside of a patient's bottom lip. *(© Lester V. Bergman/Corbis.)*

may relieve the pain. The use of corticosteroid ointments sometimes speeds healing. If an ointment is used to treat a canker sore, the parent should first dry the sore. Next, a small amount of medicine should be put on a cotton swab and applied to the sore. The child should not have anything to drink or eat for 30 minutes to keep the medicine from washing away. Parents can also try preparations of tea tree oil, goldenseal, propolis, licorice, myrrh, and lysine, products with healing nutrients that are backed by research.

There are several treatments for reducing the pain and duration of the sores. The drugs frequently recommended are anti-inflammatory steroid mouthwashes, **analgesics**, and numbing ointments containing benzocaine.

The following treatments may be effective in relieving symptoms or shortening the duration of canker sores in their children.

- for pain relief in a prescription, 2 percent viscous lidocaine, applied with a cotton swab (Q-tip) several times daily
- prescription steroid ointment, Kenalog (triamcinolone) or Orabase; also tetracycline syrup or tetracycline capsules dissolved in water
- an anti-inflammatory ointment, Aphthasol, which is only modestly effective
- over-the-counter benzocaine preparations (Anbesol and Oragel); ointments such as Orabase or Zilactin-B to coat the ulcers and provide some protection and comfort
- for older children, zinc lozenges, taking vitamin C or vitamin B complex, using a sage and chamomile mouthwash, or taking lysine supplements
- tincture of propolis, available at health food stores, the "cement" made by honeybees to make their **hives** with remarkable antiseptic and healing properties
- for symptomatic relief of sores caused from food **allergies**, mixed equal amounts of milk of magnesia and Benadryl liquid, a teaspoon of which is swished by the child in his mouth for about one minute and then spat out, every four to six hours, to reduce pain

Alternative treatment

Alternative therapies for canker sores are meant to heal existing sores and prevent their recurrence. Several herbal remedies, including calendula (*Calendula officinalis*), myrrh (*Commiphora molmol*), and goldenseal (*Hydrastis canadensis*), may be helpful in treating existing sores. Compresses soaked in teas made from these herbs are applied directly to the sores. The tannic acid in a tea bag can also help dry up the sores when the wet tea bag is used as a compress. Taking dandelion (*Taraxacum officinale*) tea or capsules may help heal sores and prevent future outbreaks. Home remedies and herbal preparations may not be readily available as over-the-counter in forms suitable for pediatric patients. Since canker sores are often brought on by stress, stress-relieving techniques such as cuddling and rocking babies until they fall asleep may help relieve the stress associated with the severity of pain.

Prognosis

There is no cure for canker sores, and they occur more often with age. Treatments are to relieve symptoms or shorten the duration of the sore. If observation suggests a secondary infection, topical application of tetracycline to the lesion, three or four times daily, shortens healing to two to four days. Left untreated, canker sores can last as long as two weeks. Sores that persist for a longer time should be checked by a doctor.

Prevention

Children should avoid trauma, such as biting the inside of their mouth and tongue, or vigorous tooth brushing. Injury to the mucous membranes of the mouth and gums leaves places where bacteria can grow. This may make canker sores worse.

To decrease the incidence canker sores, parents may consider changing to a toothpaste free of sodium laurylsulfate.

KEY TERMS

Anti-inflammatory—A class of drugs, including nonsteroidal anti-inflammatory drugs (NSAIDs) and corticosteroids, used to relieve swelling, pain, and other symptoms of inflammation.

Inflammation—Pain, redness, swelling, and heat that develop in response to tissue irritation or injury. It usually is caused by the immune system's response to the body's contact with a foreign substance, such as an allergen or pathogen.

Over-the-counter treatments—Medications that can be purchased without a prescription.

Recurrent—Tendency to repeat.

Sore—A wound, lesion, or ulcer on the skin.

Ulcer—A site of damage to the skin or mucous membrane that is characterized by the formation of pus, death of tissue, and is frequently accompanied by an inflammatory reaction.

Nutritional concerns

Eating certain foods can be painful to a child with canker sores. Parents should remove spicy foods and citrus fruit from the child's diet. These foods may aggravate the sores and cause unnecessary pain. Parents should encourage their children to eat yogurt with active lactobacillus cultures because it may prevent outbreaks.

Parental concerns

Parents are mainly concerned with the comfort of their babies and small children. They must also pay attention to the nutritional intake of infants and small children who may refuse to eat because of the pain from canker sores.

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Aliene S. Linwood, RN, DPA, FACHE

Car sickness *see* **Motion sickness**

Carbohydrate intolerance

Definition

Carbohydrate intolerance is the inability of the small intestine to completely process the nutrient carbohydrate (a classification that includes sugars and starches) into a source of energy for the body. This is usually due to deficiency of an enzyme needed for digestion. **Lactose intolerance** is the inability to digest the sugar found in milk.

Description

Carbohydrates are the primary source of energy and, along with fats and proteins, one of the three major nutrients in the human diet. Carbohydrates are classified according to their structure, based on the number of basic sugar, or saccharide, units they contain.

A monosaccharide, called a simple sugar, is the simplest carbohydrate. Simple sugars include glucose (the form in which sugar circulates in the blood), fructose (found in fruit and honey), and galactose (produced by the digestion of milk). These simple sugars are important because they can be absorbed by the small intestine.

Two simple sugars linked together form a disaccharide. Disaccharide sugars present in the diet are maltose (a product of the digestion of starch), sucrose (table sugar), and lactose (the sugar in milk). These disaccharides must be broken down by enzymes into two simple sugars so that they can be absorbed by the intestine.

Polysaccharides are much more complex carbohydrates made up of many simple sugars. The most important polysaccharides are glycogen, which is stored in the liver, and cellulose (starch).

Digestion of sugars

Digestion of food begins in the mouth, moves on to the stomach, and then into the small intestine. Along the way, specific enzymes are needed to process different types of sugars. An enzyme is a substance that acts as a catalyst to produce chemical changes without being changed itself. The enzymes lactase, maltase, and isomaltase (or sucrase) are needed to break down the disaccharides; when one or more is inadequate, the result is carbohydrate intolerance.

Types of intolerance

Carbohydrate intolerance can be congenital, primary, or secondary. Congenital deficiency is caused by an enzyme defect present at birth. Alactasia is a very rare congenital condition and the result of a genetic defect

that causes the complete absence of lactase, the enzyme needed to digest milk sugar. Primary deficiency is caused by an enzyme defect developed over time. The most common is lactose intolerance. Secondary deficiencies, often caused by a disease or disorder of the intestinal tract, disappear when the underlying cause is treated. Secondary deficiencies include protein deficiency, pancreatitis, **celiac disease**, short-bowel syndrome, and some intestinal infections. Chronic renal failure and certain medications also can cause secondary deficiencies.

Demographics

Lactose intolerance is widespread, affecting about 20 percent of American children and up to 70 percent of the world's adult population. Lactose intolerance is the most common of all enzyme deficiencies, and an estimated 30–50 million Americans have this condition. Some racial and ethnic populations are affected more than others. Lactose intolerance is found in as many as 75 percent of African Americans, Jewish Americans, Mexican Americans, and Native Americans, and in 90 percent or more of Asian Americans and some Mediterranean peoples. Descendants of Northern Europe usually do not develop the condition (incidence is less than 20 percent in these populations). Deficiencies in enzymes other than lactase are extremely rare.

Causes and symptoms

Causes

Enzymes play an important role in breaking down carbohydrates into forms that can pass through the intestine and be used by the body. Usually they are named by adding *ase* to the name of the substance they act on (for example, lactase is the enzyme needed to process lactose). In the mouth, cooked starch is broken down to a disaccharide by amylase, an enzyme in the saliva. The disaccharides maltose, sucrose, and lactose cannot be absorbed until they have been separated into simple sugar molecules by their corresponding enzymes present in the cells lining the intestinal tract. If this process is not completed, digestion is interrupted.

Although not common, a deficiency in the enzymes needed to digest lactose, maltose, and sucrose is sometimes present at birth. Intestinal lactase enzymes usually decrease naturally with age, but this occurs at varying degrees. Because of the uneven distribution of enzyme deficiency based on race and ethnic heritage, especially in lactose intolerance, genetics are believed to play a role in the cause of primary carbohydrate intolerance.

Digestive diseases such as celiac disease and tropical sprue (which affect absorption in the intestine), as well as intestinal infections and injuries, can reduce the amount of enzymes produced. In **cancer** patients, treatment with radiation therapy or **chemotherapy** may affect the cells in the intestine that normally secrete lactase, leading to intolerance.

Symptoms

The severity of the symptoms depends upon the extent of the enzyme deficiency, and ranges from a feeling of mild bloating to severe **diarrhea**. In the case of a lactase deficiency, undigested milk sugar remains in the intestine, which is then fermented by the normal intestinal bacteria. These bacteria produce gas, cramping, bloating, a "gurgly" feeling in the abdomen, and flatulence. In a growing child, the main symptoms are diarrhea and a failure to gain weight. Lactase deficiency causes gastrointestinal distress to begin about 30 minutes to two hours after eating or drinking foods containing lactose.

Food intolerances can be confused with **food allergies**, since the symptoms of **nausea**, cramps, bloating, and diarrhea are similar. Food intolerances involve an exaggerated or abnormal physical reaction to a food or food additive, and are not associated with an immune reaction. Food **allergies** involve an immune reaction.

Sugars that are not broken down into one of the simplest forms cause the body to push fluid into the intestines, which results in watery diarrhea (osmotic diarrhea). The diarrhea may sweep other nutrients out of the intestine before they can be absorbed, causing **malnutrition**.

When to call the doctor

If a child has the following symptoms, the parent should contact the child's pediatrician or gastroenterologist:

- abdominal **pain**, **vomiting**, or diarrhea that awakens the child during the night
- persistent or severe abdominal pain or diarrhea
- unexplained weight loss
- rectal bleeding
- blood or mucus in stools
- fever

Diagnosis

The doctor may recommend a lactose-free diet for two or three weeks to determine if lactose intolerance is

causing the symptoms. During the lactose-free period, the child should avoid all products containing lactose. The parent and child are asked to record the intake of all foods and beverages and note when symptoms occur after eating or drinking.

To identify other problem-causing foods or beverages, it is helpful for the parent and child to keep a diary of symptoms for two or three weeks. The doctor can then review the diary with the parent and child to identify possible problem foods.

The diagnosis of carbohydrate or lactose intolerance is supported by the presence of symptoms related to the condition. In addition, the primary pediatrician or gastroenterologist may confirm the diagnosis after questioning the child (if old enough to provide an accurate history of symptoms) or parent about his or her physical health, performing a physical examination, and ordering laboratory tests to rule out other conditions that resemble carbohydrate intolerance.

When carbohydrate intolerance is suspected, the diagnosis can be confirmed using oral tolerance tests. The carbohydrate being investigated is given by mouth in liquid form. Several blood levels are measured and compared to normal values. This helps evaluate the individual's ability to digest the sugar.

To identify lactose intolerance in children and adults, the hydrogen breath test is used to measure the amount of hydrogen in the breath. The patient drinks a beverage containing lactose and the breath is analyzed at regular intervals. If undigested lactose in the large intestine (colon) is fermented by bacteria, various gases are produced. Hydrogen is absorbed from the intestines and carried by the bloodstream into the lungs, where it is exhaled. Normally, there is very little hydrogen detectable in the breath; therefore, its presence indicates faulty digestion of lactose.

When lactose intolerance is suspected in infants and young children, many pediatricians recommend simply changing from cow's milk to soy formula and watching for improvement. If needed, a stool sample can be tested for acidity. The inadequate digestion of lactose will result in an increase of acid in the waste matter excreted by the bowels and the presence of glucose.

Treatment

Carbohydrate intolerance caused by temporary intestinal diseases disappears when the condition is successfully treated. In primary conditions, no treatment exists to improve the body's ability to produce the enzymes, but symptoms can be controlled by diet.

An over-the-counter product marketed by the brand name Beano contains the enzyme alpha-galactosidase that works with the body's digestive system to break down complex carbohydrates into simple sugars that are easily digested. Beano is taken just before consuming gas-producing foods.

Nutritional concerns

Because there is wide variance in the degree of lactose intolerance, treatment should be tailored for the individual. Milk products should be avoided in young children who have signs of lactose intolerance. The child's doctor or a registered dietitian can help in making dietary adjustments and can advise when to start gradually reintroducing milk products, if applicable.

In infants, switching to soy-based formula may help. Special formulas, such as a glucose polymer-based formula, or a casein-based formula, may be recommended in infants with severe carbohydrate intolerance or when symptoms are severe.

Older children can adjust their intake of lactose, depending on how much and what they can tolerate. For some, a small glass of milk will not cause problems, while others may be able to handle ice cream or aged cheeses such as cheddar or Swiss, but not other dairy products. Generally, small amounts of lactose-containing foods eaten throughout the day are better tolerated than a large amount consumed all at once.

For those who are sensitive to even very small amounts of lactose, the lactase enzyme supplement is available without a prescription. The supplement is available in liquid form for use with milk. The addition of a few drops to a quart of milk will reduce the lactose content by 70 percent after 24 hours in the refrigerator. Heating the milk speeds the process, and doubling the amount of lactase liquid will result in milk that is 90 percent lactose free. Chewable lactase enzyme tablets are also available. Three to six tablets taken before a meal or snack will aid in the digestion of solid foods. Lactose-reduced milk and other products are also available in stores. Lactose-reduced milk contains the same nutrients as regular milk.

Because dairy products are an important source of calcium, people who reduce or severely limit their intake of these foods and beverages may need to consider other ways to consume an adequate amount of calcium. Taking calcium supplements or choosing other foods high in calcium may be needed to meet the recommended daily requirement of calcium. In addition, foods high in vitamin A, riboflavin, and vitamin B_{12} should be included in the daily diet to compensate for the nutrients normally found in cow's milk.

KEY TERMS

Alactasia—A rare inherited condition causing the lack of the enzyme needed to digest milk sugar.

Celiac disease—A disease, occurring in both children and adults, which is caused by a sensitivity to gluten, a protein found in grains. It results in chronic inflammation and shrinkage of the lining of the small intestine. Also called gluten enteropathy or nontropical sprue.

Cellulose—The primary substance composing the cell walls or fibers of all plant tissues.

Constipation—Difficult bowel movements caused by the infrequent production of hard stools.

Defecation—The act of having a bowel movement or the passage of feces through the anus.

Diarrhea—A loose, watery stool.

Digestion—The mechanical, chemical, and enzymatic process in which food is converted into the substances suitable for use by the body.

Enzyme—A protein that catalyzes a biochemical reaction without changing its own structure or function.

Feces—The solid waste, also called stool, that is left after food is digested. Feces form in the intestines and pass out of the body through the anus.

Gastroenterologist—A physician who specializes in diseases of the digestive system.

Hydrogen breath test—A test used to determine if a person is lactose intolerant or if abnormal bacteria are present in the colon.

Lactose—A sugar found in milk and milk products.

Metabolism—The sum of all chemical reactions that occur in the body resulting in growth, transformation of foodstuffs into energy, waste elimination, and other bodily functions. These include processes that break down substances to yield energy and processes that build up other substances necessary for life.

Nutrient—Substances in food that supply the body with the elements needed for metabolism. Examples of nutrients are vitamins, minerals, carbohydrates, fats, and proteins.

Peristalsis—Slow, rhythmic contractions of the muscles in a tubular organ, such as the intestines, that move the contents along.

Sugars—Those carbohydrates having the general composition of one part carbon, two parts hydrogen, and one part oxygen.

Alternative treatment

Alternative and complementary therapies include approaches that are considered to be outside the mainstream of traditional health care. The list of alternative treatments for carbohydrate intolerance includes aromatherapy, homeopathy, hydrotherapy, juice therapy, acupuncture, chiropractic, osteopathy, naturopathic medicine, and Chinese traditional herbal medicine.

Before learning or practicing any particular technique, it is important for the parent or caregiver and child to learn about the therapy, its safety and effectiveness, potential side effects, and the expertise and qualifications of the practitioner. Although some practices are beneficial, others may be harmful to certain patients.

Relaxation techniques and dietary supplements should not be used as a substitute for medical therapies prescribed by a doctor. Parents should discuss the alternative treatments with the child's doctor to determine the techniques and remedies that may be beneficial for the child.

Prognosis

Carbohydrate intolerance has a very low mortality rate. Newborns and infants have a higher risk of chronic diarrhea and malnutrition from carbohydrate intolerance. With good dietary management, children with carbohydrate intolerance can lead normal lives.

Prevention

Since the cause of the enzyme deficiency leading to carbohydrate intolerance is unknown, there is no way to prevent this condition.

Nutritional concerns

To help prevent or decrease the child's symptoms, parents can:

- help the child identify and avoid problematic foods
- work with a registered dietitian to facilitate specific dietary changes
- incorporate changes in the child's diet gradually, giving his or her body time to adjust
- establish set times for meals, and not permit the child to skip a meal
- encourage the child to drink at least eight glasses of water per day

- encourage the child to eat more slowly
- offer smaller, more frequent meals

Parental concerns

Parents should reinforce with the child that carbohydrate intolerance is not a life-threatening condition and that dietary changes can help reduce symptoms. They should remind the child that a few months may be needed before he or she notices substantial improvement in symptoms.

See also Lactose intolerance.

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Carbon monoxide poisoning

Definition

Carbon monoxide (CO) **poisoning** occurs when carbon monoxide gas is inhaled. CO is a colorless, odorless, highly poisonous gas that is produced by incomplete combustion. It is found in automobile exhaust fumes, faulty stoves and heating systems, fires, and cigarette smoke. Other sources include wood-burning stoves, kerosene heaters, improperly ventilated water heaters and gas stoves, and blocked or poorly maintained chimney flues. CO interferes with the ability of the blood to carry oxygen. The result is **headache**, **nausea**, convulsions, and finally death by asphyxiation.

Description

Carbon monoxide, sometimes called coal gas, has been known as a toxic substance since the third century B.C. It was used for executions and suicides in early Rome.

Anyone who is exposed to CO becomes sick, and the entire body is involved in CO poisoning. A developing fetus can also be poisoned if a pregnant woman breathes CO gas. Infants, people with heart or lung disease, or those with anemia may be more seriously affected. People such as underground parking garage attendants who are exposed to car exhausts in a confined area are more likely to be poisoned by CO. Firemen also run a higher risk of inhaling CO.

Demographics

Carbon monoxide is the leading cause of accidental poisoning in the United States. Experts estimate that 1,500 Americans die each year from accidental exposure to CO and another 2,300 from intentional exposure (**suicide**). An additional 10,000 people seek medical attention after exposure to CO and recover.

Causes and symptoms

Normally when a person breathes fresh air into the lungs, the oxygen in the air binds with a molecule called hemoglobin (Hb) that is found in red blood cells. This process allows oxygen to be moved from the lungs to every part of the body. When the oxygen/hemoglobin complex reaches a muscle where it is needed, the oxygen is released. Because the oxygen binding process is reversible, hemoglobin can be used over and over again to pick up oxygen and move it throughout the body.

Inhaling carbon monoxide gas interferes with this oxygen transport system. In the lungs, CO competes with oxygen to bind with the hemoglobin molecule. Hemoglobin prefers CO to oxygen and accepts it more than 200 times more readily than it accepts oxygen. Not only does the hemoglobin prefer CO, it holds on to the CO much more tightly, forming a complex called carboxyhemoglobin (COHb). As a person breathes CO contaminated air, more and more oxygen transportation sites on

the hemoglobin molecules become blocked by CO. Gradually, there are fewer and fewer sites available for oxygen. All cells need oxygen to live. When they do not get enough oxygen, cellular metabolism is disrupted and eventually cells begin to die.

The symptoms of CO poisoning and the speed with which they appear depend on the concentration of CO in the air and the rate and efficiency with which a person breathes. Heavy smokers can start off with up to 9 percent of their hemoglobin already bound to CO, which they regularly inhale in cigarette smoke. This makes them much more susceptible to environmental CO. The Occupational Safety and Health Administration (OSHA) has established a maximum permissible exposure level of 50 parts per million (ppm) over eight hours.

With exposure to 200 ppm for two to three hours, a person begins to experience headache, fatigue, nausea, and **dizziness**. These symptoms correspond to 15 to 25 percent COHb in the blood. When the concentration of COHb reaches 50 percent or more, death results in a very short time. Emergency room physicians have the most experience diagnosing and treating CO poisoning.

The symptoms of CO poisoning in order of increasing severity include the following:

- headache
- shortness of breath
- dizziness
- fatigue
- mental confusion and difficulty thinking
- loss of fine hand-eye coordination
- nausea and vomiting
- rapid heart rate
- hallucinations
- inability to execute voluntary movements accurately
- collapse
- lowered body temperature (hypothermia)
- coma
- convulsions
- seriously low blood pressure
- cardiac and respiratory failure
- death

In some cases, the skin, mucous membranes, and nails of a person with CO poisoning are cherry red or bright pink. Because the color change does not always occur, it is an unreliable symptom to count on for diagnosis.

Although most CO poisoning is acute, or sudden, it is possible to suffer from chronic CO poisoning. This condition exists when a person is exposed to low levels of the gas over a period of days to months. Symptoms are often vague and include (in order of frequency) fatigue, headache, dizziness, **sleep** disturbances, cardiac symptoms, apathy, nausea, and memory disturbances. Little is known about chronic CO poisoning, and it is often misdiagnosed.

When to call the doctor

A healthcare professional should be consulted whenever more than passing exposure to carbon monoxide is suspected. While waiting for help to arrive, a potentially affected person should be moved outdoors.

Diagnosis

The main reason to suspect CO poisoning is evidence that fuel is being burned in a confined area, for example, a car running inside a closed garage, a charcoal grill burning indoors, or an unvented kerosene heater in a workshop. Under these circumstances, one or more persons suffering from the symptoms listed above strongly suggests CO poisoning. In the absence of some concrete reason to suspect CO poisoning, the disorder is often misdiagnosed as migraine headache, **stroke**, psychiatric illness, **food poisoning**, alcohol poisoning, or heart disease.

Concrete confirmation of CO poisoning comes from a carboxyhemoglobin test. This blood test measures the amount of CO that is bound to hemoglobin in the body. Blood is drawn as soon after suspected exposure to CO as possible.

Other tests that are useful in determining the extent of CO poisoning include measurement of other arterial blood gases and pH; a complete blood count; measurement of other blood components such as sodium, potassium, bicarbonate, urea nitrogen, and lactic acid; an electrocardiogram (ECG); and a chest x ray.

Treatment

Immediate treatment for CO poisoning is to remove the victim from the source of carbon monoxide gas and into fresh air. If the victim is not breathing and has no pulse, **cardiopulmonary resuscitation** (CPR) should be started. Depending on the severity of the poisoning,

100 percent oxygen may be given with a tight fitting mask as soon as it is available.

Taken with other symptoms of CO poisoning, COHb levels of over 25 percent in healthy individuals, over 15 percent in people with a history of heart or lung disease, and over 10 percent in pregnant women usually indicate the need for **hospitalization**. In the hospital, fluids and electrolytes are given to correct imbalances that have arisen from the breakdown of cellular metabolism.

In severe cases of CO poisoning, individuals are given hyperbaric oxygen therapy. This treatment involves placing the person in a chamber in which the person breathes 100 percent oxygen at a pressure of more than one atmosphere (the normal pressure the atmosphere exerts at sea level). The increased pressure forces more oxygen into the blood. Hyperbaric facilities are specialized and are usually available only at larger hospitals.

Prognosis

The speed and degree of recovery from CO poisoning depends on the length of exposure to the gas and the concentration of carbon monoxide. The half-life of CO in normal room air is four to five hours, which means that in four to five hours half of the CO bound to hemoglobin will be replaced with oxygen. At normal atmospheric pressures, but breathing 100 percent oxygen, the half-life for the elimination of CO from the body is 50 to 70 minutes. In hyperbaric therapy at three atmospheres of pressure, the half-life is reduced to between 20 and 25 minutes.

Although the symptoms of CO poisoning may subside in a few hours, some affected persons show memory problems, fatigue, confusion, and mood changes for two to four weeks after their exposure to the gas.

Prevention

Carbon monoxide poisoning is preventable. Particular care should be paid to situations where fuel is burned in a confined area. Portable and permanently installed carbon monoxide detectors that sound a warning similar to smoke detectors are available for under $50. Specific actions that prevent CO poisoning include the following:

- Stop **smoking**. Smokers have less tolerance to environmental CO.
- Have heating systems and appliances installed by a qualified contractor to assure that they are properly vented and meet local building codes.

KEY TERMS

Carboxyhemoglobin—Hemoglobin that is bound to carbon monoxide instead of oxygen.

Hemoglobin—An iron-containing pigment of red blood cells composed of four amino acid chains (alpha, beta, gamma, delta) that delivers oxygen from the lungs to the cells of the body and carries carbon dioxide from the cells to the lungs.

Hypothermia—A serious condition in which body temperature falls below 95°F (35 °C). It is usually caused by prolonged exposure to the cold.

pH—A measurement of the acidity or alkalinity of a solution. Based on a scale of 14, a pH of 7.0 is neutral. A pH below 7.0 is an acid; the lower the number, the stronger the acid. A pH above 7.0 is a base; the higher the number, the stronger the base. Blood pH is slightly alkaline (basic) with a normal range of 7.36–7.44.

- Inspect and properly maintain heating systems, chimneys, and appliances.
- Do not use a gas oven or stove to heat the home.
- Do not burn charcoal indoors.
- Make sure there is good ventilation if using a kerosene heater indoors.
- Do not leave cars or trucks running inside the garage.
- Keep car windows rolled up when stuck in heavy traffic, especially if inside a tunnel.

Parental concerns

Parents should not allow children to **play** in areas heated by kerosene space heaters or to use charcoal grills of any kind indoors.

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American Academy of Pediatrics. 141 Northwest Point Boulevard, Elk Grove Village, IL 60007–1098. Web site: <www.aap.org/default.htm>.

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American College of Emergency Physicians. PO Box 619911, Dallas, TX 75261–9911. Web site: <www.acep.org/>.

American College of Hyperbaric Medicine. PO Box 25914–130, Houston, Texas 77265. Web site: <www.hyperbaricmedicine.org/>.

American College of Occupational and Environmental Medicine. 55 West Seegers Road, Arlington Heights, IL 60005. Web site: <www.acoem.org/>.

American College of Osteopathic Emergency Physicians. 142 E. Ontario Street, Suite 550, Chicago, IL 60611. Web site: <www.acoep.org/>.

International Congress on Hyperbaric Medicine. 1592 Union Street, San Francisco, CA 94123. Web site: <www.ichm.net/>.

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Cardiopulmonary resuscitation

Definition

Cardiopulmonary resuscitation (CPR) is a procedure to support and maintain breathing and circulation for an infant, child, or adolescent who has stopped breathing (respiratory arrest) and/or whose heart has stopped (cardiac arrest).

Purpose

CPR is performed to restore and maintain breathing and circulation and to provide oxygen and blood flow to the heart, brain, and other vital organs. CPR can be performed by trained laypeople or healthcare professionals on infants, children, adolescents, and adults. CPR should be performed if an infant, child, or adolescent is unconscious and not breathing. Respiratory and cardiac arrest can be caused by allergic reactions, an ineffective heartbeat, asphyxiation, breathing passages that are blocked, **choking**, drowning, drug

reactions or overdoses, electric shock, exposure to cold, severe shock, or trauma. In newborns, the most common cause of cardiopulmonary arrest is respiratory failure caused by **sudden infant death syndrome** (SIDS), airway obstruction (usually from inhalation of a foreign body), sepsis, neurologic disease, or drowning. Cardiac arrest in children over one year of age is most commonly caused by shock and/or respiratory failure resulting from an accident or injury.

Description

CPR is part of the emergency cardiac care system designed to save lives. Many deaths can be prevented by prompt recognition of cardiopulmonary arrest and notification of the emergency medical system (EMS), followed by early CPR, defibrillation (which delivers a brief electric shock to the heart in attempt to get the heart to beat normally), and advanced cardiac life support measures. When performed by a layperson, CPR is designed to support and maintain breathing and circulation until emergency medical personnel arrive and take over. When performed by healthcare personnel, it is used in conjunction with other basic and advanced life support measures.

CPR must be performed within four to six minutes after cessation of breathing to prevent brain damage or death. CPR consists of rescue breathing, which delivers oxygen to the victim's lungs, and external chest compressions, which help circulate blood through the heart to vital organs.

CPR technique differs for infants, children, and adolescents. The American Heart Association and the American Red Cross, the two organizations that provide CPR training and guidelines, distinguish infants, children, and adolescents for the purposes of CPR as follows:

- "Infant" includes neonates (those in the first 28 days of life) and extends to the age of one year.
- "Child" includes toddlers aged one year to children aged eight years.
- "Adult" includes children aged eight years and older.

Because infants and children under the age of eight have smaller upper and lower airways and faster heart rates than adults, CPR techniques are different for them than for older children and adults. Children and adolescents aged eight years and older have reached a body size that can be handled using adult CPR techniques and are thus classified as adults for delivery of CPR and life support. CPR is always begun after assessing the victim and contacting EMS.

Performing CPR on an infant

For an infant, the rescuer opens the airway using a gentle head tilt/chin lift or jaw thrust, places their mouth over the infant's mouth and nose then delivers gentle breaths so that the infant's chest rises with each breath. Chest compressions are delivered by placing two fingers of one hand over the lower half of the infant's sternum slightly below the nipple line and pressing down about one half inch to one inch. Compressions are delivered at a rate of 100 times per minute, giving five chest compressions followed by one rescue breath in successive cycles.

Performing CPR on a child aged one to eight

For a child aged one to eight years, the compression rate is the same—five compressions and one rescue breath. Rescue breaths are delivered using a mouth-to-mouth seal, instead of mouth-to-mouth-and-nose. Chest compressions are delivered by placing the heel of one hand over the lower half of the sternum and depressing about one to one and one half inches per compression.

Performing CPR on a child aged eight and older

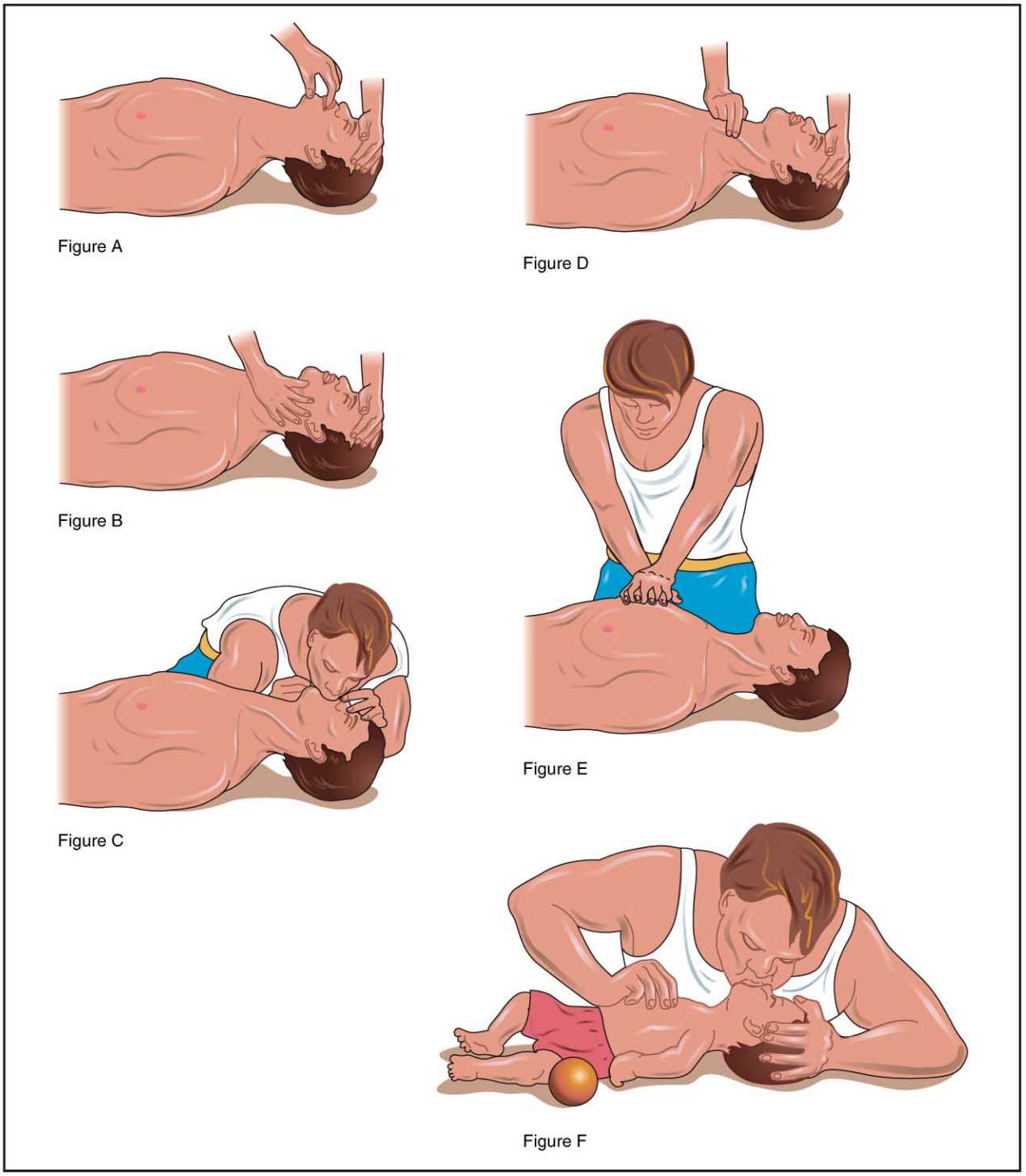
For a child aged eight years and older, and for larger children under age eight, two hands are used for compressions, with the heel of one hand on the lower half of the sternum and the heel of the other hand on top of that hand. The chest is compressed about one and one half to two inches per compression. Rescue breaths are delivered with a mouth-to-mouth seal. The compression rate is 80 to 100 per minute delivered in cycles of 15 compressions followed by two rescue breaths.

Preparation

Before administering CPR to an infant or child, laypeople should participate in hands-on training. More than 5 million Americans annually receive training in CPR through American Heart Association and American Red Cross courses. In addition to courses taught by instructors, the American Heart Association also has an interactive video called *Learning System*, which is available at more than 500 healthcare institutions. Both organizations teach CPR the same way, but they use different terms. CPR training should be retaken every two to three years to maintain skill level.

Precautions

CPR should not be performed based on the overview contained in this article. To prevent disease transmission during CPR, face masks and face shields are available to prevent direct contact during rescue breathing.



CPR in basic life support. Figure A: The victim should be flat on his back and his mouth should be checked for debris. Figure B: If the victim is unconscious, open airway, lift neck, and tilt head back. Figure C: If victim is not breathing, begin artificial breathing with four quick full breaths. Figure D: Check for carotid pulse. Figure E: If pulse is absent, begin artificial circulation by depressing sternum. Figure F: Mouth-to-mouth resuscitation of an infant. *(Illustration by Electronic Illustrators Group.)*

Aftercare

Emergency medical care is always necessary after CPR. Once a person's breathing and heartbeat have been is coming and talk positively until professionals arrive restored, the rescuer should make the person comfortable and stay there until emergency medical personnel arrive. The rescuer can continue to reassure the person that help and take over.

KEY TERMS

Cardiac arrest—Temporary or permanent cessation of the heartbeat.

Cardiopulmonary—Relating to the heart and lungs.

Defibrillation—A procedure to stop the type of irregular heart beat called ventricular fibrillation, usually by using electric shock.

Resuscitation—Bringing a person back to life or consciousness after he or she was apparently dead.

Ventricular fibrillation—An arrhythmia characterized by a very rapid, uncoordinated, ineffective series of contractions throughout the lower chambers of the heart. Unless stopped, these chaotic impulses are fatal.

Risks

CPR can cause injury to a person's ribs, liver, lungs, and heart. However, these risks must be accepted if CPR is necessary to save the person's life.

Normal results

In many cases, successful CPR results in restoration of consciousness and life. Barring other injuries, a revived person usually returns to normal functions within a few hours of being revived.

Abnormal results include injuries incurred during CPR and lack of success with CPR. Possible sites for injuries include a person's ribs, liver, lungs, and heart. Partially successful CPR may result in brain damage. Unsuccessful CPR results in death.

Parental concerns

Because most cardiopulmonary arrest in infants and children occurs in or around the home and results from SIDS, trauma, drowning, choking, or **poisoning**, all parents and child caregivers should consider becoming trained in CPR. Training is available at local schools and community centers.

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Casts *see* **Immobilization**

Cat bite infection *see* **Animal bite infections**

CAT scan *see* **Computed tomography**

Cat's cry syndrome *see* **Cri du chat syndrome**

Cat-scratch disease

Definition

Cat-scratch disease is an uncommon infection that typically results from a cat's scratch or bite. Most

sufferers experience only moderate discomfort and find that their symptoms clear up without any lasting harm after a few weeks or months. Professional medical treatment is rarely needed.

Description

Cat-scratch disease (also called cat-scratch fever) is caused by the *Bartonella henselae* bacterium, which is found in cats around the world and is transmitted from cat to cat by fleas. Researchers have discovered that large numbers of North American cats carry antibodies for the disease (meaning that the cats have been infected at some point in their lives). Some parts of North America have much higher rates of cat infection than others, however. *Bartonella henselae* is uncommon or absent in cold climates, which fleas have difficulty tolerating, but prevalent in warm, humid places such as Memphis, Tennessee, where antibodies were found in 71 percent of the cats tested. The bacterium, which remains in a cat's bloodstream for several months after infection, seems to be harmless to most cats, and normally an infected cat will not display any symptoms. Kittens (cats younger than one year old) are more likely than adult cats to be carrying the infection.

Demographics

Bartonella henselae can infect people who are scratched or (more rarely) bitten or licked by a cat. It cannot be passed from person to person. Although cats are popular pets found in about 30 percent of American households, human infection appears to be rare. One study estimated that for every 100,000 Americans there are only 2.5 cases of cat-scratch disease each year. It is also unusual for more than one **family** member to become ill; a Florida investigation discovered multiple cases in only 3.5 percent of the families studied. Children and teenagers appear to be the most likely victims of cat-scratch disease, although the possibility exists that the disease may be more common among adults than previously thought.

Causes and symptoms

The first sign of cat-scratch disease may be a small blister at the site of a scratch or bite three to ten days after injury. The blister (which sometimes contains pus) often looks like an insect bite and is usually found on the hands, arms, or head. Within two weeks of the blister's appearance, lymph nodes near the site of injury become swollen. Often the infected person develops a fever or experiences fatigue or headaches. The symptoms usually disappear within a month, although the lymph nodes may remain swollen for several months. Hepatitis, **pneumonia**, and other dangerous complications can arise, but the likelihood of cat-scratch disease posing a serious threat to health is very small. **AIDS** patients and other immunocompromised people face the greatest risk of dangerous complications.

Occasionally, the symptoms of cat-scratch disease take the form of what is called Parinaud's oculoglandular syndrome. In such cases, a small sore develops on the palpebral conjunctiva (the membrane lining the inner eyelid) and is often accompanied by **conjunctivitis** (inflammation of the membrane) and swollen lymph nodes in front of the ear. Researchers suspect that the first step in the development of Parinaud's oculoglandular syndrome occurs when *Bartonella henselae* bacteria pass from a cat's saliva to its fur during grooming. Rubbing one's eyes after handling the cat then transmits the bacteria to the conjunctiva.

Diagnosis

A family doctor should be called whenever a cat scratch or bite fails to heal normally or is followed by a persistent fever or other unusual symptoms such as long-lasting bone or joint **pain**. The appearance of painful and swollen lymph nodes is another reason for consulting a doctor. When cat-scratch disease is suspected, the doctor will ask about a history of exposure to cats and look for evidence of a cat scratch or bite and swollen lymph nodes. A blood test for *Bartonella henselae* may be ordered to confirm the doctor's diagnosis.

Treatment

For otherwise healthy people, rest and over-the-counter medications for reducing fever and discomfort (such as **acetaminophen**) while waiting for the disease to run its course are usually all that is necessary. **Antibiotics** are prescribed in some cases, particularly when complications occur or the lymph nodes remain swollen and painful for more than two or three months, but there is no agreement among doctors about when and how they should be used. If a lymph node becomes very swollen and painful, the family doctor may decide to drain it.

Prognosis

Most people recover completely from a bout of cat-scratch disease. Further attacks are rare.

Prevention

Certain common-sense precautions can be taken to guard against the disease. Scratches and **bites** should be washed immediately with soap and water, and it is never a good idea to rub one's eyes after handling a cat without first washing one's hands. Children should be told not to **play** with stray cats or make cats angry. Immunocompromised people should avoid owning kittens, which are

KEY TERMS

Acetaminophen—A drug used for pain relief as well as to decrease fever. A common trade name for the drug is Tylenol.

Acquired immunodeficiency syndrome (AIDS)—An infectious disease caused by the human immunodeficiency virus (HIV). A person infected with HIV gradually loses immune function, becoming less able to resist other infections and certain cancers.

Antibiotics—Drugs that are designed to kill or inhibit the growth of the bacteria that cause infections.

Antibody—A special protein made by the body's immune system as a defense against foreign material (bacteria, viruses, etc.) that enters the body. It is uniquely designed to attack and neutralize the specific antigen that triggered the immune response.

Bacteria—Singluar, bacterium; tiny, one-celled forms of life that cause many diseases and infections.

Hepatitis—An inflammation of the liver, with accompanying liver cell damage or cell death, caused most frequently by viral infection, but also by certain drugs, chemicals, or poisons. May be either acute (of limited duration) or chronic (continuing). Symptoms include jaundice, nausea, vomiting, loss of appetite, tenderness in the right upper abdomen, aching muscles, and joint pain. In severe cases, liver failure may result.

Immune system—The system of specialized organs, lymph nodes, and blood cells throughout the body that work together to defend the body against foreign invaders (bacteria, viruses, fungi, etc.).

Immunocompromised—A state in which the immune system is suppressed or not functioning properly.

Lymph nodes—Small, bean-shaped collections of tissue located throughout the lymphatic system. They produce cells and proteins that fight infection and filter lymph. Nodes are sometimes called lymph glands.

Pneumonia—An infection in which the lungs become inflamed. It can be caused by nearly any class of organism known to cause human infections, including bacteria, viruses, fungi, and parasites.

Pus—A thick, yellowish or greenish fluid composed of the remains of dead white blood cells, pathogens, and decomposed cellular debris. It is most often associated with bacterial infection.

more likely than adult cats to be infectious. Because cat-scratch disease is usually not a life-threatening illness and people tend to form strong emotional bonds with their cats, doctors do not recommend getting rid of a cat suspected of carrying the disease.

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Cavities, dental *see* **Tooth decay**

Celiac disease

Definition

Celiac disease is a disease of the digestive system in which the inside lining of the small intestine (mucosa) is damaged after eating wheat, rye, oats, or barley, resulting in interference with the absorption of nutrients from food.

Description

Celiac disease occurs when the body reacts abnormally to gluten, a protein found in grains, including wheat, rye, barley, and possibly oats. When someone with celiac disease eats foods containing gluten, that person's immune system causes an inflammatory response in the small intestine, which damages the tissues and results in impaired ability to absorb nutrients from foods (malabsorption). The inflammation and malabsorption create wide-ranging problems in many systems of the body. Since the body's own immune system causes the damage, celiac disease is classified as an autoimmune disorder.

Each person with celiac disease is affected differently. When food containing gluten reaches the small intestine, the immune system begins to attack a substance called gliadin, which is found in the gluten. The resulting inflammation causes damage to the delicate finger-like structures in the intestine, called villi, where food absorption actually takes place. This damage is referred to as villus atrophy. The patient may experience a number of symptoms related to the inflammation and the chemicals it releases, and/or the lack of ability to absorb nutrients from food, which can cause **malnutrition**.

Celiac disease is also called sprue, nontropical sprue, gluten sensitive enteropathy, and celiac sprue.

Demographics

Celiac disease may be discovered at any age, from infancy through adulthood. The disorder is more commonly found among white Europeans and in people of European descent. It is very unusual to find celiac disease in African or Asian people. The exact incidence of the disease is uncertain. Estimates vary from one in 5,000, to as many as one in every 300 individuals with this background. The prevalence of celiac disease seems to be different from one European country to another and between Europe and the United States. This discrepancy may be due to differences in diet and/or the possibility that the disease goes unrecognized in some areas. One study of random blood samples tested for celiac disease in the United States showed one in 250 testing positive. It is clearly underdiagnosed, probably because the symptoms are attributed to another problem, and physicians and laboratory technicians lack knowledge about celiac disease.

Because celiac disease has a hereditary influence or genetic component, close relatives (especially first-degree relatives, such as children, siblings, and parents) have a higher risk of being affected with the condition. The chance that a first-degree relative of someone with celiac disease has the disease is about 10 percent.

Causes and symptoms

The pattern of inheritance is complicated regarding this disease. The type of inheritance pattern that celiac disease follows is called multifactorial (caused by many factors, both genetic and environmental). Researchers think that several factors must exist in order for the disease to occur. The patient must have a genetic predisposition to develop the disorder. Then something in their environment acts as a stimulus, or trigger, to their immune system, causing the disease to become active for the first time. For conditions with multifactorial inheritance, people without the genetic predisposition are less likely to develop the condition with exposure to the same triggers, or they may require more exposure to the stimulus before developing the disease than someone with a genetic predisposition. Stimuli that may provoke a reaction include surgery, especially gastrointestinal surgery; a change to a low fat diet, which includes an increased number of wheat-based foods; severe emotional stress; or a viral infection. The combination of genetic susceptibility and an outside agent leads to celiac disease.

The most commonly recognized symptoms of celiac disease relate to the improper absorption of food in the gastrointestinal system. Many patients with gastrointestinal symptoms will have **diarrhea** and fatty, greasy, unusually foul-smelling stools. The patient may complain of excessive gas (flatulence), distended abdomen, weight loss, and generalized weakness. Not all people have digestive system complications; some people only have irritability or depression. Irritability is one of the most common symptoms in children with celiac disease.

Not all individuals with celiac disease exhibit typical symptoms. As more is learned about celiac disease, it has become evident that the disease has many variations that may not produce typical symptoms. Unrecognized and therefore untreated celiac disease may cause or contribute to a variety of other conditions. The decreased ability to digest, absorb, and utilize food properly (malabsorption) may cause anemia (low red blood count from iron deficiency) or easy bruising from a lack of vitamin K. Poor mineral absorption may result in osteoporosis, or brittle bones, which may lead to bone **fractures**. Vitamin D levels may be insufficient and bring about a softening of bones (osteomalacia), which produces **pain** and bony deformities, such as flattening or bending. Defects in the tooth enamel, characteristic of celiac disease, may be recognized by dentists. Celiac disease may be discovered during medical tests performed to investigate **failure to thrive** in infants or lack of proper growth in children and adolescents. People with celiac disease may also experience **lactose intolerance** because they do not produce enough of the enzyme lactase, which breaks down the sugar in milk into a form the body can absorb. Other symptoms can include **muscle cramps**, fatigue, delayed growth, tingling or **numbness** in the legs (from nerve damage), pale sores in the mouth (called aphthus ulcers), tooth discoloration, or missed menstrual periods (due to severe weight loss).

A distinctive, painful skin rash, called **dermatitis** herpetiformis, may be the first sign of celiac disease in adults but rarely occurs in children with celiac disease.

Many disorders are associated with celiac disease, although the nature of the connection is unclear. One type of epilepsy is linked to celiac disease. Once their celiac disease is successfully treated, a significant number of these patients have fewer or no seizures. Patients with **alopecia** areata, a condition in which hair loss occurs in sharply defined areas, have been shown to have a higher risk of celiac disease than the general population. There appears to be a higher percentage of celiac disease among people with **Down syndrome**, but the link between the conditions was unknown as of 2004.

Several conditions attributed to a disorder of the immune system have been associated with celiac disease. People with insulin-dependent diabetes (type I) have a much higher incidence of celiac disease. One source estimates that as many as one in 20 insulin-dependent diabetics may have celiac disease. Patients with other conditions in which celiac disease may be more commonly found include those with juvenile chronic arthritis, some thyroid diseases, and IgA deficiency.

There is an increased risk of intestinal lymphoma, a type of **cancer**, in individuals with celiac disease. Successful treatment of the celiac disease seems to decrease the chance of developing lymphoma.

When to call the doctor

A doctor should be consulted when a child exhibits symptoms characteristic of this disease.

Diagnosis

Because of the variety of ways celiac disease can manifest itself, it is often not discovered promptly. Its symptoms are similar to many other conditions including **irritable bowel syndrome**, Crohn's disease, ulcerative colitis, diverticulosis, intestinal infections, chronic fatigue syndrome, and depression. The condition may persist without diagnosis for so long that the patient accepts a general feeling of illness as normal. This acceptance leads to further delay in identifying and treating the disorder. It is not unusual for the disease to be identified in the course of medical investigations for seemingly unrelated problems.

If celiac disease is suspected, based on symptoms, physical appearance, or delayed growth, a blood test should be ordered. This test looks for the antibodies to gluten (called antigliadin, anti-endomysium, and antireticulin) that the immune system produces in celiac disease. Antibodies are chemicals produced by the immune system in response to substances such as germs and other potentially harmful substances. Some experts advocate not just evaluating patients with symptoms, but using these blood studies as a screening test for high-risk individuals, such as those with relatives (especially first-degree relatives) known to have the disorder. An abnormal result points toward celiac disease, but further tests are needed to confirm the diagnosis. Because celiac disease affects the ability of the body to absorb nutrients from food, several tests may be ordered to look for nutritional deficiencies. For example, doctors may order a test of iron levels in the blood because low levels of iron (anemia) may accompany celiac disease. Doctors may also order a test for fat in the stool, since celiac disease prevents the body from absorbing fat from food.

If these tests are suspicious for celiac disease, the next step is a biopsy (surgical removal of a tiny piece of tissue) of the small intestine. This is usually done by a gastroenterologist, a physician who specializes in diagnosing and treating bowel disorders. It is generally performed in the office or in a hospital's outpatient department. The patient remains awake but is sedated. A narrow tube, called an endoscope, is passed through the mouth, down through the stomach, and into the small intestine. A small sample of tissue is taken and sent to the laboratory for analysis. If it shows a pattern of tissue damage characteristic of celiac disease, the diagnosis is established.

Treatment

The only treatment for celiac disease is a gluten-free diet (GFD). This diet is easy for the doctor to prescribe but may be difficult for a child to follow. For most people, adhering to this diet stops symptoms and prevents damage to the intestines. Damaged villi can be functional again in three to six months. This diet must be followed permanently, however. The fact that people had symptoms that were cured by the GFD is further evidence that the diagnosis was correct.

The physician will periodically recheck the level of antibody in the child's blood. After several months, the small intestine is biopsied again. If the diagnosis of celiac disease was correct (and the child followed the rigorous diet), healing of the intestine will be apparent. Most experts agree that it is necessary to follow these steps in order to be sure of an accurate diagnosis. Disorders other than celiac disease can cause a similar type of villus atrophy, especially in children under two years of age, so rechecking the intestine is especially important for very young children. If healing is evident, then gluten is reintroduced to the diet and a third biopsy is performed

weeks to months later to see if the reintroduction of gluten results in villus atropy again. If the atrophy returns, the child has celiac disease, and a gluten-free diet should be continued for life.

A child with undiagnosed celiac disease may become very ill with severe diarrhea and malnutrition. Corticosteroids such as prednisone and intravenous (IV) fluids may be temporarily given while the child begins a GFD. Because celiac disease is diagnosed more quickly than in the past, corticosteroids are seldom required.

Nutritional concerns

Although there is no risk and much potential benefit to the use of GFD for treatment of celiac disease, the widespread use of gluten-containing grains in Western cultures makes adapting to a gluten-free diet challenging. Gluten is present in any product that contains wheat, rye, barley, or oats. It helps make bread rise and gives many foods a smooth, pleasing texture. In addition to the many obvious places gluten can be found in a normal diet, such as breads, cereals, and pasta, there are many hidden sources of gluten. Thickening agents, emulsifiers, fillers, flavor enhancers, and food stabilizers as well as products used in food packaging may contain gluten. Gluten may even be present on surfaces used for food preparation or cooking.

Fresh foods that have not been artificially processed, such as fruits, vegetables, and meats, are permitted as part of a GFD. Gluten-free foods can be found in health food stores and in some supermarkets. Mail-order food companies often have a selection of gluten-free products. Help in dietary planning is available from dieticians (healthcare professionals specializing in food and **nutrition**) or from support groups for individuals with celiac disease. There are many cookbooks on the market specifically for those on a GFD.

Prognosis

Treating celiac disease with a strict GFD is almost always completely effective. Gastrointestinal complaints and other symptoms are alleviated. Secondary complications, such as anemia and osteoporosis, resolve in almost all patients. People who have experienced lactose intolerance related to their celiac disease usually see those symptoms subside as well.

Once the diet has been followed for several years, individuals with celiac disease have similar mortality rates as the general population. However, about 10 percent of people with celiac disease develop a cancer involving the gastrointestinal tract (both carcinoma and lymphoma).

A few patients develop a refractory type of celiac disease, in which the GFD no longer seems effective. Once the diet has been thoroughly assessed to ensure no hidden sources of gluten are causing the problem, medications may be prescribed. Steroids or immunosuppressant drugs are often used to try to control the disease. It is unclear whether these efforts meet with much success.

Experts emphasize the need for lifelong adherence to the GFD to avoid the long-term complications of this disorder. They point out that although the disease may have symptom-free periods if the diet is not followed, silent damage continues to occur. Celiac disease cannot be outgrown or cured, according to medical authorities.

Prevention

There is no way to prevent celiac disease. However, the key to decreasing its impact on overall health is early diagnosis and strict adherence to the prescribed GFD.

Parental concerns

For parents used to preparing gluten-containing meals, searching for and cooking with gluten-free products may be difficult at first. Changing cooking habits will be easier if initially gluten-free recipes and food products are used. When they use any commercial food products, they must read carefully the list of ingredients. Although ingredients are listed in order of decreasing content, any product containing the smallest of amount of gluten must be avoided. Many food manufacturers are willing to provide additional information about their products. Most food labels contain addresses of the manufacturers and many include a toll-free telephone number. Some restaurants have ingredient lists for their products posted in the restaurant or available on request.

When a child with celiac disease eats at a friend's house, the friend's parent should be aware of the child's dietary limitations. The child may have to take lunch from home to eat at school, unless the school has a dietician who can ensure that gluten-free food is provided for the child.

Family support is important in ensuring acceptance of the diet. The child must not be made to feel that he/she is abnormal and a nuisance to the family. After the GFD is begun, the benefits to the child with celiac disease will initially be obvious and enthusiastically accepted. However, as the child gets older, the period of ill health may be forgotten, and the child may be reject the diet, especially during **adolescence**, when there is a desire for conformity. Unfortunately, in older children, the symptoms may not reappear immediately although intestinal damage is occurring. The child may interpret the delay

KEY TERMS

Antibody—A special protein made by the body's immune system as a defense against foreign material (bacteria, viruses, etc.) that enters the body. It is uniquely designed to attack and neutralize the specific antigen that triggered the immune response.

Antigen—A substance (usually a protein) identified as foreign by the body's immune system, triggering the release of antibodies as part of the body's immune response.

Gluten—A protein found in wheat, rye, barley, and oats.

Immune system—The system of specialized organs, lymph nodes, and blood cells throughout the body that work together to defend the body against foreign invaders (bacteria, viruses, fungi, etc.).

Villi—Tiny, finger-like projections that enable the small intestine to absorb nutrients from food.

in the return of symptoms as evidence that the child has recovered from celiac disease, but they have not, as celiac disease cannot be cured.

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ORGANIZATIONS

American Celiac Society. 58 Musano Court, West Orange, NJ 07052. Telephone: 201/325–8837.

Celiac Disease Foundation. 13251 Ventura Blvd., Suite 1, Studio City, CA 91604–1838. Web site: <http://celiac.org>.

Celiac Sprue Association/United State of America (CSA/USA). PO Box 31700, Omaha, NE 68131–0700. Web site: <www.csaceliacs.org>.

Gluten Intolerance Group of North America. PO Box 23053, Seattle, WA, 98102–0353. Web site: <www.gluten.net>.

National Center for Nutrition and Dietetics, American Dietetic Association. 216 West Jackson Boulevard, Suite 800, Chicago, IL 60606–6995. Telephone: 800/366–1655. Web site: <www.unl.edu2020/alpha/National_Center_for_Nutrition_and_Dietetics.html>.

ROCK: Raising Our Celiac Children. 216 West Jackson Boulevard, Suite 800, Chicago, IL 60606–6995. Telephone: 800/366–1655. Web site: <www.celiac.com/cgi-bin/webc.cgi/st_main.html?p_catid=8>.

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Cerebellar herniation *see* **Chiari malformation**

Cerebral palsy

Definition

Cerebral palsy (CP) is the term used for a group of nonprogressive disorders of movement and posture caused by abnormal development of, or damage to, motor control centers of the brain. CP is caused by events before, during, or after birth. The abnormalities of muscle control that define CP are often accompanied by other neurological and physical abnormalities.

Description

Voluntary movement (for example, walking, grasping, chewing) is primarily accomplished using skeletal muscles (muscles attached to bones). Control of the skeletal muscles originates in the cerebral cortex, the largest portion of the brain. Palsy means paralysis but may also be used to describe uncontrolled muscle movement. Therefore, cerebral palsy encompasses any disorder of abnormal movement and paralysis caused by abnormal function of the cerebral cortex. CP does not include

conditions due to progressive disease or degeneration of the brain. For this reason, CP is also referred to as static (nonprogressive) encephalopathy (disease of the brain). Also excluded from CP are any disorders of muscle control that arise in the muscles themselves and/or in the peripheral nervous system (nerves outside the brain and spinal cord). CP is not a specific diagnosis but is more accurately considered a description of a broad but defined group of neurological and physical problems.v

Because CP is not one disorder, it is difficult to classify. It has been divided into four general types: spastic, athetoid, ataxic, and mixed. Another general categorization describes spastic, dyskinetic, and ataxic CP as follows:

- Spastic refers to diplegic impairment of either legs or arms, quadriplegic involving all four extremities, hemiplegic or one-sided involvement of arms and legs, or double hemiplegic impairment of both sides, arms and legs. **Spasticity** means having an increased stretch reflex.
- Dyskinetic refers to abnormal movements caused by inadequate regulation of muscle tone and coordination. The category includes athetoid or choreoathetoid CP; both are hyperkinetic forms of the disease.
- Ataxic refers to disturbances in coordination of voluntary movements; it includes mixed forms of CP, with mixed characteristics and symptoms.

Muscles that receive defective messages from the brain may be constantly contracted and tight (spastic), exhibit involuntary writhing movements (athetosis), or have difficulty with voluntary movement (dyskinesia). A lack of balance and coordination with unsteady movements (ataxia) may also be present. Spastic CP and mixed CP constitute the majority of cases. Effects on the muscles can range from mild weakness or partial paralysis (paresis) to complete loss of voluntary control of a muscle or group of muscles (plegia). CP is also designated by the number of limbs affected. For instance, affected muscles in one limb is monoplegia, both arms or both legs is diplegia, both limbs on one side of the body is hemiplegia, and in all four limbs is quadriplegia. Muscles of the trunk, neck, and head may be affected.

About 50 percent of all cases of CP diagnosed are in children who are born prematurely. Advances in the medical care of premature infants since the 1980s have dramatically increased the rate of survival of these fragile newborns. However, as gestational age at delivery and birth weight of a baby decrease, the risk for CP dramatically increases. A term pregnancy is delivered at 37–41 weeks gestation. The risk for CP in a preterm infant (32–37 weeks) is increased about five-fold over the risk for an infant born at term. Survivors of extremely preterm births (less than 28 weeks) face as much as a 50-fold increase in risk.

Two factors are involved in the risk for CP associated with **prematurity**. First, premature babies are at higher risk for various CP-associated medical complications, such as intracerebral hemorrhage, infection, and difficulty in breathing, to name a few. Second, the onset of premature labor may be induced, in part, by complications that have already caused neurologic damage in the fetus. A combination of both factors may play a role in some cases of CP. The tendency toward premature delivery runs in families, but genetic mechanisms are not fully clear.

An increase in multiple births in the early 2000s, especially in the United States, is associated with the increased use of fertility drugs. As the number of fetuses in a pregnancy increases, the risks for abnormal development and premature delivery also increase. **Twins**, for example, have four times the risk of developing CP as children from singleton pregnancies, owing to the fact that more twin pregnancies are delivered prematurely. The risk for CP in one of triplets is up to 18 times greater. Furthermore, evidence suggests that a baby from a pregnancy in which its twin died before birth is at increased risk for CP.

Although CP is the leading cause of disability in children, its incidence in the United States did not changed much between the 1980s and the early 2000s. Advances in medicine have decreased the incidence from some causes. Rh disease, for example, has been controlled by the advent of anti-Rh globulin; its administration to Rh-negative mothers has reduced one risk factor for CP. The risk has still increased from other causes, however, notably prematurity and multiple-birth pregnancies. The cause of most cases of CP remains unknown, but it has become clear in the early 2000s that birth difficulties are not to blame in most cases. Rather, developmental problems before birth, usually unknown and generally undiagnosable, are largely responsible. The rate of survival for preterm infants has leveled off in the early 2000s, and methods to improve the long-term health of these at-risk babies are being sought.

Demographics

Approximately 500,000 children and adults in the United States have CP, and it is newly diagnosed in about 6,000 infants and young children each year, representing about two to three children in 1,000 live births. No particular ethnic group seems to be at higher risk for CP. However, some low income families may be at

higher risk due to poorer access to proper prenatal care and advanced medical services.

Causes and symptoms

CP can be caused by a number of different mechanisms at various times of life, ranging from several weeks after conception, through birth, to early childhood. In the twentieth century, it was accepted that most cases of CP were due to brain injuries received during a traumatic birth, a condition known as birth asphyxia. However, extensive research in the 1980s showed that only 5 to 10 percent of CP can be attributed to birth trauma. Other possible causes include abnormal development of the brain, prenatal factors that directly or indirectly damage neurons in the developing brain, premature birth, and brain injuries that occur in the first few years of life.

The causes of CP could be grouped into those that are genetic and those that are non-genetic, although most would fall somewhere in between. Grouping causes into those that occur during pregnancy (prenatal), those that happen around the time of birth (perinatal), and those that occur after birth (postnatal), is preferable. CP related to premature birth and multiple births is somewhat different and considered separately.

Prenatal causes

Although much was learned about human embryology in the last couple of decades of the twentieth century, a great deal remains unknown in the early 2000s. Studying prenatal human development is difficult because the embryo and fetus develop in a closed environment—the mother's womb. However, the development of a number of prenatal tests has opened a window on the process. Add to that more accurate and complete evaluations of newborns, especially those with problems, and a clearer picture of what can go wrong before birth is possible.

The complicated process of brain development before birth is susceptible to many chance errors that can result in abnormalities of varying degrees. Some of these errors will result in structural anomalies of the brain, while others may cause undetectable, but significant, abnormalities in how the cerebral cortex is wired. An abnormality in structure or wiring is sometimes hereditary but is most often due to chance or some unknown cause. The possible role genetics plays in a particular brain abnormality depends to some degree on the type of anomaly and the form of CP it causes.

Several maternal-fetal infections are known to increase the risk for CP, including **rubella** (German **measles**, now rare in the United States), cytomegalovirus (CMV), and **toxoplasmosis**. Each of these infections is considered a risk to the fetus only if the mother contracts it for the first time during that pregnancy. Even in those cases, most babies are born normal. Most women are immune to all three infections by the time they reach childbearing age, but a woman's immune status can be determined using the so-called TORCH (for toxoplasmosis, rubella, cytomegalovirus, and herpes) test before or during pregnancy.

Just as a **stroke** can occur in an adult and cause neurologic damage in an adult, so too can this type of event occur in the fetus. A burst blood vessel in the brain followed by uncontrolled bleeding (intracerebral hemorrhage) can cause a fetal stroke, or a clot (embolism) can obstruct a cerebral blood vessel. Infants who later develop CP, along with their mothers, are more likely than other mother-infant pairs to have **coagulation disorders** (coagulopathies) that put them at increased risk for bleeding episodes or blood clots. Certain coagulation disorders are inherited while others may be deficiencies in essential clotting factors or defects in the coagulation process.

Any substance that might affect fetal brain development, directly or indirectly, can increase the risk for CP. Likewise, any substance that increases the risk for premature delivery and low birth weight, such as alcohol, tobacco, or cocaine, among others, might indirectly increase the risk for CP. Links between a drug or other chemical exposure during pregnancy and a risk for CP are difficult to prove.

Because the fetus receives all nutrients and oxygen from blood that circulates through the placenta, anything that interferes with normal placental function might adversely affect development of the fetus, including the brain, or might increase the risk for premature delivery. Structural abnormalities of the placenta, premature detachment of the placenta from the uterine wall (abruption), and placental infections (chorioamnionitis) are thought to pose some risk for CP.

Certain conditions in the mother during pregnancy might pose a risk to fetal development leading to CP. Women with autoimmune anti-thyroid or anti-phospholipid (APA) antibodies are at slightly increased risk for CP in their children. A potentially important clue points toward high levels of cytokines in the maternal and fetal circulation as a possible risk for CP. Cytokines are proteins associated with inflammation, such as from infection or autoimmune disorders, and they may be toxic to neurons in the fetal brain.

Serious physical trauma to the mother during pregnancy could result in direct trauma to the fetus as well, or injuries to the mother could compromise the availability of nutrients and oxygen to the developing fetal brain.

Perinatal causes

Birth asphyxia that is significant enough to result in CP is uncommon in developed countries. An umbilical cord around the baby's neck (tight nuchal cord) and the cord delivered before the baby (prolapsed cord) are possible causes of birth asphyxia, as are bleeding and other complications associated with placental abruption and placenta previa (placenta lying over the cervix).

Infection in the mother is sometimes not passed to the fetus through the placenta but is transmitted to the baby during delivery. Any such infection, such as herpes, that results in serious illness in the newborn has the potential to produce some neurological damage.

Postnatal causes

The remaining 15 percent of CP cases are due to neurologic injury sustained after birth. CP that has a postnatal cause is sometimes referred to as acquired CP, but this is only accurate for those cases caused by infection or trauma.

Incompatibility between the Rh blood types of mother and child (mother Rh negative, baby Rh positive) can result in severe anemia in the baby (**erythroblastosis fetalis**). This may lead to other complications, including severe **jaundice**, which can cause CP. Rh disease in the newborn is rare in developed countries due to routine screening of maternal blood type and routine prevention of anti-Rh antibodies in Rh negative women after each birth of an Rh positive infant. The routine, effective treatment of jaundice due to other causes has also made it an infrequent cause of CP in developed countries.

Serious infections that affect the brain directly, such as **meningitis** and **encephalitis**, may cause irreversible damage to the brain, leading to CP. A **seizure disorder** early in life may cause CP or may be the product of a hidden problem that causes CP in addition to seizures. Unexplained (idiopathic) seizures are hereditary in only a small percentage of cases. Although rare in healthy infants born at or near term, intracerebral hemorrhage and brain embolism, like fetal stroke, are sometimes genetic.

Physical trauma to an infant or child resulting in brain injury, such as from abuse, accidents, or near drowning/suffocation, might cause CP. Likewise, ingestion of a toxic substance such as lead, mercury, other poisons, or certain chemicals could cause neurological damage. Accidental overdose of certain medications might also cause similar damage to the central nervous system.

Symptoms

The symptoms of CP and their severity are variable. Those who have CP may have only minor difficulty with **fine motor skills**, such as grasping and manipulating items with their hands. A severe form of CP could involve significant muscle problems in all four limbs, **mental retardation**, seizures, and difficulties with vision, speech, and hearing.

Although the defect in cerebral function that causes CP is not progressive, the symptoms of CP often change over time. Most of the symptoms relate in some way to the aberrant control of muscles. CP is categorized first by the type of movement/postural disturbance(s) present, rather than by a description of which limbs are affected. The severity of motor impairment is also a factor. Spastic diplegia, for example, refers to continuously tight muscles that have no voluntary control in both legs, while athetoid quadraparesis describes uncontrolled writhing movements and muscle weakness in all four limbs. These may describe CP symptoms generally but do not describe all people with CP. Spastic diplegia is seen in more individuals than is athetoid quadraparesis. CP can also be loosely categorized as mild, moderate, or severe, but these are subjective terms.

A muscle that is tensed and contracted is hypertonic, while excessively loose muscles are hypotonic. Spastic, hypertonic muscles can cause serious orthopedic problems, including curvature of the spine (**scoliosis**), hip dislocation, or contractures. A contracture is shortening of a muscle, aided sometimes by a weak-opposing force from a neighboring muscle. Contractures may become permanent, i.e., fixed, without some sort of intervention. Fixed contractures may cause postural abnormalities in the affected limbs. Clenched fists and contracted feet (equinus or equinovarus) are common in people with CP. Spasticity in the thighs causes them to turn in and cross at the knees, resulting in an unusual method of walking known as scissors gait. Any of the joints in the limbs may be stiff (immobilized) due to spasticity of the attached muscles.

Athetosis and dyskinesia often occur with spasticity but do not often occur alone. The same is true of ataxia. It is important to remember that mild CP or severe CP refers not only to the number of symptoms present but also to the level of involvement of any particular class of symptoms.

Other neurologically based symptoms may include the following:

- mental retardation/learning disabilities
- behavioral disorders
- seizure disorders
- visual impairment
- hearing loss
- speech impairment (dysarthria)
- abnormal sensation and perception

These problems may have a greater impact on a child's life than the physical impairments of CP, although not all children with CP are affected by other problems. Many infants and children with CP have growth impairment. About one third of individuals with CP have moderate-to-severe mental retardation, one third have mild mental retardation, and one third have normal **intelligence**.

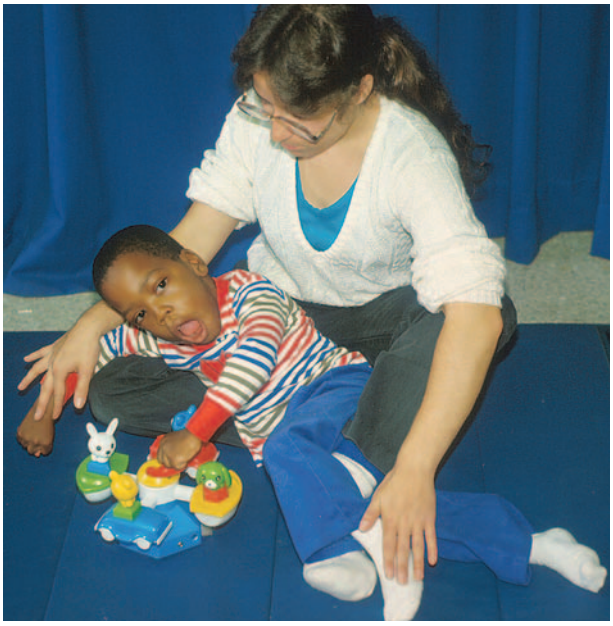
When to call the doctor

Parents should seek medical advice when they notice what seems to be slow development in movement, speech, or cognitive ability in their young child. If a child does not acquire certain skills within a normal time frame, there may be some cause for concern. However, it is known that children progress at somewhat different rates, and a slow beginning is often followed by normal development.

Normal developmental milestones with typical ages for acquiring them, include the following:

- sits well unsupported at about six months (eight to ten months)
- babbles at about six months (up to eight months)
- crawls at about nine months (up to 12 months)
- finger feeds, holds bottle at about nine months (up to 12 months)
- walks alone at about 12 months (up to 15–18 months)
- uses one or two words other than dada/mama at about 12 months (up to 15 months)
- walks up and down steps at about 24 months (24 to 36 months)
- turns pages in books and removes shoes and socks at about 24 months (to 30 months)

Children do not consistently favor one hand over the other before 12 to 18 months of age, and doing so may be a sign that the child has difficulty using the other hand. This same preference for one side of the body may show up as asymmetric **crawling** or, later on, favoring one leg while climbing stairs. Because CP is nonprogressive, continued loss of previously acquired milestones may indicate that CP is not the cause of the problem; medical evaluation is needed to determine the cause.



Young boy with cerebral palsy works with a physical therapist. *(© Custom Medical Stock Photo, Inc.)*

Diagnosis

The signs of CP are not usually noticeable at birth. Children normally progress through a predictable set of developmental milestones through the first 18 months of life. Children with CP, however, tend to develop these skills more slowly because of their motor impairments, and delays in reaching milestones are usually the first symptoms of CP. Babies with more severe cases of CP are usually diagnosed earlier than others.

No one test is diagnostic for CP, but certain factors increase suspicion. The Apgar score measures a baby's condition immediately after birth. Babies who have low Apgar scores are at increased risk for CP. Presence of abnormal muscle tone or movements may indicate CP, as may the persistence of infantile reflexes. Imaging of the brain using ultrasound, **x rays**, MRI, and/or CT scans may reveal a structural anomaly. Some brain lesions associated with CP include scarring, cysts, expansion of the cerebral ventricles (**hydrocephalus**), abnormality of the area surrounding the ventricles (periventricular leukomalacia), areas of dead tissue (necrosis), and evidence of an intracerebral hemorrhage or blood clot. Blood and urine biochemical tests, as well as genetic tests, may be used to rule out other possible causes, including muscle and peripheral nerve diseases, mitochondrial and metabolic diseases, and other inherited disorders. Evaluations by a pediatric developmental specialist and a geneticist may be of benefit.

Treatment

Cerebral palsy cannot be cured, but many of the disabilities it causes can be managed through planning and timely care. Treatment for a child with CP depends on the severity, nature, and location of the primary muscular symptoms, as well as any associated problems that might

be present. Optimal care of a child with mild CP may involve regular interaction with only a physical therapist and occupational therapist, whereas care for a more severely affected child may include visits to multiple medical specialists throughout life. With proper treatment and an effective plan, most people with CP can lead productive, happy lives.

Physical, occupational, and speech therapy

Spasticity, muscle weakness, coordination, ataxia, and scoliosis are all significant impairments that affect the posture and mobility of children and adults with CP. Physical and occupational therapists work with the patient and the **family** to maximize the patient's ability to move affected limbs, develop normal motor patterns, and maintain posture. Assistive technology, including wheelchairs, walkers, shoe inserts, crutches, and braces, are often required. A speech therapist and high-tech aids such as computer-controlled communication devices can make a tremendous difference in the life of those who have speech impairments.

Drug therapy

Before fixed contractures develop, muscle-relaxant drugs such as diazepam (Valium), dantrolene (Dantrium), and baclofen (Lioresal) may be prescribed. Botulinum toxin (Botox), a highly effective treatment, is injected directly into the affected muscles. Alcohol or phenol injections into the nerve controlling the muscle are another option. Multiple medications are available to control seizures, and athetosis can be treated using medications such as trihexyphenidyl HCl (Artane) and benztropine (Cogentin).

Surgery

Fixed contractures are usually treated with either serial casting or surgery. The most commonly used surgical procedures are tenotomy, tendon transfer, and dorsal rhizotomy. In tenotomy, tendons of the affected muscle are cut, and the limb is cast in a more normal position while the tendon regrows. Alternatively, tendon transfer involves cutting and reattaching a tendon at a different point on the bone to enhance the length and function of the muscle. A neurosurgeon performing dorsal rhizotomy carefully cuts selected nerve roots in the spinal cord to prevent them from stimulating the spastic muscles. Neurosurgical techniques in the brain such as implanting tiny electrodes directly into the cerebellum or cutting a portion of the hypothalamus have very specific uses and have had mixed results.

Prognosis

Cerebral palsy can affect every stage of maturation, from childhood through **adolescence** to adulthood. At each stage, those with CP, along with their

KEY TERMS

Asphyxia—Lack of oxygen.

Ataxia—A condition marked by impaired muscular coordination, most frequently resulting from disorders in the brain or spinal cord.

Athetosis—A condition marked by slow, writhing, involuntary muscle movements.

Cerebral palsy—A nonprogressive movement disability caused by abnormal development of or damage to motor control centers of the brain.

Coagulopathy—A disorder in which blood is either too slow or too quick to coagulate (clot).

Contracture—A tightening or shortening of muscles that prevents normal movement of the associated limb or other body part.

Cytokines—Chemicals made by the cells that act on other cells to stimulate or inhibit their function. They are important controllers of immune functions.

Diplegia—Paralysis affecting like parts on both sides the body, such as both arms or both legs.

Dorsal rhizotomy—A surgical procedure that cuts nerve roots to reduce spasticity in affected muscles.

Dyskinesia—Impaired ability to make voluntary movements.

Hemiplegia—Paralysis of one side of the body.

Hypotonia—Having reduced or diminished muscle tone or strength.

Quadriplegia—Paralysis of all four limbs and the trunk below the level of an associated injury to the spinal cord. Also called tetraplegia.

Serial casting—A series of casts designed to gradually move a limb into a more functional position.

Spastic—Refers to a condition in which the muscles are rigid, posture may be abnormal, and fine motor control is impaired.

Spasticity—Increased mucle tone, or stiffness, which leads to uncontrolled, awkward movements.

Static encephalopathy—A disease or disorder of the brain that does not get better or worse.

Tenotomy—A surgical procedure that cuts the tendon of a contracted muscle to allow lengthening.

caregivers, must strive to achieve and maintain the fullest range of experience and education consistent with their abilities. The advice and intervention of various professionals are crucial for many people with CP. Although CP itself is not considered a terminal disorder, it can affect a person's lifespan by increasing the risk for certain medical problems. People with mild cerebral palsy may have near-normal lifespan, but the lifespan of those with more severe forms may be shortened. However, over 90 percent of infants with CP survive into adulthood.

Prevention

Research in the early 2000s is focused on the possible benefits of recognizing and treating coagulopathies and inflammatory disorders in the prenatal and perinatal periods in order to reduce the incidence of CP and other congenital diseases. The use of magnesium sulfate in pregnant women with preeclampsia or threatened preterm delivery may reduce the risk of CP in very preterm infants. Finally, the risk of CP can be decreased through good maternal **nutrition**, avoidance of drugs and alcohol during pregnancy, and prevention or prompt treatment of infections.

Parental concerns

Parents of a child diagnosed with CP may not feel that they have the necessary expertise to coordinate the full range of care their child needs. Although knowledgeable and caring medical professionals are indispensable for developing a care plan, a potentially more important source of information and advice can be gained from other parents who have dealt with the same set of difficulties. Support groups for parents of children with CP can be significant sources of both practical advice and emotional support. Many cities have support groups that can be located through the United Cerebral Palsy Association, and most large medical centers have special multidisciplinary clinics for children with developmental disorders.

See also Febrile seizures; TORCH test; Seizure disorder.

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National Institute of Neurological Disorders and Stroke. 31 Center Drive, MSC 2540, Bldg. 31, Room 8806, Bethesda, MD 20814. Web site: <www.ninds.nih.gov>.

National Society of Genetic Counselors. 233 Canterbury Dr., Wallingford, PA 19086–6617. (610) 872–1192. http://www.nsgc.org/GeneticCounselingYou.asp.

United Cerebral Palsy Association Inc. (UCP). 1660 L St. NW, Suite 700, Washington, DC 20036–5602. Web site: <www.ucpa.org>.

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L. Lee Culvert
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Cerebrospinal fluid (CSF) analysis

Definition

Cerebrospinal fluid (CSF) analysis is a set of laboratory tests that examine a sample of the fluid surrounding the brain and spinal cord. This fluid is an ultrafiltrate of plasma. Plasma is the liquid portion of blood. CSF is clear and colorless. It contains glucose, electrolytes, amino acids, and other small molecules found in plasma, but it has very little protein and few cells. CSF protects the central nervous system from injury, cushions it from the surrounding bone structure, provides it with nutrients, and removes waste products by returning them to the blood.

CSF is withdrawn from the subarachnoid space through a needle by a procedure called a lumbar puncture or spinal tap. CSF analysis includes tests in clinical chemistry, hematology, immunology, and microbiology. Usually three or four tubes are collected. The first tube is used for chemical and/or serological analysis, and the last two tubes are used for hematology and microbiology tests. This method reduces the chances of a falsely elevated white cell count caused by a traumatic tap (bleeding into the subarachnoid space at the puncture

site), and contamination of the bacterial culture by skin germs or flora.

Purpose

The purpose of a CSF analysis is to diagnose medical disorders that affect the central nervous system. Some of these conditions are as follows:

- **meningitis** and **encephalitis**, which may be viral, bacterial, fungal, or parasitic infections
- metastatic tumors (e.g., leukemia) and central nervous system tumors that shed cells into the CSF
- syphilis, a sexually transmitted bacterial disease
- bleeding (hemorrhaging) in the brain and spinal cord
- Guillain-Barré, a demyelinating disease involving peripheral sensory and motor nerves

Routine examination of CSF includes visual observation of color and clarity and tests for glucose, protein, lactate, lactate dehydrogenase, red blood cell count, white blood cell count with differential, syphilis serology (testing for antibodies indicative of syphilis), Gram stain, and bacterial culture. Further tests may need to be performed depending upon the results of initial tests and the presumptive diagnosis.

GROSS EXAMINATION Color and clarity are important diagnostic characteristics of CSF. Straw, pink, yellow, or amber pigments (xanthochromia) are abnormal and indicate the presence of bilirubin, hemoglobin, red blood cells, or increased protein. Turbidity (suspended particles) indicates an increased number of cells. Gross examination is an important aid to differentiating a subarachnoid hemorrhage from a traumatic tap. The latter is often associated with sequential clearing of CSF as it is collected; streaks of blood in an otherwise clear fluid; or a sample that clots.

GLUCOSE CSF glucose is normally approximately two-thirds of the fasting plasma glucose. A glucose level below 40 mg/dL is significant and occurs in bacterial and fungal meningitis and in malignancy.

PROTEIN Total protein levels in CSF are normally very low, and albumin makes up approximately two-thirds of the total. High levels are seen in many conditions, including bacterial and fungal meningitis, tumors, subarachnoid hemorrhage, and traumatic tap.

LACTATE The CSF lactate is used mainly to help differentiate bacterial and fungal meningitis, which cause increased lactate, from viral meningitis, which does not.

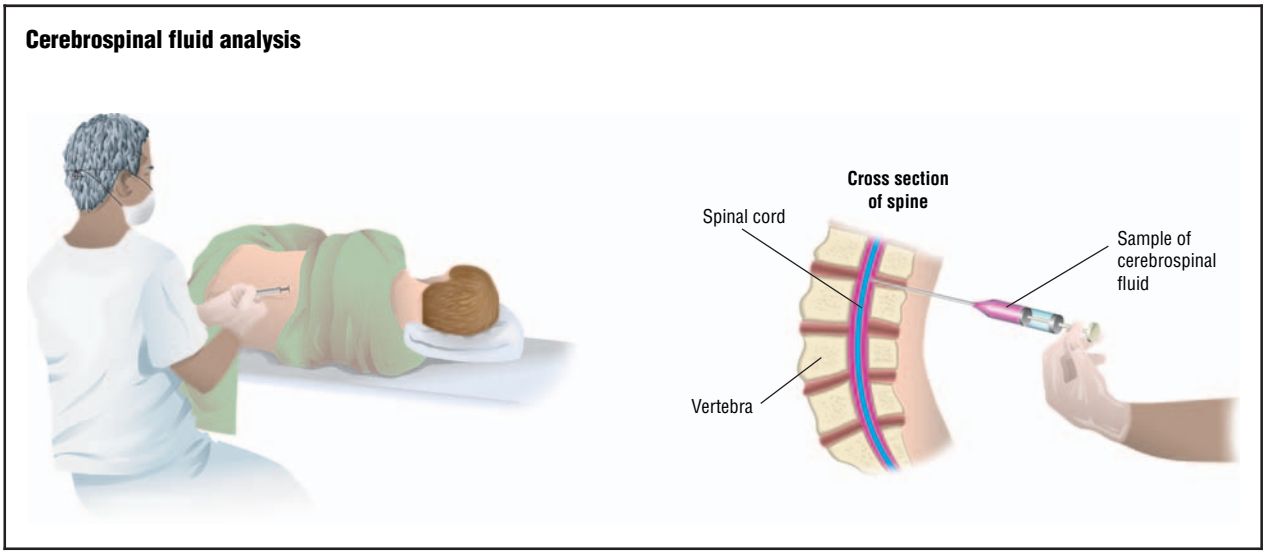
LACTATE DEHYDROGENASE This enzyme is elevated in bacterial and fungal meningitis, malignancy, and subarachnoid hemorrhage.

WHITE BLOOD CELL (WBC) COUNT The number of white blood cells in CSF is very low, usually necessitating a manual WBC count. An increase in WBCs may occur in many conditions, including infection (viral, bacterial, fungal, and parasitic), allergy, leukemia, hemorrhage, traumatic tap, encephalitis, and Guillain-Barré syndrome. The WBC differential helps to distinguish many of these causes. For example, viral infection is usually associated with an increase in lymphocytes, while bacterial and fungal infections are associated with an increase in polymorphonuclear leukocytes (neutrophils). The differential may also reveal eosinophils associated with allergy and ventricular shunts; macrophages with ingested bacteria (indicating meningitis), RBCs (indicating hemorrhage), or lipids (indicating possible cerebral infarction); blasts (immature cells) that indicate leukemia; and malignant cells characteristic of the tissue of origin. About 50 percent of metastatic cancers that infiltrate the central nervous system and about 10 percent of central nervous system tumors will shed cells into the CSF.

RED BLOOD CELL (RBC) COUNT While not normally found in CSF, RBCs will appear whenever bleeding has occurred. Red cells in CSF signal subarachnoid hemorrhage, **stroke**, or traumatic tap. Since white cells may enter the CSF in response to local infection, inflammation, or bleeding, the RBC count is used to correct the WBC count so that it reflects conditions other than hemorrhage or a traumatic tap. This is accomplished by counting RBCs and WBCs in both blood and CSF. The ratio of RBCs in CSF to blood is multiplied by the blood WBC count. This value is subtracted from the CSF WBC count to eliminate WBCs derived from hemorrhage or traumatic tap.

GRAM STAIN The Gram stain is performed on a sediment of the CSF and is positive in at least 60 percent of cases of bacterial meningitis. Culture is performed for both aerobic and anaerobic bacteria. In addition, other stains (e.g. the acid-fast stain for *Mycobacterium tuberculosis*, fungal culture, and rapid identification tests (tests for bacterial and fungal antigens) may be performed routinely.

SYPHILIS SEROLOGY Syphilis serology involves testing for antibodies that indicate neurosyphilis. The fluorescent treponemal antibody-absorption (FTA-ABS) test is often used and is positive in persons with active and treated syphilis. The test is used in conjunction with the VDRL test for nontreponemal antibodies, which is



For cerebrospinal fluid collection, the health-care provider puts a syringe between the patient's vertebrae and pulls out a sample of the fluid surrounding the spinal cord. *(Illustration by GGS Information Services.)*

positive in most persons with active syphilis, but negative in treated cases.

Description

Lumbar puncture is performed by inserting the needle between the fourth and fifth lumbar vertebrae (L4-L5). This location is used because the spinal cord stops near L2, and a needle introduced below this level will miss the cord. In rare instances, such as a spinal fluid blockage in the middle of the back, a physician may perform a spinal tap in the cervical spine.

Precautions

In some circumstances, a lumbar puncture to withdraw a small amount of CSF for analysis may lead to serious complications. Lumbar punctures should be performed only with extreme caution and only if the benefits are thought to outweigh the risks. In people who have bleeding disorders, lumbar puncture can cause hemorrhage that can compress the spinal cord. If there is increased spinal column pressure, as may occur with a brain tumor and other conditions, removal of CSF can cause the brain to herniate, compressing the brain stem and other vital structures and leading to irreversible brain damage or death. Meningitis may be caused by bacteria introduced during the puncture. For this reason, aseptic technique must be followed strictly, and a lumbar puncture should never be performed at the site of a localized skin lesion.

Specimens should be handled with caution to avoid contamination with skin flora. They should be refrigerated if analysis cannot be performed immediately.

Aftercare

After the procedure, the site of the puncture is covered with a sterile bandage. The patient should remain lying for four to six hours after the lumbar puncture. Vital signs should be monitored every 15 minutes for four hours, then every 30 minutes for another four hours. The puncture site should be observed for signs of weeping or swelling for 24 hours. The neurological status of the patient should also be evaluated for such symptoms as **numbness** and/or tingling in the lower extremities.

Risks

The most common side effect after the removal of CSF is a **headache**. This occurs in up to 40 percent of children. It is caused by a decreased CSF pressure related to a small leak of CSF through the puncture site. These headaches usually are a dull **pain**, although some people report a throbbing sensation. A stiff neck and **nausea** may accompany the headache. Lumbar puncture headaches typically begin within two days after the procedure and persist from a few days to several weeks or months.

Normal results

The normal results include the following:

- gross appearance: normal CSF, clear and colorless
- CSF opening pressure: in children older than six to eight years, 90–180 mm H_2O; in infants and younger children, 10–100 mm H_2O
- specific gravity: 1.006–1.009

KEY TERMS

Demyelination—Disruption or destruction of the myelin sheath, leaving a bare nerve. It results in a slowing or stopping of the impulses that travel along that nerve.

Encephalitis—Inflammation of the brain, usually caused by a virus. The inflammation may interfere with normal brain function and may cause seizures, sleepiness, confusion, personality changes, weakness in one or more parts of the body, and even coma.

Guillain-Barré syndrome—Progressive and usually reversible paralysis or weakness of multiple muscles usually starting in the lower extremities and often ascending to the muscles involved in respiration. The syndrome is due to inflammation and loss of the myelin covering of the nerve fibers, often associated with an acute infection. Also called acute idiopathic polyneuritis.

Meningitis—An infection or inflammation of the membranes that cover the brain and spinal cord. It is usually caused by bacteria or a virus.

Multiple sclerosis—A progressive, autoimmune disease of the central nervous system characterized by damage to the myelin sheath that covers nerves. The disease, which causes progressive paralysis, is marked by periods of exacerbation and remission.

Spinal canal—The opening that runs through the center of the spinal column. The spinal cord passes through the spinal canal. Also called the vertebral canal.

Subarachnoid—Referring to the space underneath the arachnoid membrane, the middle of the three membranes that sheath the spinal cord and brain.

Treponeme—A term used to refer to any member of the genus *Treponema*, which is an anaerobic bacteria consisting of cells, 3–8 micrometers in length, with acute, regular, or irregular spirals and no obvious protoplasmic structure.

Vertebrae—Singular, vertebra. The individual bones of the spinal column that are stacked on top of each other. There is a hole in the center of each bone, through which the spinal cord passes.

- glucose: 40–80 mg/dL
- total protein: 15–45 mg/dL
- LD: 1/10 of serum level
- lactate: less than 35 mg/dL
- leukocytes (white blood cells): 0–6/microL (adults and children); up to 19/microL in infants; up to 30/microL (newborns)
- differential: 60–80 percent lymphocytes; up to 30 percent monocytes and macrophages; other cells 2 percent or less. Monocytes and macrophages are somewhat higher in neonates, and make up as much as 80 percent or more, with only 20 percent or less being lymphocytes.
- Gram stain: negative
- culture: sterile
- syphilis serology: negative
- red blood cell count: normally, none unless the needle passes though a blood vessel on route to the CSF

Parental concerns

If the child is anxious or uncooperative, a short-acting sedative may be given. Patients receive a local anesthetic to minimize any pain in the lower back from inserting the needle.

When to call the doctor

If the child does not respond to the parents, if the puncture site continues to leak a watery fluid, or the puncture site appears red and swollen, or has other signs of infection, then the doctor should be notified.

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National Institutes of Health. 9000 Rockville Pike, Bethesda, MD 20892. Web site: <www.nih.gov>.

Mark A. Best

Cerumen impaction

Definition

Cerumen impaction refers to the buildup of layers of earwax within the ear canal to the point of blocking the canal and putting pressure on the eardrum. Ironically, cerumen impaction is often caused by misguided attempts to remove earwax.

Description

Cerumen impaction develops when earwax accumulates in the inner part of the ear canal and blocks the eardrum. It does not happen under normal circumstances because the cerumen is produced by glands in the outer part of the ear canal; it is not produced in the inner part. Cerumen traps sand or dust particles before they reach the eardrum. It also protects the outer part of the ear canal because it repels water. The slow movement of the outer layer of skin of the ear canal carries cerumen toward the outer opening of the ear. As the older cerumen reaches the opening of the ear, it dries out and falls away.

Demographics

Cerumen impaction affects between 2 percent to 6 percent of the general population in the United States. It apparently affects males and females equally.

Causes and symptoms

Causes

Cerumen is most likely to become impacted when it is pushed against the eardrum by cotton-tipped applicators, hair pins, or other objects that people put in their ears, and when it is trapped against the eardrum by a hearing aid. Less common causes of cerumen impaction include overproduction of earwax by the glands in the ear canal or an abnormally narrow ear canal that tends to trap the wax.

Symptoms

The most important symptom of cerumen impaction is partial loss of hearing. Other symptoms are **itching**, tinnitus (noise or ringing in the ears), a sensation of fullness in the ear, and otalgia, or **pain** in the ear. The pain is caused by the pressure of several layers of impacted earwax against the ear drum.

In children younger than one year, cerumen impaction is sometimes discovered during a routine check-up when the doctor finds that the earwax is blocking his or her view of the eardrum. In these cases the cerumen must be removed so that the doctor can finish checking the child's ears and sense of hearing.

When to call the doctor

Impacted cerumen is not a medical emergency. **Family** care practitioners recommend that parents try to remove the impacted wax at home before calling the doctor. Several over-the-counter products are described below under the heading of Treatment. The way to use these products is to tilt the child's head to one side and fill the ear canal with the eardrops, using an eyedropper. Allow the drops to soak in for a few minutes and then treat the other ear if needed. This home treatment method may be repeated twice a day for three or four days.

Parents should, however, take the child to the doctor in the following circumstances:

- The child complains of **dizziness** or pain in the ear.
- The impaction does not improve after several days of treatment at home.
- The child has had a **myringotomy** or ear tube placement.
- The child has a history of discharges from the ear.

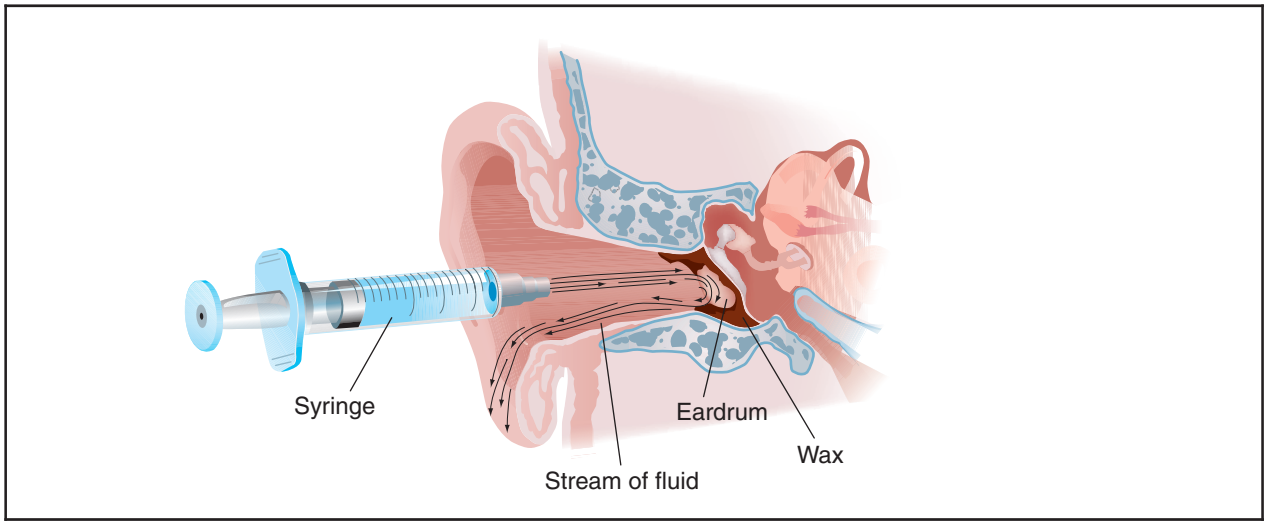
Diagnosis

The diagnosis of impacted cerumen is usually made by examining the ear canal and eardrum with an otoscope, an instrument with a light attached that allows the doctor to look into the canal.

Treatment

Irrigation is the most common method of removing impacted cerumen. It involves washing out the ear canal with water from a commercial irrigator or a syringe with a catheter attached. Although some doctors use Water Piks to remove cerumen, most do not recommend them because the stream of water is too forceful and may damage the eardrum. The doctor may add a small amount of alcohol, hydrogen peroxide, or other antiseptic. The water must be close to body temperature; if it is too cold or too warm, the child may feel dizzy or nauseated. After the ear has been irrigated, the doctor applies antibiotic ear drops to protect the ear from infection.

Irrigation should not be used to remove cerumen if the patient's eardrum is ruptured or missing; if the patient has a history of chronic **otitis media** (inflammation of the middle ear) or a myringotomy (cutting the eardrum to allow fluid to escape from the middle ear); or if the child has hearing in only one ear.



Ear wax is removed by flushing the ear canal with warm fluid. *(Illustration by Argosy, Inc.)*

If irrigation cannot be used or fails to remove the cerumen, the doctor can remove the wax with a vacuum device or a curette, which is a small scoop-shaped surgical instrument. Manual removal of the impaction is effective in 97 percent of children. The doctor holds the child's head steady with one hand while using the curette with the other hand to ease the impacted wax away from the sides of the ear canal. The doctor begins the removal in the area where the cerumen has already started to separate from the wall of the canal.

Some doctors prescribe special eardrops, such as Cerumenex, to soften the wax. The most common side effect of Cerumenex and similar products is an allergic skin reaction. Over-the-counter wax removal products include Debrox or Murine Ear Drops. A 3 percent solution of hydrogen peroxide may also be used. These products are less likely to irritate the skin of the ear.

Alternative treatment

One alternative method that is sometimes touted as a way to remove impacted cerumen is ear candling. Ear candling involves the insertion of a burning candle or a cone of wax-soaked linen or cotton into the affected ear. The person lies on his or her side with the affected ear uppermost. A collecting plate is placed on the ear to catch melted wax. The cone or candle is threaded through a hole in the plate into the ear canal and lit. A variation on this technique involves blowing herbal smoke into the ear through homemade pottery cones. Practitioners of ear candling claim that the heat from the burning candle or smoke creates a vacuum that draws out the impacted cerumen. Some also claim that ear candling improves hearing, relieves sinus infections, cures earache or swimmer's ear, stops tinnitus, or purifies the mind. None of these claims is true, however. Ear candling is not recognized as an acceptable alternative practice by naturopaths, homeopaths, practitioners of Native American medicine, or any other authority on complementary and alternative medicine.

Ear candling is not only an ineffective way to remove impacted cerumen, it can actually damage the ear. According to a 1996 survey of 122 otolaryngologists (doctors who specialize in treating ear, nose, and throat disorders) in the Spokane area, the doctors reported 21 severe ear injuries resulting from ear candling, including 13 cases of external **burns**, seven cases of ear canal obstruction from melted candle wax, and one case of eardrum perforation. Ear candles cannot legally be sold as health devices in the United States because they do not have Food and Drug Administration (FDA) approval. A similar ban is in effect in Canada.

Prognosis

In most cases, impacted cerumen is successfully removed from the child's ear by irrigation or manual extraction with no lasting side effects. Irrigation can, however, lead to infection of the outer or the middle ear if the patient has a damaged or absent eardrum. Older children or adolescents who try to remove earwax themselves with hair pins or similar objects run the risk of perforating the ear drum or damaging the fragile skin covering the ear canal, causing bleeding and the risk of infection.

Prevention

The best method of cleaning the external ear is to wipe the outer opening with a damp washcloth folded over the index finger, without going into the ear canal itself. Two techniques have been recommended to prevent cerumen from reaccumulating in the ear. The patient may

KEY TERMS

Cerumen—The medical term for earwax.

Curette—Also spelled curet; a small loop or scoop-shaped surgical instrument with sharpened edges that can be used to remove tissue, growths, or debris.

Ear candling—An alternative method for removing impacted cerumen with a lighted hollow cone of paraffin or beeswax. It does not work and is not considered an acceptable treatment for any ear problem or disorder.

Impaction—A condition in which earwax has become tightly packed in the outer ear to the point that the external ear canal is blocked.

Irrigation—Cleansing a wound with large amounts of water and/or an antiseptic solution. Also refers to the technique of removing wax (cerumen) from the ear canal by flushing it with water.

Myringotomy—A surgical procedure in which an incision is made in the ear drum to allow fluid or pus to escape from the middle ear.

Otalgia—The medical term for pain in the ear. Impacted cerumen can sometimes cause otalgia.

Otitis media—Inflammation or infection of the middle ear space behind the eardrum. It commonly occurs in early childhood and is characterized by ear pain, fever, and hearing problems.

Tinnitus—A noise, ranging from faint ringing or thumping to roaring, that originates in the ear not in the environment.

place two or three drops of mineral oil into each ear once a week, allow it to remain for two or three minutes, and rinse it out with warm water; or place two drops of Domeboro otic solution in each ear once a week after showering.

Children who wear hearing aids should have their ears examined periodically for signs of cerumen accumulation.

Parents should teach children not to use cotton swabs or other objects to remove wax from the ear, and should advise older children and adolescents against experimenting with ear candling.

Parental concerns

Removal of impacted cerumen from children's ears is a routine procedure and should not ordinarily cause parents a great deal of concern. If the child has repeated episodes of cerumen impaction, parents can discuss various preventive measures with the doctor.

See also Ear exam with an otoscope; Myringotomy and ear tubes; Otitis media.

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Rebecca Frey, PhD

Cesarean section

Definition

A cesarean section (also referred to as c-section) is the birth of a fetus accomplished by performing a

surgical incision through the maternal abdomen and uterus. It is one of the oldest surgical procedures known throughout history.

Purpose

Although Healthy People 2010 established a goal of a 15 percent rate for c-sections in the United States, the ideal rate has not been established. As of 2004, the average c-section rate is one out of every four births or approximately 26 percent of all births. A c-section allows safe and quick delivery of a baby when a vaginal delivery is not possible. The surgery is performed in the presence of a variety of maternal and fetal conditions with the most commonly accepted indications being complete placenta previa, cephalopelvic disproportion (CPD), placental abruption, active genital herpes, umbilical cord prolapse, failure to progress in labor or dystocia, proven nonreassuring fetal status, and benign and malignant tumors that obstruct the birth canal. Indications that are more controversial include breech presentation, previous c-section, major congenital anomalies, cervical cerclage, and severe Rh isoimmunization. C-sections have a higher maternal mortality rate than vaginal births with approximately 5.8 women per 100,000 live births dying, and half of these deaths are ascribed to the operation and a coexisting medical condition. Perinatal morbidity is associated with infections, reactions to anesthesia agents, blood clots, and bleeding.

Description

According to the United States Public Health Service, 35 percent of all c-sections are performed because the woman has had a previous c-section. The skin incision for a c-section is either transverse (Pfannenstiel) or vertical and does not indicate the type of incision made into the uterus. ''Once a cesarean, always a cesarean,'' is a rule that originated with the classical, vertical uterine incision. It was believed that the resulting scar weakened the uterus wall and was at risk of rupture in subsequent deliveries. As of 2004, the incision is almost always made horizontally across the lower uterine segment, called a low transverse incision. This results in reduced blood loss and a decreased chance of rupture. This kind of incision allows many women to have a vaginal birth after a cesarean (VBAC).

Failure to progress and/or dystocia is the second most common reason for a c-section and represents about 30 percent of all cases. Uterine contractions may be weak or irregular, the cervix may not be dilating, or the mother's pelvic structure may not allow adequate passage for birth. When the baby's head is too large to fit through the pelvis, the condition is called cephalopelvic disproportion (CPD). Failure to progress, however, can only be diagnosed with documentation of adequate contraction strength. The force of the contractions can be measured with an intrauterine pressure catheter (IUPC), which is a catheter that can be placed through the cervix into the uterus to measure uterine pressure during labor. Calculation of this force is determined by subtracting the baseline (resting) pressure from the peak pressure recorded for all contractions in a ten-minute period. This pressure calculation results in a force termed Montevideo units. A minimum of 200 Montevideo units are required before the forces of labor can be considered adequate. If the Montevideo units are less than this ten-minute sum and the fetal heart rate is reassuring, augmentation of labor with pitocin may be necessary.

Breech presentation occurs in about 3 percent of all births, and approximately 12 percent of c-sections are performed to deliver a baby in a breech presentation: buttocks or feet first. Breech presentations were still delivered vaginally in the 1970s, but with the advent of the malpractice climate, many doctors shied away from this practice, opting to perform a c-section. As a result, physicians who were being trained during that time period never learned how to manage a breech vaginal delivery. There was some change in this approach in the 1990s, and doctors are once again learning how to manage this situation; however, it is still uncertain whether this knowledge will be used in their practice.

Fetal distress or the more appropriate term, nonreassuring fetal heart rate, accounts for almost 9 percent of c-sections. With the introduction of **electronic fetal monitoring** (EFM) in the 1970s, doctors had more information for assessing fetal well-being. It was assumed that fetal monitoring would transmit signals of distress, thus, the EFM tracing became a legal document. There is still considerable debate as to what a non-reassuring FHR really is, but there are other parameters available to assist in this interpretation. When a fetus experiences stress, (oxygen deprivation) in utero, it may pass meconium (feces) into the amniotic fluid. The appearance of meconium in the fluid along with a questionable EFM tracing may indicate that a fetus is becoming compromised. At this point, if a woman is in early labor, a c-section may have to be performed. If, however, she is close to delivery, a vaginal delivery is often quicker. Oxygen deprivation may also be determined by testing the pH of a blood sample taken from the baby's scalp: a pH of 7.25-7.35 is normal; between 7.2 and 7.25 is suspicious; and below 7.2 is a sign of trouble. If the sample is equivocal, it can be repeated every 20 to 30 minutes.

The remaining 14 percent of c-sections occur secondary to other emergency situations, including the following:

- Umbilical cord prolapse: This situation occurs when the cord is the presenting part from the vagina. It becomes compressed and cuts off blood flow to the baby. The birth attendant must insert a hand into the vagina and relieve pressure on the cord until a c-section is performed.
- Placental abruption: The placenta separates from the uterine wall before the baby is born. If it is a complete abruption, the baby's blood flow will be cut off completely. The mother experiences severe **pain**, possible bleeding, and her abdomen feels rock hard. This situation demands an immediate c-section. Partial abruptions can occur without endangering the mother or the baby, but they need to be closely monitored. The risk of placental abruption is higher in multiple births and in women with high blood pressure.
- Placenta previa: With a complete previa, the placenta covers the cervix completely, and the mother may experience painless bleeding. With a complete previa, a c-section is mandatory as cervical dilation would cause bleeding. The baby is often in a transverse position in this case, which means it is lying horizontally across the pelvis. Women with partial previas will usually need a c-section due to bleeding problems, but those with marginal previas can often deliver vaginally.
- Active genital herpes: Any active herpetic lesions in the vaginal area can infect the baby as it passes through the birth canal. This is especially true for those with a primary outbreak, a first-time exposure.
- Mother's health status: A c-section may be necessary in women with pre-existing diseases, such as diabetes, **hypertension**, pregnancy induced hypertension (pre-eclampsia), autoimmune diseases such as lupus erythematosus, and blood incompatibilities. Each case must be evaluated on an individual basis in these instances to achieve the optimal outcome for baby and mother.

Precautions

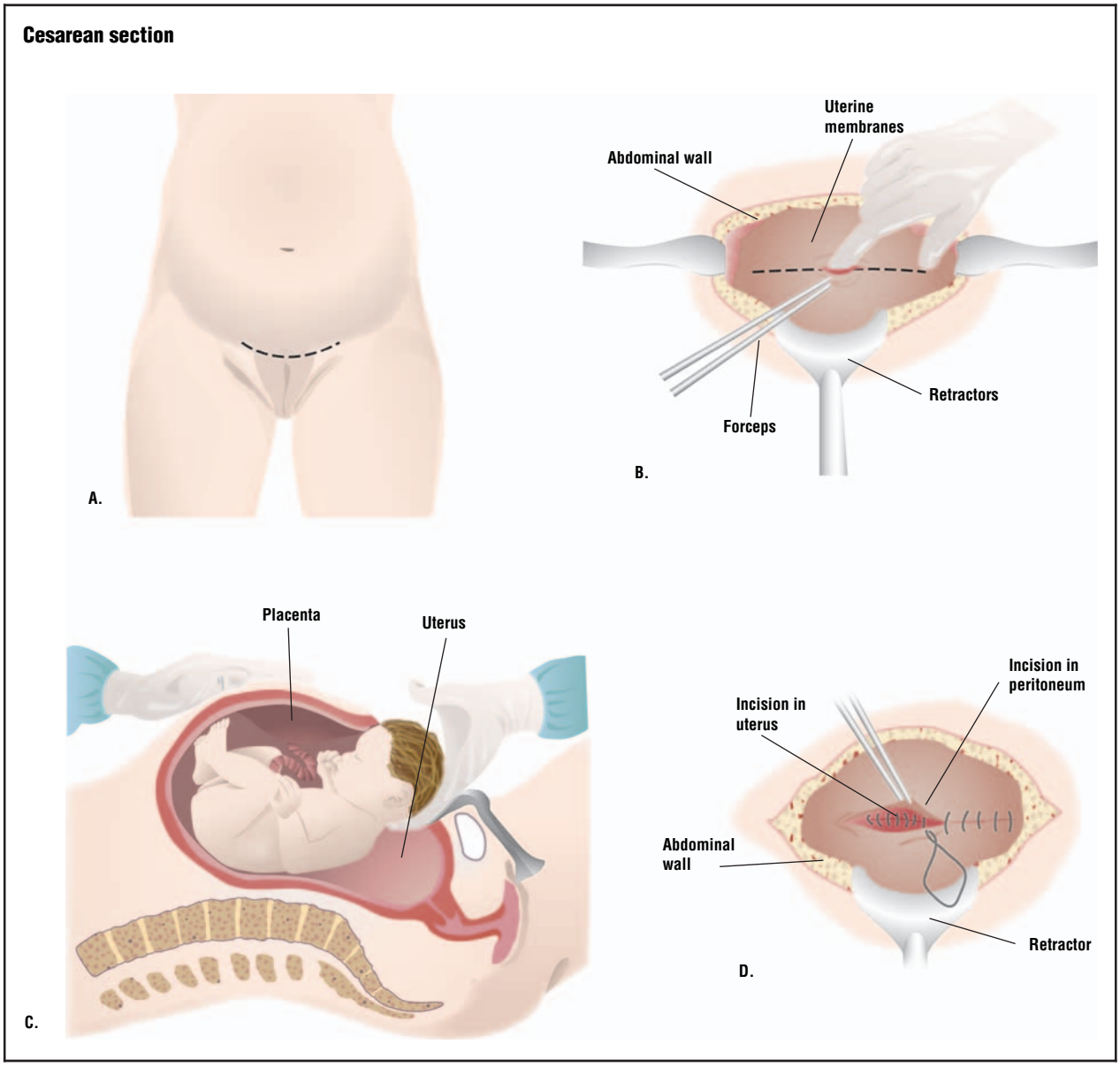
There are some precautions any pregnant woman can follow to enhance her chances of preventing a c-section. These include the following:

- She should check her doctor's c-section rate to see if it is unnecessarily high. She can ask what his/her rate is and verify it by checking with the labor and delivery nurses at the hospital or with a **childbirth** educator.
- She should not stay on her back during labor. She can walk, rock, or use a hot shower or whirlpool.
- From the beginning, she should discuss with her doctor that she wants to avoid having a c-section if at all possible and enlist his opinion on how to achieve it.
- Studies show that women who go to the hospital early have a higher c-section rate than those who do not. Therefore, when labor starts, the woman should stay home for as long as she safely can. She should not go in if contractions are further apart than four to five minutes.
- She should use a midwife since studies show that they have a higher percentage of natural childbirths without surgical intervention than obstetricians do.
- She should hire a doula to assist during labor birth. Doulas have a lower c-section rate and can offer massage, different positions, and support alternatives during the difficult phases of labor.
- She should gather as much information as possible on hospital policies to educate herself and then discuss this information with her doctor or midwife. She should keep an open mind and stay informed.

Preparation

There is no perfect anesthesia for a c-section because every choice has its advantages and disadvantages. When a c-section becomes necessary and if it is not an emergency, the mother and her significant other should take part in the choice of anesthetic by being informed of risks and side effects. The anesthesia is usually a regional anesthetic (epidural or spinal), which makes her numb from below her breasts to her toes. In some cases, a general anesthetic will be administered if the regional does not work or if it is an emergency c-section. Every effort should be made to include the significant other in the preparations and recovery as well as the surgery if at all possible. An informed consent needs to be signed, and the physician should explain the surgery at that time. The mother may already have an intravenous (IV) line of fluid running into a vein in her arm. A catheter is inserted into her bladder to keep it drained and out of the way during surgery and the upper pubic area is usually shaved. Antacids are frequently administered to reduce the likelihood of damage to the lungs should aspiration of gastric contents occur. The abdominal area is then scrubbed and painted with betadine or another antiseptic solution. Drapes are placed over the surgical area to block a direct view of the procedure.

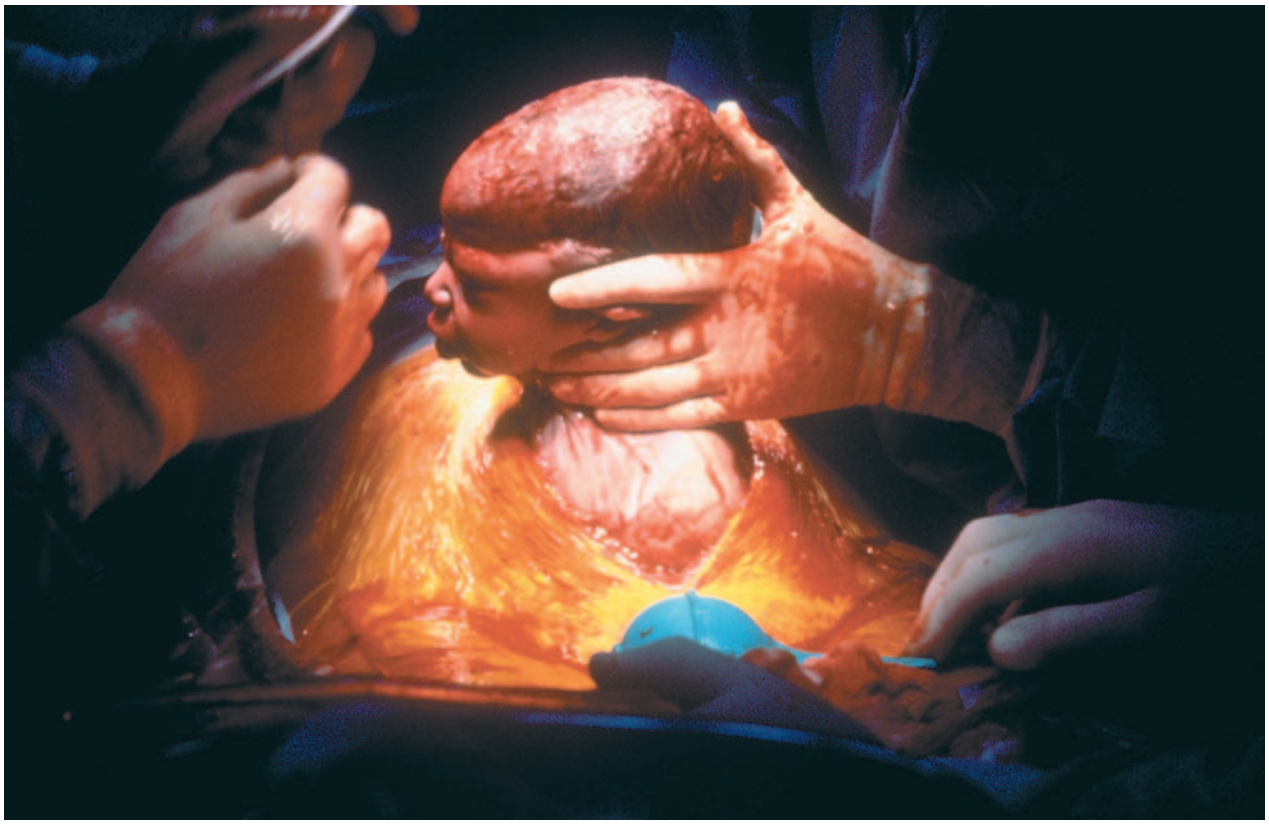
The type of skin incision, transverse or vertical, is determined by time factor, preference of mother, or physician preference. Two major locations of uterine incisions are the lower uterine segment and the upper segment of the body of the uterus (classical incision). The most common lower uterine segment incision is a transverse incision because the lower segment is the



To remove a baby by cesarean section, an incision is made into the abdomen, usually just above the pubic hairline (A). The uterus is located and divided (B), allowing for delivery of the baby (C). After all the contents of the uterus are removed, the uterus is repaired and the rest of the layers of the abdominal wall are closed (D). *(Illustration by GGS Information Services.)*

thinnest part of the pregnant uterus and involves less blood loss. It is also easier to repair, heals well, is less likely to rupture during subsequent pregnancies and makes it possible for a woman to attempt a vaginal delivery in the future. The classical incision provides a larger opening than a low transverse incision and is used in emergency situations, such as placenta previa, preterm and macrosomic fetuses, abnormal presentation, and multiple births. With the classical incision, there is more bleeding and a greater risk of abdominal infection. This incision also creates a weaker scar, which places the woman at risk for uterine rupture in subsequent pregnancies.

Once the uterus is opened, the amniotic sac is ruptured and the baby is delivered. The time from the initial incision to birth is typically five to ten minutes. The umbilical cord is clamped and cut, and the newborn is given to the nursery personnel for evaluation. Cord blood is normally obtained for analysis of the infant's blood type and pH. The placenta is removed from the mother and her uterus is closed with suture. The abdominal area may be closed with suture or surgical staple. The time from birth through suturing may take 30 to 40 minutes. The entire surgical procedure may be performed in less than one hour. Physical contact or



This baby is being delivered by cesarean section. *(Photograph by John Smith. Custom Medical Stock Photo Inc..)*

holding of the newborn may take place briefly while the mother is on the operating table if the baby is stable. The significant other can go with the baby to the nursery for the remainder of the operation.

Aftercare

Immediate postpartal care after a c-section is similar to post-operative care with the exception of palpating the fundus (top of the uterus) for firmness. If an epidural or spinal were used, Duramorph (a pain medication similar to morphine) is often administered through these catheters just prior to completion of surgery. It does very well in controlling pain but may cause **itching**, which can be managed. During recovery the mother is encouraged to turn, **cough**, and deep breathe to keep her lungs clear, and the neonate is usually brought to the mother to breastfeed if she so desires. The mother will be encouraged to get out of bed about eight to 24 hours after surgery. Walking stimulates the circulation to avoid formation of blood clots and promotes bowel movement. Once discharged home, the mother should limit stair climbing to once a day, and she should avoid lifting anything heavier than the baby. It is important to nap as often as the baby does and make arrangements for help with the housework, meals, and care of other children. Driving may be resumed after two weeks, although some doctors recommend waiting for six weeks, which is the typical recovery period from major surgery.

Risks

The maternal death rate for c-section is less than 0.02 percent (5.8 per 100,000 live births), but that is four times the maternal death rate associated with vaginal delivery. The mother is at risk for increased bleeding from two incision sites and a c-section usually has twice as much blood loss as a vaginal delivery during surgery. Complications occur in less than 10 percent of cases, but these complications can include an infection of the incision, urinary tract, or tissue lining the uterus (endometritis). Less commonly, injury can occur to the surrounding organs, i.e., the bladder and bowel.

Normal results

The after-effects of a c-section vary, depending on the woman's age, physical fitness, and overall health. Following this procedure, a woman commonly experiences gas pains, incision pain, and uterine contractions,

KEY TERMS

Breech presentation—The condition in which the baby enters the birth canal with its buttocks or feet first.

Cephalopelvic disproportion—The condition in which the baby's head is too large to fit through the mother's pelvis.

Cervical cerclage—A procedure in which the cervix of the uterus is sewn closed, it is used in cases when the cervix starts to dilate too early in a pregnancy to allow the birth of a healthy baby.

Doula—A doula is someone who undergoes special training to enable them to support women during childbirth and into the postpartum period.

Dystocia—Failure to progress in labor, either because the cervix will not dilate (expand) further or because the head does not descend through the mother's pelvis after full dilation of the cervix.

Genital herpes—A life-long, recurrent sexually transmitted infection caused by the herpes simplex virus (HSV).

Perinatal—Referring to the period of time surrounding an infant's birth, from the last two months of pregnancy through the first 28 days of life.

Pitocin—A synthetic hormone that produces uterine contractions.

Placenta previa—A condition in which the placenta totally or partially covers the cervix, preventing vaginal delivery.

Placental abruption—An abnormal separation of the placenta from the uterus before the birth of the baby, with subsequent heavy uterine bleeding. Normally, the baby is born first and then the placenta is delivered within a half hour.

Postpartal—The six-week period following childbirth.

Rh blood incompatibility—Incompatibility between the blood of a mother and her baby due the absence of the Rh antigen in the red blood cells of one and its presence in the red blood cells of the other.

Umbilical cord prolapse—A birth situation in which the umbilical cord, the structure that connects the placenta to the umbilicus of the fetus to deliver oxygen and nutrients, falls out of the uterus and becomes compressed, thus preventing the delivery of oxygen.

which are also common with vaginal delivery. The hospital stay may be three to four days. Breastfeeding the baby is encouraged, taking care that it is in a position that keeps the baby from resting on the mother's incision. As the woman heals, she may gradually increase appropriate exercises to regain abdominal tone. Full recovery may be seen in four to six weeks.

The prognosis for a successful vaginal birth after a cesarean (VBAC) may be at least 75 percent, especially when the c-section involved a low transverse incision in the uterus, and there were no complications during or after delivery.

Of the hundreds of thousands of women in the United States who undergo a c-section each year, about 500 die from serious infections, hemorrhaging, or other complications. These deaths may be related to the health conditions that made the operation necessary and not simply to the operation itself.

Parental concerns

Undergoing a c-section may inflict psychological distress on the mother, beyond hormonal mood swings and postpartum depression. The woman may feel disappointment and a sense of failure for not experiencing a vaginal delivery. She may feel isolated if the father or birthing coach is not with her in the operating room or if she is treated by an unfamiliar doctor rather than by her own doctor or midwife. She may feel helpless from a loss of control over labor and delivery with no opportunity to actively participate. To overcome these feelings, the woman needs to understand why the c-section was crucial. It is important that she be able to verbalize an understanding that she could not control the events that made the c-section necessary and recognize the importance of preserving the health and safety of both herself and her child. Women who undergo a c-section should be encouraged to share their feelings with others. Hospitals can often recommend support groups for such mothers. Women should also be encouraged to seek professional help if negative emotions persist.

See also Apgar testing; Electronic fetal monitoring.

Resources

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Association of Women's Health, Obstetric and Neonatal Nursing. 2000 L Street, NW, Suite 740, Washington, DC 20036. Web site: <www.awhonn.org>.

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Chalazion *see* **Styes and chalazia**

Charcot-Marie-Tooth disease

Definition

Charcot-Marie-Tooth disease (CMT) is the name of a group of inherited disorders of the nerves in the peripheral nervous system. These are the nerves throughout the body that communicate motor and sensory information to and from the spinal cord. CMT causes weakness and loss of sensation in the limbs.

Description

CMT is named for the three neurologists who first described the condition in the late 1800s. It is also known as hereditary motor and sensory neuropathy and is sometimes called peroneal muscular atrophy, referring to the muscles in the leg that are often affected. The age of onset of CMT can vary anywhere from young childhood to the 50s or 60s. Symptoms typically begin by the age of 20. For reasons unknown as of 2004, the severity in symptoms can also vary greatly, even among members of the same **family**.

Although CMT has been described for many years, it is only since the early 1990s that the genetic cause of many of the types of CMT have become known. Therefore, knowledge about CMT increased dramatically shortly thereafter.

The peripheral nerves

CMT affects the peripheral nerves, those groups of nerve cells carrying information to and from the spinal cord. CMT decreases the ability of these nerves to carry motor commands to muscles, especially those furthest from the spinal cord located in the feet and hands. As a result, the muscles connected to these nerves eventually weaken. CMT also affects the sensory nerves that carry information from the limbs to the brain. Therefore, people with CMT also have sensory loss. This loss causes symptoms such as not being able to tell if something is hot or cold or having difficulties with balance.

There are two parts of the nerve that can be affected in CMT. A nerve can be likened to an electrical wire, in which the wire part is the axon of the nerve and the insulation surrounding it is the myelin sheath. The job of the myelin is to help messages travel very fast through the nerves.

CMT is usually classified depending by which part of the nerve is affected. Children who have problems with the myelin have CMT type 1 and children who have abnormalities of the axon have CMT type 2. Specialized testing of the nerves, called nerve conduction testing (NCV), can be performed to determine if CMT1 or CMT2 is present. These tests measure the speed at which messages travel through the nerves. In CMT1, the messages move too slowly, but in CMT2 the messages travel at the normal speed.

Demographics

Charcot-Marie-Tooth is the most common inherited neurological disorder, affecting approximately 150,000 Americans. It is the most common type of inherited neurological condition, occurring in approximately one in 2,500 people worldwide, in all races and ethnic groups.

Causes and symptoms

Genetic causes

CMT is caused by changes (mutations) in any one of a number of genes that carry the instructions on how to make the peripheral nerves. Genes contain the instructions for how the body grows and develops before and after a person is born. There are probably at least 15 different genes that can cause CMT. However, as of 2004, not all had been identified.

CMT types 1 and 2 can be broken down into subtypes based upon the gene that causes CMT. The subtypes are labeled by letters, so there is CMT1A, CMT1B, etc. Therefore, the gene with a mutation that causes CMT1A is different from the one that causes CMT1B.

Types of CMT

CMT1A The most common type of CMT, called CMT1A, is caused by a mutation in a gene called peripheral myelin protein 22 (PMP22) located on chromosome 17. The job of this gene is to make a protein (PMP22) that makes up part of the myelin. In most people who have CMT, the mutation that causes the condition is a duplication (doubling) of the PMP22 gene. Instead of having two copies of the PMP22 gene (one on each chromosome) there are three copies. It is not known how this extra copy of the PMP22 gene causes the observed symptoms. A small percentage of people with CMT1A do not have a duplication of the PMP22 gene, but rather they have a point mutation in the gene. A point mutation is like a typo in the gene that causes it to work incorrectly.

HEREDITY NEUROPATHY WITH LIABILITY TO PRESSURE PALSIES (HNPP) HNPP is a condition that is also caused by a mutation in the PMP22 gene. The mutation is a deletion. Therefore, there is only one copy of the PMP22 gene instead of two. People who have HNPP may have some of the signs of CMT. However, they also have episodes when they develop weakness and problems with sensation after compression of certain pressure points such as at the elbows or knee. Often these symptoms resolve after a few days or weeks, but sometimes they are permanent.

CMT1B Another type of CMT, called CMT1B, is caused by a mutation in a gene called myelin protein zero (MPZ), located on chromosome 1. The job of this gene is to make the layers of myelin stick together as they are wrapped around the axon. The mutations in this gene are point mutations because they involve a change (either deletion, substitution, or insertion) at one specific component of a gene.

CMTX Another type of CMT, called CMTX, is usually considered a subtype of CMT1 because it affects the myelin, but it has a different type of inheritance than type 1 or type 2. In CMTX, the CMT-causing gene is located on the X chromosome and is called connexin 32 (Cx32). The job of this gene is to code for a class of protein called connexins that form tunnels between the layers of myelin.

CMT2 There are at least five different genes that can cause CMT type 2. Therefore, CMT2 has subtypes A, B, C, D and E. As of early 2001, scientists have narrowed in on the location of most of the CMT2-causing genes. However, the specific genes and the mutations have not yet been found as of 2004 for most types. In the early 2000s, the gene for CMT2E was found. The gene is called neurofilament-light (NF-L). Because it has just been discovered, not much is known about how mutations in this gene cause CMT.

CMT3 In the past a condition called Dejerine-Sottas disease was referred to as CMT3. This is a severe type of CMT in which symptoms begin in infancy or early childhood. It is known as of 2004 that this is not a separate type of CMT; in fact, people who have onset in infancy or early childhood often have mutations in the PMP22 or MPZ genes. Children with type 3 CMT may not develop early motor skills such as walking until they are three or four years old.

CMT4 CMT4 is a rare type of CMT in which the nerve conduction tests have slow response results. However, it is classified differently from CMT1 because it is passed through families by a different pattern of inheritance. There are five different subtypes, and each has as of 2004 only been described in a few families. The symptoms in CMT4 are often severe and other symptoms such as deafness may be present. There are three different genes that have been associated with CMT4 as of early 2001. They are called MTMR2, EGR2, and NDRG1. More research is required to understand how mutations in these genes cause CMT.

Inheritance

Children with CMT have an increased risk for passing on the genes that cause the condition when they start a family of their own.

CMT1A and 1B, HNPP, and all of the subtypes of CMT2 have autosomal dominant inheritance. Autosomal refers to the first 22 pairs of chromosomes that are the same in males and females. Therefore, males and females are affected equally in these types. In a dominant condition, only one gene of a pair needs to have a mutation in order for a person to have symptoms of the condition. Therefore, individuals who have these types have a 50 percent, or one in two, chance of passing CMT on to each of their children. This chance is the same for each pregnancy and does not change based on previous children.

CMTX has X-linked inheritance. Since males only have one X chromosome, they only have one copy of the Cx32 gene. Thus, when a male has a mutation in his Cx32 gene, he will have CMT. However, females have two X chromosomes and, therefore, have two copies of the Cx32 gene. If they have a mutation in one copy of

their Cx32 genes, they will only have mild to moderate symptoms of CMT that may go unnoticed. This is because their normal copy of the Cx32 gene does make normal myelin.

Females pass on one or the other of their X chromosomes to their children—sons or daughters. If a woman with a Cx32 mutation passes her normal X chromosome, she will have an unaffected son or daughter who will not pass CMT on to his or her children. If the woman passes the chromosome with Cx32 mutation on she will have an affected son or daughter, although the daughter will be mildly affected or have no symptoms. Therefore, a woman with a Cx32 mutation has a 50 percent, or a one in two, chance of passing the mutation to her children: a son will be affected, and a daughter may only have mild symptoms.

When males pass on an X chromosome, they have a daughter. When they pass on a Y chromosome, they have a son. Since the Cx32 mutation is on the X chromosome, a man with CMTX will always pass the Cx32 mutation on to his daughters. However, when he has a son, he passes on the Y chromosome, and the son will not be affected. Therefore, an affected male passes the Cx32 gene mutation on to all of his daughters, but to none of his sons.

CMT4 has autosomal recessive inheritance. Males and females are equally affected. In order for individuals to have CMT4, they must have a mutation in both of their CMT-causing genes, one inherited from each parent. The parents of an affected person are called carriers. They have one normal copy of the gene and one copy with a mutation. Carriers do not have symptoms of CMT. Two carrier parents have a 25 percent, or one in four, chance of passing CMT on to each of their children.

Symptoms

The onset of symptoms is highly variable, even among members of the same family. Symptoms usually progress very slowly over a person's lifetime. The main problems caused by CMT are weakness and loss of sensation mainly in the feet and hands. The first symptoms are usually problems with the feet such as high arches and problems with walking and running. Tripping while walking and sprained ankles are common. Muscle loss in the feet and calves leads to foot drop in which the foot does not lift high enough off the ground when walking. Complaints of cold legs are common, as are cramps in the legs, especially after **exercise**. Most children with CMT remain able to walk throughout their lives.

In many people, the fingers and hands eventually become affected. Muscle loss in the hands can make fine movements such as working buttons and zippers difficult. Some patients develop tremor in the upper limbs. Loss of sensation can cause problems such as **numbness** and the inability to feel if something is hot or cold.

Diagnosis

When CMT is suspected in a child, the diagnosis begins with a careful neurological exam to determine the extent and distribution of weakness. A thorough family history should be taken at this time to determine if other people in the family are affected. Testing may also be performed to rule out other causes of neuropathy.

A nerve conduction velocity test should be performed to measure how fast impulses travel through the nerves. This test may show characteristic features of CMT, but it is not diagnostic of CMT. Nerve conduction testing may be combined with electromyography (EMG), an electrical test of the muscles.

A nerve biopsy (removal of a small piece of the nerve) may be performed to look for changes characteristic of CMT. However, this testing is not diagnostic of CMT and is usually not necessary for making a diagnosis.

Definitive diagnosis of CMT is made only by genetic testing, usually performed by drawing a small amount of blood. As of early 2001, testing is available to detect mutations in PMP22, MPZ, Cx32 and EGR2. However, research is progressing rapidly and new testing is often made available every few months. All affected members of a family have the same type of CMT. Therefore, once a mutation is found in one affected member, it is possible to test other members who may have symptoms or are at risk of developing CMT.

Prenatal diagnosis

Testing during pregnancy to determine whether an unborn child is affected is possible if genetic testing in a family has identified a specific CMT-causing mutation. This can be done after ten to 12 weeks of pregnancy using a procedure called chorionic villus sampling (CVS). CVS involves removing a tiny piece of the placenta and examining the cells. Testing can also be done by **amniocentesis** after 16 weeks gestation by removing a small amount of the amniotic fluid surrounding the baby and analyzing the cells in the fluid. Each of these procedures has a small risk of miscarriage associated with it, and those who are interested in learning more should check with their doctor or genetic counselor. Couples interested in these options should obtain genetic

KEY TERMS

Axon—A long, threadlike projection that is part of a neuron (nerve cell).

Myelin—A fatty sheath surrounding nerves throughout the body that helps them conduct impulses more quickly.

Nerve condition velocity (NCV)—Technique for studying nerve or muscle disorders, measuring the speed at which nerves transmit signals.

Neuropathy—A disease or abnormality of the peripheral nerves (the nerves outside the brain and spinal cord). Major symptoms include weakness, numbness, paralysis, or pain in the affected area.

Peripheral nerves—Nerves outside the brain and spinal cord that provide the link between the body and the central nervous system.

counseling to carefully explore all of the benefits and limitations of these procedures.

Treatment

As of 2004, there was no cure for CMT. However, physical and occupational therapy are an important part of CMT treatment. Physical therapy is used to preserve range of motion and minimize deformity caused by muscle shortening, or contracture. Braces are sometimes used to improve control of the lower extremities and can help with balance. After wearing braces, children often find that they have more energy because they are using less energy to focus on their walking. Occupational therapy is used to provide devices and techniques that can assist tasks such as dressing, feeding, writing, and other routine activities of daily life. Substances such as **caffeine** and alcohol should be avoided if tremor is present. Voice-activated software can also help children who have problems with fine motor control.

It is very important that individuals with CMT avoid injury that causes them to be immobile for long periods of time. It is often difficult for people with CMT to return to their original strength after injury.

There is a long list of medications that should be avoided if possible by people diagnosed with CMT, such as the high blood pressure-reducing medication hydralazine (Apresoline), megadoses of vitamin A, B6, and D, and large intravenous doses of penicillin. Complete lists are available from the CMT support groups. Parents considering providing any of these medications to their children with CMT should weigh the risks and benefits with their physician.

Prognosis

The symptoms of CMT usually progress slowly over many years but do not usually shorten life expectancy. The majority of children with CMT do not need to use a wheelchair at any time during their lives. Most people with CMT are able to lead full and productive lives despite their physical challenges.

Prevention

As of 2004 there was no known way to prevent CMT. Genetic counseling for parents with CMT can help them understand the risk they face of having children with the disorder.

Parental concerns

The goal for children with CMT is to live as normal a life as possible. Along with seeing that children obtain proper healthcare from a specialist knowledgeable about the condition, parents can take the following precautions to help minimize problems:

- Children should be properly dressed in cold weather to avoid chilled hands and feet.
- The home should be designed to avoid the risk of falls, including installing handrails on steps, removing throw rugs, using nonskid bathmats, and arranging furniture so it does not block passage through rooms.
- Learning about CMT and discussing it with teachers, classmates, friends, and family members may help normalize the disorder and create support for the child.

Resources

ORGANIZATIONS

Charcot Marie Tooth Association (CMTA). 2700 Chestnut Parkway, Chester, PA 19013. Web site: <www.charcot-marie-tooth.org/site/content>.

CMT International. 1 Springbank Dr., St. Catherine's, ONT L2S2K1, Canada. Web site: <www.cmtint.org>.

Muscular Dystrophy Association. 3300 East Sunrise Dr., Tucson, AZ 85718. Web site: <www.mdausa.org>.

Neuropathy Association. 60 E. 42nd St., Suite 942, New York, NY 10165. Web site: <www.neuropathy.org>.

WEB SITES

"Charcot-Marie-Tooth Disease." *Health and Disease Information A to Z*, February 27, 2004. Available online at <www.hmc.psu.edu/healthinfo/c/cmt.htm> (accessed December 7, 2004).

"Charcot-Marie-Tooth Disorder Information Page." *NINDS Charcot-Marie-Tooth Disorder Information Page*, February 27, 2004. Available online at <www.ninds.nih.gov/health_and_medical/disorders/charcot_doc.htm> (accessed December 7, 2004).

Christine Kuehn Kelly

Charley horse *see* **Muscle spasms and cramps**

Chemotherapy

Definition

Chemotherapy, sometimes referred to as "chemo," is the treatment of **cancer** with anticancer drugs.

Purpose

The main purpose of chemotherapy is to kill cancer cells. It usually is used to treat patients with cancer that has spread from the place in the body where it originated (metastasized). Chemotherapy destroys cancer cells anywhere in the body. It even kills cells that have broken off from the main tumor and traveled through the blood or lymph systems to other parts of the body.

Chemotherapy can cure some types of cancer. In some cases, it is used to slow the growth of cancer cells or to keep the cancer from spreading to other parts of the body. Chemotherapy may be given before surgery or radiation therapy to shrink the tumor (neoadjuvant therapy). When a cancer has been removed by surgery or treated with radiation therapy, chemotherapy may be used to keep the cancer from coming back (adjuvant therapy).

Once a remission is achieved, consolidation chemotherapy, also called intensification chemotherapy, is given to sustain a remission. Maintenance chemotherapy is chemotherapy given in lower doses as a treatment to prolong a remission in certain types of cancer. Chemotherapy also can ease the symptoms of cancer (palliative chemotherapy), helping some patients have a better quality of life.

Description

Chemotherapy for the treatment of cancer began in the 1940s with the use of nitrogen mustard. More than 100 chemotherapy drugs are now available to treat cancer, and many more are being tested for their ability to destroy cancer cells.

Most chemotherapy drugs interfere with the ability of cells to grow or multiply. Although these drugs affect all cells in the body, many useful treatments are most effective against rapidly growing cells. Cancer cells grow more quickly than most other body cells. Other cells that grow fast are cells of the bone marrow that produce blood cells, cells in the stomach and intestines, and cells of the hair follicles. Therefore, the most common side effects of chemotherapy are linked to the treatment's effects on other fast-growing cells.

Types of chemotherapy drugs

Chemotherapy drugs are classified according to how they work. The main types of chemotherapy drugs are:

- Alkylating drugs—kill cancer cells by directly attacking DNA, the genetic material of the genes. Cyclophosphamide is an alkylating drug.
- Antimetabolites—interfere with the production of DNA and keep cells from growing and multiplying. An example of an antimetabolite is 5-fluorouracil (5-FU).
- Antitumor antibiotics—made from natural substances such as fungi in the soil. They interfere with important cell functions, including production of DNA and cell proteins. Doxorubicin and bleomycin belong to this group of chemotherapy drugs.
- Plant alkaloids—prevent cells from dividing normally. Vinblastine and vincristine are plant alkaloids obtained from the periwinkle plant.
- Steroid hormones—slow the growth of some cancers that depend on hormones. For example, tamoxifen is used to treat breast cancers whose growth depends on the hormone estrogen.
- Topoisomerase inhibitors—interfere with the action of topoisomerase enzymes, the enzymes that control the part of DNA needed to multiply. Etoposide belongs to this group.

Biological therapy

Biological therapy, also called immunotherapy, consists of treatment with substances that boost the body's own immune system against cancer. The body usually produces these substances in small amounts to fight diseases. These substances can be made in the laboratory

and given to patients to destroy cancer cells, change the way the body reacts to a tumor, or help the body repair or make new cells destroyed by chemotherapy.

Combination chemotherapy

Chemotherapy usually is given in addition to other cancer treatments, such as surgery and radiation therapy. When given with other treatments, it is called adjuvant chemotherapy. An oncologist decides on the specific chemotherapy drug or combination of drugs that will work best for each patient. The use of two or more drugs together—combination chemotherapy—often works better than using a single drug. Scientific studies of different drug combinations help doctors learn the combinations that work best for each type of cancer. For example, research in 2003 found that a combination of chemotherapy and gene therapy stopped breast cancer and its metastasis (spread to other organs or parts of the body).

Clinical trials

Some patients may be eligible to participate in clinical trials, research programs conducted with patients to evaluate a new drug, medical treatment, device, or combination of treatments. The purpose of clinical trials is to find new and improved methods of treating different diseases and special conditions. More information is available at the National Institutes of Health's clinical trials Web site, <www.clinicaltrials.gov> or by calling (888) FIND-NLM, (888) 346-3656 or (301) 594-5983. Another resource is the National Cancer Institute's Web site, <www.cancer.gov/clinicaltrials>.

Precautions

There are many different types of chemotherapy drugs. Oncologists, doctors who specialize in treating cancer, determine the drugs that are best suited for each patient. This decision is based on the type and severity of cancer, location of the cancer, patient's age and health, and other drugs the patient takes. Some patients should not be treated with certain chemotherapy drugs. Age and other conditions may affect the drugs selected for treatment. Heart disease, kidney disease, and diabetes are conditions that may limit the choice of treatment drugs. In 2003, research revealed that **obesity** appears to reduce the effectiveness of high-dose chemotherapy. Researchers said further study was needed to determine the best dosage for obese patients receiving therapy.

How chemotherapy is given

Chemotherapy is administered in different ways, depending on the drugs to be given and the type of cancer. Doctors determine the dose of chemotherapy drugs based on many factors, including the patient's height and weight.

Chemotherapy may be given by one or more of the following methods:

- orally (by mouth)
- injection
- through a catheter or port
- topically (via the skin)

Oral chemotherapy is given by mouth in the form of a pill, capsule, or liquid. This is the easiest method and can usually be done at home.

Intravenous (IV) chemotherapy is injected into a vein. A small needle is inserted into a vein on the hand or lower arm. The needle usually is attached to a small tube called a catheter, which delivers the drug to the needle from an IV bag or bottle.

Intramuscular (IM) chemotherapy is injected into a muscle. Chemotherapy given by intramuscular injection is absorbed into the blood more slowly than IV chemotherapy. Because of this, the effects of IM chemotherapy may last longer than chemotherapy given intravenously. Chemotherapy also may be injected subcutaneously (SQ or SC), which means under the skin. Injection of chemotherapy directly into the cancer is called intralesional (IL) injection.

Chemotherapy also may be given by a catheter or port permanently inserted into a central vein or body cavity. A port is a small reservoir or container that is placed in a vein or under the skin in the area where the drug will be given. These methods eliminate the need for repeated injections and may allow patients to spend less time in the hospital while receiving chemotherapy. A common location for a permanent catheter is the external jugular vein in the neck.

Intraperitoneal (IP) chemotherapy is administered into the abdominal cavity through a catheter or port. Chemotherapy given by catheter or port into the spinal fluid is called intrathecal (IT) administration. Catheters and ports also may be placed in the chest cavity, abdomen, bladder, or pelvis (intracavitary or IC catheter), depending on the location of the cancer to be treated.

Topical chemotherapy is given as a cream or ointment applied on the surface of the skin. This method is more common in treatment of certain types of skin cancers.

Treatment location and schedule

Patients may take chemotherapy at home, in the doctor's office, or as an inpatient or outpatient at the

hospital. The choice of where to have chemotherapy depends on the drugs, delivery method, and sometimes the patient's and family's personal preferences. Most patients receive chemotherapy as an outpatient. Some patients stay in the hospital when first beginning chemotherapy, so their doctor can check for any side effects and change the dose if needed.

Frequency and length of chemotherapy treatment depends on the form of cancer, types of drugs, how the patient responds to the treatment, and the patient's health and ability to tolerate the drugs. Chemotherapy administration may take only a few minutes or as long as several hours. Chemotherapy may be given daily, weekly, or monthly. A rest period may follow a course of treatment before the next course begins, allowing new cells to grow and the patient to recover and regain strength. In combination chemotherapy, more than one drug may be given at a time, or the drugs may be given alternately, one following the other.

Maintaining the prescribed treatment schedule is essential to ensuring that the drugs work properly. The doctor should be contacted as soon as possible if a treatment session is missed, or a dose of the drug is skipped, for whatever reasons. Sometimes the doctor may need to delay a treatment based on the results of certain blood tests. Specific instructions will be provided if a treatment delay becomes necessary.

Preparation

TESTS A number of medical tests are done before chemotherapy is started. The results of **x rays**, other imaging tests, and tumor samples taken during biopsy or surgery will help the oncologist determine how much the cancer has spread.

Blood tests give the doctor important information about the function of the blood cells and levels of chemicals in the blood. A complete blood count (CBC) is commonly done before and regularly during treatment. The CBC shows the numbers of white blood cells, red blood cells, and platelets in the blood. Because chemotherapy affects the bone marrow, where blood cells are made, levels of these cells often drop during chemotherapy. The white blood cells and platelets are most likely to be affected by chemotherapy. A drop in the white blood cell count means the immune system cannot function properly. Low levels of platelets can cause a patient to bleed easily from a cut or other wound. A low red blood cell count can lead to anemia (deficiency of red blood cells) and fatigue.

INFORMED CONSENT Informed consent is an educational process between health care providers, patients, and/or their legal guardians. Before any procedure is performed or any form of medical care is provided, the patient and parents (if the patient is under age 18), are asked to sign a consent form, which provides permission for the child to receive chemotherapy treatment. The health care provider will review the informed consent form with the parents before they are asked to sign it. Before signing the form, the patient and parents should understand the nature and purpose of the treatment, its risks and benefits, and alternatives, including the option of not proceeding with the treatment. During the discussion about treatment, the health care providers are available to answer the patient's and parents' questions about the consent form or course of treatment.

PREPARING FOR THE TREATMENT When a chemotherapy treatment takes a long time, the patient may prepare for it by wearing comfortable clothes. Packing a book, favorite game, or an audiotape may help pass the time and ease the stress of receiving chemotherapy.

Usually parents stay with their child during the treatment. It is necessary to drive the child home (even if he or she is old enough to drive), since the medications taken to control **nausea** and the chemotherapy treatment itself can cause drowsiness.

ANTI-EMETIC DRUGS Sometimes, patients taking chemotherapy drugs known to cause nausea are given medications called anti-emetics before chemotherapy is administered. Anti-emetic drugs help to lessen feelings of nausea. Two anti-nausea medications that may be used are Kytril and Zofran. To decrease nausea from occurring just after a chemotherapy session, the child should not eat for about two hours before the treatment appointment.

Research published in 2003 revealed that taking melatonin, a natural hormone substance, may help improve chemotherapy's effectiveness and reduce the toxic effects of the drugs.

Aftercare

Tips for helping to control side effects after chemotherapy include:

- following any instructions given by the doctor or nurse
- taking all prescribed medications
- eating small amounts of bland foods
- maintaining good **nutrition** by getting enough calories, including protein in the diet, and taking a daily multivitamin (as recommended by the child's physician)

- drinking at least eight cups of fluids per day
- getting plenty of rest
- exercising regularly

Some patients find it helpful to breathe fresh air or get mild **exercise**, such as taking a walk.

Side effects and their severity are not indicators of how well the chemotherapy is working, since they vary greatly among patients and from drug to drug. Tests and exams can help determine the effectiveness of the chemotherapy.

Risks

Chemotherapy drugs are toxic to normal cells as well as cancer cells. A dose that will destroy cancer cells will probably cause damage to some normal cells. Doctors adjust doses to do the least amount of harm possible to normal cells. Side effects are temporary, and damaged non-cancerous cells will be replaced with healthy cells.

Some patients feel few or no side effects, and others may have more serious side effects. In some cases, a dose adjustment is all that is needed to reduce or stop a side effect. The types and severity of side effects depend on the chemotherapy drugs, dose, length of therapy, the body's reaction to the drug, and the child's overall health at the start of chemotherapy.

Some chemotherapy drugs have more side effects than others. Among the most common side effects are:

- fatigue
- **nausea and vomiting**
- loss of appetite
- **diarrhea**
- hair loss
- anemia
- infection
- easy bleeding or bruising
- sores in the mouth and throat
- neuropathy and other damage to the nervous system
- kidney damage

Fatigue (feeling tired and lacking energy) is the most common side effect of cancer and chemotherapy medications. Fatigue gradually goes away as the cancer responds to treatment. To help a child cope with fatigue, parents should plan rest periods, provide nutritious meals to maintain energy and meet caloric needs, limit **caffeine**, and encourage exercise and activity.

Nausea and vomiting are common, but can usually be controlled by taking anti-nausea drugs; consuming adequate fluids; drinking fluids at least one hour before or after a meal; eating and drinking slowly, chewing food completely; eating smaller meals throughout the day; choosing high-carbohydrate, low-fat foods; and avoiding sweet, fried, or spicy foods. When vomiting episodes stop, the child may feel better after eating easy-to-digest and bland foods such as clear liquids, crackers, gelatin, and plain toast.

Loss of appetite may be due to nausea, changes in taste and smell, or the stress of undergoing cancer treatment. To help maintain the child's appetite, meals and snacks should be small rather than large. Food should be served when the child is hungry, and he or she should be offered favorite foods. It is recommended that children help select and prepare foods. Calories may be boosted by offering high-calorie and high-protein snacks and foods. Sometimes a feeding tube may be needed to maintain a child's weight or for children who cannot eat or drink.

If the child has diarrhea, high-fiber and high-fat foods, gassy foods, and carbonated beverages should be avoided. It is important for the child to continue drinking fluids throughout the day to avoid **dehydration** from diarrhea or vomiting.

Some chemotherapy drugs cause hair loss, but it is almost always temporary. The doctor can advise the parents and patients if hair loss is expected with the type of chemotherapy drug to be given. When hair loss occurs, it may begin after a few treatments, or several weeks after the first treatment. To care for the scalp and hair during chemotherapy, the child should use a mild shampoo and soft brush, and low heat for hair drying. The head should be protected from heat and sun with a hat or scarf. If desired, a wig or hair piece may be worn.

Low blood cell counts caused by the effect of chemotherapy on the bone marrow can lead to anemia, infections, and easy bleeding and bruising. Patients with anemia have too few red blood cells to deliver oxygen and nutrients to the body's tissues. Anemic patients feel tired and weak, are short of breath, and may have a rapid heartbeat. If red blood cell levels fall too low, a blood transfusion may be given.

Patients receiving chemotherapy are more likely to acquire infections because their infection-fighting white blood cells are reduced. It is important to take measures

to avoid infections. When the white blood cell count drops too low, the doctor may prescribe medications called colony stimulating factors, which help white blood cells grow. Neupogen and Leukine are two colony stimulants that help fight infection. To reduce the risk of infection, thorough and frequent hand washing and safe food preparation are essential.

Platelets are blood cells that make the blood clot. When patients do not have enough platelets, they may bleed or bruise easily, even from small injuries. Patients with low blood platelets should take precautions to avoid injuries. Medicines such as aspirin and other **pain** relievers can affect platelets and slow down the clotting process.

Chemotherapy can cause irritation and dryness in the mouth and throat. Painful sores may form that can bleed and become infected. To help avoid mouth sores and irritation, the child should have a dental cleaning before chemotherapy begins, take care of the teeth and gums by brushing and flossing after every meal with a soft brush, rinse with a solution of baking soda and water, and avoid mouth washes or rinses that contain salt or alcohol. After use, the toothbrush should be rinsed thoroughly and stored in a dry place.

To help the child cope with a dry mouth, parents should encourage him or her to drink plenty of liquids. Popsicles or lollipops offer relief. Soft foods may be prepared, and dry foods may be moistened with sauce, butter, or gravy.

Tiredness, confusion, and depression can occur from chemotherapy's effect on certain central nervous system functions. The doctor should be notified if these symptoms occur.

Tests will be performed to monitor the effects of chemotherapy medications on the patient's kidneys and liver. Monitoring kidney and liver function helps to avoid potential damage or complications.

Normal results

The main goal of chemotherapy is to cure cancer. In fact, many cancers are cured by chemotherapy. Chemotherapy may be used in combination with surgery to keep a cancer from spreading to other parts of the body. Some widespread, fast-growing cancers are more difficult to treat. In these cases, chemotherapy may slow the growth of cancer cells.

The results of medical tests provide information so doctors can tell if the chemotherapy is working. Physical examination, blood tests, and x rays are used to check the effects of treatment on the cancer.

The possible outcomes of chemotherapy are:

- Complete remission or response. The cancer completely disappears; there is no evidence of disease. The course of chemotherapy is completed and the patient is tested regularly for a recurrence.
- Partial remission or response. The cancer shrinks in size but does not disappear. The same chemotherapy may be continued or a different combination of drugs may be given.
- Stabilization. The cancer does not grow or shrink. Other therapy options may be explored. A tumor may stay stabilized for many years.
- Progression. The cancer continues to grow. Other therapy options may be explored.
- A secondary malignancy may develop from the one being treated, and that second cancer may need additional chemotherapy or other treatment.

Parental concerns

Some important questions parents can ask about their child's course of chemotherapy include:

- What specific drugs will be given?
- How will the drugs be administered, and where will they be given?
- What are the potential benefits and risks of these drugs?
- What are some other possible treatments for the child's type of cancer?
- What is the standard care for the child's type of cancer?
- Are there any applicable clinical trials currently enrolling children?
- How many treatments will be needed? How long will they last?
- What are the potential side effects? When might they occur? How can they be treated or relieved? How serious are they likely to be? What side effects should be reported to the child's doctor?
- Can the child take other prescription or over-the-counter medications while receiving chemotherapy?
- What activities should be restricted or limited during the course of treatment?
- What is the long-term effect of chemotherapy?

Most school-age children can continue to go to school while receiving chemotherapy. However, the

school schedule may need adjustment according to how the child feels and what side effects he or she experiences. During the cold and flu season, it may be best to keep the child home to prevent infection. If possible, treatments should be scheduled on a day when there is no school the next day, to provide time to recover.

To reduce the child's exposure to colds and illnesses and to help the child avoid infection:

- The child should avoid crowded areas, such as shopping malls.
- The child and entire **family** should be encouraged to wash hands frequently.
- People who are sick should be avoided, and they should be asked to refrain from visiting until they are healthy.
- The child should stay away from children who have recently received live virus vaccines such as chicken pox and oral **polio** since they may be contagious to people with a low blood cell count.
- Contact with animal litter boxes and waste, bird cages, and fish should be prevented.
- Contact with standing water, such as bird baths, flower vases, or humidifiers, should be prevented.
- Food must be safely prepared and cooked thoroughly to avoid food-borne illnesses.
- Parents should check with the child's doctor before scheduling immunizations, flu, or **pneumonia** vaccines.

Aspirin and products containing aspirin should be avoided, as they can affect platelet counts. Parents should check with the child's doctor before giving any **vitamins**, herbal supplements, and any over-the-counter medications.

The child's doctor should provide specific activity guidelines, including recommendations regarding the child's **sports** participation. Contact sports may be discouraged to reduce the risk of injury.

Treatment and care for a child with cancer can be costly, and some health insurance plans may not cover all expenses associated with a child's **hospitalization** or treatment. Help is available to cover medical expenses. The parents can discuss financial aid with the hospital. Some organizations provide financial assistance to children in need of chemotherapy or other cancer treatments.

Caring for a child with cancer is demanding. Support groups are available to help parents and caregivers cope with the challenges of providing care for children with special medical needs. It is important for parents to take care of themselves, too, by eating properly, exercising regularly, taking care of personal hygiene, keeping in contact with friends and family members for support, and managing stress by practicing relaxation techniques.

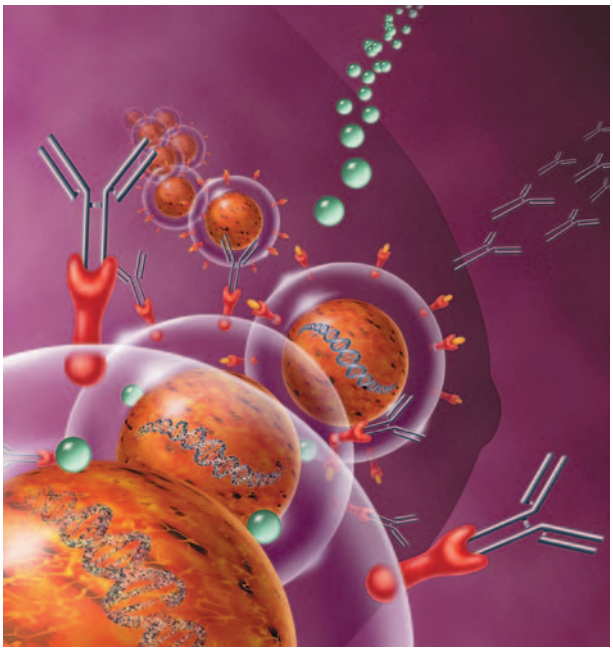

Illustration showing herceptin antibody, which inhibits DNA repair. When combined with chemotherapy (green balls), which damages DNA, the two inhibit the growth of cancer cells. *(© Art & Science/Custom Medical Stock Photo, Inc.)*

When to call the doctor

If a child has any of these symptoms, the parent or caregiver should call the child's doctor right away, as they could indicate an infection, blood clotting problem, or effect on the central nervous system:

- abdominal pain, vomiting, or diarrhea that awakens the child during the night
- persistent or severe abdominal pain, vomiting, or diarrhea
- unexplained weight loss
- fever
- chills or sweating
- frequent urgency to urinate, burning during urination, or change in color of urine
- rectal bleeding, or black or bloody bowel movements
- severe **cough** or sore throat
- redness, swelling, or tenderness, especially around a wound or sore

KEY TERMS

Adjuvant therapy—A treatment that is intended to aid primary treatment.

Alkaloid—A type of chemical commonly found in plants and often having medicinal properties.

Alkylating agent—A chemical that alters the composition of the genetic material of rapidly dividing cells, such as cancer cells, causing selective cell death; used as a chemotherapeutic agent.

Alopecia—The loss of hair, or baldness.

Anti-emetic—A preparation or medication that relieves nausea and vomiting. Cola syrup, ginger, and motion sickness medications are examples of antiemetics.

Antimetabolite—A drug or other substance that interferes with a cell's growth or ability to multiply.

Benign—In medical usage, benign is the opposite of malignant. It describes an abnormal growth that is stable, treatable, and generally not life-threatening.

Blood cell count—The number of red blood cells, white blood cells, and platelets in a sample of blood. Also called a complete blood count (CBC).

Bone marrow—The spongy tissue inside the large bones in the body that is responsible for making the red blood cells, most white blood cells, and platelets.

Catheter—A thin, hollow tube inserted into the body at specific points in order to inject or withdraw fluids from the body.

Chest x ray—Brief exposure of the chest to radiation to produce an image of the chest and its internal structures.

Infusion—Introduction of a substance directly into a vein or tissue by gravity flow.

Lymph nodes—Small, bean-shaped collections of tissue located throughout the lymphatic system. They produce cells and proteins that fight infection and filter lymph. Nodes are sometimes called lymph glands.

Malignant—Cells that have been altered such that they have lost normal control mechanisms and are capable of local invasion and spread to other areas of the body. Often used to describe a cancer.

Metastatic—The term used to describe a secondary cancer, or one that has spread from one area of the body to another.

Platelet—A cell-like particle in the blood that plays an important role in blood clotting. Platelets are activated when an injury causes a blood vessel to break. They change shape from round to spiny, "sticking" to the broken vessel wall and to each other to begin the clotting process. In addition to physically plugging breaks in blood vessel walls, platelets also release chemicals that promote clotting.

Radiation therapy—A cancer treatment that uses high-energy rays or particles to kill or weaken cancer cells. Radiation may be delivered externally or internally via surgically implanted pellets. Also called radiotherapy.

Red blood cells—Cells that carry hemoglobin (the molecule that transports oxygen) and help remove wastes from tissues throughout the body.

Remission—A disappearance of a disease and its symptoms. Complete remission means that all disease is gone. Partial remission means that the disease is significantly improved, but residual traces of the disease are still present. A remission may be due to treatment or may be spontaneous.

Tumor—A growth of tissue resulting from the uncontrolled proliferation of cells.

White blood cells—A group of several cell types that occur in the bloodstream and are essential for a properly functioning immune system; they fight infection.

- earaches, headaches, or stiff neck
- mouth sores, or blisters on the lips or skin
- sinus pain or pressure
- headaches
- changes in vision
- unexplained bleeding or bruising
- red spots under the skin
- confusion
- persistent depressed mood
- worsening overall health

Resources

BOOKS

Fischer, David A., et al. *The Cancer Chemotherapy Handbook*. London: C.V. Mosby, 2003.

McKay, Judith, and Nancee Hirano. *The Chemotherapy and Radiation Therapy Survivor's Guide,* 2nd ed. Oakland, CA: New Harbinger Publications, 1998.

Skeel, Roland T. K. *Handbook of Cancer Chemotherapy,* 6th ed. Philadelphia: Lippincott Williams & Wilkins, 2003.

PERIODICALS

"Chemotherapy and You: A Guide to Self-Help During Cancer Treatment." National Institutes of Health, National Cancer Institute. NIH Publication No. 03-1136, 2003.

"Gene Therapy and Chemotherapy Combine to Stop Breast Cancer and its Metastasis." *Gene Therapy Weekly* (Oct. 30, 2003): 2.

"Melatonin Improves the Efficacy of Chemotherapy and Quality of Life" *Biotech Week* (Sept. 10, 2003): 394.

"Obesity May Reduce Efficacy of High-Dose Chemotherapy." *Health Medicine Week* (Aug. 11, 2003): 385.

ORGANIZATIONS

American Cancer Society. 1599 Clifton Rd., NE, Atlanta, GA 30329-4251. (800) 227-2345 or (404) 320-3333. Web site:<www.cancer.org>.

Cancercare. (800) 813- 4673. Web site: <www.cancercare.org>.

National Cancer Institute. U.S. National Institutes of Health. Building 31, Room 10A31, 31 Center Drive, MSC 2580, Bethesda, MD 20892- 2580. (800) 422-6237. Web site: <www.cancer.gov>.

WEB SITES

CancerAnswers. Available online at: <www.canceranswers.com>.

Chemocare.com. Available online at: <www.chemocare.com>.

OncoLink. University of Pennsylvania Cancer Center. Available online at: <www.oncolink.upenn.edu>.

Planet Cancer. Available online at: <www.planetcancer.org>.

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Chest physical therapy

Definition

Chest physical therapy is the term for a group of treatments designed to improve respiratory efficiency, promote expansion of the lungs, strengthen respiratory muscles, and eliminate secretions from the respiratory system.

Purpose

The purpose of chest physical therapy, also called chest physiotherapy, is to help patients breathe more freely and to get more oxygen into the body. Chest physical therapy includes postural drainage, chest percussion, chest vibration, turning, deep breathing exercises, and coughing. In the early 2000s, some newer devices, such as the positive expiratory pressure (PEP) valve and the flutter device, have been added to the various chest physical therapy techniques. Chest physical therapy is normally done in conjunction with other treatments to rid the airways of secretions. These other treatments include suctioning, nebulizer treatments, and the administering expectorant drugs.

Description

Good respiratory health is not possible without efficient clearance of secretions in the airway. In a healthy person, this is normally accomplished through two mechanisms: the mucociliary clearance system (MCS) and the ability to **cough**. There are many diseases and disabilities in children linked with poor lung health and an impaired ability to clear secretions. These include **cystic fibrosis**, **asthma**, **cerebral palsy**, **muscular dystrophy**, and various **immunodeficiency** disorders. When a child is unable to clear mucus, breathing becomes hard work. He or she must expend extra effort and energy in order to get oxygen. This difficulty can lead to a vicious cycle of recurrent episodes of inflammation, respiratory infections, lung damage, increased production of excess mucus, and possibly airway obstruction. Chest physical therapy is one way to reduce the risks of an inefficient clearance of airway secretions. Depending on the specific technique and health situation, chest physical therapy may be used on children from newborns to adolescents.

Various methods of chest physical therapy have been used since the early 1900s to help manage airway clearance disorders. The techniques have been refined since that time. The procedure may be performed by a respiratory therapist, a nurse, or a trained **family** member. However, chest physical therapy presents some challenges and requires skill and training in order to be safely and effectively performed.

Chest physical therapy is a method of clearing the airway of excess mucus. It is based on the theory that when various areas of the chest and back are percussed, shock waves are transmitted through the chest wall, loosening the airway secretions. If the child is positioned appropriately, the loosened secretions will then drain

into the upper airways, where they can then be cleared using coughing and deep breathing techniques. The following techniques are all part of chest physical therapy.

Turning

Turning from side to side permits lung expansion. The child may turn on his or her own, or be turned by a caregiver. Turning should be done at a minimum of every two hours if the child is bedridden. The head of the bed can also be elevated in order to promote drainage.

Coughing

Coughing helps to break up secretions in the lungs so that the mucus can be expectorated or suctioned out if necessary. Patients sit upright and inhale deeply through the nose. They then exhale in short puffs or coughs. This procedure is repeated several times a day.

Deep breathing

Deep breathing helps expand the lungs and forces an improved distribution of the air into all sections of the lungs. The patient either sits in a chair or sits upright in bed and inhales then pushes the abdomen out to force maximum amounts of air into the lung. The abdomen is then contracted, and the patient exhales. Deep breathing exercises are done several times each day for short periods.

Because of the mind-body awareness required to perform coughing and deep breathing exercises, they are unsuitable for most children under the age of eight.

Postural drainage

Postural drainage uses the force of gravity to assist in effectively draining secretions from the smaller airways into the central airway where they can either be coughed up or suctioned out. The child is placed in a head- or chest-down position and is kept in this position for up to 15 minutes. To obtain the head-down positions, the use of a pillow, beanbag chair, or couch cushions can be helpful. Often, percussion and vibration are performed in conjunction with postural drainage.

Percussion

Percussion involves rhythmically striking the chest wall with cupped hands. It is also called cupping or clapping. The purpose of percussion is to break up thick secretions in the lungs so they can more easily be removed. Percussion is performed on each lung segment for one to two minutes at a time. Mechanical percussors are available and may be suitable for children over two years of age. The percussor is moved over one lobe of the lung for approximately five minutes, while the patient is encouraged to performing coughing and deep breathing techniques. This process is repeated until each segment of the lung is percussed.

Vibration

As with percussion, the purpose of vibration is to help break up lung secretions. Vibration can be either mechanical or manual. It is performed as the patient breathes deeply. When done manually, the person performing the vibration places his or her hands against the patient's chest and creates vibrations by quickly contracting and relaxing arm and shoulder muscles while the patient exhales. The procedure is repeated several times each day for about five exhalations.

Positive expiratory pressure (PEP)

PEP therapy has been extensively tested and is equivalent to standard chest physical therapy. It is an airway clearance method that is administered by applying a mechanical pressure device to the mouth. By breathing out with a moderate force through the resistance of the device, a positive pressure is created in the airways that helps to keep them open. This positive pressure permits airflow to reach beneath the areas of mucus obstruction and to move the mucus toward the larger airways where it can be expectorated. This technique may be suitable for alert, cooperative children over the age of four.

Flutter

The flutter valve is a hand-held mucus clearance device designed to combine positive expiratory pressure (PEP) with high frequency airway oscillations. The device looks like a pipe containing an inner cone that cradles a steel ball sealed with a perforated cover. Exhalation through the device results in a vibration of the airway walls, which in turn loosens secretions. It may be a suitable technique for children aged five years and over.

A child is considered to have responded positively to chest physical therapy if some, but not necessarily all, of the following changes occur:

- increased volume of sputum secretions
- changes in breath sounds
- improved chest x ray
- increased oxygenation of the blood as measured by arterial blood gas sampling
- the child's report of increased ease in breathing

Precautions

Chest physical therapy should not be performed on those children with the following:

- bleeding in the lungs
- head or neck injuries
- fractured ribs
- collapsed lungs
- acute asthma
- pulmonary embolism
- active hemorrhage
- some spinal injuries
- open **wounds** or burns

Preparation

The child should be taught about the necessity and rationale for chest physical therapy. It may be a challenge to get children to cooperate with the procedure. Providing a toy, watching a video, or giving a reward may be ways to encourage cooperation.

Aftercare

Many children may wish to perform **oral hygiene** measures after therapy to lessen the poor taste of the secretions they have expectorated.

Risks

The risks and complications associated with chest physical therapy are dependent upon the health of the child. Although chest physical therapy normally poses few problems, in some patients it may cause the following:

- oxygen deficiency if the head is kept lowered for drainage
- increased intracranial pressure
- temporary lowering of blood pressure
- bleeding in the lungs
- pain or injury to the ribs, muscles, or spine
- vomiting
- inhalation of secretions into the lungs
- heart irregularities

Parental concerns

Because chest physical therapy is often prescribed for childrven with chronic health problems, parents are often required to learn the techniques so the procedure can be performed regularly at home. Many parents are fearful they might hurt their child or may perform the procedure incorrectly. They should be reassured that thousands of parents have learned how to perform chest physical therapy and do so safely and effectively.

KEY TERMS

Coughing—In chest physical therapy, coughing is used to help break up secretions in the lungs so that the mucus can be suctioned out or expectorated. Patients sit upright and inhale deeply through the nose. They then exhale in short puffs or coughs.

Deep breathing—Deep breathing helps expand the lungs and forces better distribution of the air into all sections of the lung. The patient either sits in a chair or sits upright in bed and inhales, pushing the abdomen out to force maximum amounts of air into the lung. The abdomen is then contracted, and the patient exhales.

Mucociliary escalator—The coordinated action of tiny projections on the surfaces of cells lining the respiratory tract, which moves mucus up and out of the lungs.

Percussion—An assessment method in which the surface of the body is struck with the fingertips to obtain sounds that can be heard or vibrations that can be felt. It can determine the position, size, and consistency of an internal organ. It is performed over the chest to determine the presence of normal air content in the lungs, and over the abdomen to evaluate air in the loops of the intestine.

Postural drainage—The use of positioning to drain secretions from the bronchial tubes and lungs into the trachea or windpipe where they can either be coughed up or suctioned out.

Vibration—The treatment that is applied to help break up lung secretions. Vibration can be either mechanical or manual. It is performed as the patient breathes deeply. When done manually, the person performing the vibration places his or her hands against the patient's chest and creates vibrations by quickly contracting and relaxing arm and shoulder muscles while the patient exhales. The procedure is repeated several times each day for about five exhalations.

Resources

PERIODICALS

"Maintaining Healthy Lungs: The Role of Airway Clearance Therapy." *The Exceptional Parent* 31 (August 2001): 126–33.

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Chiari malformation

Definition

Chiari malformation is a congenital anomaly (a condition that is present at birth), in which parts of the brain protrude through the opening in the base of the skull into the spinal column.

Description

In order to explain the various components of Chiari malformation, it is helpful to describe a few parts of the brain and their functions. There are four major regions of the brain affected in Chiari malformation: the cerebellum, the brain stem, the ventricles, and the cerebrum. The cerebellum is located at the base of the skull and is divided into two parts or hemispheres with a third section that connects the hemispheres. Its main purpose is to coordinate body movements. The brain stem is located in front of the cerebellum and is composed of two parts. It regulates involuntary actions the body must conduct to survive, such as breathing, swallowing, and blinking the eyes. There are four ventricles in the brain. They are located above and in front of the cerebellum, and their function is to produce and circulate cerebrospinal fluid (CSF), the protective fluid that circulates through the brain and the spinal cord. The cerebrum is the largest part of the brain and is divided into two halves or hemispheres as well. It is located above the cerebellum and is responsible for the higher functions of the brain, such as thought. In Chiari malformation, one or more of these parts of the brain function improperly or are malformed. In addition to brain anomalies, Chiari malformation can also involve defects in the base of the skull and in the bony part of the spine.

There are four types of Chiari malformation. In Type I malformation, the lower portions of the cerebellum, known as the cerebellar tonsils, protrude through the opening in the skull known as the foramen magnum and into the spinal cord canal. It is often accompanied by a condition known as syringomyelia in which pockets of CSF form in the spinal cord. This type is usually diagnosed in **adolescence** or early adulthood when symptoms most commonly appear; however, with the availability of **magnetic resonance imaging** (MRI), many children are diagnosed at a much younger age.

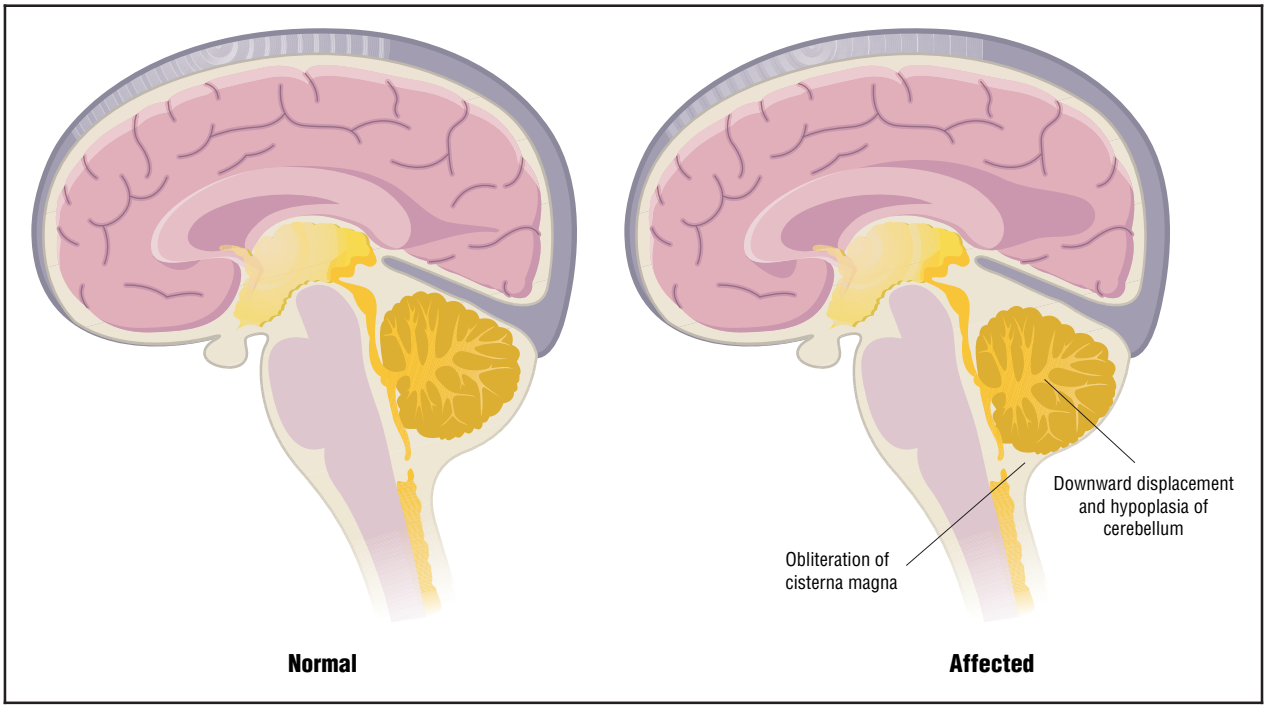
Type II malformation, sometimes called Arnold Chiari malformation, is more severe than Type I and involves herniation of a more significant part of the cerebellum, part of the fourth ventricle, and parts of the brain stem. The brain tissues protrude farther into the spinal column than in Type I. These malformations are part of a larger syndrome seen in children with **spina bifida**, a condition in which the spine and spinal cord have not formed properly. Approximately 80–90 percent of children with Chiari malformation Type II also have **hydrocephalus**, a condition in which one or more of the ventricles becomes enlarged due to an accumulation of CSF. In these children, hydrocephalus is caused by obstruction of the fourth ventricle due to its herniation into the spinal column. Type II Chiari malformation may be diagnosed prenatally by ultrasound or shortly after birth during medical evaluation of the accompanying spina bifida.

In Type III malformation, parts of the cerebellum and the brain stem protrude into a spina bifida defect located at the base of the skull, on the neck. Type III malformation occurs rarely. Some neurologists do not consider it a Chiari malformation but rather a specific type of spina bifida called an encephalocervical meningocele.

Type IV malformation consists of an underdevelopment of the cerebrum and involves no herniation of brain tissue into the spinal area. As with Type III malformation, many neurologists do not consider this a Chiari malformation but rather cerebellar hypoplasia (underdevelopment). Both Type III and IV Chiari malformations are extremely rare, and this term is not often used in diagnosis of these conditions. The remainder of this entry only discusses Chiari malformation Types I and II.

Demographics

The true incidence of Chiari malformation is unknown. Some researchers believe that there may be far



Comparison of normal brain (left) with brain affected by Arnold-Chiari malformation. *(Illustration by Argosy. Inc.)*

more cases of Type I malformation, in particular, than reported since many individuals with Type I malformation experience few if any symptoms. Most researchers agree that the rate of both Type I and Type II Chiari malformation is approximately 1 percent of all live births. Of those with Type I malformation, approximately 25 percent also have syringomyelia. However, the majority of all Chiari malformations are Type II and are almost exclusively associated with spina bifida defects. Spina bifida occurs in approximately one to two per 1,000 births. As access to imaging testing such as MRI has increased, so has the number of children diagnosed with Chiari malformation Type I. Therefore, the incidence of known Type I Chiari malformation is anticipated to increase.

Causes and symptoms

All Chiari malformations are present at birth, though symptoms may not begin until years later. The exact cause is unknown; however, it is suspected that, at some point during embryonic development, an increased pressure in the brain may cause brain structures to be displaced or moved into the spinal canal. Other possible causes for this malformation include exposure to harmful substances during fetal development or genetic factors. In general, there are several accepted theories of what may lead to problems that affect normal development of the brain: exposure to toxic or harmful substances; a lack of proper **vitamins** and nutrients in the mother's diet during pregnancy; infection; maternal use of prescription medication, illegal drugs, or alcohol; and genetic or familial factors. Chiari malformations are found in several known genetic disorders such as achondroplasia, Hadju-Cheney syndrome, and Klippel-Feil syndrome. Similarly, studies of families and identical **twins** with Chiari malformation show that the malformation occurs more often in these families than in families in which no member is affected. Another proposed cause for Chiari malformation is an abnormality in bone development. Chiari malformation may result because the cerebellum is of normal size, but the bones at the base of the skull are too small.

Symptoms of Chiari malformation vary according to the type of malformation. In Type I Chiari malformation, symptoms may begin anytime between infancy and early adulthood. Depending on when it is diagnosed, these symptoms may include blurred or double vision; involuntary eye movements; **headache**, usually at the base of the skull or upper neck which may become worse with coughing or straining; **scoliosis** or an abnormal curvature of the spine; **dizziness** and impaired muscle coordination; low muscle tone; alteration of the voice or a high-pitched cry; frequent respiratory tract infections, **vomiting** and difficulty swallowing; drop attacks (a sudden loss of muscle control that results in a collapse to the floor); and central cord syndrome, a pattern of reduced sensation and weakness in the arms.

Symptoms of Type II Chiari malformation include those that occur in Type I malformation. Victims may also exhibit vocal cord paralysis and episodes of apnea

(a cessation of breathing sometimes requiring resuscitation). Type II malformation occurs almost exclusively with spina bifida, which causes symptoms that may include paralysis of the lower extremities (and less often, the upper limbs), and bowel and bladder dysfunction. If the child has hydrocephalus, these symptoms may worsen and can be fatal unless the hydrocephalus is treated.

Diagnosis

In some children Chiari malformation is evident at birth, especially Type II malformation. Type I malformations may have no symptoms for years. When symptoms are present, they may be very subtle. A complete medical history and physical exam will be conducted by the physician. If the doctor suspects a Chiari malformation, magnetic resonance imaging (MRI) is the most helpful diagnostic tool. MRI is a diagnostic procedure in which high-powered magnets, radio frequencies, and computers are used to produce detailed images of structures within the body. It is painless, noninvasive, and allows doctors to see the brain and spinal cord from several different angles. A **computed tomography** scan (CAT scan) may also be performed. In this imaging procedure, a combination of x ray and computer technology produces cross-sectional images of the body. This procedure is most helpful in assessing abnormalities of the skull and backbone associated with Chiari malformation. Ultrasonography (ultrasound) is also routinely used to evaluate the fetus before birth and in the period shortly after birth. Ultrasonography is a diagnostic procedure in which ultrasonic waves are used to visualize internal organs. This procedure is commonly used for diagnosis and follow-up care of hydrocephalus.

Prognosis

The prognosis for Chiari malformation varies depending on which type of malformation is present. For children and adolescents with Type I malformation, corrective surgery may be highly effective at relieving symptoms related to compression of the brain, such as vision problems; headaches; difficulties with balance, coordination, and swallowing; and frequent respiratory tract infections. Symptoms related to the fluid-filled sacs in the spine may be less responsive to surgery. Scoliosis will most likely stabilize and may improve in a significant portion of patients. Central cord syndrome, however, is less successfully corrected by surgery with only about one third of patients experiencing lasting improvement. Generally, in those with Type I Chiari malformation, younger children experience the most benefit from surgical interventions, and approximately 78 percent of all patients who have surgical procedures to correct the malformation have a better outcome than those who have no surgical intervention at all. Some patients may continue to experience neurological symptoms, but the long term prognosis for children with this type of Chiari malformation is excellent, including normal development and intellectual functioning.

The majority of all Chiari malformations are Type II. Children with Chiari II malformation have a much poorer prognosis than those with Type I malformation. Because of the associated conditions of spina bifida and hydrocephalus, these children are usually quite ill. Multiple surgeries are required, and almost 50 percent of these children die at an early age. Of those who survive, many have developmental delays and impaired intellectual functioning.

Prevention

Chiari malformation is a congenital anomaly, and no method of prevention is known.

Parental concerns

Chiari malformation can have a significant impact on both the child and the **family**. The full extent of problems associated with Chiari malformation may not be evident at birth, especially for children with Type I malformation. Children with Type I malformation may experience months of subtle but progressive symptoms before a diagnosis is made. Parents must play an active role in securing appropriate health care and an accurate diagnosis for their child. The symptoms of Chiari Type I malformation when observed individually may not seem significant. However, these symptoms form the basis of the initial diagnosis. Parents should document all unusual events their child experiences such as dizziness, headaches, slurred speech, fainting spells, and **numbness** or **pain** in the arms and legs. The first step for diagnosing Chiari malformation Type I is an accurate and detailed history and physical examination. Following this, an MRI is performed. The MRI usually provides a definitive diagnosis and helps determine if the child is a candidate for decompression surgery.

Medical treatment is a multidisciplinary team effort often involving the pediatrician, a neurosurgeon, a rhematologist, and a neurologist. Once diagnosed, the child and family face the prospect of surgery and recuperation. A child will typically be hospitalized for four to seven days after surgery. The prognosis for children with Chiari malformation Type I is very good. Over 70 percent of those who have surgery to correct Type I malformation experience a significant reduction in symptoms; however, there are some symptoms that may not resolve. There may be continued discomfort in the neck and lower head, and some muscle weakness may be permanent.

KEY TERMS

Brain stem—The part of the brain that is continuous with the spinal cord and controls most basic life functions. It is the last part of the brain that is destroyed by Alzheimer's disease.

Catheter—A thin, hollow tube inserted into the body at specific points in order to inject or withdraw fluids from the body.

Cerebellum—The part of the brain involved in the coordination of movement, walking, and balance.

Cerebrospinal fluid—The clear, normally colorless fluid that fills the brain cavities (ventricles), the subarachnoid space around the brain, and the spinal cord and acts as a shock absorber.

Cerebrum—The largest section of the brain, which is responsible for such higher functions as speech, thought, vision, and memory.

Computed tomography (CT)—An imaging technique in which cross-sectional x rays of the body are compiled to create a three-dimensional image of the body's internal structures; also called computed axial tomography.

Congenital—Present at birth.

Dura mater—The strongest and outermost of three membranes that protect the brain, spinal cord, and nerves of the cauda equina.

Embryonic—Early stages of life in the uterus.

Foramen magnum—The opening at the base of the skull, through which the spinal cord and the brainstem pass.

Herniation—Bulging of tissue through opening in a membrane, muscle, or bone.

Hydrocephalus—An abnormal accumulation of cerebrospinal fluid within the brain. This accumulation can be harmful by pressing on brain structures, and damaging them.

Hypoplasia—An underdeveloped or incomplete tissue or organ usually due to a decrease in the number of cells.

Magnetic resonance imaging (MRI)—An imaging technique that uses a large circular magnet and radio waves to generate signals from atoms in the body. These signals are used to construct detailed images of internal body structures and organs, including the brain.

Neurologist—A doctor who specializes in disorders of the nervous system, including the brain, spinal cord, and nerves.

Neurosurgeon—Physician who performs surgery on the nervous system.

Rhematologist—A physician who specializes in the treatment of disorders of the connective tissue structures, such as the joints and related structures.

Scoliosis—An abnormal, side-to-side curvature of the spine.

Syringomyelia—Excessive fluid in the spinal cord.

Syrinx—A tubular fluid-filled cavity within the spine.

Ultrasonography—A medical test in which sound waves are directed against internal structures in the body. As sound waves bounce off the internal structure, they create an image on a video screen. Ultrasonography is often used to diagnose fetal abnormalities, gallstones, heart defects, and tumors. Also called ultrasound imaging.

Ventricles—Four cavities within the brain that produce and maintain the cerebrospinal fluid that cushions and protects the brain and spinal cord.

Many children may also experience a recurrence of symptoms within two years of surgery. For this reason, parents must continue follow-up treatment and care for at least two years following surgery. The availability of MRI technology has led to diagnosis of younger children with Type I malformation and improved the quality of life for these children. The earlier treatment can begin, the better outcome these children experience.

When discussing Chiari malformation and surgery with their child, parents should use words the child can understand. How illness and surgery are discussed depends on the age of the child. A younger child needs short simple answers, a school-aged child may understand more complicated explanations, and an adolescent will probably be present at all meetings with doctors and should be encouraged to talk to healthcare providers him- or herself. Regardless of the age of the child, mental health experts agree that children should be told about medical procedures and surgeries before they occur. Depending on the age of the child, parents may choose to talk to him a day before or weeks ahead. The child may have questions. Parents can answer them simply and honestly. While a younger child may ask many questions, an adolescent may be reluctant to appear ignorant and may

not express his **fear** or confusion. Parents should encourage an adolescent to discuss all concerns and should be prepared to explain and reassure. Many doctors and children's hospitals have brochures to assist parents when discussing illness and surgery with their child.

Children with Chiari II malformation have a much poorer prognosis than those with Type I malformation and will usually be quite ill. Multiple surgeries are required, and many of these children die at an early age. All of these factors have a significant impact on the family. These children require multiple services, including surgery, physical and occupational therapy, and **special education**. Many have developmental delays and impaired intellectual functioning in addition to the physical limitations often caused by the accompanying spina bifida. Parents may need support services in addition to the team of healthcare providers. Many parents find it helpful to participate in a support group of other families of children with special needs. Most states and children's hospitals offer these services and know of organizations in the area that may help these families. Parents can work closely with the hospital's social work department to learn more about available resources.

See also Hydrocephalus; Spina bifida.

Resources

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Chickenpox

Definition

Chickenpox (also called varicella) is a common, extremely infectious, rash-producing childhood disease that also affects adults on occasion.

Description

Chickenpox is caused by the varicella-zoster virus (a member of the herpes virus family), which is spread through the air or by direct contact with an infected person. It produces an itchy, blistery rash that typically lasts about a week and is sometimes accompanied by a **fever** or other symptoms. A single attack of chickenpox almost always brings lifelong immunity against the disease. Because the symptoms of chickenpox are easily recognized and in most cases merely unpleasant rather than dangerous, treatment can almost always be carried out at home. Severe complications can develop, however, and professional medical attention is essential in some circumstances.

Once someone has been infected with the virus, an incubation period of about 10 to 21 days passes before symptoms begin. The period during which infected people are able to spread the disease is believed to start one or two days before the rash breaks out and to continue until all the blisters have formed scabs, which usually happens four to seven days after the rash breaks out but may be longer in adolescents and adults. For this reason, doctors recommend keeping children with chickenpox away from school for about a week. It is not necessary, however, to wait until all the scabs have fallen off.

Prior to the use of the varicella vaccine, chickenpox was a typical part of growing up for most children in the industrialized world. The disease can strike at any age, but throughout the twentieth century by ages nine or ten about 80 to 90 percent of American children had already been infected. U.S. children living in rural areas and many foreign-born children were less likely to be immune. Study results reported by the Centers for Disease Control and Prevention (CDC) indicate that more than 90 percent of American adults are immune to the chickenpox virus. Adults, however, are much more likely than children to suffer dangerous complications. More than half of all chickenpox deaths occur among adults.

Demographics

Before the varicella vaccine (Varivax) was released for use in 1995, nearly all of the 4 million children born

each year in the United States contracted chickenpox, resulting in **hospitalization** in five of every 1,000 cases and 100 deaths. Because almost every case of chickenpox, no matter how mild, leads to lifelong protection against further attacks, adults account for less than 5 percent of all cases in the United States.

Causes and symptoms

A case of chickenpox usually starts without warning or with only a mild fever and a slight feeling of unwellness. Within a few hours or days small red spots begin to appear on the scalp, neck, or upper half of the trunk. After another 12 to 24 hours the spots typically become itchy, fluid-filled bumps called vesicles, which continue to appear in crops for the next two to five days. In any area of skin, lesions of a variety of stages can be seen. These blisters can spread to cover much of the skin, and in some cases also may be found inside the mouth, nose, ears, vagina, or rectum. Some people develop only a few blisters, but in most cases the number reaches 250 to 500. The blisters soon begin to form scabs and fall off. Scarring usually does not occur unless the blisters have been scratched and become infected. Occasionally a minor and temporary darkening of the skin (called hyperpigmentation) is noticed around some of the blisters. The degree of itchiness can range from barely noticeable to extreme. Some chickenpox sufferers also have headaches, abdominal **pain**, or a fever. Full recovery usually takes five to ten days after the first symptoms appear. Again, the most severe cases of the disease tend to be found among older children and adults.

Although for most people chickenpox is no more than a matter of a few days' discomfort, some groups are at risk for developing complications, the most common of which are bacterial infections of the blisters, **pneumonia**, **dehydration**, **encephalitis**, and hepatitis. Some of the groups at greater risk are:

- Infants: Complications occur much more often among children younger than one year old than among older children. The threat is greatest to newborns, who are more at risk of death from chickenpox than any other group. Under certain circumstances, children born to mothers who contract chickenpox just prior to delivery face an increased possibility of dangerous consequences, including brain damage and death. If the infection occurs during early pregnancy, there is a small (less than 5%) risk of congenital abnormalities.
- Immunocompromised children: Children whose immune systems have been weakened by a genetic disorder, disease, or medical treatment usually experience the most severe symptoms of any group. They have the second-highest rate of death from chickenpox.
- Adults and children 15 and older: Among this group, the typical symptoms of chickenpox tend to strike with greater force, and the risk of complications is much higher than among young children.

Immediate medical help should always be sought when anyone in these high-risk groups contracts the disease.

Diagnosis

Where children are concerned, especially those with recent exposure to the disease, diagnosis can usually be made at home, by a school nurse, or by a doctor over the telephone if the child's parent or caregiver is unsure that the disease is chickenpox.

Treatment

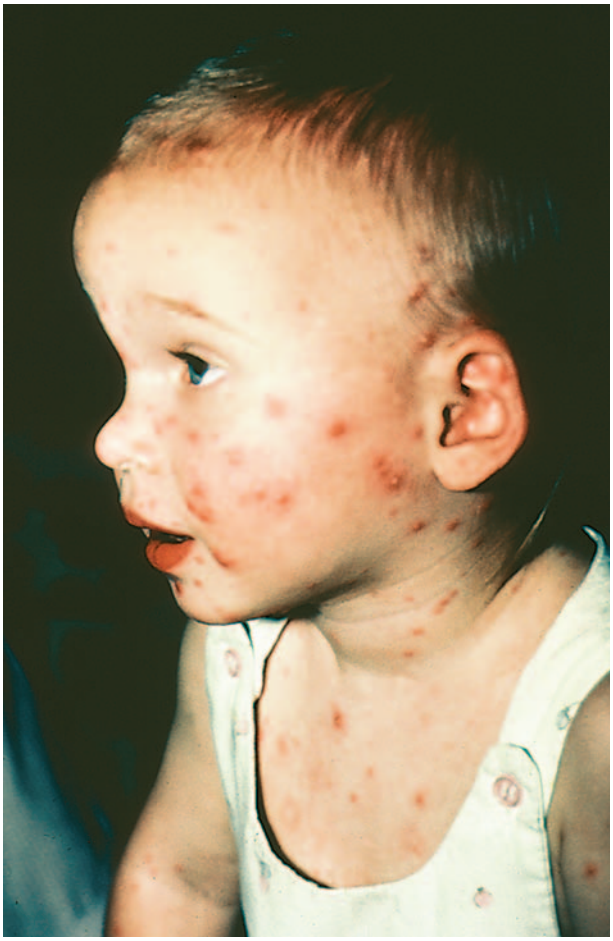
With children, treatment usually takes place in the home and focuses on reducing discomfort and fever. Because chickenpox is a viral disease, **antibiotics** are ineffective against it.

Applying wet compresses or bathing the child in cool or lukewarm water once a day can help the itch. Adding four to eight ounces of baking soda or one or two cups of oatmeal to the bath is a good idea (oatmeal bath packets are sold by pharmacies). Only mild soap should be used in the bath. Patting, not rubbing, is recommended for drying the child off, to prevent irritating the blisters. Calamine lotion (and some other kinds of lotions) also reduces itchiness. Because scratching can cause blisters to become infected and lead to scarring, the child's nails should be cut short. Of course, older children need to be warned not to scratch. For babies, light mittens or socks on the hands can help guard against scratching.

If mouth blisters make eating or drinking an unpleasant experience, cold drinks and soft, bland foods can ease the child's discomfort. Painful genital blisters can be treated with an anesthetic cream recommended by a doctor or pharmacist. Antibiotics often are prescribed if blisters become infected.

Fever and discomfort can be reduced by **acetaminophen** or another medication that does not contain aspirin. Aspirin and any medications that contain aspirin or other salicylates must not be used with chickenpox, for they appear to increase the chances of developing **Reye's syndrome**. The best idea is for a parent to consult a doctor or pharmacist to confirm which medications are safe.

Immunocompromised chickenpox sufferers are sometimes given an antiviral drug called acyclovir (Zovirax). Studies have shown that Zovirax also lessens the symptoms of otherwise healthy children and adults who contract chickenpox, but the notion that it should be



Child with chickenpox on the face and chest. *(Photograph by John D. Cunningham. Visuals Unlimited.)*

used to treat the disease among the general population, especially in children, is controversial.

Prognosis

Most cases of chickenpox run their course within a week without causing lasting harm. However, there is one long-term consequence of chickenpox that strikes about 20 percent of the population, particularly people 50 and older. Like all herpes viruses, the varicella-zoster virus never leaves the body after an episode of chickenpox. It lies dormant in the nerve cells, where it may be reactivated years later by disease or age-related weakening of the immune system. The result is shingles (also called herpes zoster), a painful nerve inflammation, accompanied by a rash that usually affects the trunk or the face for ten days or more. Especially in the elderly, pain, called postherpetic neuralgia, may persist at the site of the shingles for months or years. As of 2004, two relatively newer drugs for treatment of shingles are valacyclovir (Valtrex) and famciclovir (Famvir), both of which stop the replication of herpes zoster when administered within 72 hours of appearance of the rash. The effectiveness of these two drugs in immunocompromised patients has not been established, and Famvir was not recommended for patients under 18 years.

Prevention

A substance known as varicella-zoster immune globulin (VZIG), which reduces the severity of chickenpox symptoms, is as of 2004 available to treat immunocompromised children and others at high risk of developing complications. It is administered by injection within 96 hours of known or suspected exposure to the disease and is not useful after that. VZIG is produced as a gamma globulin from blood of recently infected individuals.

A vaccine for chickenpox became available in the United States in 1995 under the name Varivax. Varivax is a live, attenuated (weakened) virus vaccine. It has been proven to be 85 percent effective for preventing all cases of chickenpox and close to 100 percent effective in preventing severe cases. Side effects are normally limited to occasional soreness or redness at the injection site. CDC guidelines state that the vaccine should be given to all children (with the exception of certain high-risk groups) at 12 to 18 months of age, preferably when they receive their measles-mumps-rubella vaccine. For older children, up to age 12, the CDC recommends **vaccination** when a reliable determination that the child in question has already had chickenpox cannot be made. Vaccination also is recommended for any older child or adult considered susceptible to the disease, particularly those, such as healthcare workers and women of childbearing age, who face a greater likelihood of severe illness or transmitting infection. A single dose of the vaccine was once thought sufficient for children up to age 12; older children and adults received a second dose four to eight weeks later. However, an outbreak at a day-care center in 2000 brought concern in the medical community about a second vaccination for younger children, since many of the affected children had been vaccinated. Researchers began recommending a second vaccination in 2002. In 1997, the cost of two adult doses of the vaccine in the United States was about $80. Although this cost was not always covered by health insurance plans, children up to age 18 without access to the appropriate coverage could be vaccinated free of charge through the federal Vaccines for Children program. Varivax is not given to patients who already have overt signs of the disease. It was once thought unsafe for children with chronic kidney disease, but a 2003 report said the vaccination was safe in these children. The finding is important, since even chickenpox can be a serious complication in children who must undergo a kidney transplant.

KEY TERMS

Acetaminophen—A drug used for pain relief as well as to decrease fever. A common trade name for the drug is Tylenol.

Acyclovir—An antiviral drug, available under the trade name Zovirax, used for combating chickenpox and other herpes viruses.

Dehydration—An excessive loss of water from the body. It may follow vomiting, prolonged diarrhea, or excessive sweating.

Encephalitis—Inflammation of the brain, usually caused by a virus. The inflammation may interfere with normal brain function and may cause seizures, sleepiness, confusion, personality changes, weakness in one or more parts of the body, and even coma.

Hepatitis—An inflammation of the liver, with accompanying liver cell damage or cell death, caused most frequently by viral infection, but also by certain drugs, chemicals, or poisons. May be either acute (of limited duration) or chronic (continuing). Symptoms include jaundice, nausea, vomiting, loss of appetite, tenderness in the right upper abdomen, aching muscles, and joint pain. In severe cases, liver failure may result.

Immune system—The system of specialized organs, lymph nodes, and blood cells throughout the body that work together to defend the body against foreign invaders (bacteria, viruses, fungi, etc.).

Immunocompromised—A state in which the immune system is suppressed or not functioning properly.

Pneumonia—An infection in which the lungs become inflamed. It can be caused by nearly any class of organism known to cause human infections, including bacteria, viruses, fungi, and parasites.

Pus—A thick, yellowish or greenish fluid composed of the remains of dead white blood cells, pathogens, and decomposed cellular debris. It is most often associated with bacterial infection.

Reye's syndrome—A serious, life-threatening illness in children, usually developing after a bout of flu or chickenpox, and often associated with the use of aspirin. Symptoms include uncontrollable vomiting, often with lethargy, memory loss, disorientation, or delirium. Swelling of the brain may cause seizures, coma, and in severe cases, death.

Salicylates—A group of drugs that includes aspirin and related compounds. Salicylates are used to relieve pain, reduce inflammation, and lower fever.

Shingles—An disease caused by an infection with the *Herpes zoster* virus, the same virus that causes chickenpox. Symptoms of shingles include pain and blisters along one nerve, usually on the face, chest, stomach, or back.

Trunk—That part of the body that does not include the head, arms, and legs. Also called the torso.

Varicella zoster—The virus that causes chickenpox (varicella).

Varicella-zoster immune globulin—A substance that can reduce the severity of chickenpox symptoms.

Varivax—The brand name for varicella virus vaccine live, an immunizing agent used to prevent infection by the *Herpes (Varicella) zoster* virus. The vaccine works by causing the body to produce its own protection (antibodies) against the virus.

Virus—A small infectious agent consisting of a core of genetic material (DNA or RNA) surrounded by a shell of protein. A virus needs a living cell to reproduce.

The vaccine also is not recommended for pregnant women, and women should delay pregnancy for three months following a complete vaccination. The vaccine is useful when given early after exposure to chickenpox and, if given in the midst of the incubation period, it can be preventative. The Infectious Diseases Society of America stated in 2000 that immunization is recommended for all adults who have never had chickenpox.

While there was initial concern regarding the vaccine's safety and effectiveness when first released, the vaccination is in the early 2000s gaining acceptance as numerous states require it for admittance into daycare or public school. In 2000, 59 percent of toddlers in the United States were immunized; up from 43.2 percent in 1998. A study published in 2001 indicates that the varicella vaccine is highly effective when used in clinical practice. Although evidence has not ruled out a booster shot later in life, all research addressing the vaccine's effectiveness throughout its six-year use indicates that chickenpox may be the first human herpes virus to be wiped out. Although initial concerns questioned if the vaccination might make shingles more likely, studies are

beginning in the early 2000s to show the effectiveness of the vaccine in reducing cases of that disease.

Parental concerns

A doctor should be called immediately if any of the following occur:

- The child's fever goes above 102°F (38.9°C) or takes more than four days to disappear.
- The child's blisters appear infected. Signs of infection include leakage of pus from the blisters or excessive redness, warmth, tenderness, or swelling around the blisters.
- The child seems nervous, confused, unresponsive, or unusually sleepy; complains of a stiff neck or severe **headache**; shows signs of poor balance or has trouble walking; finds bright lights hard to look at; is having breathing problems or is coughing a lot; is complaining of chest pain; is **vomiting** repeatedly; or is having convulsions. These may be signs of Reye's syndrome or encephalitis, two rare but potentially dangerous conditions.

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Chickenpox vaccine

Definition

Chickenpox vaccine or varicella zoster vaccine (VZV) is an injection that protects children from contracting chickenpox (varicella), one of the most common childhood diseases.

Description

VZV consists of living but attenuated (weakened) varicella zoster, the virus that causes chickenpox. The weakened virus induces a child's immune system to develop antibodies against the varicella virus without causing the disease. Thus it prevents children from contracting chickenpox. Prior to the introduction of VZV, approximately 4 million Americans contracted chickenpox each year, and 95 percent of children contracted the disease before the age of 18. The vaccine first became available in the United States in 1995 and is produced by Merck & Company under the trade name Varivax.

Vaccine development

A sample of the varicella zoster virus was isolated from the blood of a three-year-old Japanese boy in 1972. A Japanese researcher, Michiaki Takahashi, attenuated the virus by growing it in various animal and human cell cultures. He then tested it on children and found that it was effective in preventing chickenpox. This "Oka" varicella strain, named after the original infected child, was licensed by Merck in 1981 and used to develop Varivax. The vaccine was clinically tested for safety and effectiveness.

Producing sufficient quantities of the vaccine to immunize all children against chickenpox has proven to be a major obstacle. Weakened viruses for vaccines are grown in cell cultures. However unlike other weakened viruses, varicella zoster remains in the cell rather than being secreted from the cell and collected from the culture medium. Thus the infected cells must be collected and broken open by ultrasound. The released virus is extremely sensitive to heat caused by ultrasound, and Merck scientists had to determine the precise conditions for opening the cells and releasing the virus unharmed. The company built a new production facility for Varivax that uses robots to strictly control the ultrasound procedure.

In addition to the live attenuated varicella virus, Varivax contains:

- bovine (cow) albumin or serum
- sodium ethylenediamine tetraacidic acid (EDTA)

- gelatin
- monosodium glutamate
- protein from the human cell line MRC-5
- neomycin, an antibiotic
- phosphate buffers
- sodium chloride
- sucrose

Effectiveness

VZV is considered to be safe and 70 to 90 percent effective. Vaccinated children who do contract chickenpox usually have milder symptoms. The vaccine also prevents chickenpox in children exposed to the virus three to five days prior to **vaccination**.

The Centers for Disease Control and Prevention (CDC), the American Academy of Pediatrics, and the American Academy of Family Physicians all recommend that healthy children be vaccinated against chickenpox. In 2001 child-care facilities and public schools began phasing in a varicella vaccination requirement for enrollment. By 2002, some 81 percent of American children had been vaccinated with VZV, and the CDC determined that the number of chickenpox cases had declined substantially. The CDC expects that widespread childhood vaccination against chickenpox will further reduce the incidence of the virus in the general population. This, in turn, will reduce the incidence of chickenpox among those who cannot receive VZV, including children who are most at risk for serious complications from the disease.

Breakthrough infections

As of 2004 it was unclear whether VZV provided life-long immunity to chickenpox. The U.S. Food and Drug Administration (FDA) required Merck to follow several thousand children for 15 years, to determine the long-term effects of the vaccine and whether additional booster shots of VZV would be necessary. It is possible that vaccinated children obtain booster immune effects through repeated contact with the virus from infected children.

Early evidence suggested that the rate of breakthrough chickenpox infections (infections in previously vaccinated children) was about 2 percent annually and that the likelihood of such infections did not increase with time after vaccination. Breakthrough infections in vaccinated children usually are very mild. They last only a few days and there are fewer than 50 lesions on the child's body and little or no **fever**. It is not clear whether breakthrough chickenpox infections are less contagious than infections in unvaccinated children.

Some physicians remain reluctant to vaccinate against an usually mild childhood disease such as chickenpox. Some also are concerned that vaccinated children may contract chickenpox as adults when it can be a much more serious disease with a 20 percent higher risk of death.

Shingles

Although children who have had chickenpox are immune to the disease and cannot contract it a second time, the varicella zoster virus can remain inactive in the human body. These dormant viruses are concentrated in nerve cells near the spinal cord and may reactivate in adults, causing the disease herpes zoster or shingles. The reactivated virus further infects nerve cells, causing severe **pain**, burning, or **itching**. Shingles usually occurs in people over the age of 50 and may be associated with a weakening immune system.

It is not known whether the weakened virus used for VZV can remain dormant in the body, eventually causing shingles in the same way that the naturally occurring varicella virus can. In 1998 the CDC found 2.6 cases of post-vaccination herpes zoster for every 100,000 distributed doses of VZV. In contrast there were 68 cases of herpes zoster in healthy children under age 20, following natural infection with varicella. However, as of 2004, it is too early to determine whether vaccinated children are more or less likely to develop shingles in adulthood as compared with adults who were naturally infected with chickenpox as children.

A 2002 study indicated that exposure to varicella is much higher in adults living with children and that such exposure substantially boosts immunity against shingles. The authors of the study predicted that mass vaccination against varicella will create an epidemic of herpes zoster, affecting as many as 50 percent of those who were between the ages of ten and 44 at the time that the vaccine was introduced.

General use

Consequences of chickenpox

Chickenpox is highly contagious and easily transmitted among children through personal contact, coughing, or sneezing. The disease is characterized by red spots on the face, chest, back, and other body parts. These spots fill with fluid, rupture, and crust over. Symptoms of chickenpox may not appear for as long as two to three weeks following infection. The virus is contagious

from one or two days before the first rash appears until the blisters have formed complete scabs and no new rash has appeared for 24 hours. This may take from five days to two weeks. Thus the varicella virus can spread very rapidly within families and among groups of children in school and daycare.

In most instances chickenpox is not a serious disease, although the itchy lesions and fever and other mild flu-like symptoms may cause a week or two of discomfort. However the disease can have serious complications. Scratching the pox can cause bacterial infection that can lead to permanent scars. In rare cases chickenpox can lead to the following:

- muscle aches
- sore throat
- ear infections
- **pneumonia**
- arthritis
- neurological symptoms, including shakiness
- **encephalitis**, an inflammation of the brain

In the United States more children die of chickenpox than of any other disease that can be prevented by a vaccine. Prior to the introduction of VZV, there were about 100 deaths and 12,000 hospitalizations annually as a result of chickenpox infections. Approximately 40 percent of the deaths and 60 percent of the hospitalizations occurred in children under age ten. Teenagers and adults, as well as children with leukemia or other cancers or with impaired immune systems, are at particular risk for severe chickenpox and its complications. Babies whose mothers contracted chickenpox during pregnancy are at risk for multiple birth defects. Babies whose mothers contract chickenpox shortly before or after giving birth are at risk of developing a severe form of the disease. As many as 5 percent of these babies die. Most high-risk children and non-immune adults contract chickenpox from unvaccinated children.

Children with chickenpox miss an average of five to six days of school and their parents miss an average of three to four days of work while caring for them. The CDC estimates that, including direct medical costs and indirect societal costs, $5.40 is saved for every $1.00 spent on childhood VZV immunization.

Vaccine administration

It is recommended that babies receive a single-dose injection of Varivax between the ages of 12 and 18 months, usually at the same time that they receive their first **measles**, **mumps**, and **rubella** (MMR) vaccine. Children and adolescents who have not already had chickenpox can be vaccinated at any time. However, adolescents aged 13 or older, as well as adults, require two doses of Varivax, four to eight weeks apart, to obtain the same level of immunity as children under 13. The reason for this is not known.

VZV usually is covered by health insurance. In the United States the Vaccines for Children program covers the cost of chickenpox vaccination for children without health insurance and for specific other groups of children, including Native Americans.

Precautions

In rare instances it is possible to contract the weakened vaccine strain of varicella from a recently vaccinated child.

Healthy children

Children on long-term steroids for any reason, including **asthma**, should consult their physician about the timing of the vaccination. Children should not receive VZV if the following applies:

- They are allergic to gelatin or the antibiotic neomycin.
- They have had a serious reaction to a previous varicella vaccination.
- They are taking aspirin or other salicylates that have the remote possibility of causing Reye's syndrome.

In addition, infants under one year and pregnant teenagers should not receive VZV. Females should not become pregnant within one month of receiving VZV.

Children at high risk for severe chickenpox or its complications, including newborns and premature infants exposed to chickenpox after birth, often are given varicella-zoster immune globulin (VZIG). VZIG is made from the blood serum of people with high antibody levels against the varicella virus. It must be administered within 96 hours of exposure to chickenpox, and it results in a passive immunity against the disease for about three months.

Additional CDC precautions for administrating VZV pertain to the following groups of children:

- those with a family history of immunodeficiency
- those who have had a blood transfusion or received other blood or serum products within the past five months

- those who have received antibody-containing products, including VZIG or other immune globulin, within the past 11 months

Children with medical conditions

Medical conditions that preclude vaccination against chickenpox include active, untreated **tuberculosis** and any other moderate or serious illness. Moreover, children with weakened immune systems should not receive a live virus vaccine such as VZV. This restriction applies to children who have the following situations:

- They have leukemia or other cancers.
- They have had **cancer** treatments, including radiation or drugs.
- They have received organ transplants or hematopoietic stem cell transplants.
- They have a weakened immune system due to HIV/AIDS.

Children with leukemia in remission or HIV-infected children with normal immune function may be eligible for VZV. However, chickenpox can cause serious complications in HIV-infected children with compromised immune systems. Therefore, the National Institute of Allergy and Infectious Diseases (NIAID) and the National Institute of Child Health and Human Development (NICHD) are as of 2004 sponsoring a clinical study of the safety and effectiveness of Varivax in HIV-infected children. In the initial phase of the study, HIV-infected children who were without symptoms tolerated Varivax well. Since shingles is very common in HIV-infected children, the NIAID and NICHD also launched a clinical study to determine whether Varivax can prevent shingles in HIV-infected children who have had chickenpox.

Side effects

Reactions to VZV are usually mild and may include:

- pain, rash, hardness, and/or swelling at the injection site in about 20 percent of children and about one in three adolescents
- small chickenpox lesions one to two weeks after vaccination
- generalized mild **rashes** or small bumps up to a month after vaccination in 1–4 percent of VZV recipients

Moderate or severe reactions to VSV have been reported very rarely. These reactions include: high fever or seizures one to six weeks after vaccination in fewer than one out of 1,000 children; pneumonia; and **anaphylaxis**, an allergic reaction that may include weakness, wheezing, breathing difficulties, **hives**, a fast heart rate, **dizziness**, or behavior changes, within a few minutes to a few hours after the injection. Other reactions, such as a low blood count or brain involvement, including encephalitis, occur so rarely that they may not be associated with VZV.

Following the distribution of the first 10 million doses of VZV, it was determined that severe reactions occurred with a frequency of approximately one in 50,000. This is far lower than the risks associated with chickenpox. There is no evidence that healthy children who have had chickenpox or who received VZV previously are at a greater for adverse effects from an additional dose of Varivax.

The National Vaccine Injury Compensation Program helps pay for medical expenses resulting from vaccine reactions. In case of a serious reaction to VZV, parents should do the following:

- A doctor should be consulted immediately.
- The date, time, and type of reaction should be recorded.
- Medical personnel or the local health department should file a Vaccine Adverse Event Report.

Interactions

VZV is not known to interact with any foods or drugs. However, **antiviral drugs** for treating herpes viruses, including acyclovir or valacyclovir, should not be administered within 24 hours of Varivax, because these drugs can reduce the effectiveness of the vaccine.

Parental concerns

Most children are afraid of injections; however, there are simple methods for easing a child's **fear**. Prior to the vaccination, parents should do the following:

- Tell children that they will be getting a shot and that it will feel like a prick; however, it will only sting for a few seconds.
- Explain to children that the shot will prevent them from becoming sick.
- Have older siblings comfort and reassure a younger child.
- Bring along the child's favorite toy or blanket.
- Never threaten children by telling them they will get a shot.
- Read the vaccination information statement (VIS) and ask the medical practitioner questions.

During the vaccination, parents should follow these steps:

- Hold the child.
- Make eye contact with the child and smile.
- Talk softly and comfort the child.
- Distract the child by pointing out pictures or objects or by using a hand puppet.
- Sing or tell the child a story.
- Have the child tell a story.
- Teach the child how to focus on something other than the shot.
- Help the child to take deep breaths.
- Allow the child to cry.
- Stay calm.

Parents may choose to use a comforting restraint method while the child is receiving the injection. These methods enable the parent to control and steady the child's arm while not holding the child down. With toddlers, the positions are as follows:

- The child is held on the parent's lap.
- The child's arm is behind the parent's back, held under the parent's arm.
- The parent's arm and hand control the child's other arm.
- The child's feet are held between the parent's thighs and steadied with the parent's other arm.

With older children, the parent and child can assume the following positions:

- The child is held on the parent's lap or stands in front of the seated parent.
- The parent's arms embrace the child.
- The child's legs are between the parent's legs.

Following the vaccination, parents should do the following:

- Hold and caress or breastfeed the child.
- Talk soothingly and reassuringly.
- Hug and praise the child for doing well.
- Review the VIS for possible side effects.
- Use a cool, wet cloth to reduce soreness or swelling at the injection site.
- Check the child for rashes over the next few days.

KEY TERMS

Antibody—A special protein made by the body's immune system as a defense against foreign material (bacteria, viruses, etc.) that enters the body. It is uniquely designed to attack and neutralize the specific antigen that triggered the immune response.

Booster shot—An additional dose of a vaccine to maintain immunity to the disease.

Breakthrough infection—A disease that is contracted despite a successful vaccination against it.

Herpes zoster virus—Acute inflammatory virus that attacks the nerve cells on the root of each spinal nerve with skin eruptions along a sensory nerve ending. It causes chickenpox and shingles. Also called varicella zoster virus.

Immunity—Ability to resist the effects of agents, such as bacteria and viruses, that cause disease.

Varicella zoster—The virus that causes chickenpox (varicella).

Varicella-zoster immune globulin—A substance that can reduce the severity of chickenpox symptoms.

In addition, parents should anticipate that their children may eat less during the first 24 hours after the injection, and they should receive plenty of fluids. The medical practitioner may suggest a non-aspirin-containing pain reliever.

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Child abandonment *see* **Abandonment**

Child abuse

Definition

Child abuse is the blanket term for four types of child mistreatment: physical abuse, sexual abuse, emotional abuse, and neglect.

Description

Prevalence of abuse

Child abuse was once viewed as a minor social problem affecting only a handful of U.S. children. However, in the late 1990s and early 2000s it has received close attention from the media, law enforcement, and the helping professions, and with increased public and professional awareness has come a sharp rise in the number of reported cases. Because abuse is often hidden from view and its victims too young or fearful to speak out, however, experts suggest that its true prevalence is possibly much greater than the official data indicate. An estimated 896,000 children across the country were victims of abuse or neglect in 2002, according to national data released by the U.S. Department of Health and Human Services (HHS) in April 2004. Parents were the abusers in 77 percent of the confirmed cases, other relatives in 11 percent. Sexual abuse was more likely to be committed by males, whereas females were responsible for the majority of neglect cases. The data show that child protective service agencies received about 2,600,000 reports of possible maltreatment in 2002. About 1,400 children died of abuse or neglect, a rate of 1.98 children per 100,000 children in the population. In many cases children are the victims of more than one type of abuse. The abusers can be parents or other **family** members, caretakers such as teachers and **babysitters**, acquaintances (including other children), and (in rare instances) strangers.

Although experts are quick to point out that abuse occurs among all social, ethnic, and income groups, reported cases usually involve poor families with little education. Young mothers, **single-parent families**, and parental alcohol or drug abuse are also common in reported cases. Statistics show that more than 90 percent of abusing parents have neither psychotic nor criminal personalities. Rather they tend to be lonely, unhappy, angry, young, and single parents who do not plan their pregnancies, have little or no knowledge of child development, and have unrealistic expectations for child behavior. From 10 percent to perhaps as many as 40 percent of abusive parents were themselves physically abused as children, but most abused children do not grow up to be abusive parents.

Types of abuse

PHYSICAL ABUSE Physical abuse is the non-accidental infliction of physical injury to a child. The abuser is usually a family member or other caretaker and is more likely to be male. One fourth of the confirmed cases of child abuse in the United States involve physical abuse. A rare form of physical abuse is **Munchausen syndrome** by proxy, in which a caretaker (most often the mother) seeks attention by making the child sick or appear to be sick.

EMOTIONAL ABUSE Emotional abuse is the rejecting, ignoring, criticizing, isolating, or terrorizing of children, all of which have the effect of eroding their **self-esteem**. Emotional abuse usually expresses itself in verbal attacks involving rejection, scapegoating, belittlement, and so forth. Because it often accompanies other types of abuse and is difficult to prove, it is rarely reported and accounts for only about 6 percent of the confirmed cases.

SEXUAL ABUSE Psychologists define child sexual abuse as any activity with a child, before the age of legal consent, that is for the sexual gratification of an adult or a significantly older child. It includes, among other things, sexual touching and penetration, persuading a child to expose his or her sexual organs, and allowing a child to view pornography. In most cases the child is related to or knows the abuser, and about one in five abusers are themselves underage. Sexual abuse accounts for 12 to 15 percent of confirmed abuse cases. In multiple surveys, 20 to 25 percent of females and 10 to 15 percent of males report that they were sexually abused by age 18.

NEGLECT Neglect, the failure to satisfy a child's basic needs, can assume many forms. Physical neglect is

the failure (beyond the constraints imposed by poverty) to provide adequate food, clothing, shelter, or supervision. Emotional neglect is the failure to satisfy a child's normal emotional needs, or behavior that damages a child's normal emotional and psychological development (such as permitting drug abuse in the home). Failing to see that a child receives proper schooling or medical care is also considered neglect. Slightly more than half of all reported abuse cases involve neglect.

Infancy and toddlerhood

Infants who are premature, mentally retarded, or have physical handicaps are more likely to provoke abuse from their caregiver than are infants without such problems. Similarly, nonhandicapped infants who are nonrhythmic (that is, have uneven **sleep** and eating patterns) are more likely to be abused. It appears that the child's tendency to learn slowly, to be less coordinated, or less affectionate—rather than any physical problem—that promotes abuse. Infants, because of their fragility, are more susceptible to injury from physical **discipline** than older children. Infants are especially susceptible to **head injury** from shaking or being thrown. A baby can be fatally injured by being thrown even onto a soft mattress. The baby's brain hits the back of the skull if the child is thrown with even mild force and intracranial bleeding can result.

Shaken baby syndrome (SBS) is the leading cause of death in child abuse cases in the United States. The syndrome results from injuries caused by someone vigorously shaking an infant, usually for five to 20 seconds, which causes brain damage. In some cases, the shaking is accompanied by a final impact to the baby's head against a bed, chair, or other surface. Although SBS is occasionally seen in children up to four years of age, the vast majority of incidents occur in infants who are younger than one year; the average age of victims is between three and eight months. Approximately 60 percent of shaken babies are male, and children of families who live at or below the poverty level are at an increased risk for SBS (and any other type of child abuse).

Preschool

Typically, abused children show developmental delays by **preschool** age. It is unclear whether these delays occur due to cumulative neurological damage or due to inadequate stimulation and uncertainty in the child about the learning environment and the absence of positive parental interactions that would stimulate language and motor processes. These delays, in concert with their parents' higher-than-normal expectations for their children's self-care and self-control abilities, may provoke additional abuse. Abused preschoolers respond to peers and other adults with more aggression and anger than do non-abused children. A coercive cycle frequently develops in which parents and children mutually control one another with threats of negative behavior.

School age

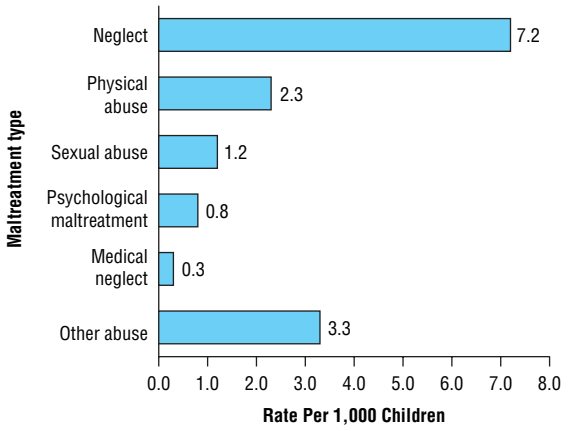
School-aged children who are abused typically have problems academically and have poorer grades and performance on standardized achievement tests. Studies of abused children's intellectual performance find lower scores in both verbal and math and visual-spatial areas. Abused children also tend to be distracted and overactive, making school a very difficult environment for them. With their peers, abused children are often more aggressive and more likely to be socially rejected than nonabused children. Less mature socially, abused children show difficulty in developing trusting relationships with others. The anger that is often instilled in such children is likely to be incorporated into their personality structures. Carrying an extra load of anger makes it difficult for them to control their behavior and increases their risk for resorting to violent action. To control their fears, children who live with violence may repress feelings. This defensive maneuver takes its toll in their immediate lives and can lead to further pathological development. It can interfere with their ability to relate to others in meaningful ways and to feel empathy. Individuals who cannot empathize with others' feelings are less likely to curb their own aggression and more likely to become insensitive to brutality in general.

As adolescents, abused children are more likely to be in contact with the juvenile justice system than nonabused children of comparable family constellation and income level. Many of these children are labeled "ungovernable" for committing offenses such as **running away** and **truancy**. A higher proportion of abused than nonabused delinquent youth are also involved in crimes of assault. Follow-up studies on abused children in later **adolescence** show that in addition to having problems with the law, they are also more likely to be substance abusers or to have emotional disturbances such as depression.

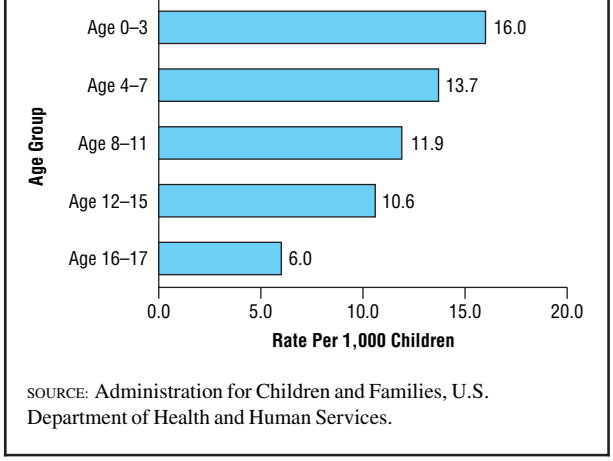
Common problems

Physical abuse

The usual physical abuse scenario involves a parent who loses control and lashes out at a child. The trigger may be normal child behavior such as crying or dirtying a diaper. Unlike nonabusive parents, who may become angry at or upset with their children from time to time



This graph of 2002 data on child abuse in the United States shows that neglect is by far the most common type of abuse. *(Graph by GGS Information Services.)*



This graph of 2002 data on child abuse in the United States shows that younger children are more likely to be vicims of abuse than older children. *(Graph by GGS Information Services.)*

but are genuinely loving, abusive parents tend to harbor deep-rooted negative feelings toward their children. Unexplained or suspicious **bruises** or other marks on the skin are typical signs of physical abuse, as are **burns**. Skull and other bone **fractures** are often seen in young abused children, and in fact, head injuries are the leading cause of death from abuse. Children less than one year old are particularly vulnerable to injury from shaken baby syndrome. Physical abuse also causes a wide variety of behavioral changes in children.

Emotional abuse

Emotional abuse can happen in many settings: at home, at school, on **sports** teams, and so on. Some of the possible symptoms include loss of self-esteem, sleep disturbances, headaches or stomach aches, school avoidance, and running away from home.

Sexual abuse

The two prerequisites for this form of maltreatment are sexual arousal towards children and the willingness to act on this arousal. Factors that may contribute to this willingness include alcohol or drug abuse, poor impulse control, and a belief that the sexual behaviors are acceptable and not harmful to the child. The chances of abuse are higher if the child is developmentally handicapped or vulnerable in some other way. Genital or anal injuries or abnormalities (including the presence of **sexually transmitted diseases**) can be signs of sexual abuse, but often there is no physical evidence for a doctor to find. In fact, physical examinations of children in cases of suspected sexual abuse supply grounds for further suspicion only 15 to 20 percent of the time. **Anxiety**, poor academic performance, and suicidal conduct are some of the behavioral signs of sexual abuse but are also found in children suffering other kinds of stress. Excessive **masturbation** and other unusually sexualized kinds of behavior are more closely associated with sexual abuse itself.

Neglect

Many cases of neglect occur because the parent experiences strong negative feelings toward the child. At other times, the parent may truly care about the child but lacks the ability or strength to adequately provide for the child's needs because handicapped by depression, drug abuse, **mental retardation**, or some other problem. Neglected children often do not receive adequate nourishment or emotional and mental stimulation. As a result, their physical, social, emotional, and mental development is hindered. They may, for instance, be underweight, develop language skills less quickly than other children, and seem emotionally needy.

Parental concerns

When children reach age three, parents should begin teaching them about "bad touches" and about confiding in a suitable adult if they are touched or treated in a way that makes them uneasy. Parents also need to exercise

caution in hiring babysitters and other caretakers. Anyone who suspects abuse should immediately report those suspicions to the police or their local child protection services agency, which is usually listed in the blue pages of the telephone book under Rehabilitative Services or Child and Family Services, or in the yellow pages. Round-the-clock crisis counseling for children and adults is offered by the Childhelp USA/IOF Foresters National Child Abuse Hotline. The National Committee to Prevent Child Abuse is an excellent source of information on the many support groups and other organizations that help abused and at-risk children and their families. One of these organizations, National Parents Anonymous, sponsors 2,100 local self-help groups throughout the United States, Canada, and Europe. Telephone numbers for its local groups are listed in the white pages of the telephone book under Parents Anonymous or can be obtained by calling the national headquarters.

When to call the doctor

Physical signs of abuse may include bruises, especially those in different stages of healing, bruises in the shape of an object, such as fingers, a ring, or a belt buckle; unexplained burns, black eyes, or broken bones; vaginal or rectal bleeding, **pain**, **itching**, swelling or discharge; a vacant stare or dazed appearance; frequent attempts to run away; and sexual promiscuity.

Behavioral signs of child abuse include: low self esteem; flinching or ducking from motion or people moving towards them; eating disorders or loss of appetite; self mutilation such as "cutting," biting oneself or pulling out hair; unusual habits like rocking, sucking cloth; extreme changes in behavioral patterns; poor interpersonal relationships or a lack of self-confidence; clinginess, withdrawal or aggressiveness; regressing to infantile behavior such as bedwetting, **thumb sucking** or excessive crying; recurrent **nightmares**, disturbed sleep patterns, or a sudden **fear** of the dark; unexplained fear of a particular person; unusual knowledge of sexual matters; acting much younger or older than chronological age; frequent **lying**, or a fall in grades at school; and depression.

It is important to remember that some of these symptoms of child abuse can be normal manifestations of **play** and activity. Other symptoms could be the result of a traumatic event that is not necessarily abuse, such as **divorce**. Still, others are definitely "red flag" symptoms of abuse. If any physical signs of abuse appear, get medical help immediately. Talk frankly with the doctor and share any concerns about possible abuse. If there is physical proof of abuse, get a doctor's report in writing. Any behavioral signs of abuse are cause for concern to a good parent, teacher, or caregiver. A good first move is to open and nurture trusting lines of communication. The parent should increase the time spent with the child and increase the attention given to the child. The parent should show more interest in the child's life and ask more questions. The parent needs to assure the child of the parent's unqualified love and support, and make sure the children know that the parent wants them to feel happy and confident. Children need to know that no matter what has happened, their parents will always love them.

KEY TERMS

Munchausen syndrome by proxy—A form of abuse in which a parent induces symptoms of disease in a child.

Nonrhythmic—Having uneven sleep and eating patterns.

Shaken baby syndrome—Injuries caused by someone vigorously shaking an infant, usually for five to twenty seconds, which causes brain damage.

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Child care *see* **Day care**

Child custody laws

Definition

Child custody laws are federal and state laws that govern a parent's legal authority to make decisions affecting a child (legal custody) and to maintain physical control over the child (physical custody). Child custody laws also pertain to the visitation rights of the non-custodial parent.

Purpose

Child custody laws exist to provide a legal structure for relationships between children and their divorced parents. Ideally, divorced parents should work together to have an amicable relationship and shared custody, but bitterness between divorced spouses and tendencies to involve children in marital and **divorce** disputes require child custody laws. Child custody laws help to define the **family** situation in terms of the best interests of the child or children involved in the divorce. Child custody laws can also be applied in cases when unmarried parents claim custody based on a biological relationship, when grandparents dispute the competence of the child's parents, and when same-sex couples with adopted children separate. In some cases, custody may be granted to an individual or individuals not related (e.g., foster parents).

Description

In the United States, responsibilities for a child's care and decision-making related to that care are governed by federal and state laws. In general, custody laws and custody decisions favor continued and frequent contact between the child and both parents, as well as an ongoing role for both parents in the raising of their children. However, custody decisions are strongly influenced by the circumstances of each individual case, the welfare of the involved child or children, and the perceived effect of each parent on the child.

In almost all custody cases, courts consider a value called the "best interests of the child" as the highest priority when rendering a custody decision. The best interests of a child are determined by considering a number of factors, including the following:

- child's age, sex, and mental/physical health
- mental and physical health of both parents
- child's established lifestyle (home, school, church, etc.)
- lifestyle of both parents, including any history of child abuse
- emotional bonds between each parent and child, and the ability of each parent to provide emotional support and guidance
- ability of each parent to provide physical necessities (e.g., food, home, clothing, healthcare)
- impact of change on the child
- ability and willingness of each parent to encourage a healthy relationship and communication between the child and the other parent
- child's preferences

Most courts use the above factors to determine which parent can provide the child with a stable home environment and continuity of lifestyle.

Child custody laws address several different types of parenting situations and custody circumstances. For the purposes of custody, legal definitions of parenthood are as follows:

- Biological parents: The mother and father responsible for conception and birth of the child.

- Stepparent: A non-biological parent who marries or cohabitates with a biological parent.
- Stepchild: A non-biological child brought into the family by marriage or cohabitation with the biological parent.
- Custodial parent: The parent awarded primary custody by a court during divorce proceedings.
- Non-custodial parent: The parent awarded part-time custody or visitation rights by a court during divorce proceedings.

Custody decisions involve physical and legal custody. Physical custody refers to the responsibility of taking care of the children (food, clothing, housing, etc.). Legal custody refers to the responsibility for decisions that affect the child's interests (medical, educational, and religious decisions, etc.). In 20 states, custody is divided into physical custody and legal custody; in the remaining states, physical and legal custody are not considered separately, and the term "custody" refers to both responsibilities. In states that do not distinguish between physical and legal custody, the term "custody" implies both types of responsibilities. Custody decisions by a court of law designate joint custody between two parents or primary custody for one parent (the custodial parent) and visitation rights for the non-custodial parent. Custody decisions are described as follows:

- Joint physical custody: Children split their time between parents, spending a substantial amount of time with each parent.
- Joint legal custody: Parents share in decision-making regarding medical, educational, and religious issues involving the children.
- Joint legal and physical custody: Parents share both time and decision-making responsibilities.
- Primary (sole) custody: One parent is designated the primary physical and legal custodian of the child or children, and the other parent is granted visitation rights.

Courts in every state are willing to order joint legal custody; however, about half the states are reluctant to order joint physical custody unless both parents agree to it, the child's lifestyle is not substantially disrupted (e.g., parents live within the same school district), and parents appear to be able to effectively and amicably cooperate with each other regarding their children. Two states (New Mexico and New Hampshire) require joint custody to be awarded, except when the children's best interests or a parent's health or **safety** are compromised.

Primary or sole custody is usually awarded when parents live a significant distance from one another, when one parent can provide clear benefits for the child over the other parent, or when one parent is deemed unfit to care for the child. In some cases, neither parent is judged fit to retain custody, usually due to substance abuse problems, mental health issues, or prolonged absence or incarceration. In such cases, an individual or individuals other than the parents are granted custody or given a temporary guardianship or **foster care** arrangement by a court. In general, courts would prefer that a child remain with family members than be placed in foster care.

Common problems

Unfortunately, children are often involved in divorce and custody battles and, as a result, may suffer from psychological and emotional damage that will require counseling and therapy. In some cases, a child has the legal right to choose which parent he/she wants to live with. However, placing the responsibility of making such a decision on the child can cause internal conflict and emotional stress related to feeling that they have to choose one parent over the other. Parents should make every effort to keep bitter feelings between themselves and not involve children in their divorce conflict. Having the child attend regular therapy or counseling sessions can help the child's adjustment to the divorce and changes in the living situation. Group therapy with other children in similar circumstances can be especially helpful.

Although child custody laws were established to protect the best interests of the child, final custody decisions are not always best for the child. In some cases, the parent with the best legal representation, not necessarily the parent who will provide the best care, wins custody. Parents may misrepresent their ability to properly care for children or provide false information about the other parent in order to win custody. An independent custody evaluator, usually appointed by the court, can help by conducting psychological evaluations of both parents and children to determine the custody arrangement that will be in the best interests of the children.

Parental concerns

In the early 2000s one parent often lives out of state for a variety of reasons, including employment, extended family relationships, and standard of living. All states and the District of Columbia follow the Uniform Child Custody Jurisdiction Act, which sets standards for when a court can determine custody and when a court must defer to an existing determination from another state. In general, a state court can decide custody about a child if

the state is the child's home state; the child has significant connections with individuals (grandparents, doctors, teachers) in the state concerning the child's care, protection, and personal relationships; the child is in the state and has been either abandoned or is in danger of being abused or neglected if sent back to the other state; or no other state can meet one of the above three tests; or a state can meet at least one of the tests but has declined to make a custody decision. Parents who wrongfully remove or retain a child in order to create a home state or significant connections will usually be denied custody. In cases where more than one state meets the above standards, the law requires that only one state award custody. The Uniform Child Custody Jurisdiction Act has helped establish consistency in the treatment of custody decisions and helped solve problems created by kidnapping or disagreements over custody between parents living in different states.

Divorce and custody battles can create stress and worries for parents. In making custody decisions, courts look for responsible parents who are actively involved in the children's lives. Liberal, unrestricted visitation for the non-custodial parent is often based on the relationship with children and the degree of involvement in the children's everyday activities.

During the twentieth century, custody laws often mandated that custody be automatically given to mothers, especially for younger children. In the early 2000s, in most states, this ruling has been rejected or revised, and no state as of 2004 required that a child be awarded to the mother without regard for the fitness of both parents as primary caregivers. Fathers who desire physical custody should not assume that gender stereotypes will result in a mother automatically being given custody. As of 2004, when both mother and father often work full-time, parents enter a custody case as equals with regard to physical custody. Parents who present themselves as flexible and willing to assume physical custody are more likely to be granted custody.

Parents in same-sex relationships may have concerns regarding custody issues due to their sexual orientation. However, in a few states, including Alaska, California, New Jersey, New Mexico, Pennsylvania, and the District of Columbia, a parent's sexual orientation cannot in and of itself prevent a parent from being given custody or visitation rights. However, gay and lesbian parents may still be denied custody or visitation because many judges may be motivated by personal or community prejudices. Stepparents may face a similar situation, since stepparents, unless they legally adopt a stepchild, have no legal rights with regard to custody or visitation. In cases in which a stepparent may provide a more stable environment for a child than the biological parents, judges may still favor biological parents due to personal and societal beliefs about what constitutes a "normal" family.

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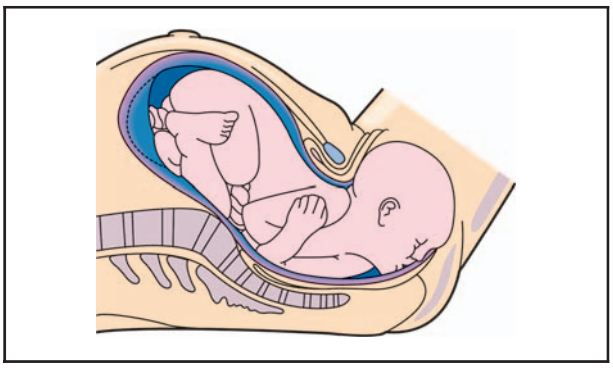
Childbirth

Definition

Childbirth is formally divided by the medical field into three stages. The first stage is labor, which has three phases: early, active, and transitional. The first stage ends with complete dilatation (opening) of the cervix. The second stage is delivery, which involves pushing and the actual birth of the baby. The third stage is delivery of the placenta or afterbirth.

Description

A full-term pregnancy is considered to be 280 days, nine calendar months or ten lunar months calculated from the first day of the last menstrual period. This is a fairly arbitrary number that may, in fact, vary with genetic differences and depends on a normal menstrual cycle, which varies considerably from woman to woman. The average actual length from conception to birth is estimated as 267 days. Childbirth is a natural process,



Childbirth in stage 2. The baby's head is crowning and about to emerge from the vagina. *(Illustration by Hans & Cassidy.)*

and it, too, varies among women. Despite what the obstetrical texts say about what to expect, there are many variations that make each woman's experience hers alone. The whole process averages about 14 hours for first-time mothers and about eight hours for mothers in their subsequent pregnancies.

Labor can be described in terms of a series of stages.

First stage of labor

During the first stage of labor, the cervix dilates (opens) from 0 to 10 centimeters (cm). This stage has an early, or latent, phase, an active phase, and a transitional phase. The latent phase usually lasts the longest and is the least intense phase of labor. This phase is characterized by dilatation (opening) of the cervix to 3–4 cm along with the thinning out of the cervix (effacement). It can take place over a period of days without being noticed or over a period of two to six hours with distinctive contractions. Most women are relatively comfortable during the latent phase, and walking around is encouraged, since it naturally stimulates the process.

With the initiation of labor, the muscular wall of the uterus begins to contract causing the cervix to open (dilatation) and thin out (efface). For a first-time mother the cervix must completely efface before dilatation continues. Effacement is reported in percentages as 50 percent or 100 percent, which is completely thinned out. The amniotic sac may or may not break during labor, and the birth attendant may rupture the bag with an amniohook, which looks a little like a large crochet hook. There is no **pain** involved with the breaking the bag of waters, although the contractions may intensify. During a contraction, the infant experiences pressure that pushes it against the cervix to assist with the dilatation. During this first phase, a woman's contractions typically increase in frequency and duration. Periodic vaginal exams are performed by the physician or nurse to determine progress. As pain and discomfort increase, however, the woman may be tempted to request pain medication. The administration of pain medication or anesthetics should be delayed until the active phase of labor begins, at which point the medication will not act to slow down or stop the labor.

The active phase of labor is usually shorter than the first, lasting an average of two to four hours. The contractions are more intense and accomplishing more in less time. They may be three to four minutes apart lasting 40–60 seconds even though the pattern may not be regular. During the active phase, dilatation continues to 7 cm. Relaxing between contractions is essential for coping because these contractions are more intense. Breathing exercises learned in childbirth classes can help the woman cope with the discomfort experienced during this phase. Pain medication offered at this point consists of either a short-term medication, such as Nubain or Stadol, or long-term such as epidural anesthesia.

The transitional phase continues dilation 7–10 cm. It is the most exhausting and demanding phase of labor. The contractions become very strong, are two to three minutes apart, and last 60–90 seconds. It may feel as if the contractions never stop, and there is no time to relax between them. Dilatation of the final 3 cm to 10 cm takes, on average, 15 minutes to an hour. Strong rectal pressure, with or without an urge to push or move the bowels, may cause the woman to grunt involuntarily. If it is a natural labor and delivery, the laboring woman at this phase becomes very inwardly focused and can lose control. It is important to breathe with her through contractions as this keeps her attention on what she needs to do.

Second stage of labor

Up to this point, the woman may feels as if her participation is small, because all she has done is breathe. Active involvement can now begin along with some emotional relief that it is almost over. Without anesthesia, there is often an overwhelming urge to push, and the mother gets a second wind. The baby's head is through the cervix and on its way down the birth canal. The uterine contractions get stronger, and the infant passes along the vagina helped by contractions of the uterus and the mother's pushing. If an epidural anesthetic is being used, many practitioners recommend decreasing the dosage so the mother has better control of her pushing. Research has shown, however, that the contractions will continue to push the baby down the birth canal without mother's help. If a woman is numb from an epidural, she cannot push effectively, and it is usually better to let the contractions work alone. This is called "laboring down."

When the top of the baby's head appears at the opening of the vagina, the birth is nearing completion. First

A newborn baby sits crying on the mother's stomach. *(© Jules Perrier/Corbis.)*

the head passes under the pubic bone. It fills the lower vagina and stretches the perineum (the tissues between the vagina and the rectum). This position is called "crowning," since only the crown of the head is visible. When the entire head is out, the shoulders follow. The attending practitioner suctions the baby's mouth and nose to ease the baby's first breath. The rest of the baby usually slips out easily, and the umbilical cord is cut.

Episiotomy

Many practitioners argue that it is better to cut the perineum than to let it tear. This cut is called an episiotomy. In reality, it is more difficult to repair a straight cut than a small tear in much the same way it is harder to put together a puzzle with straight edges; it is more difficult to match evenly and can result in vaginal discomfort once healed. Instead, the perineum can be massaged and gently stretched to prevent tearing as the baby's head crowns. There is also less pain associated with a tear than an episiotomy. If the woman has not had an epidural or pudendal block, she will get a local anesthetic to numb the area for repair.

Third stage

In the final stage of labor, the placenta is expelled by the continuing uterine contractions. The placenta is pancake shaped and about 10 cm (25 cm) in diameter. During pregnancy, it is attached to the wall of the uterus and served to exchange needed nourishment from the mother to the fetus and simultaneously to remove waste products from the fetus. Generally, there is a rise in the uterus due to a contraction and a gush of blood as the placenta is expelled. The placenta should be examined to make sure it is intact. Retained placenta can cause severe uterine bleeding after delivery, and it must be removed.

Breech presentation

Approximately 4 percent of babies present in the breech position when labor begins. In this presentation, the baby's bottom is the presenting part instead of the head, which is called a vertex presentation. Using a technique called a version, an obstetrician may attempt to turn the baby to a head down position. This is only successful approximately half the time, and there are possible complications with the procedure, such as umbilical cord entanglement and separation of the placenta. However, some practitioners are very successful with versions, and it does make a vaginal delivery safer.

The risks of vaginal delivery with breech presentation are much higher than with a head-first (vertex) presentation. The mother and attending practitioner need to weigh the risks to make a decision on whether to deliver via a **cesarean section** or attempt a vaginal birth. The degree of risk depends to a great extent on which one of the three types of breech presentations it is. In a frank breech the baby's legs are folded up against its body. This is the most common breech presentation and the safest for vaginal delivery. The others include complete breech, in which the baby's legs are crossed under and in front of the body, and footling breech, in which one leg or both legs are positioned to enter the birth canal. Neither of these is considered safe enough for a vaginal delivery.

Even with a complete breech, there are other factors to consider for a vaginal birth. An ultrasound examination should be done to determine that the baby's head is not too large and that it is tilted forward (flexed) rather than back (hyperextended). Fetal monitoring and close observation of the progress of labor are also important. A slowing of labor or any indication of difficulty in the body passing through the pelvis should be an indication that it is safer to consider a cesarean section.

Forceps delivery

Although not used as much in the early 2000s as in earlier times, forceps can be used if the baby's head is very low in the birth canal. Also, if there is some sudden change in the maternal-fetal status, the doctor may opt for a forceps delivery if it would be faster than a cesarean section. Forceps are spoon-shaped devices that can be placed around the baby's head while the doctor gently pulls the baby out of the vagina.

Before placing the forceps around the baby's head, pain medication or anesthesia may be given to the mother. The doctor may use a catheter to empty the mother's bladder and may clean the perineal area with soapy water. Often an episiotomy is done before a forceps birth, although tears can still occur. The use of forceps can cause vaginal lacerations in the mother.

Half of the forceps are slid into the vagina and around the side of the baby's head to gently grasp the head. When both forceps are in place, the doctor pulls on them to help the baby through the birth canal during a uterine contraction. The frequency of forceps delivery varies from one hospital to the next, depending on the experience of staff and the types of anesthesia offered at the hospital. Some obstetricians accept the need for a forceps delivery as a way to avoid cesarean birth while other obstetrical services do not use forceps at all. Complications from forceps deliveries can occur, such as nerve damage or temporary **bruises** to the baby's face. When used by an experienced physician, forceps can save the life of a baby in distress.

Vacuum-assisted birth

This method of delivering a baby was developed as a gentler alternative to forceps. Similar to forceps deliveries, vacuum-assisted births can only be used with a fully dilated cervix and a well-descended head. In this procedure, a device called a vacuum extractor is used by placing a large rubber or plastic cup against the baby's head. A pump then creates suction that gently pulls on the cup to ease the baby out the birth canal. The force of the suction may cause a bruise or swelling on the baby's head, but it resolves in a day or two.

The vacuum extractor is less likely to injure the mother than forceps, and it allows more space for the baby to pass through the pelvis. There can be problems in maintaining the suction during the vacuum-assisted birth, however, so forceps might be a better choice if the delivery needs to be expedited.

Cesarean sections

A cesarean section, also called a c-section, is a surgical procedure in which an incision is made through a woman's abdomen and uterus to deliver her baby. This procedure is performed whenever abnormal conditions complicate labor and vaginal delivery that threaten the life or health of the mother or the baby. The procedure is performed in the United States on nearly one in every four women resulting in more than 900,000 babies each year being delivered by c-section. The procedure is often used in women who have had a previous c-section, but if the incision on the uterus is not vertical, the woman can try a vaginal birth after cesarean (VBAC).

Dysfunctional labor is commonly caused by one of the three following conditions: maternal structural abnormalities; abnormal fetal presentations; failure to

progress. Non-reassuring fetal heart rate tracings represent a condition in which the fetus may not be tolerating labor and oxygen deprivation can occur. Other conditions which might indicate a need for c-section include: vaginal herpes, **hypertension** (high blood pressure), and uncontrolled diabetes in the mother.

Causes and symptoms

Childbirth usually begins spontaneously, but it may be started by artificial means if the pregnancy continues past 41 weeks gestation. There are three signs that labor may be starting: rhythmical contractions of the uterus; leaking of the bag of waters (amniotic sac); and bloody show. The importance of the sign of contractions is in the rhythm and not the contractions. True labor contractions may start once every ten or 15 minutes or even at longer intervals, but gradually the interval decreases until they come every three to four minutes. The most important thing a woman can do at this phase is to remain relaxed. The bag of waters may leak slowly or may suddenly burst, and there is a gush of fluid. There is no pain when the water breaks, although it may be startling. If contractions are not ongoing prior to this, they are likely to start soon after. If they do not, it may be necessary to stimulate labor as the womb is now open to possible infection. The bloody show is a slight discharge of blood and mucus. It usually occurs after the cervix has started to dilate slightly and the mucus plug that keeps the cervix sealed from potential pathogens becomes dislodged.

Diagnosis

The diagnosis of true labor can only be determined by a vaginal exam to determine if the cervix has changed in dilatation (opening). True labor is determined by whether the contractions are, in fact, changing the cervix. If a woman is experiencing contractions and makes no cervical change, then this is false labor. Dilatation is measured in centimeters and it goes from zero to ten centimeters, which is complete dilatation. Although the woman having the contractions may feel like she is really experiencing labor, true labor is determined by cervical change. Many women may experience Braxton-Hicks contractions (practice contractions) in preparation for true labor, and these can become uncomfortable at times, which prevents the woman from resting. A warm bath or warm drink may help her to relax and **sleep**. Inevitably she will wake up in true labor with effective contractions. Palpating contractions as they occur can assist in determining whether they are strong. A very strong contraction cannot be indented and will feel as hard as the forehead. A moderate contraction will palpate like the feel of the chin and an easy contraction feels like the end of the nose. If the contractions can be indented, they probably do not constitute true labor.

Electronic fetal monitoring

Electronic fetal monitoring (EFM) involves the use of an electronic fetal heart rate (FHR) monitor to record the baby's heart rate. The FHR is picked up by means of an ultrasound transducer and the movement of the heart valves. Elastic belts are used to hold sensors against the pregnant woman's abdomen. The sensors are connected to the monitor and detect the baby's heart rate as well as the uterine contractions. The monitor then records the FHR and the contractions as a pattern on a strip of paper, called a tracing. Electronic fetal monitoring is frequently used during labor to assess fetal well-being. EFM can be used either externally or internally. Internal monitoring does not use ultrasound, is more accurate than electronic monitoring, and provides continuous monitoring for the high-risk mother. An internal monitor requires that the bag of waters be broken and that the woman is at least two to three centimeters dilated. It is used in high-risk situations or when it is difficult to obtain an accurate FHR tracing.

Telemetry monitoring has been available since the early 1990s but is not used in many hospitals as of 2004. Telemetry uses radio waves transmitted from an instrument on the mother's thigh, which allows the mother to remain mobile. It provides continuous monitoring and does not require the patient to be in bed continuously.

Besides EFM and telemetry, which is usually continuous, there is intermittent monitoring using a handheld Doppler to assess the FHR. This method gives the mother freedom of movement during labor. Prior to electronic gadgetry a special stethoscope was used, called a fetoscope, which is rarely seen as of 2004 because it requires more skill to use. Research on the use of intermittent monitoring and continuous monitoring found no difference in fetal outcomes with intermittent monitoring. The use of continuous monitoring does result in a higher c-section rate partly because the tracing can be misinterpreted or because the mother usually requires more interventions when she cannot be mobile.

Treatment

Many women choose some type of pain relief during childbirth, ranging from relaxation and imagery to drugs. The specific choice may depend on what is available, the woman's preferences, her doctor's recommendations, and how the labor is proceeding. All drugs have some risks and some advantages.

Regional anesthetics

Regional anesthetics include epidurals and spinals. With this procedure, medication is injected into the space surrounding the spinal nerves. Depending on the type of medications used, this type of anesthesia can block nerve signals, causing temporary pain relief or a loss of sensation from the waist down. An epidural or spinal block can provide complete pain relief during cesarean birth.

An epidural is placed with the woman lying on her side or sitting up in bed with the back rounded to allow more space between the vertebrae. Her back is scrubbed with antiseptic, and a local anesthetic is injected in the skin to numb the site. The needle is inserted between two vertebrae and through the tough tissue in front of the spinal column. A catheter is put in place that allows continuous doses of anesthetic to be given.

This type of anesthesia provides complete pain relief and can help conserve a woman's energy, since she can relax or even sleep during labor. This type of anesthesia does require an IV and fetal monitor. It may be harder for a woman to bear down when it comes time to push, although the amount of anesthesia can be adjusted as this stage nears.

Spinal anesthesia operates on the same principle as epidural anesthesia and is used primarily in cases of c-section delivery. It is administered in the same way as an epidural, but the catheter is not left in place following the surgery. The amount of anesthetic injected is large, since it must be injected at one time. Spinals provide quick and strong anesthesia and allow for major abdominal surgery with almost no pain.

Narcotics

Short-acting narcotics can ease pain and not interfere with a woman's ability to push. However, they can cause sedation, **dizziness**, **nausea**, and **vomiting**. Narcotics cross the placenta and can affect the baby.

Natural childbirth and preparation for childbirth

There are several methods available to prepare for childbirth. The one selected often depends on what is available through the healthcare provider. Overall, **family** involvement is receiving increased attention by the healthcare systems, and the majority of hospitals now offer birthing rooms and maternity centers to accommodate the entire family.

Lamaze, or Lamaze-Pavlov, is the most commonly used method in the United States as of 2004. It became the first popular natural childbirth method in the 1960s. Various breathing techniques, cleansing breath, panting and blowing, are used for different phases together with the use of a focal point to enable the laboring woman to maintain control. A partner helps by coaching the mother throughout the birthing process.

The Read method, named for Dr. Grantly Dick-Read (who published his book *Childbirth Without Fear* in 1944) involves primarily remaining relaxed and breathing normally. Dr. Dick-Read promoted this method in the 1930s to help mothers deal with apprehension and tension associated with childbirth. He emphasized the practice of tensing and relaxing muscles so that complete relaxation occurs between contractions in labor. This action also serves to promote good oxygenation to the muscles.

KEY TERMS

Amniotic sac—The membranous sac that contains the fetus and the amniotic fluid during pregnancy.

Breech birth—Birth of a baby bottom-first, instead of the usual head-first delivery. This can add to labor and delivery problems because the baby's bottom doesn't mold a passage through the birth canal as well as does the head.

Cervix—A small, cylindrical structure about an inch or so long and less than an inch around that makes up the lower part and neck of the uterus. The cervix separates the body and cavity of the uterus from the vagina.

Embryo—In humans, the developing individual from the time of implantation to about the end of the second month after conception. From the third month to the point of delivery, the individual is called a fetus.

Gestation—The period from conception to birth, during which the developing fetus is carried in the uterus.

Perineum—The area between the opening of the vagina and the anus in a woman, or the area between the scrotum and the anus in a man.

Placenta—The organ that provides oxygen and nutrition from the mother to the unborn baby during pregnancy. The placenta is attached to the wall of the uterus and leads to the unborn baby via the umbilical cord.

Vertex—The top of the head or highest point of the skull.

The Bradley method is called father-coached childbirth, because it focuses on the father serving as the coach throughout the process. It encourages normal activities during the first stages of labor without interventions and focuses on breathing and relaxation.

HypnoBirthing is becoming increasingly popular in the United States in the early 2000s and has proven to be quite effective. Based upon the work of Grantly Dick-Read, it teaches the mother to understand and release the fear-tension-pain syndrome, which so often is the cause of pain and discomfort during labor. When people are afraid, their bodies divert blood and oxygen from non-essential defense organs to large muscle groups in their extremities. Unfortunately, the body considers the uterus to be a non-essential organ. HypnoBirthing explores the myth that pain is a necessary accompaniment to a normal birthing. When a laboring woman's mind is free of **fear**, the muscles in her body, including her uterine muscles, relax, thus facilitating an easier, stress-free birth. In many cases, first stage labor shortens, which diminishes fatigue during labor leaving the mother stronger for pushing. The founder of HypnoBirthing, Marie Mongan, promotes the philosophy that eliminating fear allows the woman's body to work like it is supposed to.

The LeBoyer method stresses a relaxed delivery in a quiet, dimly lit room. It strives to avoid overstimulation of the baby and to foster mother-child **bonding** by placing the baby on the mother's abdomen and having the mother massage him or her immediately after the birth. This is followed by the father giving the baby a warm bath.

See also Apgar testing; Electronic fetal monitoring; Cesarean section.

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Childbirth Enhancement Foundation. 1004 George Avenue, Rockledge, Fl 32955. Web site: <www.cefcares.org/>.

HypnoBirthing Institute. PO Box 810, Epsom, NH 03234. Web site: <www.joes.com/home/HYPNOBIRTHING/>.

International Association of Parents and Professionals for Safe Alternatives in Childbirth. Rte. 1, Box 646, Marble Hill, MO 63764. Web site: <www.napsac.org/default.htm>.

International Childbirth Education Association. PO Box 20048, Minneapolis, MN 55420. Web site: <www.icea.org/>.

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Childhood Vaccine Injury Act

Definition

The Childhood Vaccine Injury Act established a federal program for compensating victims of vaccine-related injuries or death.

Purpose

During the early 1980s childhood immunization programs fell into chaos. Vaccine manufacturers and healthcare providers were overwhelmed with liability lawsuits from parents who believed that their children had been harmed by the **DTP vaccine**, which protects against **diphtheria**, **tetanus**, and pertussis (**whooping cough**). Companies that developed and produced vaccines halted or threatened to halt production and serious vaccine shortages developed. Childhood immunization rates fell.

To address this problem, physicians, public health agencies, the pharmaceutical industry, government representatives, and the parent-founded and -operated National Vaccine Information Center called for a no-fault alternative to litigation for resolving vaccine injury claims.

In response the U.S. Congress passed the Childhood Vaccine Injury Act of 1986 (PL 99-660). The purpose of the act was to do the following:

- establish the National Vaccine Injury Compensation Program (VICP) as a no-fault alternative to suits

against vaccine manufacturers and healthcare providers

- provide victims with an accessible and efficient means of obtaining compensation
- reduce the costs of litigation
- ensure adequate supplies of vaccine
- stabilize the cost of vaccines
- help prevent vaccine injuries through education and a vaccine adverse event reporting system (VAERS)
- create incentives for developing safer vaccines

The Childhood Vaccine Injury Act was part of an initiative to immunize all children against potentially life-threatening diseases. The act was amended several times after its original passage.

Description

In addition to establishing the VICP, the Childhood Vaccine Injury Act requires that **vaccination** records be included in a patient's permanent medical record and that they include the following:

- date of vaccine administration
- vaccine manufacturer and lot number
- name, address, and title of the healthcare provider

The act also requires that doctors report all adverse events occurring within 30 days of vaccination to the VAERS. About 12,000 vaccine-related adverse reactions are reported annually; however, it is estimated that less than 10 percent of doctors file such reports.

The Vaccine Injury Compensation Program (VICP)

The VICP took effect on October 1, 1988. The program is administered jointly by the Division of Vaccine Injury Compensation within the Health Resources and Services Administration (HRSA) of the U.S. Department of Health and Human Services (HHS), by the U.S. Court of Federal Claims, and by the U.S. Department of Justice (DOJ). A nine-member Advisory Commission on Childhood Vaccines oversees the VICP. This commission performs the following services:

- recommends improvements to the program
- proposes changes to the Vaccine Injury Table that specifies qualifying vaccines and injuries or deaths
- proposes legislation
- proposes new, safer vaccines for inclusion in the VICP
- collects information about vaccine-related injuries from federal, state, and local immunization programs
- revises vaccine information statements
- revises adverse reaction reporting requirements

Vaccine Injury Table

The VICP applies to all vaccines recommended by the U.S. Centers for Disease Control and Prevention (CDC) for routine administration to children. However, to qualify for compensation, the petitioner must prove one of the following:

- that an injury occurred that is specified in the Vaccine Injury Table
- that a vaccine significantly aggravated a pre-existing condition
- that a vaccine caused a condition

As of August 26, 2002, the Vaccine Injury Table included the following vaccines:

- DTP for diphtheria, tetanus, and pertussis, as well as other tetanus- and pertussis-containing vaccines
- MMR for **measles**, **mumps**, and **rubella** (German measles), as well as other measles- and rubella-containing vaccines
- OPV and IPV for **polio**, including cases in which polio was contracted from a child vaccinated with OPV
- hepatitis B
- Hib conjugate vaccines for haemophilus influenzae type B that causes meningitis
- varicella (chickenpox)
- pneumococcal conjugate vaccines
- live, oral, rhesus-based rotavirus vaccines for **gastroenteritis**, administered on or before August 26, 2002 (This vaccine was administered routinely only between October 1, 1998 and October 15, 1999.)
- new vaccines

These vaccines are covered regardless of whether they were administered individually or in combination or whether they were administered by public or private healthcare providers. When a new vaccine is added to the Vaccine Injury Table, coverage is retroactive for eight years. When the CDC recommends a new vaccine for routine administration to children, it may be automatically added to the table. Claims can be filed for other vaccines; however, the claimant must prove that the injury was caused by the vaccine. Since this can be very

difficult to prove, most VICP claims fall within the Vaccine Injury Table.

The Vaccine Injury Table contains guidelines for evaluating whether the injury or death was vaccine-related. For example, a claim that a child's seizures were triggered by a vaccine must include proof that the child's first seizure occurred within three days of the vaccine administration.

Furthermore, claims for vaccine-related injuries are only valid if the effects continued for at least six months following the vaccination or resulted in **hospitalization** or surgical intervention. The claim must be filed within 36 months of the appearance of the first symptoms. Claims for vaccine-related deaths must be filed within 24 months of the death or within 48 months of the onset of the injury that caused the death.

VICP procedures

Claims must be filed through the VICP before civil litigation can be pursued. Claims can be filed by the injured individual or by a parent, legal guardian, or trustee on behalf of a child or incapacitated victim. The procedural steps are as follows:

- A claimant files a petition for compensation with the U.S. Court of Federal Claims in Washington, DC, and with the secretary of HHS, along with a $150-filing fee.
- A physician from within the Division of Vaccine Injury Compensation reviews the petition and makes a recommendation.
- The physician's recommendation is included in a report filed with the court by the DOJ.
- Hearings are held before a "special master," an attorney appointed by the court judges; a DOJ attorney represents the HHS position; hearings usually last one or two days.
- The special master decides whether the claimant should be compensated.
- A hearing determines the amount of compensation.
- Decisions may be appealed first to a judge of the court, then to the Federal Circuit Court of Appeals, and finally to the Supreme Court.

Although claimants are not required to be represented by an attorney, most petitioners find that they need one. The law provides for compensation for reasonable attorney fees and costs, regardless of the outcome of the claim. An attorney filing a VICP petition must be a member of the bar of the U.S. Court of Federal Claims.

When the victim is a child, the following medical records must accompany the original petition to the court and the HHS secretary:

- all prenatal and birth records and newborn hospital records
- all medical records prior to and including vaccination records
- all post-injury hospital/emergency treatment records
- all post-injury outpatient records
- the VAERS form, if available
- long-term records, including school records, reports, and evaluations, and educational and psychological testing results
- police/ambulance records
- death and autopsy records

Compensation

Compensation for a vaccine-related injury may include the following:

- past and future non-reimbursable medical and custodial care and rehabilitation costs
- a maximum of $250,000 for **pain**, suffering, and emotional distress
- lost earnings or potential lost earnings
- reasonable attorney's fees and costs

Compensation for approved claims have averaged $824,462. The highest compensation award was for $9.13 million. Compensation for a vaccine-related death is limited to $250,000 for the deceased's estate plus attorney's fees and costs. Compensation is paid from the Vaccine Injury Compensation Trust Fund that is funded by a 75-cent excise tax on every purchased dose of a covered vaccine.

Petitions cannot be filed under VICP if a civil action for damages related to a vaccine injury is pending or if damages have been awarded by a court or in a settlement against the manufacturer or vaccine administrator. Civil litigation cannot be pursued if the petitioner accepts an award under the VICP.

Victims may file a civil suit against a vaccine manufacturer or the vaccine administrator given the following:

- The VICP petition is dismissed or ruled non-compensable.

KEY TERMS

Autism—A developmental disability that appears early in life, in which normal brain development is disrupted and social and communication skills are retarded, sometimes severely.

Thimerosal—A mercury-containing preservative used in some vaccines.

Vaccine—A substance prepared from a weakened or killed microorganism which, when injected, helps the body to form antibodies that will prevent infection by the natural microorganism.

Vaccine Adverse Event Reporting System (VAERS)—A federal government program for reporting adverse reactions to the administration of a vaccine.

Vaccine Injury Compensation Program (VICP)—A program through which victims of vaccine-induced injury or death can be awarded financial compensation.

Vaccine Injury Table—The guidelines by which claims to the VICP are evaluated; includes the vaccines, injuries or other conditions, and the allowable time periods for coverage by the VCIP.

- The VICP compensation offer is rejected by the claimant.
- The vaccine is not covered by VICP.

Common problems

Some lawyers and parents argue that there are serious problems with the VICP. They claim the following:

- The VICP is much too complicated and time-consuming.
- Victims are not necessarily able to recover damages.
- The VICP does not consider the emotional and psychological effects of a vaccine-related injury on the child's **family**.
- The VICP greatly underestimates the amount of legal work required for a reasonable chance of a successful claim.
- There is no real difference between the VICP and civil litigation except that the defendant is the U.S. Government rather than the vaccine manufacturer.
- The death-benefit limit of $250,000 does not take into account instances in which prolonged hospitalization preceded death.
- The death-benefit limit does not take into account an adult, family wage earner with a vaccine-related injury or death resulting from exposure to a newly-vaccinated child. (However, a claimant affected by the death benefit limitation may file a civil suit.)

Parental concerns

Outcomes

Between 1988 and 2004, 6,506 petitions were filed with the VICP, of which 4,246 were claims for **autism** resulting from the mercury-containing vaccine preservative thimerosal. Between 1989 and 2004, 140 of the thimerosal claims were dismissed and the rest of these claims were found to be not compensable. Of the non-autism/thimerosal claims, 916 were dismissed and 680 were compensated. Between 1990 and 2004, the government paid out over $588 million in compensation, fees, and costs for 676 claims. Nearly $10 million were paid out for attorney fees.

The VCIP has been credited with helping to stabilize the American vaccine market by providing the following services:

- providing a faster and easier method for resolving claims
- providing liability protection to vaccine manufacturers and healthcare providers
- encouraging research and development of new, safer vaccines

See also Chickenpox vaccine; Hepatitis B vaccine; Meningitis vaccine.

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Childproofing

Definition

About 2 million children are injured or killed by hazards in the home each year. Many of these incidents are preventable simply by taking precautions and by using simple, relatively inexpensive child **safety** products widely available. The practice of altering an environment in order to maximize the safety of small children is called childproofing.

Description

According to the National Safety Council, more than 20,000 accidental deaths and approximately 25 million accidental injuries occur each year to those under the age of 25. Most of these events occur in the home. The leading causes of injuries to children at home are **burns**, drowning, **poisoning**, cuts, and falls. Safety experts state that most of these accidents are preventable with planning and foresight.

Pediatricians advise parents to begin thinking about home safety when their children are around six months old. As soon as the child is even slightly mobile, childproofing the home is a wise practice. The following suggestions are common precautions that may be taken to avoid accidents in the home.

In the kitchen

The kitchen is one of the most potentially dangerous rooms in the household. In order to avoid the possible hazards, parents should follow these guidelines:

- Always buy the least hazardous products possible and keep toxic items out of sight, out of reach, and under lock and key.
- Post the numbers of the doctor, hospital, emergency service, and poison-control center on or near telephone.
- Teach children never to run with sharp implements (such as knives). Store knives in a secured drawer, in slotted knife blocks, or in trays attached to the wall, out of reach.
- Remove stove knobs or put covers over the burners when the stove is not in use.
- Never transfer household products to a container that once held food. Use the original containers.
- Never store snacks over the stove where a child might be tempted to try to reach them.

In the bathroom

The bathroom also contains a variety of potential dangers for children. These measures may help prevent accidental injuries in this room:

- Unplug electrical appliances when not in use and store them in a locked cabinet or drawer.
- Place nonslip mats or decals in the tub or shower and place a padded spout cover on the faucet.
- To prevent scald burns, set the temperature on the hot-water heater to a maximum of 120°F (49°C).
- Install safety locks on the medicine cabinet to prevent children from reaching prescriptions, **vitamins**, soaps, and other toxic substances. All medications should have child-resistant caps.
- Keep razors and scissors stored in a locked cabinet.
- Install toilet locks to prevent the lid from being lifted by young children. Children can easily fall into the toilet and may drown in as little as 1 in (2.5 cm) of water.

In children's rooms

Each child's room poses certain dangers, and the following are steps to assure children's safety in their bedrooms:

- Keep unopened windows locked and move furniture away from windows. Consider putting guards even on ground-floor windows. Though screens offer the illusion of safety, they are not strong enough to prevent a child from falling through the window.
- Make certain toy boxes are either without a lid or have a feature that prevents the lid from closing on a child's head or fingers.
- Make sure baby furniture meets current safety standards. There should be no sharp edges, and crib slats should be no more than 2.5 inches (6 cm) apart. If the distance is wider than this, a child may be caught or strangled between the bars.
- The crib mattress should fit snugly within the crib frame, with no more than two fingers' distance between the mattress and the crib railing, to avoid strangulation.
- Consider securing heavier pieces of furniture to walls, as children are often injured by pulling heavy furniture down on top of themselves.

In the living room

The living room can be dangerous for children. The following are some safety measures to take:

- Cover all unused electrical outlets with safety plugs.
- Pad sharp tables, and remove glass-topped tables, if possible.
- Pad the corners on a raised hearth or cover the edges with heat-resistant padding.
- Place a protective screen around the fireplace, and never leave children unsupervised when the fireplace is in use.

Hallways and stairs

Hallways and steps can be dangerous for **family** members. The following are some measures to take to make them less so:

- Install a smoke detector on every level of the home, as well as in the hallways outside every sleeping area. Change the batteries yearly.
- Use a carbon monoxide detector outside each sleeping area and near appliances which burn fuel.
- Teach children to use the handrail on stairways and to walk, not run, on the steps. Keep stairways cleared of all objects.
- Use safety gates that screw to the wall at the tops of stairways.

In the yard

The property around the house poses some dangers for children. Here are some steps parents can take to make the yard a safer place:

- Check to see if any plants in the yard are poisonous, and remove them or isolate them with fencing.
- Remove rotting or loose branches from trees promptly.
- Inspect gates and fences regularly to be certain there are no rusty nails or splintered wood.
- If there is a pool, it should be fenced, and all windows and doors providing access to the pool area should be locked. Alarms may be placed on these doors, to alert adults if children do slip into the pool area. Install a phone or keep a charged, waterproof cordless phone near the pool with emergency numbers posted on it.

In the garage

The garage can be a lethal place for small children. Here are some steps to take to secure it for the whole family:

- Petroleum products and any other poisonous substances should be stored up high, in appropriate containers, and locked.
- Garage doors should have electric openers that have an automatic reversing mechanism to prevent the door from closing on a child.

Child safety devices

The following child safety devices can help prevent injuries to young children and are commonly available in stores:

- Safety latches and locks. Latches and locks that are easy to install and use but sturdy enough to withstand pulls and tugs from children are good choices. Safety latches do not guarantee protection, but they can make it more difficult for children to reach dangerous substances.
- Safety gates. Safety gates can help to keep children away from stairs or rooms that contain hazards. Gates that cannot be dislodged easily by a child, but that adults can open and close without difficulty are best. Newer safety gates that meet safety standards have a

certification seal from the Juvenile Products Manufacturers Association (JPMA). Check older gates to be sure they do not have V-shapes large enough for a child's head and neck to fit into.

- Door knob covers and door locks. These devices help prevent children from entering rooms with possible hazards. Be certain that the door knob cover is sturdy enough so that it does not break but will still permit a door to be opened quickly by an adult in the case of an emergency.
- Anti-scald devices. These devices help regulate water temperature in order to prevent burns from water that is too hot.
- Smoke detectors. Used on every level of the home and especially near bedroom areas, smoke detectors should be checked monthly to make certain they are working.
- Window guards. To help prevent falls from windows, decks, and balconies, install window guards. However, remember that adults should be able to open at least one window in every room easily in case of fire.
- Corner and edge bumpers. Devices like corner and edge bumpers can be used on sharp edges to prevent injuries from falls.
- Outlet covers. Outlet covers should be inserted in all unused outlets as a way to prevent electrocution or electrical shock. Make sure they cannot be easily removed and that they are large enough so as not to pose a **choking** hazard for young children.
- Carbon monoxide detectors. Parents should place carbon monoxide detectors near all sleeping areas in the home, especially if the house uses gas or oil heat or if it has an attached garage.
- Door stops and door holders. These aid in preventing finger and hand injuries by preventing small fingers and hands from being pinched in doors or hinges.

Common problems

One of the most common childproofing challenges in a home is making certain that **toys** are age-appropriate and that they are in good working order. It can be difficult to keep toys that are geared for an older child away from a younger sibling, but parents should make an attempt to do so. The primary hazard presented by toys meant for older children is the possibility that a smaller child will choke on smaller parts. Parents can instruct their older children why it is important to keep these types of toys away from their siblings who can possibly be harmed by them.

Parents should also be aware if any toy or equipment made for children has been recalled by the manufacturer. This information is easily obtained by checking the Consumer Product Safety Commission (CPSC) recall list (available online at <www.cpsc.gov/cpscpub/prerel/prerel.html>).

Parental concerns

Parents are responsible for providing a safe home environment for their children. However, no amount of childproofing can replace vigilant supervision as a means of maintaining child safety. On the other hand, taking these simple and common sense precautions can help prevent many potential accidents.

See also Safety.

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Children's Apperception Test

Definition

The Children's Apperception Test (CAT) is a projective personality test used to assess individual variations in children's responses to standardized stimuli presented in the form of pictures of animals (CAT-A) or humans (CAT-H) in common social situations. In a supplement to the CAT—the CAT-S—the stimuli include pictures of children in common **family** situations such as

prolonged illnesses, births, deaths, and separations from parental figures.

Purpose

The CAT is used to assess personality, level of maturity, and, often, psychological health. The theory is that a child's responses to a series of **drawings** of animals or humans in familiar situations are likely to reveal significant aspects of a child's personality. Some of these dimensions of personality include level of reality testing and judgment, control and regulation of drives, defenses, conflicts, and level of autonomy.

Description

The CAT, developed by psychiatrist and psychologist Leopold Bellak and Sonya Sorel Bellak and first published in 1949, is based on the picture-story test called the **Thematic Apperception Test** (TAT). The TAT, created by psychologist Henry A. Murray for children (ten years old and older) as well as adults, uses a standard series of 31 picture cards in assessing perception of interpersonal relationships. The cards, which portray humans in a variety of common situations, are used to stimulate stories or descriptions (orally or in writing) about relationships or social situations and can help identify dominant drives, emotions, sentiments, conflicts and complexes. The examiner summarizes and interprets the stories in light of certain common psychological themes.

In creating the original CAT, animal figures were used instead of the human figures depicted in the TAT because it was assumed that children from three to ten years of age would identify more easily with drawings of animals. The original CAT consisted of ten cards depicting animal (CAT-A) figures in human social settings. The Bellaks later developed the CAT-H, which included human figures, for use in children who, for a variety of reasons, identified more closely with human rather than animal figures. A supplement to the CAT (the CAT-S), which included pictures of children in common family situations, was created to elicit specific rather than universal responses.

Like the TAT and the Rorschach inkblot test, the CAT is a type of personality **assessment** instrument known as a projective test. The term projective refers to a concept originated by Sigmund Freud. In Freud's theory, unconscious motives control much of human behavior. Projection is a psychological mechanism by which a person unconsciously projects inner feelings onto the external world, then imagines those feelings are being expressed by the external world toward him or herself.

As opposed to cognitive tests, which use intellectual and logical problems to measure what an individual knows about the world, projective assessments such as the CAT are designed to be open-ended and to encourage free expression of thoughts and feelings, thereby revealing how an individual thinks and feels.

Administration

The CAT, which takes 20–45 minutes to administer, is conducted by a trained professional—psychiatrist, psychologist, social worker, teacher or specially trained pediatrician—in a clinical, research, or educational setting. The test may be used directly in therapy or as a **play** technique in other settings.

After carefully establishing rapport with the child, the examiner shows the child one card after another in a particular sequence (although fewer than ten cards may be used at the examiner's discretion) and encourages the child to tell a story—with a beginning, middle, and end—about the characters. The examiner may ask the child to describe, for example, what led up to the scene depicted, the emotions of the characters, and what might happen in the future.

Scoring

In a projective test such as the CAT, there is no right or wrong answer. Thus there is no numerical score or scale for the test. The test administrator records the essence of each of the stories told and indicates the presence or absence of certain thematic elements on the form provided. As in the TAT, each story is carefully analyzed to uncover the child's underlying needs, conflicts, emotions, attitudes, and response patterns. The CAT's creators suggest a series of ten variables to consider when interpreting the results. These variables include the story's major theme, the major character's needs, drives, anxieties, conflicts, fears, and the child's conception of the external world.

Reliability and validity

Although responses in projective tests are believed to reflect personality characteristics, many experts have called into question the reliability, validity, and hence, usefulness of these tests as diagnostic techniques.

The CAT, as well as other projective measures, has been criticized for its lack of a standardized method of administration as well as the lack of standard norms for interpretation. Studies of the interactions between examiners and test subjects have found, for example, that the race, gender, and social class of both participants influence the stories that are told as well as the way the stories are interpreted by the examiner.

KEY TERMS

Apperception—The process of understanding through linkage with previous experience.

Projective test—A type of psychological test that assesses a person's thinking patterns, observational ability, feelings, and attitudes on the basis of responses to ambiguous test materials. Projective tests are often used to evaluate patients with personality disorders.

Rorschach test—A well-known projective test in which subjects are asked to describe a series of black or colored inkblots. The inkblots allow the patient to project his or her interpretations, which can be used to diagnose particular disorders. Also known as the Rorschach Psychodiagnostic Test.

Suggested uses

The CAT, which is designed for use in clinical, educational, and research settings, provides the examiner with a source of data, based on the child's perceptions and imagination, for use in better understanding the child's current needs, motives, emotions, and conflicts, both conscious and unconscious. Its use in clinical assessment is generally part of a larger battery of tests and interview data.

Parental concerns

Although it can provide useful information about a child's personality, the CAT, as a projective measure, relies heavily on the interpretations of the test administrator and is often referred to as an assessment tool rather than a test.

In addition to questioning the general reliability and validity of all projective tests, some experts maintain that cultural and language differences among children tested may affect CAT test performance and may produce inaccurate test results.

Parents need to keep in mind that **psychological tests** such as the CAT, which should be administered only by well-trained professionals, are only one element of a child's psychological assessment. These tools should never be used as the sole basis for a diagnosis. A detailed review of psychological, medical, educational, or other relevant history are required to lay the foundation for interpreting the results of any psychological measurement.

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Genevieve Slomski, Ph.D.

Choking

Definition

Choking is a condition caused by inhalation of a foreign object that partially or fully blocks the airway.

Description

Choking is a major cause of respiratory emergencies and cardiac arrest in infants and children. Choking occurs when a foreign object, such as food, buttons, coins, or toy parts, are inhaled and partially or completely block the airway, preventing adequate breathing. In many cases of choking, particularly in adults, the individual actively coughs and is able to expel the foreign object with no assistance or medical attention. However, children and infants are at increased risk of choking and foreign body airway obstruction due to immature airway and dental anatomy, distraction and **play** during eating, and a natural tendency to put objects into their mouths.

A 1995 study of the characteristics of objects known to commonly cause choking deaths in children found that round objects are most dangerous. For example, a small ball or marble can completely seal a child's or infant's trachea (windpipe). Round or cylindrical foods, hard candy, chewing gum, and balloons also present choking hazards. In infants, choking usually results from inhalation of small objects (coins, small **toys**, deflated balloons, buttons) that they place in their mouths.

Demographics

Each year, more than 17,000 infants and children are treated in hospital emergency departments for choking-related incidents, and more than 80 percent of these occur in children aged four years and younger. Airway obstruction death and injury are especially prevalent in children under age four due to anatomy (small airway), natural curiosity and tendency to put objects in their mouths, and incomplete chewing. According to statistics from the Centers for Disease Control and Prevention, choking rates in 2001 were highest in infants.

Causes and symptoms

Choking is a major cause of death for children under three and is a hazard for older children as well. Young children naturally explore the world with their mouths, and they will readily put in their mouths anything that fits. If a small object slips back into the throat and blocks the trachea, the child may become unable to breathe, and unless the child is helped to eject the object quickly, the child may asphyxiate and die. Food is also a choking hazard, especially for children under three who do not know how to chew food thoroughly.

According to statistics from the Centers for Disease Control and Prevention, of the 17,000 or so cases of pediatric choking in 2001, 60 percent were related to food, 31 percent were related to non-food substances, and 9 percent were related to undetermined objects. Of the food-related choking incidents, 19 percent resulted from candy or gum. Of the choking incidents resulting from non-food objects, 13 percent were related to coins.

Food-related choking usually occurs because infants and young children do not chew their food well, and larger pieces can become stuck in their throat. The following foods have been identified by the American Academy of Pediatrics as presenting choking hazards:

- hot dogs
- hard candy
- chewing gum
- nuts and seeds
- chunks of meat or cheese
- whole grapes
- popcorn
- chunks of peanut butter
- raw vegetables
- raisins

The following objects have been identified as presenting choking hazards:

- coins
- buttons
- marbles
- small balls
- deflated balloons
- watch batteries
- jewelry
- ball point pen caps and paper clips
- arts and crafts supplies
- small toys and toys with small detachable parts

When to call the doctor

All infants, children, and adolescents who have a choking incident should see a doctor, since complications can occur even if the object causing the choking is successfully expelled. Sometimes, pieces can be aspirated into the lungs, and even though breathing returns to normal, wheezing, chest **pain**, persistent **cough**, and **pneumonia** can develop within a few days due to the foreign body in the lung. Foreign bodies may require removal by bronchoscopy or surgery.

Vomiting may occur after being treated with the **Heimlich maneuver**. All infants and children who experience a choking episode severe enough to require the Heimlich maneuver should be taken to the hospital emergency room to be examined for airway injuries.

Diagnosis

Choking is diagnosed by observation of the choking victim. Children able to actively cough should be watched to make sure they expel the object on their own and that their airways do not become blocked. Indications that a choking victim's airway is blocked include the following:

- inability to cough, cry, or speak
- blue or purple face color from lack of oxygen
- grabbing at throat
- weak cough and labored breathing that produces a high-pitched noise
- all of the above, followed by loss of consciousness

When the actual choking incident is not observed, choking can be diagnosed by observing the above symptoms. For children, infants, and adolescents who are unconscious, choking and foreign body obstruction can be diagnosed by attempting to give rescue breaths. If a breath administered to the victim does not inflate the chest, rescuers should assume that the airway is blocked and take steps to clear the airway.

Treatment

An emergency choking incident is treated using the Heimlich maneuver, usually administered by parents, caregivers and teachers, or bystanders. Children who have a choking incident that requires the Heimlich maneuver should be examined by a physician for potential injuries to their airway and aftereffects of oxygen deprivation.

The Heimlich maneuver is used when a child is choking on a foreign object to the extent that he/she cannot breathe. Oxygen deprivation from a foreign body airway obstruction can result in permanent brain damage or death in four minutes or less. Using the Heimlich maneuver can save a choking victim's life. The Heimlich maneuver is not performed on infants under one year of age; rather, a series of back blows and chest thrusts are used to attempt to dislodge the foreign object.

If the foreign body cannot be expelled from the child's airway using the Heimlich maneuver, cardiac and/or respiratory arrest may occur, and the child may stop breathing. If this happens, **cardiopulmonary resuscitation** (CPR) is performed to restore and maintain breathing and circulation and to provide oxygen and blood flow to the heart, brain, and other vital organs. CPR can be performed by trained laypeople or healthcare professionals on infants, children, adolescents, and adults. CPR should be performed if an infant, child, or adolescent is unconscious and not breathing.

Prognosis

Incorrectly applied, the Heimlich maneuver can break bones or damage internal organs. Following the Heimlich maneuver, dysphagia (swallowing difficulty) and obstructive pulmonary edema (fluid accumulation in the lungs) may occur and require medical treatment.

KEY TERMS

Bronchoscopy—A procedure in which a hollow tube (bronchoscope) is inserted into the airway to allow visual examination of the larynx, trachea, bronchi, and bronchioles. It is also used to collect specimens for biopsy or culturing, and to remove airway obstructions.

Cardiopulmonary resuscitation (CPR)—An emergency procedure designed to stimulate breathing and blood flow through a combination of chest compressions and rescue breathing. It is used to restore circulation and prevent brain death to a person who has collapsed, is unconscious, is not breathing, and has no pulse.

Diaphragm—The thin layer of muscle that separates the chest cavity containing the lungs and heart from the abdominal cavity containing the intestines and digestive organs. This term is also used for a dome-shaped device used to cover the back of a woman's vagina during intercourse in order to prevent pregnancy.

Heimlich maneuver—An emergency procedure for removing a foreign object lodged in the airway that is preventing the person from breathing. To perform the Heimlich maneuver on a conscious adult, the rescuer stands behind the victim and encircles his waist. The rescuer makes a fist with one hand and places the other hand on top, positioned below the rib cage and above the waist. The rescuer then applies pressure by a series of upward and inward thrusts to force the foreign object back up the victim's trachea.

Trachea—The windpipe. A tube composed of cartilage and membrane that extends from below the voice box into the chest where it splits into two branches, the bronchi, that lead to each lung.

In many cases the foreign material is dislodged from the throat, and the choking victim suffers no permanent effects of the episode. If the foreign material is not removed, the choking victim may suffer permanent brain damage from lack of oxygen or may die.

Prevention

Choking is easily preventable by taking the following steps:

- supervising infants and children while they eat and play

- childproofing play areas by removing small objects
- cutting foods into very small pieces
- avoiding serving foods listed above as choking hazards to children under age four
- monitoring older children to make sure they do not give younger children hazardous foods or objects
- following age and **safety** guidelines on toys
- learning CPR and the Heimlich maneuver
- not letting children and infants play with coins

Parental concerns

Because most choking incidents occur in the home, all parents and infant/child caregivers should be trained in the Heimlich maneuver. Training is available through the American Red Cross and American Heart Association at local schools, YMCAs, and community centers.

The likelihood of choking incidents can be reduced by closely supervising infants and children while they eat and play. Most choking incidents are associated with food items, especially hot dogs, candies, grapes, nuts, popcorn, and carrots. Common non-food items that present choking hazards include deflated balloons, buttons, coins, small balls, small toys, and toy parts. All toys should be examined to make sure they are age-appropriate and do not have loose parts.

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Cholesterol, high

Definition

High cholesterol (hypercholesterolemia or hyperlipidemia) refers to the presence of higher than normal amounts of total cholesterol circulating in the bloodstream. Cholesterol is a fatty substance (lipid) that is essential to the body as protection for the walls of the vasculature (veins and arteries) and linings of body organs, a component in the manufacture of hormones, and a factor in the digestion of consumed fats in foods. It is manufactured in the liver and carried throughout the body in the bloodstream. Cholesterol is also a component of animal tissue and can be consumed in products such as meat, eggs, fish, milk, and milk products such as butter and cheese. Elevated cholesterol levels can result in the accumulation of fatty deposits on blood vessel walls, narrowing veins and arteries and impeding blood flow to the heart, brain, and other organs.

Description

Cholesterol has both a good form and a bad form that add up to total cholesterol when measured together. The body needs cholesterol to produce bile acids that help digest fats ingested in food, make hormones, protect cell walls, and participate in other processes that help maintain health. Ironically, cholesterol can also be a problem, if too much is manufactured by the liver or consumed through the diet and not metabolized or used. The utilization of fat in the body, or fat metabolism, is a complex process, complicated even more by abnormally high levels of cholesterol found circulating in the blood. Although high cholesterol is not often found in young children, it may begin to develop in adolescents or young adults either as an inherited condition or through

unhealthy eating habits and can continue into adulthood, creating potentially serious health problems. High cholesterol levels and fatty deposits in veins and arteries (atherosclerosis) have been found during autopsies of children who have died of accidents and other causes.

The liver metabolizes cholesterol, including the cholesterol obtained from foods in the diet. The components of cholesterol are then carried into the bloodstream bound to the surface of certain lipoproteins. Low-density lipoproteins or LDLs carry about 75 percent of the cholesterol into the blood and high-density lipoproteins carry the other 25 percent. LDL is the lipoprotein known as bad cholesterol because it consists primarily of cholesterol and is most associated with the development of vascular disease. Cholesterol is not the major part of HDL, the so-called good cholesterol, and the presence of higher amounts of HDL in the blood actually helps reduce the more harmful LDL levels. Another lipoprotein, very low-density lipoprotein (VLDL), carries harmful fats known as triglycerides but does not carry a significant amount of cholesterol. Triglycerides are also measured as part of a lipid profile and high levels are associated with vascular disease and heart disease. Cholesterol levels in blood serum vary considerably from day to day and even from one time of a day to another related to the consumption of fats in the diet.

High LDL (low-density lipoprotein) is a major precursor of vascular disease and heart disease. This form of cholesterol combines with triglycerides, cellular waste, calcium, and scar tissue to form a waxy deposit (plaque) on the inner walls of large and medium-sized arteries, causing a condition called hardening of the arteries (atherosclerosis or arteriosclerosis). Plaque typically builds up as people get older, more in some people than others depending on lifestyle (diet, **exercise**, alcohol consumption, and **smoking**) and heredity. The result may be a narrowing (stenosis) or blockage of blood vessels, interrupting the essential flow of blood and oxygen to the heart, brain, abdominal organs, and peripheral circulation to the arms and legs. Eventually this can lead to heart attack or **stroke**, permanent damage to the heart or brain, and life-threatening complications.

The population as a whole is at some risk of developing high LDL cholesterol. Specific risk factors include a **family** history of high cholesterol, **obesity**, coronary artery disease (atherosclerosis), stroke, **alcoholism**, diabetes, high blood pressure, and lack of regular exercise. The chances of developing high cholesterol increase after the age of 45. One of the primary causes of high LDL cholesterol is a combination of too much fat and sugar in the diet, especially through the consumption of fast foods and refined or packaged foods, a problem that has been especially true in the United States since the advent of manufactured foods. A renewed interest in whole foods may help to alter the prevalence of high cholesterol and vascular disease.

An increased serum cholesterol may be found in familial hyperlipidemia or hypercholesterolemia, underactive thyroid (**hypothyroidism**), untreated diabetes, a high-fat diet, pregnancy, heart attack, stress, and certain liver conditions (cirrhosis). A decreased level may be found in liver dysfunction, overactive thyroid (**hyperthyroidism**), malabsorption, **malnutrition**, or advanced **cancer**, among other conditions.

Although high cholesterol has been shown to be a risk factor for developing atherosclerosis in adults, with associated increased morbidity and mortality, studies have not indicated that high cholesterol in children and adolescents is related to the development of specific illness or increasing mortality in adulthood. There is strong evidence in numerous research studies, however, that a family history of high cholesterol, atherosclerosis, heart attack, or stroke increases the risk of a child developing high cholesterol levels.

High cholesterol is often diagnosed and treated by general practitioners or family practice physicians. In some cases, the condition is treated by an endocrinologist or cardiologist. Pediatricians will generally refer affected children to the appropriate specialist.

Demographics

The U.S. Food and Drug Administration (FDA) estimates that 90 million American adults, roughly half the adult population, have elevated cholesterol levels. This estimate does not indicate that as many children are candidates for high cholesterol levels; however, about 2 percent of the U.S. population has a family history of hypercholesterolemia in parents or grandparents, and this history is the most common predictor of high cholesterol levels in children and adolescents. Before **puberty**, average total and LDL cholesterol levels are higher in girls than in boys. Both LDL and HDL levels are higher in non-Hispanic black children than in non-Hispanic whites and Mexican-American children.

Causes and symptoms

The causes of high cholesterol may be genetic or hereditary factors in the manufacture of cholesterol by the liver or in fat metabolism, a diet high in saturated fats and trans-fatty acids, obesity, alcoholism, smoking, and lack of exercise.

There are no readily apparent symptoms that indicate high cholesterol, high LDL, high triglycerides, or low HDL. Obesity is a general indication of possible

high cholesterol levels. Labored breathing or general feelings of sluggishness and lack of energy may warrant examination by a physician and testing of cholesterol. Families or individuals who regularly consume a high-fat diet consisting of animal products, fast foods, and refined foods may also benefit from being tested for abnormal cholesterol levels.

When to call the doctor

Excess weight may be the only sign of possible high cholesterol in children. It is wise for parents to consult a physician if a child is consistently overweight and diet or exercise does not seem to make a significant difference. Sluggishness may also be noted if a child's veins and arteries are consistently filled with higher than normal amounts of fatty substances that are not being metabolized by the body.

Diagnosis

Total serum cholesterol is the cholesterol most often measured and reported in medical office tests, home tests, and blood cholesterol screening clinics; people who quote their cholesterol level as high may be talking about a total cholesterol of over 200mg/dL. A definitive diagnosis of high cholesterol, however, ideally includes measuring LDL, HDL, total cholesterol, and triglyceride levels, as well as the cholesterol to HDL ratio. This combination of tests performed in the clinical laboratory is called a lipid panel or lipid profile. Most physicians want to know the results of a lipid panel before diagnosing high cholesterol and recommending treatment. Screening for lipid levels in all children is not usually recommended. It is recommended that children whose parents have a total cholesterol level over 200mg/dL or whose family history includes heart disease or stroke in either parents or grandparents have a cholesterol screening performed. If the fasting blood level of cholesterol is 170 to 199 mg/dL, total cholesterol should be repeated and the two tests averaged. A final result of 200 mg/dL or over indicates that the entire lipid panel should be done to determine if hyperlipidemia is present.

In most adults the recommended levels for cholesterol and triglycerides, measured as milligrams per deciliter (mg/dL) of blood, are: total cholesterol, less than 200; LDL, less than 130; HDL, more than 35; triglycerides, 30–200; and cholesterol to HDL ratio, four to one. However, the recommended cholesterol levels may vary from person to person, depending on other risk factors such as a family history of heart disease or stroke or the presence of **hypertension**, diabetes, advanced age, alcoholism, or smoking.

The physician may recommend nuclear magnetic resonance (NMR) lipoprofile testing for individuals whose lipid measurements, history, and risk factors are not diagnostic, that is they are not revealing why an individual has coronary artery disease. Doctors have always been puzzled by why some people develop heart disease while others with identical HDL and LDL levels do not. Research studies in the early 2000s indicate that it may be due to the size of the cholesterol particles in the bloodstream. Nuclear magnetic resonance (NMR) lipoprofile exposes a blood sample to a magnetic field to determine the size of the cholesterol particles. Particle size also can be determined by a centrifugation test, in which blood samples are spun very quickly to allow particles to separate and move at different distances. The smaller the particles, the greater the chance of developing heart disease. It allows physicians to treat patients who have normal or close to normal results from a lipid panel but abnormal particle size.

Treatment

The primary goal of cholesterol treatment is to lower LDL to under 160 mg/dL in people without heart disease and who are at lower risk of developing it. The goal in people with higher risk factors for heart disease is less than 130 mg/dL. In patients who already have heart disease, the goal is under 100 mg/dL, according to FDA guidelines. Also, since low HDL levels increase the risk of developing heart disease, the goal for all individuals is to maintain an HDL of more than 35 mg/dL. These values apply to children and adolescents as well as adults.

First-line treatment of high cholesterol for all ages includes diet, exercise, and weight loss. The National Cholesterol Education Program recommends that children over age two eat a variety of foods for healthy development and ideal weight, consuming no more than 30 percent total fat in the diet and no more than 10 percent saturated fat as in animal foods. The American Heart Association Step 2 diet has been tested as a diet-alone treatment and in conjunction with drug therapy for children with high cholesterol, with good results. Regular exercise through aerobic activity is recommended.

In addition to diet and exercise, a variety of prescription medicines are available to help reduce cholesterol levels in the blood; these medications may not always be recommended for children, except for those whose parents or grandparents have high cholesterol and coronary artery disease. A class of drugs called statins is known to help lower LDL in combination with dietary changes and exercise, and studies have shown that they have no adverse effects in children. A class of drugs called fibric acid derivatives is sometimes recommended to lower triglycerides and raise HDL. Doctors decide which drug is most effective for an individual based on the cause and

the severity of the cholesterol problem and other health conditions that may be present, as well as possible side effects of the drug. Diet and exercise remain important factors in reducing elevated cholesterol levels, even if drug therapy is prescribed.

Alternative treatment

Alternative treatment of high cholesterol may include high doses of garlic, niacin, soy protein, algae, or other fatty acids, and the Chinese medicine supplement Cholestin (a red yeast fermented with rice).

GARLIC A number of clinical studies have indicated that garlic can offer modest reductions in cholesterol. A 1997 study by **nutrition** researchers at Pennsylvania State University found that men who took garlic capsules for five months reduced their total cholesterol by 7 percent and LDL by 12 percent. Another study showed that seven cloves of fresh garlic a day significantly reduced LDL, as did a daily dose of four garlic extract pills.

CHOLESTIN Cholestin has been available since 1997 as a cholesterol-lowering dietary supplement. It is a processed form of red yeast fermented with rice, a traditional herbal remedy used for centuries in China. Two studies released in 1998 showed Cholestin lowered LDL cholesterol by 20 to 30 percent. It also appeared to raise HDL and lower triglyceride levels. Although the supplement contains hundreds of compounds, the major active LDL-lowering ingredient is lovastatin, a chemical also found in the prescription drug Mevacor. The product is available as a dietary supplement, not a drug; its actual mechanism is not known. No serious side effects have been reported, but minor side effects, including bloating and heartburn, have been noted.

OTHER TREATMENTS A study released in 1999 indicated that blue-green algae contains polyunsaturated fatty acids that lower cholesterol. The algae, known as *Aphanizomenon flos-aquae* (AFA) is available as an over-the-counter dietary supplement. Flax seed oil is another source of fatty acids known to reduce cholesterol levels. Niacin, also known as nicotinic acid or vitamin B3, has been shown to reduce LDL levels by 10 to 20 percent and raise HDL levels by 15 to 35 percent. It also reduces triglycerides. Because an extremely high dose of niacin is needed to treat cholesterol problems, it should only be taken under a doctor's supervision to monitor possible toxic side effects. Niacin can also cause flushing when taken in high doses. Soy protein with high levels of isoflavones also has been shown to reduce LDL levels by up to 10 percent. In 2003, a Cuban research study revealed that policosanol, a substance made from sugar cane wax or beeswax, lowered LDL cholesterol nearly 27 percent in study subjects.

Nutritional concerns

Several specific diet options have been shown to be beneficial for reducing cholesterol. A vegetarian diet provides up to 100 percent more fiber and up to 50 percent less cholesterol from food than a meat-based diet. A balanced vegetarian diet consists of at least six servings of whole grain foods, three or more servings of green leafy vegetables, two to four servings of fruit, two to four servings of legumes (protein source), and one or two servings of non-fat dairy products daily. The macrobiotic diet is similar, with brown rice being a staple, but with the addition of other protein sources such as fish and fowl, tofu, and other soy products (miso, tempeh). The low glycemic or diabetic diet is known to raise the HDL (good cholesterol) level by as much as 20 percent in three weeks. Low glycemic foods promote a slow but steady rise in blood glucose levels following a meal, which increases the level of HDL. They also lower total cholesterol and triglycerides. Low glycemic foods include certain fruits, vegetables, beans, and whole grains. Processed (packaged foods) and refined foods (white flour products, white rice) and refined sugars (white sugar, brown sugar, molasses, and products made with them) should be avoided in all diets. Soy protein can be added to the daily diet to help replace animal sources of protein and reduce cholesterol; a diet containing 62 mg of isoflavones in soy protein is recommended and can be incorporated into other diet regimens, including vegetarian, macrobiotic, and low glycemic.

Prognosis

High cholesterol is one of the key risk factors for heart disease and has been shown to be treatable. Left untreated, high levels of LDL and total cholesterol can lead to the formation of plaque, the narrowing of blood vessels, vascular disease, and subsequent heart attacks and stroke.

Prevention

Since a large number of people with high cholesterol are overweight, a healthy diet and regular exercise are probably the most beneficial ways to control cholesterol levels. Exercise is an extremely important part of burning calories obtained by eating fats and helps maintain lower bad cholesterol and higher levels of good cholesterol. Exercise should consist of 20 to 30 minutes of vigorous aerobic exercise at least three times a week. Exercises that cause the heart to beat faster include fast

KEY TERMS

Atherosclerosis—A disease process whereby plaques of fatty substances are deposited inside arteries, reducing the inside diameter of the vessels and eventually causing damage to the tissues located beyond the site of the blockage.

Fatty acid—The primary component of lipids (fats) in the body. The body requires some, called essential fatty acids, to form membranes and synthesize important compounds.

Glycemic—The presence of glucose in the blood.

Hypertension—Abnormally high arterial blood pressure, which if left untreated can lead to heart disease and stroke.

Legumes—A family of plants, including beans, peas, and lentils, that bear edible seeds in pods. These seeds are high in protein, fiber, and other nutrients.

Lipids—Organic compounds not soluble in water, but soluble in fat solvents such as alcohol. Lipids are stored in the body as energy reserves and are also important components of cell membranes. Commonly known as fats.

Polyunsaturated fat—A non-animal oil or fatty acid rich in unsaturated chemical bonds. This type of fat is not associated with the formation of cholesterol in the blood.

Trans-fatty acid—A type of fat created by hydrogenating polyunsaturated oils. This changes the double bond on the carbon atom from a cis configuration to a trans configuration, making the fatty acid saturated, and a greater health concern. For example, stick margarines are known to contain more trans-fatty acids than liquid oils.

walking, bicycling, jogging, roller skating, swimming, and walking up stairs.

Nutritional concerns

In general, the nutritional goals for preventing high levels of cholesterol are to substantially reduce or eliminate foods high in animal fat, including meat, shellfish, eggs, and dairy products. The use of polyunsaturated fats in cooking is also recommended, including cold pressed oils such as olive oil, canola oil, and sesame oil. Many vegetable oils are hydrogenated or extracted at high temperatures and are best avoided. Trans-fatty acids found in solid shortenings, most margarines, and hydrogenated oils or products containing them should also be avoided because they are known to increase levels of LDL.

Parental concerns

Parents need not be concerned about high cholesterol levels in their children unless the child is obese or there is a family history of high cholesterol, heart attack, or stroke. Parents who have cholesterol levels over 200 mg/dL themselves may want to have their children's cholesterol levels tested. Much information is available from public health sources and family physicians about diet and exercise recommendations to help people of all ages reduce the risk of vascular disease and related illnesses, such as heart disease and stroke.

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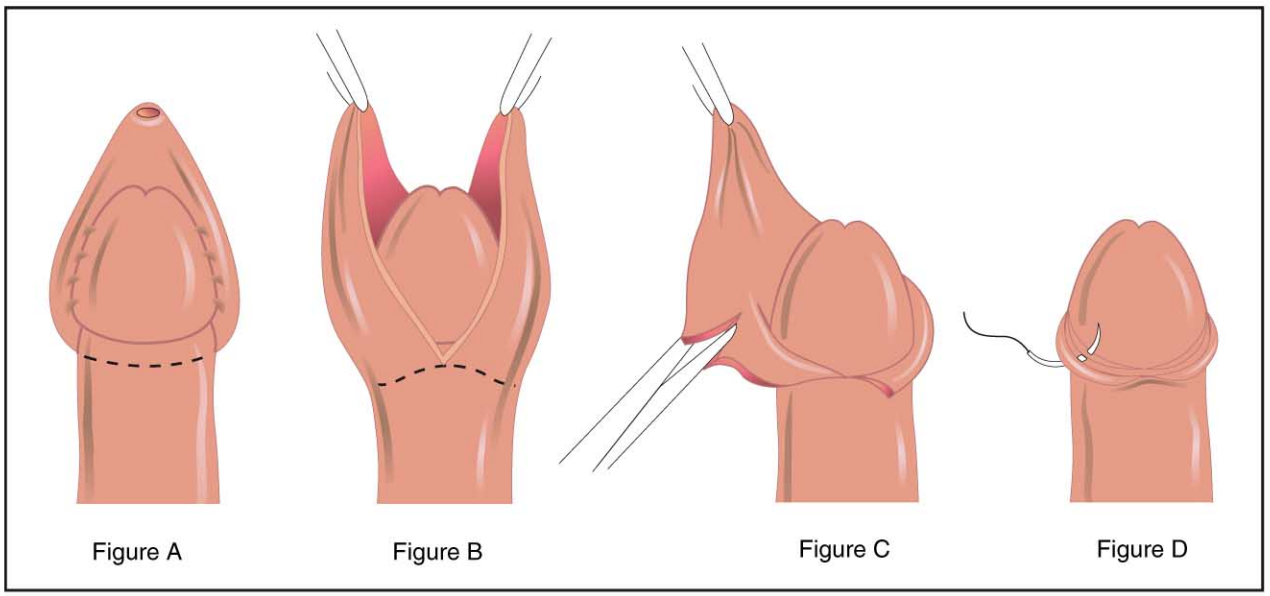
Chordee *see* **Hypospadias**

Chorea *see* **Movement disorders**

Circumcision

Definition

The surgical removal of the foreskin of the penis in a male or the prepuce of a clitoris in a female.



A typical circumcision procedure involves the following steps: Figure A: The surgeon makes an incision around the foreskin. Figure B: The foreskin is then freed from the skin covering the penile shaft. Figure C: The surgeon cuts the foreskin to the initial incision, lifting the foreskin from the mucous membrane. Figure D: The surgeon sutures the top edge of the skin that covers the penile shaft and the mucous membrane. *(Illustration by Electronic Illustrators Group.)*

Purpose

In the United States, circumcision in infant boys is performed for social, medical, cultural, or religious reasons. Once a routine operation urged by pediatricians and obstetricians for newborns in the middle of the twentieth century, circumcision has become an elective option that parents make for their sons on an individual basis. Families who practice Judaism or Islam may select to have their sons circumcised as a religious practice. Others may elect circumcision for medical reasons.

Female circumcision (also known as female genital mutilation) is usually performed for cultural and social reasons by **family** members and others who are not members of the medical profession, with no anesthesia. Not only is the prepuce of the clitoris removed but often the vaginal opening is sewn to make it smaller. This practice is supposed to ensure the virginity of a bride on her wedding day. It also prevents the woman from achieving sexual pleasure during coitus. Female circumcision is illegal in most countries of the world. It is considered by most people to be a human rights violation.

Though the incidence of male circumcision has decreased from 90 percent in 1979 to 60 percent in 2002, it is still the most common surgical procedure in the United States. Circumcision rates are much lower for the rest of the industrialized world. In Britain, it is only performed for religious practices or to correct a specific medical condition of the penis.

Parents may choose circumcision because they believe the surgery protects against infections of the urinary tract and the foreskin, prevents **cancer**, lowers the risk of getting **sexually transmitted diseases**, and prevents phimosis (a tightening of the foreskin that may close the opening of the penis). Though studies indicate that uncircumcised boys under the age of five are 20 times more likely than circumcised boys to have urinary tract infections (UTIs), the rate of incidence of UTIs is quite low. There are also indications that circumcised men are less likely to suffer from penile cancer, inflammation of the penis, or have many sexually transmitted diseases. Here again, the rate of incidence is low. Good hygiene usually prevents most infections of the penis. Phimosis and penile cancer are very rare, even in men who have not been circumcised. Education and good safe-sex practices can prevent sexually transmitted diseases in ways that a surgical procedure cannot because these are diseases acquired through risky behaviors.

With these factors in mind, the American Academy of Pediatrics issued a policy pronouncement that states although there is existing scientific evidence that support the medical benefits of circumcision, the benefits are not strong enough to recommended circumcision as a routine practice.

Description

The foreskin of the penis safeguards the sensitivity of the glans and shields it from irritation by urine, feces,

and foreign materials. It also protects the urinary opening against infection and accidental injury.

Despite a long-standing belief that infants do not experience serious **pain** from circumcision, most authorities in the early 2000s believe that some form of local anesthesia is necessary. The physician injects local anesthesia at the base of the penis or under the skin around the penis (subcutaneous ring block). Both anesthetics block key nerves. EMLA cream, a topical formula of several anesthetics, can also be used.

In circumcision of infant boys, the foreskin is pulled tightly into a specially designed clamp. Pressure is applied and the foreskin pulls away from the broadened tip of the penis. Pressure from the clamp stops bleeding from blood vessels that supplied the foreskin. In older boys or adults, an incision is made around the base of the foreskin, the foreskin is pulled back, and then it is cut away from the tip of the penis. Stitches are usually used to close the skin edges.

After circumcision, the wound should be washed daily. An antibiotic ointment or petroleum jelly may be applied to the site. If there is an incision, a wound dressing will be present and should be changed each time the diaper is changed. Sometimes a plastic ring is used instead of a bandage. The ring usually falls off in five to eight days. The penis heals in seven to 10 days.

Infants who undergo circumcision may be fussy for some hours afterward, so parents should be prepared for crying, feeding problems, and **sleep** problems. Generally these go away within a day. In older boys, the penis may be painful, but the pain goes away gradually. A topical anesthetic ointment or spray may be used to relieve this temporary discomfort. There may also be a bruise on the penis, which typically goes away with no particular attention.

The clitoral prepuce of infant girls is drawn up and away from the body before being removed. In this procedure, the clitoris is also frequently removed. The vaginal opening may be partially closed. Healing is slower in girls than for boys. Most female circumcisions are performed under unhygienic conditions using primitive, homemade implement such as rusty razor blades and thorns. Infections are common.

Risks

Complications following newborn circumcision appear in one out of every 500 procedures. Most complications are minor. Bleeding occurs in half of the complications and is usually easy to control. Infections are rare and present with **fever** and signs of inflammation. Uneven healing of skin may lead to laterally curving erections in adulthood.

KEY TERMS

Foreskin—A covering fold of skin over the tip of the penis.

Glans penis—The cone-shaped tip of the penis.

Hernia—A rupture in the wall of a body cavity, through which an organ may protrude.

Hydrocele—A collection of fluid between two layers of tissue surrounding the testicle; the most common cause of painless scrotal swelling.

Hypospadias—A congenital abnormality of the penis in which the urethral opening is located on the underside of the penis rather than at its tip.

Phimosis—A tightening of the foreskin that may close the opening of the penis.

Prepuce—A fold of skin, such as the foreskin of the penis or the skin that surrounds the clitoris.

There may be injuries to the body of the penis that may be difficult to repair. In 2000, there were reports that the surgical clamps used in circumcision were at fault in over 100 injuries reported between July 1996 and January 2000. In nearly all cases, the clamps were assumed to be in working order but had been repaired with replacement parts that were not of the manufacturer's specifications. Physicians were urged to inspect the clamps before use and ensure that their dimensions fit the infant's body parts.

Circumcised girls have a high incidence of infertility and sexual dysfunction as adults. Most experience infections immediately after the procedure.

Normal results

Among boys who are circumcised, most have no penile, urologic, or sexual dysfunction as adults. A majority of girls who are circumcised experience urologic, reproductive, and sexual dysfunction as adults.

Parental concerns

The only medical justification for male circumcision is to correct a health problem or condition. There are no medical justifications for female circumcision. The only other justification for male or female circumcision is religious or cultural. Leaders of the vast majority of religions throughout the world condemn any form of female circumcision.

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American Academy of Pediatrics. 141 Northwest Point Boulevard, Elk Grove Village, IL 60007–1098. Web site: <www.aap.org/>.

American College of Surgeons. 633 North St. Clair Street, Chicago, IL 60611–32311. Web site: <www.facs.org/>.

American Medical Association. 515 N. State Street, Chicago, IL 60610. Web site: <www.ama-assn.org/>.

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L. Fleming Fallon Jr., MD, DrPH

Cleft lip and palate

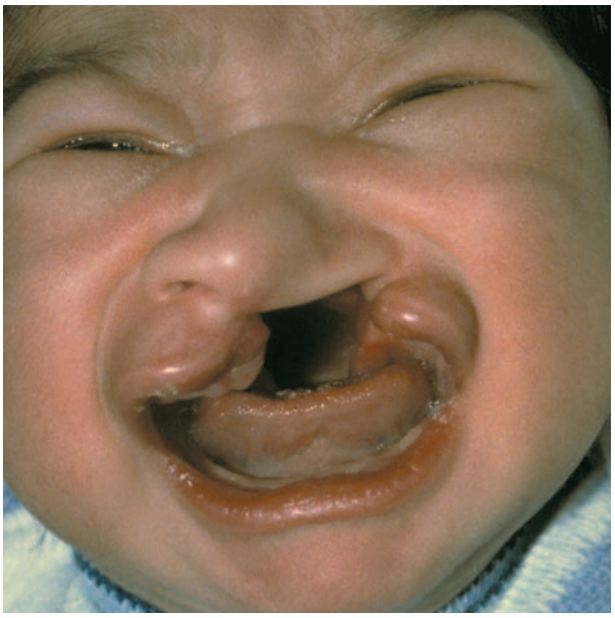
Definition

A cleft lip and/or palate is a birth defect (congenital) of the upper part of the mouth. A cleft lip creates an opening in the upper lip between the mouth and nose and a cleft palate occurs when the roof of the mouth has not joined completely.

Description

Cleft means split or separated. During the first months of pregnancy, separate areas of the face—such as bony and muscular parts, mouth, and throat, develop individually and then join together. If some parts do not join properly the result is a cleft, the type and severity of which can vary. During the fifth through ninth weeks of pregnancy genetic and environmental factors are most likely to affect lip and palate development. Cleft palate occurs when the right and left segments of the palate fail to join properly. The back of the palate (toward the throat) is called the soft palate, and the front section (toward the mouth opening) is known as the hard palate. A cleft palate can range from just an opening at the back of the soft palate to a nearly complete separation of the roof of the mouth (soft and hard palate). In some cases, an infant with a cleft palate may also have a small lower jaw and have difficulty breathing. This condition is called Pierre Robin sequence.

Cleft lip occurs when the lip elements fail to come together during fetal development, thus creating an opening in the upper lip between the mouth and nose. The lip looks split. A cleft lip may be complete, meaning that



Infant with a unilateral cleft lip and palate. *(Custom Medical Stock Photo Inc.)*

there is complete separation in one or both sides of the lip extending up and into the nose, or it may be incomplete, in which case there is only a notch in the fleshy portion of the lip. The incomplete cleft lip results in less facial distortion because the connected parts of muscle and tissue have a stabilizing effect. In a complete cleft lip, the muscles pull away from the center of the face, resulting in distortion of the nose and mouth. A cleft on one side is called a unilateral cleft. If a cleft occurs on both sides, it is called a bilateral cleft.

Demographics

Over 5,000 infants are born each year in the United States with a cleft lip or palate (about one in every 700 births). Cleft lip without cleft palate is the third most common congenital malformation among newborns in the United States and is estimated to occur roughly twice as often in males than in females. Cleft palate without cleft lip is fifth most common, and it affects roughly twice as many girls as boys. Clefts may affect the left or right side of the mouth only (unilateral) or both sides (bilateral). Left-side clefts represent 70 percent of all unilateral clefts. In the United States, clefting seems to be at least in part related to ethnicity, occurring most often among Asians, Latinos, and Native Americans (one in 500), next most often among persons of European ethnicity (one in 700), and least often among persons of African ethnicity (one in 1,000).

Causes and symptoms

The causes of clefts are as of 2004 still poorly understood. Most scientists believe that clefting occurs as a result of a combination of genetic and environmental factors. In the United States and western Europe, researchers report that a **family** history of facial clefts is present in approximately 40 percent of all cases. The likelihood of a baby being born with a facial cleft increases if a first-degree relative (mother, father, or sibling) has a cleft. Mothers who abuse alcohol and drugs, lack **vitamins** (especially **folic acid**) during the first weeks of pregnancy, or have diabetes are more likely to have a child with facial clefts.

Clefts may occur alone or with other abnormalities that may be hidden or obvious. Up to 13 percent of infants with cleft lip or palate have other birth defects. Some cases involve genetic syndromes that may result in specific problems for the infant and may have a high risk of affecting others in the family. For this reason, newborns with clefts should be thoroughly examined by a specialized physician soon after birth.

When to call the doctor

Families with a history of cleft lip or palate or any other syndrome or condition associated with clefting should discuss the chances of recurrence with a genetic counselor.

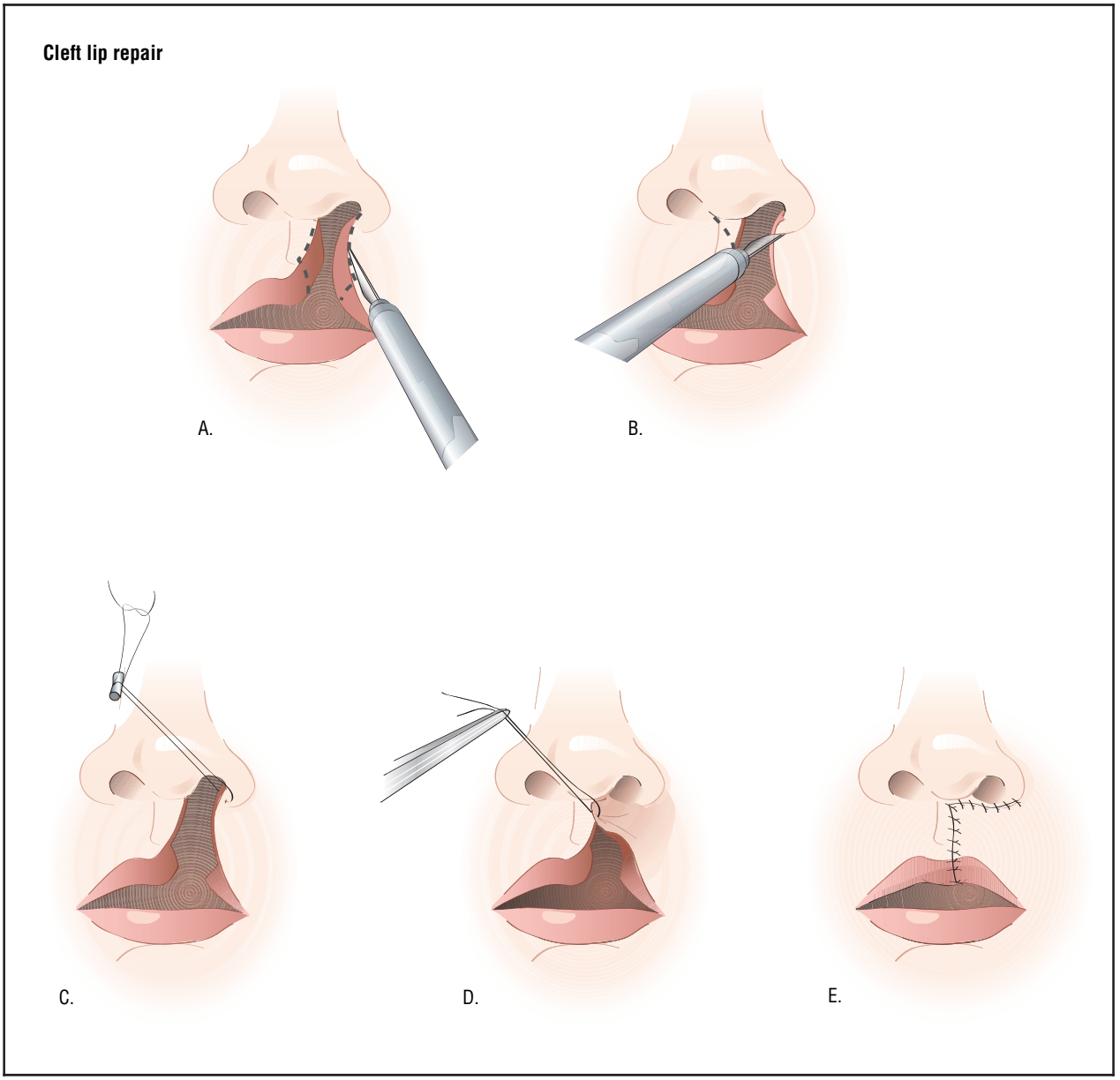
Diagnosis

Because clefting causes specific physical manifestations, it is easy to diagnose. Although some types of clefts can be detected during pregnancy by an ultrasound test, many are not discovered until birth.

Treatment

A cleft lip and/or palate can be repaired with corrective surgery, performed in a hospital under general anesthesia. Generally, within the first few days following birth of an infant with a facial cleft, a team is assembled to prepare a plan for treatment of the cleft. The treatment team usually includes representatives from several medical or psychological specialties, including pediatrics, plastic surgery, otolaryngology, orthodontia, prosthodontics, oral surgery, speech and language pathology, audiology, nursing, and psychology. It is common for one team member to coordinate service and communication between the team members and the family.

Surgical repair of a cleft lip is carried out at about three to four months of age. The whole emphasis in repairing the lip is on the muscle repair in order to mold the distorted front central section of the upper gum



Cleft lip repair. The edges of the cleft between the lip and nose are cut (A and B). The bottom of the nostril is formed with suture (C). The upper part of the lip tissue is closed (D), and the stitches are extended down to close the opening entirely (E). *(Illustration by Argosy, Inc.)*

containing the four upper front teeth (premaxilla) back into its proper position. Cleft lip often requires only one reconstructive surgery, especially if the cleft is unilateral. The surgeon makes an incision on each side of the cleft from the lip to the nostril, drawing the two sides of the cleft together and suturing them together. Bilateral cleft lips are usually repaired in two surgeries, about a month apart. The first surgery is performed when a baby is between six and 10 weeks old and usually requires a one-night stay in the hospital.

Cleft palate can require several surgical procedures during the course of a child's first 18 years. The first surgery to repair the palate usually occurs when the infant is between six and 12 months old. It usually involves palatal lengthening and drawing tissue from either side of the mouth to rebuild the palate. The procedure usually requires two or three nights in the hospital, the first night in the intensive care unit. The purpose of this surgery is to create a functional palate, reduce the chances that fluid will develop in the middle ears, and help the proper

development of the child's teeth and facial bones. In addition, the functional palate helps the child's speech development and feeding abilities. In both types of surgery, the necessity for more operations depends on the skill of the surgeon as well as the severity of the cleft, its shape, and the thickness of available tissue that can be used to create the palate. About 20 percent of children with a cleft palate require further surgical procedures to help improve their speech. Additional surgeries may also improve the appearance of the lip and nose, close openings between the mouth and nose, help breathing, and stabilize and realign the jaw.

Nutritional concerns

Infants with cleft lip or cleft soft palate generally have few feeding problems. However, when the cleft involves the hard palate, the infant is usually not able to suck efficiently. For these infants, caregivers must experiment with various feeding techniques, such as special nipples or alternate feeding positions. The infant with a cleft should be held in a nearly sitting position during feeding to prevent the breast or formula milk from flowing back into the nose. In addition, the infant should be burped frequently, approximately every three or four minutes. The sucking reflex is strong in all infants and should be encouraged in infants with facial clefts even if the sucking is inefficient, since the reflex seems to help the later development of speech. It is important to keep the cleft clean and not to allow formula, mucus, or other matter to collect in the cleft.

Prognosis

Both cleft lip and cleft palate are treatable birth defects. Most children born with either or both of these conditions undergo reconstructive surgery while they are still infants to correct the defect and significantly improve facial appearance. With advances in surgical techniques and with more complete repair of facial clefts, about 80 percent of affected children have normal speech development by the time they enter school. Continuation of speech therapy results in continuous improvement for most common speech problems.

Prevention

While little is known about how to prevent clefts, researchers from the California Birth Defects Monitoring Program found that women considering pregnancy may be able to reduce the risk of facial clefts (and possibly other birth defects) in their offspring by taking a multivitamin containing folic acid for one month prior to becoming pregnant. Other studies have shown that fetuses with certain predisposing genes may be at increased risk for cleft palate if their mothers smoke. Because some types of medications (for example some drugs used to treat epilepsy) have been linked to increased risk of clefts, women who take medications for chronic illnesses should check with their doctors before they become pregnant.

KEY TERMS

Bilateral cleft lip—A cleft that occurs on both sides of the lip.

Cleft—An elongated opening or slit in an organ.

Complete cleft—A cleft that extends through the entire affected mouth structure.

Congenital—Present at birth.

Palatal lengthening (palatal pushback)—A surgical procedure in which tissue from the front part of the mouth is moved back to lengthen it.

Palate—The roof of the mouth.

Premaxilla—The front central section of the upper gum, containing the four upper front teeth.

Unilateral cleft—A cleft that occurs on only the right or left side of the lip.

Parental concerns

Parents of a newborn baby with a cleft lip or palate are often confused and afraid of the impact the defect will have on their child's life. These feelings can be alleviated by learning about the cleft and treatment options. They also must communicate what they are learning about clefts to family, siblings, and friends. It is important for people who come into contact with the child to realize that a cleft is not a wound, although it may give the impression that it is tender or sore. Parents can help others understand that the cleft does not hurt and that it will be repaired. To ensure normal psychological and speech development, parents should interact with their infant as they would with any newborn in the family and should encourage others to do the same.

As the child with a cleft grows and develops, he or she will certainly experience many good and bad reactions from adults and children. Other children may tease the child or use the term "harelip." It may be helpful for parents of a child with a facial cleft to meet with his classmates and teachers to explain the history of the term

harelip. Although a facial cleft was once referred to as a harelip to reflect its similarity to the mouth of a rabbit, the term is considered insulting today. Educating adults and children about cleft lip and palate is the best way to relieve others' **anxiety** about the defect and lessen any negative psychological effects that bad reactions might have on the child.

See also Language development.

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About Face USA. PO Box 969, Batavia, IL 60510–0969. Web site: <www.aboutfaceusa.org/default.htm>.

American Cleft Palate: Craniofacial Association (ACPCA)/ Cleft Palate Foundation (CPF). 1504 East Franklin Street, Suite 102, Chapel Hill, NC 27514–2820. Web site: <www.cleftline.org>.

American Speech-Language-Hearing Association. 10801 Rockville Pike, Rockville, MD 20852. Web site: <www.asha.org>.

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Clotting disorders *see* **Coagulation disorders**

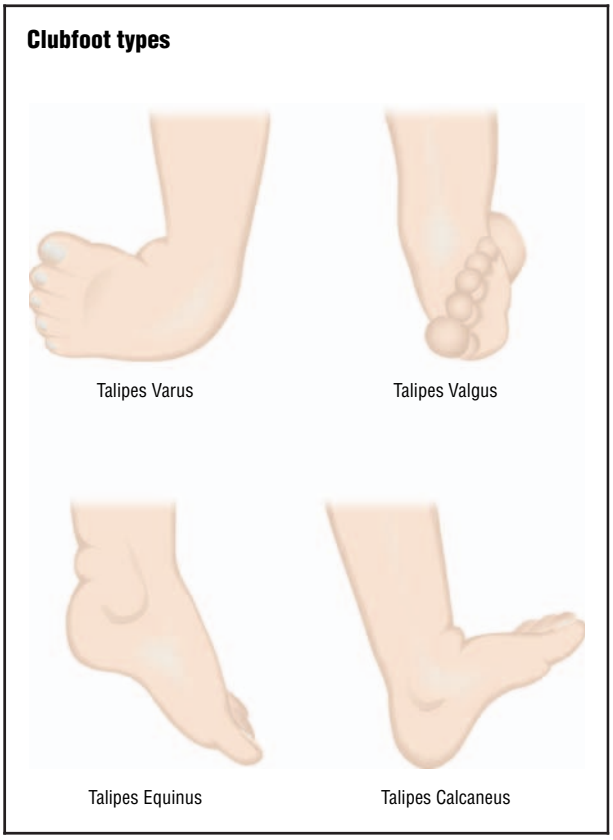
Clubfoot

Definition

Clubfoot is a condition in which one or both feet are twisted into an abnormal position at birth. The condition is also known as talipes.

Description

True clubfoot is characterized by abnormal bone formation in the foot. There are four variations of clubfoot: talipes varus, talipes valgus, talipes equines, and talipes calcaneus. In talipes varus, the most common form of clubfoot, the foot generally turns inward so that the leg and foot look somewhat like the letter J (when looking at the left foot head-on). In talipes valgus, the foot rotates outward like the letter L. In talipes equinus, the foot points downward, similar to that of a toe dancer. In talipes calcaneus, the foot points upward, with the heel pointing down.



The four varieties of clubfoot. Talipes varus is by far the most common type. *(Illustration by GGS Information Services.)*

Clubfoot can affect one foot or both feet. Sometimes the feet of an infant appear abnormal at birth because of the intrauterine position of the fetus prior to birth. If there is no anatomic abnormality of the bone, this is not true clubfoot, and the problem can usually be corrected by applying special braces or casts to straighten the foot.

True clubfoot is usually obvious at birth because a clubfoot has a typical appearance of pointing downward and being twisted inwards. Since the condition starts in the first trimester of pregnancy, the abnormality is quite well established at birth, and the foot is often very rigid. Uncorrected clubfoot in an adult causes only part of the foot, usually the outer edge or the heel or the toes, to touch the ground. For a person with clubfoot, walking becomes difficult or impossible.

Demographics

The ratio of males to females with clubfoot is 2.5 to 1. The incidence of clubfoot varies only slightly. In the United States, the incidence is approximately one in every 1,000 live births. A 1980 Danish study reported an overall incidence of 1.2 in every 1,000 children. By 1994, that number had doubled to 2.41 in every 1,000 live births. No reason was offered for the increase.

A **family** history of clubfoot has been reported in 24.4 percent of families in a single study. These findings suggest the potential role of one or more genes being responsible for clubfoot.

Causes and symptoms

Experts do not agree on the precise cause of clubfoot. Some experts feel that clubfoot may begin early in pregnancy, probably in the 10th to 12th weeks of gestation. The exact genetic mechanism of inheritance has been extensively investigated using family studies and other epidemiological methods. As of 2004, no definitive conclusions had been reached, although a Mendelian pattern of inheritance is suspected. This may be due to the interaction of several different inheritance patterns, different patterns of development appearing as the same condition, or a complex interaction between genetic and environmental factors. The MSX1 gene has been associated with clubfoot in animal studies. But, as of 2004, these findings had not been replicated in humans.

Several environmental causes have been proposed for clubfoot. Many obstetricians feel that intrauterine crowding causes clubfoot. This theory is supported by a significantly higher incidence of clubfoot among **twins** compared to singleton births. Intrauterine exposure to the drug misoprostol has been linked with clubfoot. Misoprostol is commonly used when trying, usually unsuccessfully, to induce abortion in Brazil and in other countries in South and Central America. Researchers in Norway have reported that males who are in the printing trades have significantly more offspring with clubfoot than men in other occupations. For unknown reasons, **amniocentesis**, a prenatal test, has also been associated with clubfoot. The infants of mothers who smoke during pregnancy have a greater chance of being born with clubfoot than are offspring of women who do not smoke.

The physical appearance of a clubfoot may vary. However, at birth, an affected foot usually turns inward and points downward. It resists realignment. The calf muscle may be smaller and less well developed than normal. One or both feet may be affected.

When to call the doctor

An pediatrician should be consulted at birth, the usual time clubfoot is initially diagnosed. While there is no immediate urgency, the condition should be evaluated by a pediatrician or an orthopedic surgeon in the first weeks of life so that treatment can be started.

KEY TERMS

Intrauterine—Situated or occuring in the uterus.

Orthopedist—A doctor specializing in treatment of the musculoskeletal system.

Diagnosis

Clubfoot is diagnosed by physician inspection. This is most often completed immediately after birth. Clubfoot may be suspected during the latter stages of pregnancy, especially in a mother of shorter or smaller than normal stature, a large fetus, or multiple infants.

Treatment

Clubfoot is corrected by casting or surgery. To have the best chances for successful resolution without resorting to surgery, treatment as soon after birth as possible. The Ponseti method of stretching and casting has been used with increasing success since the 1990s. The Ponseti method requires that a doctor stretch the child's affected foot toward its anatomically correct position and hold it in place with a cast. The foot is realigned and a new cast applied weekly for several weeks. Once the correct position has been achieved, a brace must be worn during periods of **sleep** to maintain the correction. To be successful, the method requires active parental involvement.

When casting and bracing are not successful, surgery may be required to realign the tendons, ligaments, and joints in the foot and ankle. Such a procedure is usually completed between nine and 12 months of age. After surgery, a cast holds the foot in the desired position.

Prognosis

The prognosis for successfully treating clubfoot is good at this time. Persons with clubfoot that is corrected by surgery may notice some increased stiffness in their affected feet as they age. A corrected clubfoot is often a shoe size smaller than normal and may be somewhat less flexible. The calf muscles in an affected clubfoot leg may be slightly smaller than an unaffected leg. However, without treatment, clubfoot will result in a functional disability.

Prevention

At the present time, there is no way to prevent clubfoot. Pregnant women can reduce the risk of clubfoot by refraining from **smoking**.

Parental concerns

Parents of an infant with clubfoot should be prepared to participate in treatment for two or more years. They should seek prompt treatment from a qualified health care provider.

Resources

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CMV *see* **Cytomegalovirus infection**

CO poisoning *see* **Carbon monoxide poisoning**

Coagulation disorders

Definition

Coagulation disorders (coagulopathies) are disruptions in the body's ability to control blood clotting, an essential function of the body designed to prevent blood loss. The most commonly known coagulation disorder is **hemophilia**, a condition in which a critical component of blood coagulation is missing, causing individuals to bleed for long periods of time before clotting occurs. There are numerous other coagulation disorders stemming from a variety of causes.

Description

Coagulation, or clotting, is a complex process (called the coagulation cascade) that involves 12 coagulation factors (designated by Roman numerals as factors I through XII) found in blood plasma and several other blood components. The factors include prothrombin, thrombin, and fibrin. Each has a precise role in coagulation. Besides the factors, which are all proteins, plasma (the fluid component of the blood) carries a number of other proteins that regulate bleeding. Platelets, tiny colorless cells in the blood, initiate contraction of damaged blood vessels so that less blood is lost. They also help plug damaged blood vessels and work with other constituents in plasma to accelerate blood clotting. A deficiency in clotting factors or a disorder that affects platelet production or one of the many steps in the entire process can disrupt clotting and severely complicate blood loss from injury, **childbirth**, surgery, and specific diseases or conditions in which bleeding can occur.

Coagulation disorders arise from different causes and involve different complications. Some common coagulation disorders are:

- Hemophilia or hemophilia A (factor VIII deficiency) is an inherited coagulation disorder, affecting about 20,000 Americans. This genetic disorder is carried by females but most often affects male offspring. It is characterized by spontaneous musculoskeletal bleeding. Christmas disease or hemophilia B (factor IX deficiency) is less common than hemophilia A with similar symptoms. Factor IX is produced in the liver and is dependent on interaction with vitamin K in order to function properly. Deficiency in the vitamin can affect the clotting factor's performance as well as deficiency in the factor itself.
- Disseminated intravascular coagulation, also known as consumption coagulopathy, is not a disease in itself but a clinical emergency that occurs as a result of other diseases and conditions. This condition accelerates clotting, which ironically can result in hemorrhage when the clotting factors are exhausted.
- Thrombocytopenia, the most common cause of coagulation disorder, is characterized by reduced numbers of circulating platelets in the blood. This disease also includes idiopathic thrombocytopenia.
- Von Willebrand's disease, a hereditary disorder with prolonged bleeding time, is due to a clotting factor deficiency and impaired platelet function. It is the most common inherited coagulation disorder.
- Hypoprothrombinemia is a congenital deficiency of clotting factors that can lead to hemorrhage.
- Other coagulation disorders include factor XI deficiency (hemophilia C), and factor VII deficiency. Hemophilia C afflicts one in 100,000 people and is the second most common bleeding disorder among women. Factor VII is also called serum prothrombin conversion accelerator (SPCA) deficiency. One in 500,000 people may be afflicted with this disorder that is often diagnosed in newborns because of bleeding into the brain as a result of traumatic delivery.

Demographics

Hemophilia, or hemophilia A (factor VIII deficiency) affects about 20,000 Americans and one out of every 5000 males worldwide; Christmas disease, or hemophilia B, is less common than hemophilia A. Von Willebrand's disease affects both males and females and is often diagnosed in children. Thrombocytopenia is the most common coagulation disorder. Factor XI deficiency, or hemophilia C, afflicts one in 100,000 people and is the second most common bleeding disorder among women; it occurs more frequently among certain ethnic groups, with an incidence of about one in 10,000 among Ashkenazi Jews. A deficiency of factor VII, also called serum prothrombin conversion accelerator (SPCA) deficiency, affects one in 500,000 people and is often diagnosed in newborns.

Causes and symptoms

Some coagulation disorders present symptoms such as severe bruising. Others show no apparent symptoms but carry the threat of severe internal bleeding.

Hemophilia

Because of its hereditary nature, hemophilia A may be suspected before symptoms occur. Some signs of hemophilia A are numerous large, deep **bruises** and painful, swollen joints caused by internal bleeding. Individuals with hemophilia do not bleed faster, just longer. A person with mild hemophilia may first discover the disorder with prolonged bleeding following a surgical procedure or injury. If there is bleeding into the neck, head, or digestive tract, or bleeding from an injury, emergency measures may be required. Bleeding can be spontaneous, occurring with no obvious trauma.

Mild and severe hemophilia A are inherited through a complex genetic system that passes a recessive gene on the female chromosome. Women usually do not show signs of hemophilia but are carriers of the disease. Each male child of the carrier has a 50 percent chance of having hemophilia, and each female child has a 50 percent chance of passing the gene on.

Christmas disease

Christmas disease, or hemophilia B, is also hereditary but less common than hemophilia A. The severity of Christmas disease varies from mild to severe, although mild cases are more common. The severity depends on the degree of deficiency of factor IX. Hemophilia B symptoms are similar to those of hemophilia A, including numerous, large, and deep bruises and prolonged bleeding. The more dangerous symptoms are those that represent possible internal bleeding, such as swelling of joints or bleeding into internal organs upon trauma. Hemophilia most often occurs in families with a known history of the disease, but occasionally, new cases occur in families with no apparent history.

Disseminated intravascular coagulation

Disseminated intravascular coagulation (DIC) occurs when the malfunction of clotting factors causes platelets to form clots in small blood vessels throughout the body. This action leads to depletion of clotting factors and platelets, which are then not available at a site of injury where clotting is needed. When DIC occurs, the individual bleeds abnormally even though there is no history of coagulation abnormality. Symptoms may include minute spots of hemorrhage on the skin, and purple patches or hematomas caused by bleeding under the skin. Bleeding may occur at a surgical site or intravenous injection (IV) sites. Related symptoms include **vomiting**; seizures; shortness of breath; severe **pain** in the back, muscles, abdomen, or chest; and, if prolonged or uncorrected, shock and coma or death.

Not inherited and not a disease, DIC results from vascular complications during pregnancy or delivery, surgery, overwhelming infections, acute leukemia, metastatic **cancer**, extensive **burns**, liver disease, pancreatitis, trauma, snakebites, and other causes. As of 2004 it was not precisely understood why or how these various disorders lead to uncontrolled intravascular coagulation. What the underlying causes of DIC have in common is a dysfunction that involves proteins, platelets, or other clotting factors and processes. For example, uterine tissue can enter the mother's circulation during prolonged labor, introducing foreign proteins into the blood, or the venom of some exotic snakes can activate one of the clotting factors. Severe head trauma can expose blood to brain tissue. Regardless of the specific cause of DIC, the results are a malfunction of thrombin (an enzyme) and prothrombin (a glycoprotein), which activate the fibrinolytic system, releasing clotting factors in the blood. DIC can alternate from hemorrhage to thrombosis, and both can exist, which further complicates diagnosis and treatment.

Thrombocytopenia

Thrombocytopenia may be acquired or congenital (existing at birth). It represents a defective or decreased production of platelets. Symptoms include sudden onset of small bruises or spots of hemorrhage on the skin or bleeding into mucous membranes (such as nosebleeds). The disorder may also be evident as blood in vomit or stools, bleeding during surgery, or heavy menstrual flow.

Some patients show none of these symptoms but complain of fatigue and general weakness. There are several causes of thrombocytopenia, which is more commonly acquired as a result of another disorder. Common underlying disorders include leukemia, drug toxicity, or aplastic anemia, all of which lead to decreased or defective production of platelets in the bone marrow. Other diseases may destroy platelets outside the marrow. These include severe infection, disseminated intravascular coagulation, and cirrhosis of the liver. The idiopathic form most commonly occurs in children and is most likely the result of production of antibodies that cause destruction of platelets in the spleen and to a lesser extent the liver.

Von Willebrand's disease is caused by a defect in the von Willebrand clotting factor, often accompanied by a deficiency of factor VIII as well. It is a hereditary disorder that affects both males and females. In rare cases, it may be acquired. Symptoms include easy bruising, bleeding in small cuts that stops and starts, abnormal bleeding after surgery, and abnormally heavy menstrual bleeding. Nosebleeds and blood in the stool with a black, tarlike appearance are also signs of von Willebrand's disease.

Hypoprothrombinemia

Hypoprothrombinemia is an inherited or acquired deficiency in prothrombin, or factor II, a glycoprotein formed and stored in the liver. Prothrombin, under the right conditions, is converted to thrombin, which activates fibrin and begins the process of coagulation. Some individuals may show no symptoms, and others may suffer severe hemorrhaging. Easy bruising, profuse nosebleeds, postpartum hemorrhage, excessively prolonged or heavy menstrual bleeding, and postsurgical hemorrhage may also result. Acquired hypoprothrombinemia usually arises from a vitamin K deficiency caused by liver disease, newborn hemorrhagic disease, or other causes.

Other coagulation disorders

Factor XI deficiency, or hemophilia C, is a bleeding disorder that occurs among certain ethnic groups. Nearly 50 percent of individuals with this disorder experience no symptoms, but others may notice blood in their urine, nosebleeds, or bruising. Some factor XI deficiencies may result in bleeding long after an injury, and some women experience prolonged bleeding after childbirth. A deficiency of factor VII may cause varying levels of bleeding severity in those affected. Women may experience heavy menstrual bleeding, bleeding from the gums or nose, bleeding deep within the skin, and episodes of bleeding into the stomach, intestines, and urinary tract. Bleeding into the joints is rare but may also occur in some individuals.

When to call the doctor

Coagulation disorders are usually discovered when an injury or surgery initiates bleeding and the bleeding does not stop. Any signs of prolonged bleeding, even from a small cut, should be reported to a physician or emergency service. Bleeding under the skin (hematoma), which looks like a severe bruise, should also be reported and medical care sought. The sooner bleeding is controlled the better. A diagnostic work up is indicated to reveal any coagulopathy that exists, whether inherited or acquired.

Diagnosis

Diagnostic blood tests are performed in the clinical laboratory, including assays of the specific clotting factors, to help detect various coagulation disorders. Measured parameters are compared with known normal values to detect deficiencies or defects. Additionally, a choice of hundreds of diagnostic tests can be ordered by the physician to identify causative conditions, deficiencies, or diseases underlying the coagulopathy. Physicians also complete a medical history and physical examination. If acquired coagulation disorders are suspected, information such as prior or current diseases and medications are important to help determine the cause of the blood disorder. Each possible coagulopathy has specific criteria for diagnosis, including the following:

- Hemophilia A is diagnosed with laboratory tests that can detect the presence of clotting factor VIII, factor IX, and others, as well as the presence or absence of clotting factor inhibitors. Christmas disease involves an investigation of bleeding and clotting times, as well as determining factor IX deficiency. Other tests may include prothrombin time and thromboplastic generation. Gene carriers for both forms of hemophilia can be detected through DNA studies in conjunction with results from factor VIII assays.
- As of 2004 there was no one test or group of tests that can reliably diagnose DIC because it is a clinical event that occurs without warning, arising from another event such as surgery, childbirth, snakebite, and certain disease conditions. Diagnosis usually requires a number of laboratory tests that measure concentrations of platelets and fibrinogen in the blood along with measuring prothrombin time. Other supportive data include measuring levels of factors V and VIII, fibrinogen, hemoglobin, and platelets, any of which may be diminished or entirely depleted. Serial tests may also be

recommended, because a single coagulation parameter measured at any one moment may not reveal the rapidly progressive intravascular process.

- Tests for thrombocytopenia include coagulation tests that may reveal a decreased **platelet count** and prolonged bleeding time. Other coagulation factors may be measured. If these tests indicate that platelet destruction is causing the disorder, the physician may order a bone marrow biopsy.
- Von Willebrand's disease is diagnosed by ordering laboratory tests that reveal a prolonged bleeding time, absent or reduced levels of factor VIII, and a normal platelet count. Other tests are likely be done to confirm a diagnosis.
- Hypothrombinemia is diagnosed based on **family** history and the use of tests that measure vitamin K deficiency, deficiency of prothrombin, and measurements of clotting factors V, VII, IX, and X.
- Factor XI deficiency is determined by measuring the specific coagulation factor as well as other coagulation tests including prothrombin time and clotting time. It is diagnosed most often after injury-related bleeding.

Treatment

In mild coagulopathies, treatment may involve the use of drugs that stimulate the release of deficient clotting factors. In severe cases, bleeding may only stop if the clotting factor that is missing is replaced through infusion of human blood components containing concentrated amounts of specific clotting factors. These may be prepared in the form of fresh frozen plasma or cryoprecipitate. Cryoprecipitate was invented in 1965 to replace the need for whole plasma transfusions, which introduced more volume than needed and carried the threat of exposure to hepatitis or AIDs. More sensitive testing methods have virtually eliminated this risk. Commercial preparations of freeze-dried clotting factors have also made it possible for people to infuse themselves as directed by their physicians. This aspect of self-care made life easier for those with coagulation problems; in every other respect as of 2004, bleeding or coagulation disorders should not be self-managed. Comprehensive care addresses children's needs by providing various types of counseling to help deal with the psychosocial aspects of diseases such as von Willebrand's and hemophilia.

With mild bleeding episodes in persons afflicted with hemophilia A, infusions of a drug called desmopressin (DDAVP) may be administered. Severe bleeding episodes require transfusions of human blood clotting factors. Hemophiliacs are encouraged to receive physical therapy to help damaged joints and to **exercise** through non-contact **sports** such as swimming, bicycle riding, or walking, to avoid injury that may lead to bleeding.

Christmas disease is treated similarly to hemophilia A, with a mix of synthetic products and human blood products to provide coagulation factors as needed. Superficial **wounds** can be cleaned and bandaged. When hemophiliac children are to receive immunizations, parents should inform medical personnel in advance so that bleeding problems can be avoided. These children should probably not receive intramuscular injections.

When disseminated intravascular coagulation occurs, progression can be rapid, and treatment is complicated by the large variety of possible underlying causes. If at all possible, the physician first treats the underlying disorder. If the patient is not already bleeding, this supportive treatment may correct DIC. However, if bleeding is already occurring, a combination of transfused blood, platelets, fresh frozen plasma, or other blood products may be needed. Heparin, an anticoagulant, has been controversial in treating DIC, but it is often used as a last resort to stop hemorrhage. However, heparin has not proven useful in treating patients with DIC resulting from heat stroke, exotic snakebites, trauma, incompatible transfusions, and acute problems resulting from obstetrical complications.

Secondary acquired thrombocytopenia is best alleviated by treating the underlying cause or disorder. The specific treatment may depend on the underlying cause. Sometimes, corticosteroids or immune globulin may be given to improve platelet production.

Von Willebrand's disease is treated by several methods to reduce bleeding time and to replace factor VIII, which then replaces the von Willebrand factor. This may include infusion of cryoprecipitate or fresh frozen plasma. Desmopressin may also help raise levels of the von Willebrand factor.

Hypoprothrombinemia may be treated with concentrates of prothrombin. Vitamin K may also be given to stimulate coagulation, and in bleeding episodes, fresh plasma products may be transfused.

Factor XI (hemophilia C) deficiency is most often treated with plasma, since there are no commercially available concentrates of factor XI in the United States. Factor VII deficiency may be treated with prothrombin complex concentrates; as of 2004 factor VII is not licensed in the United States.

Prognosis

The prognosis for individuals with mild forms of coagulation disorders is normally good. Many people can lead normal lives and achieve normal life

KEY TERMS

Clotting factors—Substances in the blood, also known as coagulation factors, that act in sequence to stop bleeding by triggering the formation of a clot. Each clotting factor is designated with a Roman numeral I through XIII.

Coagulopathy—A disorder in which blood is either too slow or too quick to coagulate (clot).

Enzyme—A protein that catalyzes a biochemical reaction without changing its own structure or function.

Hemorrhage—Severe, massive bleeding that is difficult to control. The bleeding may be internal or external.

Heparin—An organic acid that occurs naturally in the body and prevents blood clots. Heparin is also made synthetically and can be given as an anticoagulant treatment.

Idiopathic—Refers to a disease or condition of unknown origin.

Metastatic—The term used to describe a secondary cancer, or one that has spread from one area of the body to another.

Thrombosis—The formation of a blood clot in a vein or artery that may obstruct local blood flow or may dislodge, travel downstream, and obstruct blood flow at a remote location. The clot or thrombus may lead to infarction, or death of tissue, due to a blocked blood supply.

expectancy. Without treatment of bleeding episodes, severe muscle and joint pain and eventually permanent damage can occur. Any incident that causes blood to collect in the head, neck, or digestive system can be very serious and requires immediate attention. DIC is an emergency situation that can be severe enough to cause stroke, coma, and death. The prognosis depends on early intervention and treatment of the underlying condition. Hemorrhage from a coagulation disorder, particularly into the brain or digestive track, can prove fatal.

Prevention

Inherited disorders cannot be prevented; they must be managed when detected. Acquired bleeding disorders are caused by a variety of conditions, some related to other diseases. There is no single prevention method although treatment of the underlying disorder or disease may prevent episodes of bleeding and subsequent coagulation problems. Episodes of bleeding can be prevented by avoiding injury. People who have hemophilia A or B and other bleeding disorders are advised to avoid activities and contact sports that can cause severe injury.

Parental concerns

Knowledge that a child has an inherited or acquired coagulation disorder that may lead to potentially dangerous bleeding episodes is of great concern to parents. Effective management of coagulation disorders by physicians can help the child to lead a relatively normal life with some cautions about avoiding injury. Counseling is available to help children handle the psychosocial aspects of living with a coagulation disorder.

See also Hemophilia.

Resources

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National Hemophilia Foundation. 116 West 32nd St., 11th Floor, New York, NY 10001. Web site: <www.hemophilia.org>.

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Coarctation of the aorta

Definition

Coarctation of the aorta (COA) is a congenital heart defect that develops in the fetus. It involves a constriction of the aorta, the main artery that delivers blood from the

left ventricle of the heart to the rest of the body. In a constriction or coarctation, the sides (walls) of the aorta press together abnormally, impeding the flow of blood. COA can produce symptoms of congestive heart failure or high blood pressure (**hypertension**) as early as the first week of life or may produce no symptoms until later in life.

Description

In the fetus, blood from the heart to the lungs is delivered into the aorta through a short blood vessel called the ductus arteriosis. This duct or shunt normally closes at birth or shortly after. In symptomatic children with COA, the descending aorta receives blood from the right side of the heart through the ductus arteriosus, an abnormal condition often accompanying other heart abnormalities such as a duct that does not close normally (**patent ductus arteriosus** or PDA), mitral valve defects, and other types of **congenital heart disease**. In asymptomatic children with COA, the descending aorta receives left ventricle blood through the ascending aorta; these children have fewer, if any, associated cardiac abnormalities. Approximately 10 percent of newborns with congenital heart disease have symptomatic coarctation of the aorta. About 85 percent of all children and adults with COA have a double aortic valve (bicuspid aortic valve) in the heart.

Blood normally leaves the heart by way of the left ventricle and is distributed to the body through the arteries. The aortic arch is the first artery to carry blood as it leaves the heart. Other arteries to the head and arms branch off the aortic arch. Constriction of the aorta, as in COA, produces resistance to the flow of blood, resulting in raising the blood pressure above the narrowing and reducing blood pressure below or downstream from the narrowing. High blood pressure (hypertension) affects parts of the body supplied by arteries that branch off the aortic arch above the narrowing. By contrast, most of the lower body does not receive enough blood supply. To compensate for this, the heart works harder, and blood pressure rises.

About half of all infants with COA are diagnosed within the first three months of life. Frequently, other congenital cardiac complications are also present. Thirty percent of infants with **Turner syndrome**, for example, also have coarctation. Evidence exists that at least some cases of coarctation may be inherited.

Demographics

Coarctation of the aorta is present in about 8 to 10 percent of infants born with other congenital heart defects, occurring approximately twice as many males as females.

Causes and symptoms

COA is congenital, that is it develops while the baby is in the womb and may appear in newborns along with other signs of congenital heart disease. Among the consequences of COA is an enlargement of the left ventricle (ventricular hypertrophy) in response to increased backpressure of the blood and the demand for more blood by the lower body. Symptoms in infants may include shortness of breath (dyspnea), difficulty in feeding, and poor weight gain. Children can also have no symptoms at all at birth (asymptomatic) and develop mild symptoms as older children. The older child with COA may display fatigue, shortness of breath, or a feeling of weakness or lameness in the legs.

When to call the doctor

COA is typically diagnosed within the first three months of life because of circulatory problems that produce symptoms. Some children have surgery, and some children are managed with drug therapy alone. Parents learn to recognize symptoms of high blood pressure or insufficient blood supply to the lower extremities. Medical attention is needed at the first sign of shortness of breath. Even when a child has had surgery or is on medication, the doctor should be contacted when any abnormal symptoms arise, such as shortness of breath, difficulty in feeding (with infants), and poor weight gain. Older children generally have fewer symptoms but may appear to be easily fatigued or to experience shortness of breath or weakness or lameness in their legs.

Diagnosis

Physical examination may reveal a pale child with some degree of dyspnea. On examination of the heart rhythm using a stethoscope, infants with coarctation of the aorta usually have an abnormal "gallop" heart rhythm, and 50 percent of children also have **heart murmurs**. Sometimes excessive arterial pulses can be seen in the arteries of the neck (carotid and suprasternal notch arteries), indicating increased pressure in these arteries, while the major artery of the legs (femoral artery) may have a weak pulse or none can be detected. The systolic pressure is higher in the arms than in the legs. Enlargement of the heart can be seen in x rays and congestion of the blood vessels in the lungs. Similar symptoms may be seen in older children and adults. A 10 mm Hg (mercury) pressure difference between the upper and lower extremities is diagnostic for coarctation of the aorta. For some individuals, the systolic pressure difference is observed only during **exercise**. Infants frequently have an abnormal electrocardiogram (ECG) that indicates that the right or both ventricles are enlarged, while in older children

the ECG may be normal or show that the left ventricle is enlarged. The site and the extent of coarctation may also be detected using color-flow Doppler ultrasonography (echocardiology).

Asymptomatic children may have a normal heart size or only slight enlargement. Differences in blood pressure between the arms and legs may be noted. Hypertension is less likely and, if noted, may be less marked than in symptomatic children with other heart defects. The bicuspid aortic valve is usually present. Color-flow Doppler studies may show a reduced blood flow below the coarctation.

Treatment

The goal of treatment in children is to reopen the ductus arteriosus and restore blood flow to the descending aorta. Congestive heart failure may be treated simultaneously with anticongestive medications. Drug therapy is used first to treat hypertension and heart failure in children and adults who have coarctation of the aorta. Surgery may be required for infants who have severe coarctation of the aorta and is usually recommended for those who have associated cardiac defects or those infants who do not respond to drug therapy. Surgery may also be indicated for infants whose early symptoms do not indicate immediate surgery but who develop severe hypertension during the first several months of life. Older children and adults are advised to avoid vigorous exercise prior to surgical correction of the coarctation. Surgery may involve resection of the coarctation segment or opening and patching the aorta where the coarctation occurred. Balloon angioplasty is sometimes performed on infants who are not ideal candidates for repair surgery because of higher risk; this procedure involves passing a catheter with an attached deflated balloon through the femoral artery in the groin and inflating the balloon to open the coarctation segment of the aorta. Recoarctation can occur in some individuals, even after corrective surgery. Recurrence is higher with balloon angioplasty than with repair surgery.

Prognosis

Approximately half of all infants diagnosed with coarctation of the aorta have no other cardiac defects and respond well to medical management, growing and developing normally. These infants are generally asymptomatic and will eventually outgrow the condition after several years of life. Although hypertension may increase for several months early in life, it eventually decreases as the circulatory system develops and vessels become larger.

Symptomatic children who have other heart defects generally respond well to repair surgery, and COA symptoms are reduced. The mortality rate for COA infants is about 5 percent. The average life span of children who have coarctation of the aorta is 34 years of age, reduced primarily due to complications and to the presence of other heart problems. The most common complications following repair surgery are postoperative renal failure and recoarctation. Complications in children who have not had surgery are hypertension, aortic rupture, intracranial bleeding, and congestive heart failure. Undetected or untreated COA can also lead to early adulthood death due to congestive heart failure, systemic hypertension, coronary artery disease, and aortic aneurysm. Women who have an uncorrected coarctation of the aorta have a mortality rate of 10 percent during pregnancy and a 90 percent rate of complications.

Parental concerns

Because congenital coarctation of the aorta is unpredicted, parents may be unprepared for the diagnosis and need careful, sensitive medical explanation by the pediatrician or surgeon. The birth of a child with this condition may raise parents' concerns about their child's ability to lead a normal life. If COA is detected, with or without the child's presenting symptoms, medical and surgical treatment usually corrects the condition and reduces symptoms. Over half of children overcome the condition and grow and develop normally without severe restrictions. Children who have other heart problems may have the repair surgery and other procedures to reduce symptoms and improve blood flow and with continued drug therapy and some restrictions on activity can lead relatively normal lives into adulthood.

See also Congenital heart disease; Patent ductus arteriosus.

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KEY TERMS

Aneurysm—A weakened area in the wall of a blood vessel which causes an outpouching or bulge. Aneurysms may be fatal if these weak areas burst, resulting in uncontrollable bleeding.

Angioplasty—A medical procedure in which a catheter, or thin tube, is threaded through blood vessels. The catheter is used to place a balloon or stent (a small metal rod) at a narrowed or blocked area and expand it mechanically.

Aorta—The main artery located above the heart that pumps oxygenated blood out into the body. The aorta is the largest artery in the body.

Dyspnea—Difficulty in breathing, usually associated with heart or lung diseases.

Electrocardiagram (ECG, EKG)—A record of the electrical activity of the heart, with each wave being labeled as P, Q, R, S, and T waves. It is often used in the diagnosis of cases of abnormal cardiac rhythm and myocardial damage.

Hypertension—Abnormally high arterial blood pressure, which if left untreated can lead to heart disease and stroke.

Patent ductus arteriosus—A congenital defect in which the temporary blood vessel connecting the left pulmonary artery to the aorta in the fetus doesn't close after birth.

Turner syndrome—A chromosome abnormality characterized by short stature and ovarian failure caused by an absent X chromosome. It occurs only in females.

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Cocaine *see* **Stimulant drugs**

Cochlear implants

Definition

A cochlear implant is a surgical treatment for hearing loss that works like an artificial human cochlea in the inner ear, helping to send sound from the ear to the brain. It is different from a hearing aid, which simply amplifies sound.

Purpose

A cochlear implant bypasses damaged hair cells in the child's cochlea and helps establish some degree of hearing by stimulating the hearing (auditory) nerve directly.

Description

Hearing loss is caused by a number of different problems that occur either in the auditory nerve or in parts of the middle or inner ear. The most common type of deafness is caused by damaged hair cells in the cochlea. The cochlea is a fluid-filled canal in the inner ear that is shaped like a snail shell. Inside are thousands of tiny hairs called cilia. As sound vibrates the fluid in the cochlea, the cilia move. This movement stimulates the auditory nerve and sends messages about sound to the brain. When these hair cells stop functioning, the auditory nerve is not stimulated, and the child cannot hear. Hair cells can be destroyed by many things, including infection, trauma, loud noise, aging, and birth defects.

The first piece of a cochlear implant is the microphone. It is usually worn behind the ear, and it picks up sound and sends it along a wire to a speech processor. The speech processor is usually worn in a small shoulder pouch, pocket, or on a belt. The processor boosts the sound, filters out background noise, and turns the sound into digital signals. Then it sends these digital signals to a transmitter worn behind the ear. A magnet holds the transmitter in place through its attraction to the receiver-stimulator, a part of the device that is surgically attached beneath the skin in the skull. The receiver picks up digital information forwarded by the transmitter and converts it into electrical impulses. These electrical impulses flow through electrodes contained in a narrow, flexible tube that has been threaded into the cochlea during surgery and stimulate the auditory nerve. The auditory nerve carries the electrical impulses to the brain, which interprets them as sound.

Despite the benefits that the implant appears to offer, some hearing specialists and members of the deaf community still believe that the benefits may not outweigh the risks and limitations of the device. Because

the device must be surgically implanted, it carries some surgical risk. Also, it is impossible to be certain how well any individual child will respond to the implant. After getting an implant, some people say they feel alienated from the deaf community, while at the same time not feeling fully a part of the hearing world.

The sounds heard through an implant are different from those sounds heard normally, and have been described as artificial or "robot-like." This is because the implant's limited number of electrodes cannot hope to match the complexity of a human's 15,000 hair cells. Cochlear implants are, however, becoming more advanced and providing even better sound resolution.

Surgical procedure

During the procedure, the surgeon makes an incision behind the ear and opens the mastoid bone (the ridge on the skull behind the ear) leading into the middle ear. The surgeon then places the receiver-stimulator into a well made in the bone and gently threads the electrodes into the cochlea. This operation takes between an hour-and-a-half and five hours. It is performed using general anesthesia.

Precautions

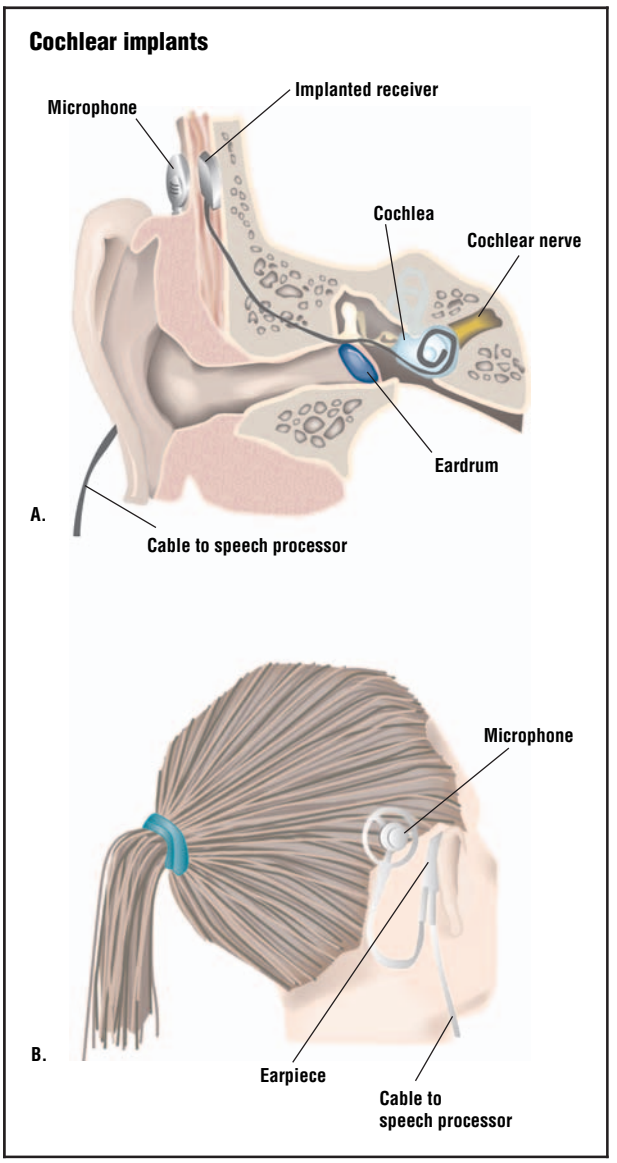
Because the implants are controversial, very expensive, and have uncertain results, the United States Food and Drug Administration (FDA) has limited the implants to people for whom the following is true:

- individuals who get no significant benefit from hearing aids
- individuals who are at least 12 months old
- individuals with severe to profound hearing loss

Preparation

Before a child gets an implant, specialists at an implant clinic conduct a careful evaluation, including extensive hearing tests to determine how well the child can hear.

First, candidates undergo a trial with a powerful hearing aid. If the hearing aid cannot improve hearing enough, a physician then performs a physical examination and orders a scan of the inner ear, because some patients with a scarred cochlea are not good candidates for cochlear implants. A doctor may also order a psychological exam to better understand the person's expectations. Patients and their families need to be highly motivated and have a realistic understanding of what an implant can and cannot do.



A cochlear implant has a microphone outside the ear that transmits sounds to an implanted receiver. In turn, the receiver transmits electrical impulses to the cochlea and cochlear nerve, which is stimulated in normal hearing. *(Illustration by GGS Information Services.)*

Aftercare

The child may remain in the hospital for a day or two after the surgery, although with improving technology and techniques some children may go home the same day. After about a month, the surgical **wounds** will have healed, and the child returns to the implant clinic to be fitted with the external parts of the device (the speech processor, microphone, and transmitter). A clinician tunes the speech processor and sets levels of stimulation for each electrode from soft to loud.

The child is then trained in how to interpret the sounds heard through the device. The length of the training varies

KEY TERMS

Cochlea—The hearing part of the inner ear. This snail-shaped structure contains fluid and thousands of microscopic hair cells tuned to various frequencies, in addition to the organ of Corti (the receptor for hearing).

Hair cells—Sensory receptors in the inner ear that transform sound vibrations into messages that travel to the brain.

Inner ear—The interior section of the ear, where sound vibrations and information about balance are translated into nerve impulses.

Middle ear—The cavity or space between the eardrum and the inner ear. It includes the eardrum, the three little bones (hammer, anvil, and stirrup) that transmit sound to the inner ear, and the eustachian tube, which connects the inner ear to the nasopharynx (the back of the nose).

from days to years, depending on how well the child can interpret the sounds heard. With the new approval for using cochlear implants in children as young as 12 months of age, the toddler may not be trained specifically to interpret the sounds in the same way an older child would. The specific therapy that is recommended is highly dependent on the age of the child.

Risks

As with all operations, there are a few risks of surgery. These include the following:

- dizziness
- facial paralysis (which is rare and usually temporary)
- infection at the incision site

Scientists are not sure about the long-term effects of electrical stimulation on the nervous system. It is also possible that the implant's internal components may be damaged by a blow to the head. This may cause the device to stop working. In general the failure rate of the implants is only 1 percent after one year.

Parental concerns

There is increasing debate about the use of cochlear implants in infants. This is considered by some to be desirable because, if the implantation is done before a child has begun to significantly acquire language, there is some evidence that the child may be able to develop at a pace similar to hearing children of the same age. Making a decision about whether or not a child, especially a very young one, should have a cochlear implant can be very difficult. The child's doctor may be able to provide parents with resources or put them in contact with other parents who have had to make the same decision whom they can consult.

See also Hearing impairment.

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Codeine *see* **Narcotic drugs**

Cognitive development

Definition

Cognitive development is the construction of thought processes, including remembering, problem

solving, and decision-making, from childhood through **adolescence** to adulthood.

Description

It was once believed that infants lacked the ability to think or form complex ideas and remained without cognition until they learned language. It is now known that babies are aware of their surroundings and interested in exploration from the time they are born. From birth, babies begin to actively learn. They gather, sort, and process information from around them, using the data to develop perception and thinking skills.

Cognitive development refers to how a person perceives, thinks, and gains understanding of his or her world through the interaction of genetic and learned factors. Among the areas of cognitive development are information processing, **intelligence**, reasoning, **language development**, and memory.

Historically, the cognitive development of children has been studied in a variety of ways. The oldest is through intelligence tests, such as the widely used Stanford Binet Intelligence Quotient (IQ) test first adopted for use in the United States by psychologist Lewis Terman (1877–1956) in 1916 from a French model pioneered in 1905. IQ scoring is based on the concept of "mental age," according to which the scores of a child of average intelligence match his or her age, while a gifted child's performance is comparable to that of an older child, and a slow learner's scores are similar to those of a younger child. IQ tests are widely used in the United States, but they have come under increasing criticism for defining intelligence too narrowly and for being biased with regard to race and gender.

In contrast to the emphasis placed on a child's native abilities by intelligence testing, learning theory grew out of work by behaviorist researchers such as John Watson (1878–1958) and B. F. Skinner (1904–1990), who argued that children are completely malleable. Learning theory focuses on the role of environmental factors in shaping the intelligence of children, especially on a child's ability to learn by having certain behaviors rewarded and others discouraged.

Piaget's theory of cognitive development

The most well-known and influential theory of cognitive development is that of French psychologist Jean Piaget (1896–1980). Piaget's theory, first published in 1952, grew out of decades of extensive observation of children, including his own, in their natural environments as opposed to the laboratory experiments of the behaviorists. Although Piaget was interested in how children reacted to their environment, he proposed a more active role for them than that suggested by learning theory. He envisioned a child's knowledge as composed of schemas, basic units of knowledge used to organize past experiences and serve as a basis for understanding new ones.

Schemas are continually being modified by two complementary processes that Piaget termed assimilation and accommodation. Assimilation refers to the process of taking in new information by incorporating it into an existing schema. In other words, people assimilate new experiences by relating them to things they already know. On the other hand, accommodation is what happens when the schema itself changes to accommodate new knowledge. According to Piaget, cognitive development involves an ongoing attempt to achieve a balance between assimilation and accommodation that he termed equilibration.

At the center of Piaget's theory is the principle that cognitive development occurs in a series of four distinct, universal stages, each characterized by increasingly sophisticated and abstract levels of thought. These stages always occur in the same order, and each builds on what was learned in the previous stage. They are as follows:

- Sensorimotor stage (infancy): In this period, which has six sub-stages, intelligence is demonstrated through motor activity without the use of symbols. Knowledge of the world is limited, but developing, because it is based on physical interactions and experiences. Children acquire object permanence at about seven months of age (memory). Physical development (mobility) allows the child to begin developing new intellectual abilities. Some symbolic (language) abilities are developed at the end of this stage.
- Pre-operational stage (toddlerhood and early childhood): In this period, which has two sub stages, intelligence is demonstrated through the use of symbols, language use matures, and memory and imagination are developed, but thinking is done in a non-logical, non-reversible manner. Egocentric thinking predominates.
- Concrete operational stage (elementary and early adolescence): In this stage, characterized by seven types of conservation (number, length, liquid, mass, weight, area, and volume), intelligence is demonstrated through logical and systematic manipulation of symbols related to concrete objects. Operational thinking develops (mental actions that are reversible). Egocentric thought diminishes.
- Formal operational stage (adolescence and adulthood): In this stage, intelligence is demonstrated through the logical use of symbols related to abstract concepts.

Early in the period there is a return to egocentric thought. Only 35 percent of high school graduates in industrialized countries obtain formal operations; many people do not think formally during adulthood.

The most significant alternative to the work of Piaget has been the information-processing approach, which uses the computer as a model to provide new insight into how the human mind receives, stores, retrieves, and uses information. Researchers using information-processing theory to study cognitive development in children have focused on areas such as the gradual improvements in children's ability to take in information and focus selectively on certain parts of it and their increasing attention spans and capacity for memory storage. For example, researchers have found that the superior memory skills of older children are due in part to memorization strategies, such as repeating items in order to memorize them or dividing them into categories.

Infancy

As soon as they are born, infants begin learning to use their senses to explore the world around them. Most newborns can focus on and follow moving objects, distinguish the pitch and volume of sound, see all colors and distinguish their hue and brightness, and start anticipating events, such as sucking at the sight of a nipple. By three months old, infants can recognize faces; imitate the facial expressions of others, such as smiling and frowning; and respond to familiar sounds.

At six months of age, babies are just beginning to understand how the world around them works. They imitate sounds, enjoy hearing their own voice, recognize parents, **fear** strangers, distinguish between animate and inanimate objects, and base distance on the size of an object. They also realize that if they drop an object, they can pick it up again. At four to seven months, babies can recognize their names.

By nine months, infants can imitate gestures and actions, experiment with the physical properties of objects, understand simple words such as "no," and understand that an object still exists even when they cannot see it. They also begin to test parental responses to their behavior, such as throwing food on the floor. They remember the reaction and test the parents again to see if they get the same reaction.

At 12 months of age, babies can follow a fast moving object; can speak two to fours words, including "mama" and "papa"; imitate animal sounds; associate names with objects; develop attachments to objects, such as a toy or blanket; and experience **separation anxiety** when away from their parents. By 18 months of age, babies are able to understand about 10–50 words; identify body parts; feel a sense of ownership by using the word "my" with certain people or objects; and can follow directions that involve two different tasks, such as picking up **toys** and putting them in a box.

Toddlerhood

Between 18 months to three years of age, toddlers have reached the "sensorimotor" stage of Piaget's theory of cognitive development that involves rudimentary thought. For instance, they understand the permanence of objects and people, visually follow the displacement of objects, and begin to use instruments and tools. Toddlers start to strive for more independence, which can present challenges to parents concerned for their **safety**. They also understand **discipline** and what behavior is appropriate and inappropriate, and they understand the concepts of words like "please" and "thank you."

Two-year-olds should be able to understand 100 to 150 words and start adding about ten new words per day. Toddlers also have a better understanding of emotions, such as love, trust, and fear. They begin to understand some of the ordinary aspects of everyday life, such as shopping for food, telling time, and being read to.

Preschool

Preschoolers, ages three to six, should be at the "preoperational" stage of Piaget's cognitive development theory, meaning they are using their imagery and memory skills. They should be conditioned to learning and memorizing, and their view of the world is normally very self-centered. Preschoolers usually have also developed their social interaction skills, such as playing and cooperating with other children their own age. It is normal for preschoolers to test the limits of their cognitive abilities, and they learn negative concepts and actions, such as talking back to adults, **lying**, and bullying. Other cognitive development in preschoolers are developing an increased attention span, learning to read, and developing structured routines, such as doing household chores.

School age

Younger school-age children, six to 12 years old, should be at the "concrete operations" stage of Piaget's cognitive development theory, characterized by the ability to use logical and coherent actions in thinking and solving problems. They understand the concepts of permanence and conservation by learning that volume, weight, and numbers may remain constant despite changes in outward appearance. These children should be able to build on past experiences, using them to explain why some things happen. Their attention span should increase with age, from

being able to focus on a task for about 15 minutes at age six to an hour by age nine.

Adolescents, ages 12 through 18, should be at the "formal operations" stage of Piaget's cognitive development theory. It is characterized by an increased independence for thinking through problems and situations. Adolescents should be able to understand pure abstractions, such as philosophy and higher math concepts. During this age, children should be able to learn and apply general information needed to adapt to specific situations. They should also be able to learn specific information and skills necessary for an occupation. A major component of the passage through adolescence is a cognitive transition. Compared to children, adolescents think in ways that are more advanced, more efficient, and generally more complex. This ability can be seen in five ways.

First, during adolescence individuals become better able than children to think about what is possible, instead of limiting their thought to what is real. Whereas children's thinking is oriented to the here and now—that is, to things and events that they can observe directly—adolescents are able to consider what they observe against a backdrop of what is possible; they can think hypothetically.

Second, during the passage into adolescence, individuals become better able to think about abstract ideas. For example, adolescents find it easier than children to comprehend the sorts of higher-order, abstract logic inherent in puns, proverbs, metaphors, and analogies. The adolescent's greater facility with abstract thinking also permits the application of advanced reasoning and logical processes to social and ideological matters. This is clearly seen in the adolescent's increased facility and interest in thinking about interpersonal relationships, politics, philosophy, religion, and morality.

Third, during adolescence individuals begin thinking more often about the process of thinking itself, or metacognition. As a result, adolescents may display increased introspection and self-consciousness. Although improvements in metacognitive abilities provide important intellectual advantages, one potentially negative byproduct of these advances is the tendency for adolescents to develop a sort of egocentrism, or intense preoccupation with the self.

A fourth change in cognition is that thinking tends to become multidimensional, rather than limited to a single issue. Whereas children tend to think about things one aspect at a time, adolescents can see things through more complicated lenses. Adolescents describe themselves and others in more differentiated and complicated terms and find it easier to look at problems from multiple perspectives. Being able to understand that people's personalities are not one-sided or that

Cognitive development

Age	Activity
One month	Watches person when spoken to.
Two months	Smiles at familiar person talking. Begins to follow moving person with eyes.
Four months	Shows interest in bottle, breast, familiar toy, or new surroundings.
Five months	Smiles at own image in mirror. Looks for fallen objects.
Six months	May stick out tongue in imitation. Laughs at peekaboo game. Vocalizes at mirror image. May act shy around strangers.
Seven months	Responds to own name. Tries to establish contact with a person by cough or other noise.
Eight months	Reaches for toys out of reach. Responds to "no."
Nine months	Shows likes and dislikes. May try to prevent face-washing or other activity that is disliked. Shows excitement and interest in foods or toys that are well-liked.
Ten months	Starts to understand some words. Waves bye-bye. Holds out arm or leg for dressing.
Eleven months	Repeats performance that is laughed at. Likes repetitive play. Shows interest in books.
Twelve months	May understand some "where is…?" questions. May kiss on request.
Fifteen months	Asks for objects by pointing. Starting to feed self. Negativism begins.
Eighteen months	Points to familiar objects when asked "where is…?" Mimics familiar adult activities. Know some body parts. Obeys two or three simple orders.
Two years	Names a few familiar objects. Draws with crayons. Obeys found simple orders. Participates in parallel play.
Two-and-a-half years	Names several common objects. Begins to take interest in sex organs. Gives full names. Helps to put things away. Peak of negativism.
Three years	Constantly asks questions. May count to 10. Begins to draw specific objects. Dresses and undresses doll. Participates in cooperative play. Talks about things that have happened.
Four years	May make up silly words and stories. Beginning to draw pictures that represent familiar things. Pretends to read and write. May recognize a few common words, such as own name.
Five years	Can recognize and reproduce many shapes, letters, and numbers. Tells long stories. Begins to understand the difference between real events and make-believe ones. Asks meaning of words.

SOURCE: *Miller-Keane Encyclopedia and Dictionary of Medicine, Nursing, and Allied Health, 5th ed.* and Child Development Institute, http://www.childdevelopmentinfo.com.

(Table by GGS Information Services.)

social situations can have different interpretations depending on one's point of view permits the adolescent to have far more sophisticated and complicated relationships with other people.

Finally, adolescents are more likely than children to see things as relative, rather than absolute. Children tend to see things in absolute terms—in black and white. Adolescents, in contrast, tend to see things as relative. They are more likely to question others' assertions and less likely to accept facts as absolute truths. This increase in relativism can be particularly exasperating to parents, who may feel that their adolescent children question everything just for the sake of argument. Difficulties often arise, for example, when adolescents begin seeing their parents' values as excessively relative.

Common problems

Cognitive impairment is the general loss or lack of development of cognitive abilities, particularly **autism** and learning disabilities. The National Institutes of Mental Health (NIMH) describes learning disabilities as a disorder that affects people's ability to either interpret what they see and hear or to link information from different parts of the brain. These limitations can show up in many ways, such as specific difficulties with spoken and written language, coordination, self-control, or attention. Such difficulties extend to schoolwork and can impede learning to read or write or to do math. A child who has a learning disability may have other conditions, such as hearing problems or serious emotional disturbance. However, learning disabilities are not caused by these conditions, nor are they caused by environmental influences such as cultural differences or inappropriate instruction.

Parental concerns

As of 2004 it is widely accepted that a child's intellectual ability is determined by a combination of heredity and environment. Thus, although a child's genetic inheritance is unchangeable, there are definite ways that parents can enhance their child's intellectual development through environmental factors. They can provide stimulating learning materials and experiences from an early age, read to and talk with their children, and help children explore the world around them. As children mature, parents can both challenge and support the child's talents. Although a supportive environment in early childhood provides a clear advantage for children, it is possible to make up for early losses in cognitive development if a supportive environment is provided at some later period, in contrast to early disruptions in physical development, which are often irreversible.

KEY TERMS

Autism—A developmental disability that appears early in life, in which normal brain development is disrupted and social and communication skills are retarded, sometimes severely.

Cognition—The act or process of knowing or perceiving.

Egocentric—Limited in outlook to things mainly relating to oneself or confined to one's own affairs or activities.

Learning disabilities—An impairment of the cognitive processes of understanding and using spoken and written language that results in difficulties with one or more academic skill sets (e.g., reading, writing, mathematics).

Metacognition—Awareness of the process of cognition.

Schemas—Fundamental core beliefs or assumptions that are part of the perceptual filter people use to view the world. Cognitive-behavioral therapy seeks to change maladaptive schemas.

Stanford-Binet intelligence scales—A device designed to measure somebody's intelligence, obtained through a series of aptitude tests concentrating on different aspects of intellectual functioning. An IQ score of 100 represents "average" intelligence.

When to call the doctor

If, by age three, a child has problems understanding simple directions or is perplexed when asked to do something simple, the parents or primary caregiver should consult a physician or pediatrician. The child may have a delay in cognitive development. Parents should also consult a healthcare professional if, after age three, their child's cognitive development appears to be significantly slower than their peers.

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Cohabitation *see* **Stepfamilies**

Cold sore

Definition

Cold sores are small red blisters, filled with clear fluid, that form on the lip and around the mouth. Rarely, they form on the roof of the mouth. Cold sores are caused by the **herpes simplex** virus type 1 (HSV-1), which lives inside nerve tissue. Despite their name, cold sores have nothing to do with colds. The herpes simplex virus type 1 should not be confused with the herpes simplex virus type 2 (HSV-2), which most often causes genital herpes.

Description

There are eight different kinds of human herpes viruses. Only two of these, herpes simplex types 1 and 2, can cause cold sores. It is commonly believed that herpes simplex virus type 1 infects above the waist and herpes simplex virus type 2 infects below the waist. This is not true. Both herpes virus type 1 and type 2 can cause herpes lesions on the lips or genitals, but recurrent cold sores are almost always type 1.

The sores can appear within days or weeks or even years following the first exposure to the virus. The first time symptoms appear they are usually more intense than later outbreaks. For example, some children experience more **pain** at the blister site or even flu-like symptoms, including swollen glands, **fever**, or **sore throat**. Medical names for cold sores include oral herpes, labial herpes, herpes labialis, and herpes febrilis.

Transmission

Herpes simplex virus is transmitted by infected body fluids (such as saliva) when they contact breaks in another person's skin or mucous membranes. Newborns may become infected during delivery through an infected birth canal. HSV-1 can be passed to children by parents, nurses, and caregivers who fail to practice careful hand washing. Children with **burns**, eczema, or **diaper rash** or those who are immunosuppressed are highly susceptible to the herpes virus.

VIRUS VERSUS BACTERIA Viruses behave differently than bacteria. While bacteria are independent and can reproduce on their own, viruses enter human cells and force them to make more viruses. The infected human cell dies and releases thousands of new viruses. The cell death and resulting tissue damage causes the cold sores. In addition, the herpes virus can infect a cell, and instead of making the cell produce new viruses, it hides inside the cell and waits. The herpes virus hides in the nervous system. This action is called latency. A latent

virus can wait inside the nervous system for days, months, or even years. At some future time, the virus "awakens" and causes the cell to produce thousands of new viruses that cause an active infection.

Latent and active infection is understood by considering the cold sore cycle. The first infection is the primary infection. The primary infection is controlled by the body's immune system and the sores heal. Between active infections, the virus is latent. At some point in the future, latent viruses become activated and once again cause sores or recurrent infections. Although it is unknown what triggers latent virus to activate, several conditions bring on infections. These include stress, illness, fever, fatigue, exposure to sunlight, **menstruation**, and diet.

Demographics

The herpes simplex virus is widespread in children. Thirty-five percent of five-year olds carry HSV-1 antibodies. A primary infection commonly occurs by the time a child reaches age five. These viral infections are more common in lower socioeconomic groups.

Nearly 80 percent of the general population carry the herpes simplex virus that causes cold sores, and 60 million have outbreaks once or more in a year. Most infants and children harbor the herpes virus before age ten. Interestingly enough, only 20 percent to 25 percent of those carrying the virus ever gets symptoms (break out in cold sores).

Causes and symptoms

While anyone can have the herpes virus infection, not everyone will show symptoms. The first symptoms of herpes occur within two to 20 days after contact with an infected person. Symptoms of the primary infection are usually more severe than those of recurrent infections. The primary infection can cause symptoms like other viral infections, including fatigue, **headache**, fever, and swollen lymph nodes in the neck.

Typically, 50 to 80 percent of children with oral herpes experience a prodrome (symptoms of oncoming disease) of pain, burning, **itching**, or tingling at the site where blisters will form. This prodrome stage may last a few hours or one to two days. The herpes infection prodrome occurs in both the primary infection and recurrent infections.

In 95 percent of the patients with cold sores, the blisters occur at the outer edge of the lips, which is called the vermillion border. Less often, blisters form on the nose, chin, or cheek. Following the prodrome, the disease process is rapid. First, small red bumps appear. These quickly form fluid-filled blisters.

Causes of cold sores in children

Infants are most likely to get a cold sore because someone with an active virus kisses them. The cause can even be a kiss from someone who did not have a visible sore but had the virus in his or her saliva. A baby may also get the herpes virus passing through the birth canal if the mother has genital herpes.

Once a child gets the virus, it stays in his body permanently, hiding in nerve cells near the ear. In some children, the virus lies dormant and never causes harm. In others, it periodically wakes up and triggers cold sores. Nobody knows what stirs the virus into action, but stress, fever, colds, and **sunburn** encourage outbreaks. Rich foods such as chocolate, **food allergies**, or hormonal changes may also cause outbreaks in children and adolescents.

Symptoms of cold sores in infants

The first symptoms of cold sores in infants are swollen gums and sore mouth. A few days later, there is a cluster of small blisters on or near the lips that turn into a shallow, painful sore. The breakout is often accompanied with fever and swollen lymph glands in the neck. In a few days the sore will crust and slowly disappear. The whole flare-up lasts about seven to ten days. The next time the infant has an outbreak the blister is the first symptom, not the swollen gums and painful mouth. If not treated, recurring outbreaks may last longer.

Stages of cold sores in children and adolescents

Cold sores, untreated, can last up to 14 days. Children often feel the tingling at the site before the sore appears. Parents should begin the treatment at the first sign of tingling or redness, which can reduce the time to two to four days. Beginning treatment after the blister appears can also significantly reduce the time and degree of pain. The following describes the stages of a cold sore:

- Day 1: Prodrome (tingle) stage: Before a blister appears, the child usually feels a tingling, itching, or burning sensation beneath the skin, usually around the mouth or the base of the nose. This is the best time to start treatment.
- Days 2 to 3: The blister or blisters appear. Children usually have one or several blisters on or around the moth, most often at the border of the lip and the skin on the face. Cold sores occasionally occur on the roof of the mouth.
- Day 4: Ulcer or weeping stage: The blister opens, revealing a reddish area. The child is most contagious and in the most pain at this point.

- Days 5 to 8: Crusting stage: The blisters dry up, and a crust forms which is yellow or brown in color. Children should be told not to pick at this crust.
- Days 9 to 14: Healing stage: A series of scabs form over the sore, each smaller than the previous one until the cold sore is healed.

When to call the doctor

The HSV-1 virus can cause ocular herpes, a serious eye infection affecting the cornea (the clear window) of the eye, which can threaten vision and needs immediate medical attention and treatment. When a baby or child has a cold sore, parents should do all they can to keep them from touching their eyes. If a painful sore appears on the child's eyelid, eye surface, or on the end of his nose, call the pediatrician right away. The child may need **antiviral drugs** to keep the infection from scarring the cornea. Ocular herpes can weaken vision and even cause blindness.

Some children have a serious primary (first episode) herpes infection called gingivostomatitis, which causes fever, swollen lymph glands, and several blisters inside the mouth and on the lips and tongue that may form large, open sores. These painful sores may last up to three weeks and can make eating and drinking difficult. Because of this problem, young children with gingivostomatitis are at risk for **dehydration**. Children with this condition should be seen by a doctor.

Most infants have protection for at least six months from HSV-1 by antibodies they received from their mothers. But if a newborn gets a cold sore, the pediatrician should be called right away.

If the immune system of children is compromised because of **cancer** treatment or **AIDS**, they could have more serious problems with the herpes virus. Parents should let the doctor know if these health conditions exist.

Children with a history of frequent herpes flare-ups who spend time skiing or on the beach should call a doctor for a prescription for starting oral anti-herpes medication (pills) before such outings and then using sunscreen while they are outside. These precautions can prevent most outbreaks.

Diagnosis

Cold sores are diagnosed by review of symptoms, physical examination, and history. The diagnosis is confirmed by various viral tests. A Tzanck or Papanicolau smear may be done. A positive smear cannot distinguish between varicella zoster virus and HSV-1, and a negative smear does not rule out HSV infection. Tissue culture provides a more reliable method of diagnosis. Cells killed by the herpes virus have a certain appearance under the microscope. Laboratory blood test looks for the virus or to confirm the presence of antibodies that fight the virus.

Approximately 85 percent of active herpes infections are without symptoms. When the symptoms do appear, they have the following sequence:

- burning, itching, or tingling at the site before the sore appears
- clusters of fluid-filled vesicles ulcerate, dry, and crust
- lesions dry and crust within seven to ten days
- usually one or two lesions present on the lips, tongue, gingival, or buccal mucosa
- puritis (itching) and pain

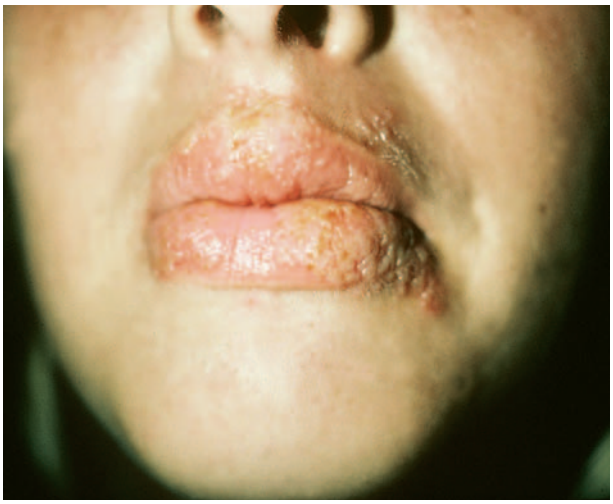
Treatment

There is no cure for herpes virus infections. Antiviral drugs are available that have some effect on lessening the symptoms and decreasing the length of herpes outbreaks. There is evidence that some may also prevent future outbreaks. These antiviral drugs work by interfering with the reproduction of the viruses and are most effective when taken as early in the infection as possible. For the best results, drug treatment should begin during the prodrome stage before blisters are visible. Depending on the length of the outbreak, drug treatment could continue for up to ten days.

Antiviral pills such as acyclovir (Zovirax), famciclovir, (Famvir), and valacyclovir (Valtrex) can cancel an outbreak and help prevent recurrent outbreaks. Acyclovir (Zovirax) is the drug of choice for herpes infection and can be given intravenously or taken by mouth. A liquid form for children is also available. Acyclovir is effective in treating both the primary infection and recurrent outbreaks. When taken by mouth, acyclovir reduces the frequency of herpes outbreaks.

Antiviral creams Zovirax and Denavir should be applied within the first 24 hours of feeling the tingling or discomfort, before the plaster erupts. The duration of the outbreak can be shortened by a day or two. Antiviral creams are not as effective as the pills.

During an outbreak, sores should be washed once or twice a day with warm, soapy water, and gently patted dry. Over-the-counter lip products that contain the chemical phenol (such as Blistex Medicated Lip Ointment) and numbing ointments (Anbesol) help to relieve cold sores. A bandage over the sores protects them and prevents spreading the virus to other sites on the lips or face.



Close-up view of a cold sore, caused by herpes simplex virus, on a patient's mouth. *(© Kenneth Greer/Visuals Unlimited.)*

Acetaminophen (Tylenol) or ibuprofen (Motrin, Advil) may be necessary to reduce pain and fever.

Alternative treatment

Cold sores in infants and children will go away on their own, but there are measures that will help the child feel better:

- To ease the pain, apply ice to the sore, or if the doctor approves, give the baby a mild pain reliever, such as ibuprofen or acetaminophen. (Do not give aspirin to a baby; it could trigger Reye's syndrome. This is a rare but potentially life-threatening illness.) Choices for pain relief include medication, ointments, or mouth rinses recommended by the pediatrician. Parents may also use Orageł, an ointment often used for **canker sores** or teething.
- Avoid giving the child spicy, salty, or sour foods, and foods with acid (oranges and grapefruits), which can irritate the open sore.
- Apply a water-based zinc ointment such as Desitin. It helps dry out the sore so it can heal faster.
- Extra **sleep** and plenty of liquids to drink can help.

Adolescents can learn to manage their own outbreaks. They can take the following steps:

- Use over-the-counter pain relievers, such as acetaminophen (Tylenol) or ibuprofen (Motrin, Advil).
- Avoid squeezing or picking the blister.
- Apply ice to ease the pain.
- Acidophilus pills may be helpful in treating cold sores. L-lysine is an amino acid widely advertised to treat cold sores. Conflicting medical opinions exist about its effectiveness.

Nutritional concerns

If children are prone to recurrent viral infections, parents should review their eating habits. The children should avoid foods and drinks that are proven suppressors of immunity (foods high in sugar, **caffeine**, and alcohol content) and have regular meals with plenty of fresh vegetables and fruits and whole-grain products. Some foods may be increased or reduced for specific types of viral infections. For herpes, foods containing the amino acid arginine (nuts and seeds) need to be reduced, and those containing the amino acid lysine (yogurt and cottage cheese) need to be increased. Immune system health in children can also benefit from a wide range of **vitamins** and **minerals** as nutritional supplements. Vitamin B complex and vitamins A, C, and E are the most important supplements.

Prognosis

Anyone can become infected with the herpes virus. Once infected, the virus remains latent in the body for life. It lives in nerve cells where the immune system cannot find it. The herpes virus lies dormant in the nerves of the face and is reactivated by sunburn, a recent viral illness (cold or flu), and periods of stress. Cold sores spread from person to person by direct skin-to-skin contact.

The highest risk for spreading the virus is the time period beginning with the blisters and ending with scab formation. However, infected people need not have visible blisters to spread the infection since the virus may be present in the saliva without obvious oral lesions.

Most children experience fewer than two recurrent outbreaks of cold sores each year; some have frequent outbreaks and others never experience outbreaks. Most blisters form in the same area each time and are triggered by the same reasons (such as stress, sun exposure, etc.).

Prevention

Cold sores are contagious. As of 2004 there were no herpes vaccines available, although herpes vaccines were in research and development. Avoiding close contact with people shedding the virus is the best way to prevent primary herpes. Several practices can reduce reoccurring cold sores and the spread of virus:

- Parents should teach children not to share drinks, food, or eating utensils, and not to exchange kisses with someone with mouth sores.

- When parents have an outbreak, they should avoid kissing their children (and other adults).
- Parents should be watchful of infected children and not allow them to share **toys** that may be put into the mouth. Toys that have been in the child's mouth should be disinfected before other children **play** with them.
- Parents should be especially careful with infants; they should not kiss the eyes or lips of a baby who is under six months old. The child's eyes and genitals are particularly susceptible to the herpes virus. They should keep the child from picking at a cold sore as much as possible.
- To keep from spreading the infection to other parts of the child's body or giving the virus to someone else, parents and caregivers should wash their hands and the child's hands often, especially after feedings and diaper changes.
- The child should be protected from the sun, since exposure to sunlight can trigger an outbreak. If the child is outside on a sunny day, generously apply sunscreen and lip balm that contains sun block before prolonged exposure to the sun. Sun block should be used during both winter and summer months, to help prevent cold cores. Wearing a hat with a large brim is also helpful.
- Parents should wear gloves when applying ointment to a child's sore. They should remove gloves and wash hands before and after changing diapers.
- Antibiotic ointments may be used to treat secondary bacterial infection of lesions. Parent should avoid using steroid creams or gels on a suspected herpes infection because these agents could make the infection worse.
- Parents should maintain good general health. A healthy diet, plenty of sleep, and **exercise** help to minimize the chance of getting a cold or the flu, which may bring on cold sores. Also, good general health keeps the immune system strong; this helps to keep the virus in check and prevents outbreaks.

Parental concerns

The child with a herpes infection is usually miserable and needs generous cuddling and comfort in spite of the infection. Parents of children who get cold sores should be aware of early symptoms and be ready to treat the sore in the first couple days. They should also keep in mind the prevention tips mentioned above. Parents should call the child's pediatrician if the child has not had a cold sore before, especially in the case of babies.

KEY TERMS

Antiviral drug—A medication that can destroy viruses and help treat illnesses caused by them.

Contagious—The movement of disease between people. All contagious disease is infectious, but not all infections are contagious.

Herpetic gingivostomatitis—A severe oral infection that affects children under five years of age; vesicles and ulcerations, edematous throat, enlarged painful cervical lymph nodes occur; chills, fever, malaise, bed breath, and drooling.

Latent virus—A nonactive virus that is in a dormant state within a cell. The herpes virus is latent in the nervous system.

Oral lesions—A single infected sore in the skin around the mouth or mucus membrane inside of the oral cavity.

Prodrome—Early symptoms that warn of the beginning of disease. For example, the herpes prodrome consists of pain, burning, tingling, or itching at a site before blisters are visible while the migraine prodrome consists of visual disturbances.

Recurrence—The return of an active infection following a period of latency.

Reye's syndrome—A serious, life-threatening illness in children, usually developing after a bout of flu or chickenpox, and often associated with the use of aspirin. Symptoms include uncontrollable vomiting, often with lethargy, memory loss, disorientation, or delirium. Swelling of the brain may cause seizures, coma, and in severe cases, death.

Secretion—A substance, such as saliva or mucus, that is produced and given off by a cell or a gland.

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Belinda Rowland, PhD

Colds *see* **Common cold**

Colic

Definition

Colic is defined as when a baby cries for longer than three hours every day for more than three days a week. It is the extreme end of normal crying behavior. The condition is harmless, even though it is distressing for parents or caregivers.

Description

Almost all babies go through a fussy period, but when crying lasts longer than about three hours a day and is not caused by a specific medical problem, it is considered colic. Pediatricians will tell parents that babies do not exhibit colic symptoms until around three weeks of age, but there are many parents who would disagree. The physician may also tell the parents that it will be at its worst around six weeks of age and then usually stops around three or four months of age. Some parents might disagree with that, too. It can be acknowledged as a relatively short period of time in a baby's life, but it seems like an eternity to the parents. It frequently, but not always, starts at the same time of day, and for most babies that is in the evening. The inconsolable crying begins suddenly; the legs may be drawn up, and the belly distended. The hands may be clenched. It seems as if it goes on forever and winds down when the baby is exhausted or when gas or stool is passed. Some babies continue crying for longer than three hours.

Demographics

Anywhere from 20 to 25 percent of babies cry enough to meet the definition of colic. There are approximately 4 million babies born every year in the United States, so that means almost a million babies have symptoms of colic.

Causes and symptoms

The baby with colic tends to be unusually sensitive to stimulation. Some babies experience greater discomfort from intestinal gas. Some cry from hunger. Some cry from overfeeding. **Fear**, frustration, or even excitement can lead to abdominal discomfort and colic. The situation may become a viscous cycle: the people caring for the baby become worried, anxious, or depressed, and the baby can sense their emotions and cries more. There are two theories regarding the cause of colic, and the first is that it is due to an immature nervous system. The majority of babies with colic can be classified with this condition to some degree. A small percentage of babies with colic may have milk **allergies**, reflux, and silent reflux. Formula changes or changes in diet for the breastfeeding mother can contribute to the problem. One recent study noted that the babies of mothers who smoke have a higher incidence of colic. The culprit is likely nicotine, which increases blood levels of a gut protein involved in digestion, according to Brown University epidemiologist Edmond Shenassa. This situation could result in painful cramping that makes babies cry.

When to call the doctor

Parents should call the pediatrician if they are concerned. A careful physical exam is prudent to insure the baby does not have a medical problem that needs attention. It is imperative not to misdiagnose a serious condition and call it colic. Should the behavior pattern of crying suddenly change and be associated with **fever**, **vomiting**, **diarrhea**, or other abnormal symptoms, parents should call the doctor immediately.

Diagnosis

Diagnosis occurs mostly by elimination. If the physical exam demonstrates nothing else is wrong, the pediatrician may diagnose colic by the parent's description of the crying.

Treatment

Parents should remember that colic is a benign condition, and the only treatment is through a matter of experimentation and observation. If a trigger for colic can be identified, that is a big start. Possible triggers include:

- Foods: Avoid stimulants such as **caffeine** and chocolate if breastfeeding.
- Formula: Switching formula works for some babies but is not at all helpful for others.
- Medicine: Medication that a breastfeeding mother takes may affect the baby.
- Feeding: If a bottle feeding takes less than 20 minutes, the hole in the nipple may be too large. Avoid overfeeding the infant or feeding too quickly.

Other strategies that can be tried include:

- movement and vibration
- using an infant swing
- rocking in a rocking chair
- going for a car ride
- holding the baby close in an upright position
- swaddling in a blanket

Nutritional concerns

The primary nutritional concerns are related to the breastfeeding mother's diet by avoiding the intake of stimulants. For those who are bottle feeding, a switch in the formula may be beneficial.

Prognosis

Colic is a benign condition. The infant outgrows it. Moreover, in spite of apparent abdominal **pain**, colicky infants eat well and gain weight normally.

Prevention

Very little can be done to prevent colic, other than trying to discover triggers that cause the baby to cry and to not smoke.

Parental concerns

It is natural for parents to be concerned when a baby cries, and their concern only heightens if it seems they can do nothing to stop the crying. Once a physical exam has been performed and medical causes have been ruled out, parents can accept the fact that the baby has colic and try to work with it the best way possible. They may want to take breaks from the baby by dividing childcare between them. A parent can be more loving to a baby when the parent has a chance to refresh.

See also Crying and fussing in an infant.

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Color blindness

Definition

Color blindness is an abnormal condition characterized by the inability to clearly distinguish different colors of the spectrum. The difficulties can range from mild to severe. It is a misleading term because people with color blindness are not blind. Rather, they tend to see colors in a limited range of hues; a rare few may not see colors at all.

Description

Normal color vision requires the use of specialized receptor cells called cones, which are located in the retina of the eye. There are three types of cones, red, blue, and green, which enable people to see a wide spectrum of colors. An abnormality, or deficiency, of any of the types of cones will result in abnormal color vision.

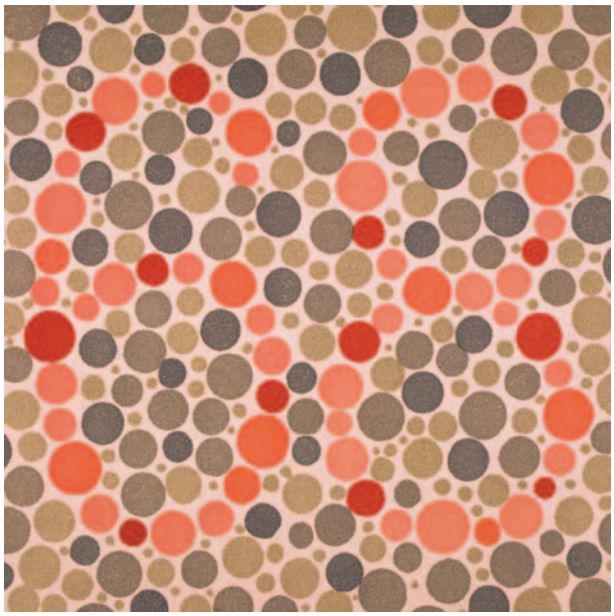
There are three basic variants of color blindness. Red/green color blindness is the most common deficiency, affecting 8 percent of Caucasian males and 0.5 percent of Caucasian females. The prevalence varies with culture.

Blue color blindness is an inability to distinguish both blue and yellow, which are seen as white or gray. It is quite rare and has equal prevalence in males and females. It is common for young children to have blue/green confusion that becomes less pronounced in adulthood. Blue color deficiency often appears in people who have physical disorders such as liver disease or **diabetes mellitus**.

A total inability to distinguish colors (achromatopsia) is exceedingly rare. These affected individuals view the world in shades of gray. They frequently have poor visual acuity and are extremely sensitive to light (photophobia), which causes them to squint in ordinary light.

Demographics

Researchers studying red/green color blindness in the United Kingdom reported an average prevalence of only 4.7 percent in one group. Only 1 percent of Eskimo males are color blind. Approximately 2.9 percent of boys from Saudi Arabia and 3.7 percent from India were found to have deficient color vision. Red/green color blindness may slightly increase an affected person's chances of contracting leprosy. Pre-term infants exhibit an increased prevalence of blue color blindness. Achromatopsia has a prevalence of about one in 33,000 in the United States and affects males and females equally.



Common test used to detect red-green color blindness. Those with normal color vision should see the number 68. *(© Lester V. Bergman/Corbis.)*

Causes and symptoms

Red/green and blue color blindness appear to be located on at least two different gene locations. The majority of affected individuals are males. Females are carriers but are not normally affected. This indicates that the X chromosome is one of the locations for color blindness. Male offspring of females who carry the altered gene have a 50 percent chance of being color-blind. The rare female that has red/green color blindness, or rarer still, blue color blindness, indicates there is an involvement of another gene. As of 2004, the location of this gene was not yet identified.

Achromatopsia, the complete inability to distinguish color, is an autosomal recessive disease of the retina. Thus, both parents have one copy of the altered gene but do not have the disease. Each of their children has a 25 percent chance of not having the gene, a 50 percent chance of having one altered gene (and, like the parents, being unaffected), and a 25 percent risk of having both the altered gene and the condition. In 1997, the achromatopsia gene was discovered to reside on chromosome 2.

The inability to correctly identify colors is the only sign of color blindness. It is important to note that people with red/green or blue varieties of color blindness use other cues such as color saturation and object shape or location to distinguish colors. They can often distinguish red or green if they can visually compare the colors. However, most have difficulty accurately identifying colors without any other references. Most people with any impairment in color vision learn colors, as do other young children. These individuals often reach **adolescence** before their visual deficiency is identified.

Color blindness is sometimes acquired. Chronic illnesses that can lead to color blindness include Alzheimer's disease, diabetes mellitus, glaucoma, leukemia, liver disease, chronic **alcoholism**, macular degeneration, multiple sclerosis, Parkinson's disease, **sickle cell anemia**, and retinitis pigmentosa. Accidents or strokes that damage the retina or affect particular areas of the brain eye can lead to color blindness. Some medications such as **antibiotics**, barbiturates, anti-tuberculosis drugs, high blood pressure medications, and several medications used to treat nervous disorders and psychological problems may cause color blindness. Industrial or environmental chemicals such as carbon monoxide, carbon disulfide, fertilizers, styrene, and some containing lead can cause loss of color vision. Occasionally, changes can occur in the affected person's capacity to see colors after age 60.

When to call the doctor

An ophthalmologist should be consulted at the time color blindness is first suspected.

Diagnosis

There are several tests available to identify problems associated with color vision. The most commonly used is the American Optical/Hardy, Rand, and Ritter Pseudoisochromatic Test. It is composed of several discs filled with colored dots of different sizes and colors. A person with normal color vision looking at a test item sees a number that is clearly located somewhere in the center of a circle of variously colored dots. A color-blind person is not able to distinguish the number.

The Ishihara Test is comprised of eight plates that are similar to the American Optical Pseudoisochromatic Test plates. The individual being tested looks for numbers among the various colored dots on each test plate. Some plates distinguish between red/green and blue color blindness. Individuals with normal color vision perceive one number. Those with red/green color deficiency see a different number. Those with blue color vision see yet a different number.

A third analytical tool is the Titmus II Vision Tester Color Perception Test. The subject looks into a stereoscopic machine. The test stimulus most often used in professional offices contains six different designs or numbers on a black background, framed in a yellow border. Titmus II can test one eye at a time. However, its value is limited because it can only identify red/green deficiencies and is not highly accurate.

Treatment

As of 2004 there is no treatment or cure for color blindness. Most color vision deficient persons compensate well for their abnormality and usually rely on color cues and details that are not consciously evident to persons with typical color vision.

Inherited color blindness cannot be prevented. In the case of some types of acquired color deficiency, if the cause of the problem is removed, the condition may improve with time. But for most people with acquired color blindness, the damage is usually permanent.

Prognosis

Color blindness that is inherited is present in both eyes and remains constant over an individual's entire life. Some cases of acquired color vision loss are not severe, may appear in only one eye, and last for only a short time. Other cases tend to become worse with time.

Prevention

There is no way to prevent genetic color blindness. There is no way to prevent acquired color blindness that is associated with Alzheimer's disease, diabetes mellitus, leukemia, liver disease, macular degeneration, multiple sclerosis, Parkinson's disease, sickle cell anemia, and retinitis pigmentosa.

Some forms of acquired color blindness may be prevented. Limiting use of alcohol and drugs such as antibiotics, barbiturates, anti-tuberculosis drugs, high blood pressure medications, and several medications used to treat nervous disorders and psychological problems to levels that are required for therapeutic benefit may limit acquired color blindness.

Parental concerns

Parents can inquire about other **family** members who have experienced color blindness. If such family members exist, parents can have their children tested for color perception at an early age. Screening for color perception is usually performed in grade school.

KEY TERMS

Achromatopsia—The inability to distinguish any colors.

Cones—Receptor cells, located in the retina of the eye, that allow the perception of colors.

Photophobia—An extreme sensitivity to light.

Retina—The inner, light-sensitive layer of the eye containing rods and cones. The retina transforms the image it receives into electrical signals that are sent to the brain via the optic nerve.

Rods—Photoreceptors, located in the retina of the eye, that are highly sensitive to low levels of light.

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Common cold

Definition

The common cold, also called a rhinovirus or coronavirus infection, is a viral infection of the upper respiratory system, including the nose, throat, sinuses, eustachian tubes, trachea, larynx, and bronchial tubes. Over 200 different viruses can cause a cold. Almost all colds clear up in less than two weeks without complications.

Description

Cold season in the United States begins in early autumn and extends through early spring. Although it is not true that getting wet or being in a draft causes a cold (a person has to come in contact with the virus to catch a cold), certain conditions may lead to increased susceptibility. These include:

- fatigue and overwork
- emotional stress
- poor nutrition
- smoking
- living or working in crowded conditions

Although most colds resolve on their own without complications, they are a leading cause of visits to the doctor and of time lost from work and school. Treating symptoms of the common cold has given rise in the United States to a multi-million dollar industry in over-the-counter medications.

Colds make the upper respiratory system less resistant to bacterial infection. Secondary bacterial infection may lead to middle ear infection (**otitis media**), **bronchitis**, **pneumonia**, sinus infection, or **strep throat**. People with chronic lung disease, **asthma**, diabetes, or a weakened immune system are more likely to develop these complications.

Transmission

People with colds are contagious during the first two to four days of the infection. Colds pass from person to person in several ways. When an infected person coughs, sneezes, or speaks, tiny fluid droplets containing the virus are expelled. If these are breathed in by other people, the virus may establish itself in their noses and airways.

Colds may also be passed through direct contact. If a person with a cold touches his runny nose or watery eyes, then shakes hands with another person, some of the virus is transferred to the uninfected person. If that person then touches his mouth, nose, or eyes, the virus is transferred to an environment where it can reproduce and cause a cold.

In addition, cold viruses can be spread through inanimate objects (door knobs, telephones, **toys**) that become contaminated with the virus. This is a common method of transmission in childcare centers. If a child with a cold touches his runny nose, then plays with a toy, some of the virus may be transferred to the toy. When another child plays with the toy a short time later, he may pick up some of the virus on his hands. The second child then touches his contaminated hands to his eyes, nose, or mouth and transfers some of the cold virus to himself.

Demographics

Colds are the most common illness to strike any part of the body, with over one billion colds in the United States each year. Anyone can get a cold, although preschool and grade school children catch them more frequently than adolescents and adults. Children average six to ten colds a year. In families with children in school, the number can be as high as 12 per year. Women, especially those aged 20 to 30 years old, have more colds than men, possibly because of their closer contact with children. Individuals older than 60 usually have fewer than one cold per year. Repeated exposure to viruses causing colds creates partial immunity.

Causes and symptoms

Colds are caused by more than 200 different viruses. The most common groups are rhinoviruses and coronaviruses. Different groups of viruses are more infectious at different seasons of the year, but knowing the exact virus causing the cold is not important in treatment.

Once acquired, the cold virus attaches itself to the lining of the nasal passages and sinuses. This condition causes the infected cells to release a chemical called histamine. Histamine increases the blood flow to the infected cells, causing swelling, congestion, and increased mucus production. Within one to three days the infected person begins to show cold symptoms.

The first cold symptoms are a tickle in the throat, runny nose, and sneezing. The initial discharge from the nose is clear and thin. Later it changes to a thick yellow

or greenish discharge. Most adults do not develop a **fever** when they catch a cold. Young children may develop a low fever of up to 102°F (38.9°C).

In addition to a runny nose and fever, signs of a cold include coughing, sneezing, nasal congestion, **headache**, muscle ache, chills, **sore throat**, hoarseness, watery eyes, tiredness, and lack of appetite. The **cough** that accompanies a cold is usually intermittent and dry.

Most people begin to feel better four to five days after their cold symptoms become noticeable. All symptoms are generally gone within ten days, except for a dry cough that may linger for up to three weeks.

When to call the doctor

Colds make people more susceptible to bacterial infections such as strep throat, middle ear infections, and sinus infections. People who have colds that do not begin to improve within a week or who experience chest **pain**, fever for more than a few days, difficulty breathing, bluish lips or fingernails, a cough that brings up greenish-yellow or grayish sputum, skin rash, swollen glands, or whitish spots on the tonsils or throat should consult a doctor to see to determine if they have acquired a secondary bacterial infection that needs to be treated with an antibiotic.

Children who have chronic lung disease, diabetes, or a weakened immune system—either from diseases such as **AIDS** or leukemia or as the result of medications, (corticosteroids, **chemotherapy** drugs)—should consult their doctor if they get a cold. Children with these health problems are more likely to get a secondary infection. For children with asthma, colds are a common trigger of asthma symptoms.

Diagnosis

Colds are diagnosed by observing a child's symptoms. There are no laboratory tests as of 2004 for detecting the cold virus. However, a doctor may do a **throat culture** or blood test to rule out a secondary infection.

Influenza is sometimes confused with a cold, but flu causes much more severe symptoms, as well as a fever. **Allergies** to molds or pollens also can make the nose run. Allergies are usually more persistent than the common cold. An allergist can do tests to determine if the cold-like symptoms are being caused by an allergic reaction. Also, some people get a runny nose when they go outside in winter and breathe cold air. This type of runny nose is not a symptom of a cold.

Treatment

There are no medicines that will cure the common cold. Given time, the body's immune system makes antibodies to fight the infection, and the cold is resolved without any intervention. **Antibiotics** are useless against a cold. However, there are many products that have been developed by pharmaceutical companies in the United States designed to relieve cold symptoms. These products usually contain **antihistamines**, **decongestants**, and/or pain relievers.

Antihistamines block the action of the chemical histamine that is produced when the cold virus invades the cells lining the nasal passages. Histamine increases blood flow and causes the cells to swell. Antihistamines are taken to relieve the symptoms of sneezing, runny nose, itchy eyes, and congestion. Side effects are dry mouth and drowsiness, especially with the first few doses. Antihistamines should not be taken by people who are driving or operating dangerous equipment. Some people have allergic reactions to antihistamines. Common over-the-counter antihistamines are Chlor-Trimeton, Dimetapp, Tavist, and Actifed. The generic name for two common antihistamines are chlorpheniramine and diphenhydramine.

Decongestants work to constrict the blood flow to the vessels in the nose. They can shrink the tissue, reduce congestion, and open inflamed nasal passages, making breathing easier. Decongestants can make people feel jittery or keep them from sleeping. They should not be used by people with heart disease, high blood pressure, or glaucoma. Some common decongestants are Neo-Synepherine, Novafed, and Sudafed. The generic names of common decongestants include phenylephrine, phenylpropanolamine, pseudoephedrine, and in nasal sprays naphazoline, oxymetazoline, and xylometazoline.

Many over-the-counter medications are combinations of both antihistamines and decongestants; an ache and pain reliever, such as **acetaminophen** (Datril, Tylenol, Panadol) or ibuprofen (Advil, Nuprin, Motrin, Medipren); and a cough suppressant (dextromethorphan). Common combination medications include Tylenol Cold and Flu, Triaminic, Sudafed Plus, and Tavist D. Aspirin should not be given to children with a cold because of its association with a risk of **Reye's syndrome**.

Nasal sprays and nose drops are other products promoted for reducing nasal congestion. These usually contain a decongestant, but the decongestant in the nasal preparations can act more quickly and strongly than ones found in pills or liquids because it is applied directly in the nose. Congestion returns after a few hours. People can become dependent on nasal sprays and nose drops. If used for a long time, users may suffer withdrawal symptoms when these products are discontinued. The label on the preparation should be checked for recommendations on length and frequency of use, since nasal sprays and nose drops should not be used for more than a few days.

People react differently to different cold medications and may find some more helpful than others. A medication may be effective initially then lose some of its effectiveness. Children sometimes react differently than adults. Over-the-counter cold remedies should not be given to infants without consulting a doctor first.

Care should be taken not to exceed the recommended dosages, especially when combination medications or nasal sprays are taken. These medicines do not shorten or cure a cold; at best they can only help a person feel more comfortable.

In addition to the optional use of over-the-counter cold remedies, there are some self-care steps that can be taken to ease discomfort. These include:

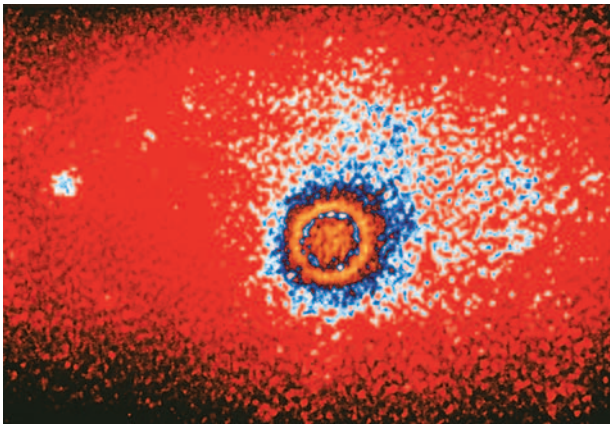
- drinking plenty of fluids, but avoiding acidic juices, which may irritate the throat
- gargling with warm salt water—made by adding one teaspoon of salt to 8 oz of water—for a sore throat
- avoiding second-hand smoke
- getting plenty of rest
- using a cool-mist room humidifier to ease congestion and sore throat
- rubbing Vaseline or other lubricant under the nose to prevent irritation from frequent nose blowing
- for babies too young to blow their noses, the mucus should be suctioned gently with an infant nasal aspirator (It may be necessary to soften the mucus first with a few drops of salt water.)

Alternative treatment

Alternative practitioners emphasize that people get colds because their immune systems are weak. They point out that everyone is exposed to cold viruses, but not everyone gets every cold. The difference seems to be in the ability of the immune system to fight infection. Prevention focuses on strengthening the immune system by eating a healthy diet low in sugars and high in fresh fruits and vegetables, practicing meditation or using other means to reduce stress, and getting regular moderate **exercise**.

Once cold symptoms appear, some naturopathic practitioners believe the symptoms should be allowed to run their course without interference. Others suggest the following:

- Aromatherapy remedy: Inhaling a steaming mixture of lemon oil, thyme oil, eucalyptus, and tea tree oil (*Melaleuca* spp.).
- Ayurvedic medicinal remedy: Gargling with a mixture of water, salt, and turmeric powder or astringents, such as alum, sumac, sage, and bayberry to ease a sore throat.
- Herbal remedies: Taking coneflower (*Echinacea* spp.) or goldenseal (*Hydrastis canadensis*). Other useful herbs to reduce symptoms are yarrow (*Achillea millefolium*), eyebright (*Euphrasia officinalis*), garlic (*Allium sativum*), and onions (*Allium cepa*).
- Homeopathic remedies: Microdoses of *Viscue album*, *Natrum muriaticum*, *Allium cepa*, or *Nux vomica*.
- Chinese traditional medicinal remedies: Taking yin chiao (sometimes transliterated as yinquiao) tablets that contain honeysuckle and forsythia when symptoms appear as well as using natural herb loquat syrup for cough and sinus congestion.
- Nutritional therapy: The use of zinc lozenges every two hours along with high doses of vitamin C as well as eliminating dairy products for the duration of the cold.



Rhinovirus, cause of the common cold, magnified. *(© 1991 CHSP. Custom Medical Stock Photo, Inc.)*

Prognosis

Given time, the body produces antibodies to cure itself of a cold. Most colds last a week to ten days. Most people start feeling better within four or five days. Occasionally a cold will lead to a secondary bacterial infection that causes strep throat, bronchitis, pneumonia, sinus infection, or a middle ear infection. These conditions usually clear up rapidly when treated with an antibiotic.

Prevention

It is not possible to prevent colds because the viruses that cause colds are common and highly infectious. However, there are some steps individuals can take to reduce their spread. These include:

KEY TERMS

Bronchial tubes—The major airways to the lungs and their main branches.

Coronavirus—A genus of viruses that cause respiratory diseases and gastroenteritis.

Corticosteroids—A group of hormones produced naturally by the adrenal gland or manufactured synthetically. They are often used to treat inflammation. Examples include cortisone and prednisone.

Eustachian tube—A thin tube between the middle ear and the pharnyx. Its purpose is to equalize pressure on either side of the ear drum.

Reye's syndrome—A serious, life-threatening illness in children, usually developing after a bout of flu or chickenpox, and often associated with the use of aspirin. Symptoms include uncontrollable vomiting, often with lethargy, memory loss, disorientation, or delirium. Swelling of the brain may cause seizures, coma, and in severe cases, death.

Rhinovirus—A group of small RNA viruses that infects the upper respiratory system and causes the common cold.

- washing hands well and frequently, especially after touching the nose or before handling food
- using instant hand sanitizers, which are antiseptics and not antibiotics
- covering the mouth and nose when sneezing
- disposing of used tissues properly
- avoiding close contact with someone who has a cold during the first two to four days of their infection
- not sharing food, eating utensils, or cups
- using paper towels rather than shared cloth towels
- avoiding crowded places where cold germs can spread
- eating a healthy diet and getting adequate sleep
- using a daycare facility with six or fewer children, to dramatically reduce germ contact

Parental concerns

The over-use of antibiotics has led to the development of antibiotic-resistant stains of bacteria. For these bacteria, antibiotics may be ineffective. Therefore, parents should not press the doctor to prescribe antibiotics when their children only have a cold.

Also, a parent should not give a child aspirin during a cold, because aspirin has been linked to the development of Reye's syndrome in children recovering from viral illnesses, especially influenza (flu) or **chickenpox**. Reye's syndrome can lead to permanent brain damage or death.

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Common variable immunodeficiency

Definition

Common variable **immunodeficiency** (CVID) is a disorder of the immune system characterized by low levels of specific immunoglobulins, antibodies produced by the immune system to fight infection or disease. In CVID, immunoglobulin G (IgG) antibodies, one of several classes of antibodies, are either absent or produced in lower than normal numbers. Children who have this disorder are subject to recurring infections and may not respond appropriately to immunization. In some children, levels of the four types of IgG may be out of balance, a condition that has been associated with autoimmune diseases.

Description

The function of the immune system is to respond to organisms and substances that invade the body, such as bacteria, viruses, fungi, parasites, and toxins, by producing antibodies against them. Antibodies are specific proteins (immunoglobulins) manufactured by the immune system to bind to corresponding molecules (antigens) on the cell surfaces of foreign organisms in an attempt to make them harmless. This antigen/antibody reaction is the body's way of protecting itself from invasion and possible illness. Immunodeficiency means that the immune system is deficient in one or more of its components and is unable to respond effectively to

disease-producing organisms that invade the body. IgG antibodies, the specific immunoglobulins absent or reduced in CVID, are targeted at bacterial organisms, viruses, and certain toxins.

Individuals with CVID will typically have frequent infections, especially repeat infections caused by the same organism. Recurring infections are an indication that the immune system is not responding normally and that immunity to reinfection has not developed. Surprisingly, people with CVID will usually have a normal number of B cells, the type of white blood cells (B-cell lymphocytes) that make antibodies to fight infection. However, the B cells will either be lacking one of the necessary IgG antibodies (IgG subclasses IgG1, IgG2, IgG3, and IgG4) on their surfaces or will have reduced amounts of one or more subclasses, making the B cells incapable of responding appropriately to microorganisms. Although the total IgG level may be normal, the imbalance in the types of IgG antibodies makes the B cells unprepared to fight all types of infection. The toxin associated with **tetanus**, for example, is attacked by IgG1 and IgG3 antibodies; reduced percentages of either immunoglobulin subclass on a child's cells will leave the child unprotected against that specific toxin. Similarly, frequent sinus infections may result from deficiencies of IgG2 and IgG3.

CVID may include deficiencies in other immunoglobulins as well, such as IgA and IgM deficiencies, although these deficiencies are more frequently associated with a group of other primary immunoglobulin deficiencies (agammaglobulinemia, **severe combined immunodeficiency**, and others). Other components of the immune system may be normal in CVID. T-cell lymphocytes, the type of white cells responsible for cellular immunity, are usually manufactured at normal levels in the same individuals who have CVID, although certain cell signal components may be lacking.

Autoimmune diseases such as autoimmune hemolytic anemia (AIHA), immune thrombocytopenia purpura (ITP), rheumatoid arthritis, autoimmune thyroiditis, and systemic lupus erythematosus are sometimes associated with CVID. These conditions develop in CVID as a result of the production of autoantibodies (antibodies directed against the body's own tissue). The term variable applies to this range of possible complications, which also includes gastrointestinal disorders as well as certain cancers, such as lymphomas and leukemias.

Demographics

Common variable immunodeficiency is believed to affect one in 50,000 to 200,000 individuals although it is not always diagnosed, and exact numbers of cases in the population cannot be accurately determined.

Causes and symptoms

The cause of common variable immunodeficiency was as of 2004 not known, although some forms seem to be inherited. The group of deficiencies is believed to be heterogeneous, that is, having widely varying characteristics among those affected.

CVID usually appears in children after the age of ten. The primary symptom is the presence of recurring infections that tend to be chronic rather than acute. Most children have had at least one episode of **pneumonia** caused by *Streptococcus pneumoniae*. Some children may also have frequent digestive disturbances and **diarrhea** that can lead to improper absorption of nutrients and malnourishment, occurring most commonly in IgA deficiency.

When to call the doctor

Young children and teenagers who are having recurrent infections, particularly infections of the same type such as frequent upper respiratory infection or chronic chest symptoms, ear infections, **sinusitis**, **asthma**, or pneumonia should be evaluated by a pediatrician or **family** practitioner.

Diagnosis

Children are typically diagnosed after age ten, but some immunoglobulin subclass deficiencies appear between ages one and three and are diagnosed after repeat cases of sinusitis, pneumonia, bacteremia, bronchiectasis, or diarrhea and malabsorption. A history of the child's illnesses and immunizations will be obtained, and the doctor will determine the child's general pattern of growth and development. Diagnostic testing may include routine blood tests such as a complete blood count (CBC) and differential (peripheral blood smear) to evaluate overall health and determine the type and number of red cells, white cells, and platelets in the blood. B lymphocytes and T lymphocytes may be quantified. An erythrocyte sedimentation rate (ESR) may be done to determine if inflammation is present. Blood chemistries may be performed to evaluate overall organ system functioning. If immunodeficiency is suspected, the primary diagnostic test that will distinguish common variable immunodeficiency from other types of immune system dysfunction is a reduced level of IgG immunoglobulins or IgG antibody subclasses, despite a relatively normal number of B cells. Serum immunoglobulin levels are measured in the clinical laboratory by a procedure called electrophoresis. This procedure both quantifies the amount of each antibody present and identifies the various classes and subclasses of antibodies. Deficiencies

may be noted in one class or subclass or in combinations of IgG, IgM and IgA antibodies. Genetic testing may be done to rule out other types of immunodeficiency disease.

Not all children who have repeat infections are immunodeficient. Doctors tell the difference by evaluating the child's history and development. A normal child who most likely does not need further examination or diagnostic testing will have the following characteristics:

- no history of deep infection at multiple sites, even though repeat upper respiratory infections or ear infections may occur
- overall normal growth and body functions
- generally good health and normal functioning between infections
- no known family history of immune system deficiencies

The type of organism causing repeat infection can be a clue to which immunoglobulins are deficient. Therefore, when infection is present in suspected cases of common variable immunodeficiency, it may be important to identify the bacteria or virus causing the illness. Diagnostic tests may include performing a culture on material from the nose, throat, a wound, blood, or urine of the affected child.

Treatment

As of 2004 no specific treatment cured common variable immunodeficiency; each child is treated according to the individual clinical condition, the symptoms presented, and the antibody subclasses shown to be absent or deficient. Treatment is aimed generally at boosting the body's immune response and preventing or controlling infections. Immune serum, obtained from donated blood that contains adequate levels of IgG antibodies, may sometimes be transfused as a source of antibodies to boost the immune response, even though it may not contain all the antibodies the child needs and may lack antibodies specific for some of the recurring infections. The preferred treatment is to give immunoglobulins intravenously (immunoglobulin intravenous therapy or IVIG) or intramuscularly (IMIG) if specific antibody deficiencies are found; this is not usually done to boost IgA levels, however, because of the possible presence of anti-IgA antibodies that could cause an unwanted reaction. Immunization against frequent infection can be achieved in some children by administering polysaccaride-protein conjugate vaccines shown to improve immune response in certain types of infection. **Antibiotics** are used routinely at the first sign of an infection to help eliminate infectious organisms.

Alternative treatment

Several nutritional supplements are reported to help build the immune system. These include garlic (contains the essential trace element germanium), essential fatty acids (abundant in flax seed oil, evening primrose oil, and fish oils), sea vegetables such as kelp, acidophilus to supply natural bacteria in the digestive tract, and **vitamins** A and C, both powerful antioxidants that improve immune function and increase resistance to infection. Zinc is another nutrient essential to immune system functioning. Green drinks made with young barley are believed to cleanse the blood and supply chlorophyll and nutrients for tissue repair. Alcohol, medications, drugs, coffee, and other **caffeine** drinks should be avoided. Stress is known to produce biochemicals that reduce white blood cell functioning; therefore, it is important for the child to get sufficient **sleep** and reduce stress to help improve immune system functioning. Therapeutic massage, **yoga**, and other types of stress reduction programs are available in most communities.

Prognosis

Regular medical observation, treatment of symptoms, and appropriate immune system boosting usually produces a good result in children with common variable immunodeficiency. In some children, delayed maturation of certain IgG subclasses will make the condition a temporary one that corrects itself as more typical levels of the IgG antibodies develop. In other children, prognosis is related to the immune system's ability to produce specific antibodies. Individuals with common variable immunodeficiency usually have a normal life span although a variety of complications can occur, including autoimmune, gastrointestinal, granulomatous, and malignant conditions as a result of progressive immune deficiency.

Prevention

The disorder cannot be prevented, but parents can take precautions to prevent the recurrent infections commonly associated with immunodeficiency. For example, practicing good hygiene and providing optimum **nutrition** are important for helping children avoid contact with infectious organisms and to develop resistance against them. Avoiding crowds and staying away from other children or relatives who have active infections is another important way to avoid challenges to the immune system.

Nutritional concerns

Maintaining a healthy immune system requires essential nutrients that can be provided through a good

KEY TERMS

Antibody—A special protein made by the body's immune system as a defense against foreign material (bacteria, viruses, etc.) that enters the body. It is uniquely designed to attack and neutralize the specific antigen that triggered the immune response.

Antigen—A substance (usually a protein) identified as foreign by the body's immune system, triggering the release of antibodies as part of the body's immune response.

Bacteria—Singular, bacterium; tiny, one-celled forms of life that cause many diseases and infections.

Culture—A test in which a sample of body fluid is placed on materials specially formulated to grow microorganisms. A culture is used to learn what type of bacterium is causing infection.

Immunization—A process or procedure that protects the body against an infectious disease by stimulating the production of antibodies. A vaccination is a type of immunization.

Immunoglobulin G (IgG)—Immunoglobulin type gamma, the most common type found in the blood and tissue fluids.

Vaccination—Another word for immunization.

Vaccine—A substance prepared from a weakened or killed microorganism which, when injected, helps the body to form antibodies that will prevent infection by the natural microorganism.

Virus—A small infectious agent consisting of a core of genetic material (DNA or RNA) surrounded by a shell of protein. A virus needs a living cell to reproduce.

diet and regular supplementation. A diet to improve immune system functioning includes fresh fruits and vegetables, as many eaten raw as possible to provide necessary enzymes; whole grain cereals, brown rice, and whole grain pasta for essential vitamins, **minerals**, and fiber; and non-meat sources of protein such as nuts, seeds, tofu, legumes (beans), and eggs. Fish, fowl, and lean meats can be consumed in small amounts. Sweets, especially if sweetened with refined sugars, should be reduced or avoided altogether. Vitamin supplements should include vitamins A, C, and E, which are all valuable parts of the body's defense system, helping to increase the production of healthy white blood cells and to fight infection.

Parental concerns

Parents are aware that school-age children and teenagers are in frequent contact with their peers in school and at **play**, and infections commonly spread. In this situation, when infection occurs frequently, it is important to remember that not all children or teens who have repeat infections are immunodeficient and that the pediatrician or family practitioner will have specific criteria and diagnostic tests to rule out common variable immunodeficiency.

See also Immunodeficiency syndromes; HIV infection and AIDS.

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L. Lee Culvert
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Communication skills

Definition

Communication is the process by which information is exchanged between individuals. It requires a shared understanding of symbol systems, such as language and mathematics.

Description

Communication is much more than words going from one person's mouth to another's ear. In addition to the words, messages are transferred by the tone and quality of voice, eye contact, physical closeness, visual cues, and overall body language.

Experts in child development agree that all babies develop skills for spoken and written language according to a specific developmental schedule, regardless of

which language the child is exposed to. Although the milestones follow one another in roughly the same sequence, there is significant variability from child to child on when the first word is spoken and the first sentence is composed.

Language employs symbols—words, gestures, or spoken sounds—to represent objects and ideas. Communication of language begins with spoken sounds combined with gestures, relying on two different types of skills. Children first learn to receive communications by listening to and understanding what they hear (supported by accompanying gestures); next, they experiment with expressing themselves through speaking and gesturing. Speech begins as repetitive syllables, followed by words, phrases, and sentences. Later, children learn to read and write. Many children begin speaking significantly earlier or later than the milestone dates. Parents should avoid attaching too much significance to deviations from the average. When a child's deviation from the average milestones of development causes the parents concern, a pediatrician or other professional may be contacted for advice.

Infancy

Touch can be a positive, encouraging communication technique from birth through adulthood. In infancy messages of love, security, and comfort are transferred through holding, cuddling, gentle stroking, and patting. Infants cannot understand the meaning of words they hear, but they can feel, interpret, and respond to gentle, loving supportive hands caring for them.

The development of language in infants follows this progression: crying, babbling, cooing, single words (mama and daddy), and simple names of some objects.

Toddlerhood

Toddlers one and two years of age experience the world through the physical senses. **Language development** for toddlers includes: using two-word combinations, taking turns speaking and listening, using the word no frequently, and using gestures to express needs and desires.

Preschool

Preschool children from three to five years of age develop further. They expand their word combinations and are able to speak in sentences, use correct grammatical patterns, use pronouns, articulate sounds clearly, and rapidly increase their working vocabulary. Preschool children may also understand words they do not use themselves.

School-age

School-age children and adolescents appreciate giving and receiving hugs as well as getting a reassuring pat on the back or a gentle hand resting on their hand. Asking permission from a child is recommended for any contact beyond a casual touch.

School-age children six to 11 years of age learn to communicate their own thoughts, as well as understand viewpoints of others. They can understand words with multiple meanings, however, words describing what they have not experienced are not thoroughly understood. School-age children have expanding vocabularies, enabling them to describe ideas, thoughts, and feelings. Their conversational skills refine.

Adolescents 12 years of age and older are able to communicate theories and explain them like adults would. Adolescents are able to talk about and understand most adult ideas.

Privacy is sometimes necessary for good communication. Space should be available for private conversations away from roommates, friends, certain **family** members, and visitors. This is especially important when communicating with adolescents. There may be sensitive topics adolescents will not want to discuss with parents present, or will only want to discuss with one parent.

Messages must be received for communication to be complete. Listening is an essential part of communication. Children and parents need to develop active listening skills to be effective listeners. As children enter the teen years, reflective communication skills are invaluable for them and for their parents. Active listening skills involve the following:

- paying attention without distractions and maintaining eye contact
- clarifying through reflecting what is heard (This involves using similar words to express back to the speaker what was understood about the content of the message.)
- showing empathy by identifying with the other's feelings
- listening with an open mind in order to understand another person's point of view

Children's receptive communication skills are more advanced than their verbal communication skills. They understand more than people often expect, based on their verbal skills. Effective parents talk with their children, not to them. To engage children in conversation, parents can ask open-ended questions and not judge what their children say.

KEY TERMS

Active listening—Listening with undivided attention and an open mind and being able to summarize the message accurately.

Communication—The act of transmitting and receiving information.

Empathy—A quality of the client-centered therapist, characterized by the therapists conveying appreciation and understanding of the client's point of view.

Visual communication

People communicate with eyes as well as ears. Communication occurs with cues of body language and facial expression. Eye contact is a communication connector. Making eye contact helps confirm attention and interest between the individuals communicating.

Some people are visual learners. They learn best when they can see or read instructions, demonstrations, diagrams, or information. Using various methods of presenting and shaping information increases understanding. Photographs, videotapes, dolls, computer programs, charts, and graphs can as effectively communicate information as written or spoken words can.

Tone of voice

Because infants' conception of verbal language is limited, their impressions are based on tone and quality of voice. Infants are able to discriminate parental voices from those of strangers and are more responsive to familiar voices. Soft, smooth voice quality is more comforting and soothing to infants than loud, startling, harsh voices. Infants can sense their caregiver's emotional state by the person's tone of voice. Awareness of infant's sensitivity to these messages is gained by watching their body language. They are relaxed when they hear a calm, happy caregiver and tense and rigid when they hear an angry, frustrated, or frightened one.

Verbal communication extends beyond words. Audible sounds transfer meaning. In addition, tone or attitude communicates sometimes a different meaning than the words used. Effective communicators do not send mixed messages. They say what they mean without sarcasm or equivocation.

Body language

Open body stance and positioning invite communication and interaction, whereas a closed body stance and positioning impedes communication. Using an open body posture improves communication with children. Both parents and children learn to read each other's body language.

Timing

Recognizing the right time to communicate is a skill. A distraught child whose parents have left for work is not ready to hear a story. The time will be more productive and the information better received if the child has a chance to make an emotional transition.

Common problems

Parents should strive to make words and intended meanings match when communication with their children. Children who are given mixed messages are confused and uncertain. Sometimes parents unwittingly attempt to control their children with double or mixed messages; doing so is unhealthy for their relationship to one another.

There are various kinds of spoken language problems, delays, and disabilities. In general, experts distinguish between those children who are slow in developing spoken language (**language delay**) and those who have difficulty achieving a milestone of spoken language (**language disorders**). Language disorders include **stuttering**; articulation disorders, such as substituting one sound for another (tandy for candy); omitting a sound (canny for candy); or distorting a sound (shlip for slip). Voice disorders include inappropriate pitch, volume, or quality. The causes for language problems have been linked to hearing, nerve, and muscle disorders; **head injury**; viral diseases; **mental retardation**; drug abuse; and cleft lips or palate.

Parental concerns

In the past, most parents, pediatricians, and educators recommended giving a child time to outgrow a difficulty with spoken language. From the late 1990s, research had shown that early speech and language disorders could lead to later difficulties in learning to read, write, and spell. Thus, many professionals recommended evaluation by a speech-language pathologist for toddlers who displayed language delay. However, not all speech-language specialists agree on early evaluation and therapy. Researchers have found that about two-thirds of children who were not talking at age two showed continued delays until age three, and one half were still behind the typical language development schedule at age four. But by kindergarten, only one-fourth of those children had not caught up with their peers.

See also Language development.

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Computed tomography

Definition

Computed tomography (CT), formerly referred to as computerized axial tomography (CAT), is a common diagnostic imaging procedure that uses **x rays** to generate images (slices) of the anatomy.

Purpose

Computed tomography (CT) is an x-ray imaging procedure used for a variety of clinical applications. CT is used for spine and head imaging, gastrointestinal imaging, vascular imaging (e.g., detection of blood clots), **cancer** staging and radiotherapy treatment planning, screening for cancers and heart disease, rapid imaging of trauma, imaging of musculoskeletal disorders, detection of signs of infectious disease, and guidance of certain interventional procedures (e.g., biopsies). CT is the preferred imaging exam for diagnosing several types of cancers, and along with the chest x ray, CT is the most commonly performed procedure for imaging the chest. CT is also used to perform noninvasive angiographic imaging to assess the large blood vessels.

CT may be performed on newborns, infants, children, and adolescents. In children, CT is most frequently used in the hospital emergency department to evaluate the effects of trauma, especially to the head, face, brain, and spine, and to diagnose or rule out **appendicitis** and other abdominal disorders because a scan can be completed in less than 20 seconds. Chest CT examinations are used to assess complications from infectious diseases, such as **pneumonia** and **tuberculosis**, inflammation of the airways, and birth defects. CT scans of the pelvic area are used to image ovarian cysts and tumors, bladder abnormalities, urinary tract stones, kidney disease, and bone disease. Head CT scans are used to examine the brain and sinuses. For children with cancer, CT is used to assist in treatment planning and to monitor cancer progression and response to treatment. For children requiring complex surgeries (e.g., brain, spine), CT is often used to produce images of the anatomy that help surgeons plan the surgery. Newer CT scanners, called multislice or multidetector CT, are used to rapidly image newborns to assess congenital heart defects.

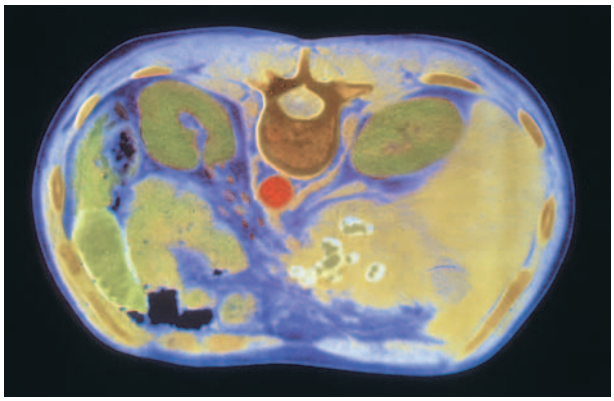
Description

CT is performed using a specialized scanner, an x-ray system, a patient table, and a computer workstation. The CT scanner is shaped like a large square with a hole in the center or round like a doughnut. X rays are produced in the form of a beam that rotates around the patient. During a CT scan, the patient table is moved through the center hole as x-ray beams pass through the patient's body. The x rays are converted into a series of black-and-white images, each of which represents a "slice" of the anatomy.

CT scans are conducted by a technologist with specialized training in x rays and CT imaging. During scanning, the technologist operates the CT scanner using a computer located in an adjacent room. Because movement during the scan can cause inaccurate images, the technologist instructs the patient via an intercom system to hold their breath and not move during the x rays. The scan itself may only take five to 15 minutes, but total examination time may be up to 30 minutes, since the patient must be prepped and positioned. Abdominal CT scans usually require that the child drink a solution that contains a dye, called oral contrast, that shows up on the CT images to help better define internal organs. For pelvic scans, contrast material may be delivered via the rectum. Some CT scans also require the injection of contrast material into the vein to help define the blood vessels and surrounding tissue.

The images from CT examination are called slices because they are acquired in very small (millimeter-size) sections of the body. The image slices are displayed on a computer monitor for viewing or printed as a film. A radiologist interprets the x-ray images produced during the CT examination. For emergency CT scans, images are interpreted immediately so that the child can be treated as soon as possible. For non-urgent outpatient CT scans, the radiologist interprets the images and sends a report to the referring physician within a few days.

For emergency situations, CT scans are performed in a hospital radiology department in conjunction with



False color computed tomography scan through the abdomen, showing the liver (larger yellow organ) and spleen (smaller yellow organ). The abdominal aorta is colored red and located above the spine and between the kidneys. *(Photo Researchers, Inc.)*

the emergency department. For non-urgent conditions, CT scans can be performed on an outpatient basis in a hospital radiology department or outpatient imaging center. In small hospitals or hospitals in rural areas, a CT scanner may not be permanently located in the hospital; rather, a mobile imaging service will be contracted to bring a specially designed trailer with CT equipment to the hospital on prescheduled days.

Precautions

CT scans expose the child to radiation, and overuse of CT scanning has received attention from organizations that regulate medical radiation exposure. Although no side effects have been linked to radiation exposure from CT imaging, the Food and Drug Administration has issued guidance to physicians regarding levels of radiation during pediatric CT examinations. New CT scanners have preset imaging features that allow scanning at the lowest radiation dose for the child's weight and age.

Oral contrast may be unpleasant tasting, although chocolate, vanilla, and fruit flavors may be available. Injected contrast can cause sensations of heat or cold through the body. Some children may have allergic reactions to the contrast material, although severe reactions are rare. Parents should inform CT staff if their child has ever had a reaction to any medication, contrast material, or anesthesia. Because contrast material may contain iodine, sensitivity to contrast material may occur if the child has other **allergies** to iodine or seafood, and CT staff should be informed if the child has such allergies. Also, because CT contrast material can affect kidney function, parents should notify CT staff if their child has kidney disease.

Preparation

Abdominal CT examinations usually require fasting for at least 12 hours before the scan. If the intestines will also be imaged, a laxative before the scan is required. Parents should alert CT staff if children are diabetic and taking insulin, since **hypoglycemia** can occur with missed meals.

Before the CT scan, the patient has to change into a hospital gown. When oral contrast is necessary, patients need to arrive at least one hour before the scan to drink the contrast solution. During the scan, the child is asked to lie on the CT table. Positioning devices, such as head cradles or knee rests, may be used. For very young or very active children, foam or Velcro restraints may be used to minimize movement during imaging. Or sedation may be used if children cannot remain still. After positioning the child, the technologist inserts an intravenous catheter to inject contrast material.

CT scanners may frighten young children, so prior to the imaging examination, the basic procedure should be explained to help reduce **fear**. Some radiology departments offer special patient education booklets for children that help explain imaging procedures.

Aftercare

No special aftercare is required following CT scans, unless sedation or general anesthesia was used during the scan. In these cases, children are required to remain in a supervised recovery area for an hour or more following the procedure to be monitored for reactions to anesthesia. If injected contrast material is used, some minor first aid (small bandage, **pain** relief) for the injection site may be necessary.

Risks

Radiation exposure is a risk during CT examinations. However, the radiation from a CT scan is usually less than that from regular x rays, and the benefits of the examination far outweigh the minor radiation dose received during the scan.

Some children may have reactions to anesthesia or sedation, including headaches, shivering, or **vomiting**. Rarely, severe anaphylactic reactions can occur that require emergency treatment.

Parental concerns

Younger children may be frightened of the CT scanner, and a parent or other **family** member may be required to be present in the scanning room. To help alleviate fear, taking the child into the CT room to see the

KEY TERMS

Anaphylaxis—Also called anaphylactic shock; a severe allergic reaction characterized by airway constriction, tissue swelling, and lowered blood pressure.

Radiologist—A medical doctor specially trained in radiology, the branch of medicine concerned with radioactive substances and their use for the diagnosis and treatment of disease.

equipment prior to the procedure may be helpful. To reduce risk of radiation exposure, anyone remaining in the scanning room during x-ray delivery will have to wear a lead apron on shield.

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American College of Radiology. 1891 Preston White Dr., Reston, VA 20191. Web site: <www.acr.org>.

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Jennifer Sisk, MA

Concussion

Definition

Concussion is a trauma-induced change in mental status, with confusion and amnesia, and with or without a brief loss of consciousness.

Description

A concussion occurs when the head hits or is hit by an object, or when the brain is jarred against the skull with sufficient force to cause temporary loss of function in the higher centers of the brain. The injured person may remain conscious or lose consciousness briefly and is disoriented for some minutes after the blow.

Demographics

According to the Centers for Disease Control and Prevention, approximately 300,000 people sustain mild to moderate sports-related brain injuries each year, most of them young men between 16 and 25 years of age.

The risk of concussion from football is extremely high, especially at the high school level. Studies show that approximately one in five players suffer concussion or more serious brain injury during their brief high-school careers. The rate at the collegiate level is approximately one in 20. Rates for hockey players are not known as certainly but are believed to be similar.

Causes and symptoms

Causes

Most concussions are caused by motor vehicle accidents and **sports injuries**. In motor vehicle accidents, concussion can occur without an actual blow to the head. Instead, concussion occurs because the skull suddenly decelerates or stops, which causes the brain to be jarred against the skull. Contact **sports**, especially football, hockey, and boxing, are among those most likely to lead to concussion. Other significant causes include falls, collisions, or blows due to bicycling, horseback riding, skiing, and soccer.

Concussion and lasting brain damage is an especially significant risk for boxers, since the goal of the sport is, in fact, to deliver a concussion to the opponent. For this reason, the American Academy of Neurology has called for a ban on boxing. Repeated concussions over months or years can cause cumulative **head injury**. The cumulative brain injuries suffered by most boxers can lead to permanent brain damage. Multiple blows to the head can cause punch-drunk syndrome or dementia pugilistica, as evidenced by Muhammad Ali, whose Parkinson's is a result of his career in the ring.

Young children are likely to suffer concussions from falls or collisions on the playground or around the home. **Child abuse** is, unfortunately, another common cause of concussion.

Symptoms

Symptoms of concussion include the following:

- headache
- disorientation as to time, date, or place
- confusion
- dizziness
- vacant stare or confused expression
- incoherent or incomprehensible speech
- lack of coordination or weakness
- amnesia for the events immediately preceding the blow
- nausea or **vomiting**
- double vision
- ringing in the ears

These symptoms may last from several minutes to several hours. More severe or longer-lasting symptoms may indicate more severe brain injury. The person with a concussion may or may not lose consciousness from the blow; if he does lose consciousness, it will be for several minutes at the most. Prolonged unconsciousness indicates more severe brain injury.

The severity of concussion is graded on a three-point scale, used as a basis for treatment decisions.

- Grade 1: no loss of consciousness, transient confusion, and other symptoms that resolve within 15 minutes
- Grade 2: no loss of consciousness, transient confusion, and other symptoms that require more than 15 minutes to resolve
- Grade 3: loss of consciousness for any period

Days or weeks after the accident, the person may show signs of the following:

- headache
- poor attention and concentration
- memory difficulties
- anxiety
- depression
- sleep disturbances
- light and noise intolerance

The occurrence of such symptoms is called "post-concussion syndrome."

When to call the doctor

A doctor should be consulted whenever a head injury causes any of the symptoms noted above.

Diagnosis

It is very important for those attending an individual with a concussion to pay close attention to the person's symptoms and progression immediately after the accident. The duration of unconsciousness and degree of confusion are very important indicators of the severity of the injury and help guide the diagnostic process and treatment decisions.

A doctor, nurse, or emergency medical technician may make an immediate **assessment** based on the severity of the symptoms; a **neurologic exam** of the pupils, coordination, and sensation, and brief tests of orientation, memory, and concentration. Those with very mild concussions may not need to be hospitalized or have expensive diagnostic tests. Questionable or more severe cases may require **computed tomography** scan (CT) or **magnetic resonance imaging** (MRI) scans to look for brain injury.

Treatment

The symptoms of concussion usually clear quickly and without lasting effect, if no further injury is sustained during the healing process. Guidelines for returning to sports activities are based on the severity of the concussion.

A grade 1 concussion can usually be treated with rest and continued observation alone. The person may return to sports activities that same day, but only after examination by a trained professional, and after all symptoms have completely resolved. If the person

sustains a second concussion of any severity that same day, he or she should not be allowed to continue contact sports until he or she has been symptom-free, during both rest and activity, for one week.

A person with a grade 2 concussion must discontinue sports activity for the day, should be evaluated by a trained professional, and should be observed closely throughout the day to make sure that all symptoms have completely cleared. Worsening of symptoms or continuation of any symptoms beyond one week indicates the need for a CT or MRI scan. Return to contact sports should only occur after one week with no symptoms, both at rest and during activity, and following examination by a physician. Following a second grade 2 concussion, the person should remain symptom-free for two weeks before resuming contact sports.

A person with a grade 3 concussion (involving any loss of consciousness, no matter how brief) should be examined by a medical professional either on the scene or in an emergency room. More severe symptoms may warrant a CT or MRI scan, along with a thorough neurological and physical exam. The person should be hospitalized if any abnormalities are found or if confusion persists. Prolonged unconsciousness and worsening symptoms require urgent neurosurgical evaluation or transfer to a trauma center. Following discharge from professional care, the person is closely monitored for neurological symptoms that may arise or worsen. If headaches or other symptoms worsen or last longer than one week, a CT or MRI scan should be performed. Contact sports are avoided for one week following unconsciousness of only seconds, and for two weeks for unconsciousness of a minute or more. A person receiving a second grade 3 concussion should avoid contact sports for at least a month after all symptoms have cleared and then engage in the sport only with the approval of a physician. If signs of brain swelling or bleeding are seen on a CT or MRI scan, the athlete should not return to the sport for the rest of the season, or even indefinitely.

For someone who has sustained a concussion of any severity, it is critically important that he or she avoid the possibility of another blow to the head until well after all symptoms have cleared to prevent second-impact syndrome. The guidelines above are designed to minimize the risk of this syndrome.

Prognosis

Concussion usually leaves no lasting neurological problems. Nonetheless, symptoms of **post-concussion syndrome** may last for weeks or even months.

Studies of concussion in contact sports have shown that the risk of sustaining a second concussion is even greater than it was for the first if the person continues to engage in the sport.

While concussion usually resolves on its own without lasting effect, it can set the stage for a much more serious condition. Second impact syndrome occurs when a person with a concussion, even a very mild one, suffers a second blow before fully recovering from the first. The brain swelling and increased intracranial pressure can lead to a potentially fatal result. More than 20 such cases have been reported since the syndrome was first described in 1984.

Prevention

Many cases of concussion can be prevented by using appropriate protective equipment. This includes seat belts and air bags in automobiles and helmets in all contact sports. Helmets should also be worn while bicycling, skiing, or horseback riding. Soccer players should avoid heading the ball when it is kicked at high velocity from close range. The surfaces immediately below and surrounding playground equipment should be covered with soft material, either sand or special matting.

Parental concerns

The value of high-contact sports such as boxing, football, or hockey should be weighed against the high risk of brain injury during a young person's participation in the sport. Steering a child's general enthusiasm for sports into activities less apt to produce head impacts may reduce the likelihood of brain injury. Children participating in any contact sport or activity that can cause brain injury should always wear a helmet.

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KEY TERMS

Amnesia—A general medical term for loss of memory that is not due to ordinary forgetfulness. Amnesia can be caused by head injuries, brain disease, or epilepsy, as well as by dissociation. Includes: 1) Anterograde amnesia: inability to retain the memory of events occurring after the time of the injury or disease which brought about the amnesic state. 2) Retrograde amnesia: inability to recall the memory of events which occurred prior to the time of the injury or disease which brought about the amnesic state.

Parkinson's disease—A slowly progressive disease that destroys nerve cells in the basal ganglia and thus causes loss of dopamine, a chemical that aids in transmission of nerve signals (neurotransmitter). Parkinson's is characterized by shaking in resting muscles, a stooping posture, slurred speech, muscular stiffness, and weakness.

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American Academy of Family Physicians. 11400 Tomahawk Creek Parkway, Leawood, KS 66211–2672. Web site: <www.aafp.org/>.

American Academy of Neurology. 1080 Montreal Avenue, St. Paul, MN 55116. Web site: <www.aan.com/>.

American Academy of Pediatrics. 141 Northwest Point Boulevard, Elk Grove Village, IL 60007–1098. Web site: <www.aap.org/default.htm>.

American College of Emergency Physicians. PO Box 619911, Dallas, TX 75261–9911. Web site: <www.acep.org/>.

Brain Injury Association. 105 North Alfred Street, Alexandria, VA 22314. Web site: <www.biausa.org/Sportsfs.htm>.

International Brain Injury Association. 1150 South Washington Street, Suite 210, Alexandria, VA 22314. Web site: <www.internationalbrain.org/>.

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L. Fleming Fallon, Jr., MD, DrPH

Condom

Definition

A condom is a device, usually made of latex, used to avoid pregnancy and/or **sexually transmitted diseases** such as gonorrhea, syphilis, and human **immunodeficiency** virus (HIV). Condoms are also known as prophylactics, as well as the popular slang term "rubbers." There are male and female versions of condoms.

Description

Condoms were originally used as a contraceptive to prevent unwanted pregnancies. In the early 2000s, however, condoms are just as important as a device for preventing the spread of sexually transmitted diseases (STDs), especially **HIV**, the virus that causes acquired immune deficiency syndrome (AIDS).

Male condoms have been in use in varied forms for at least three thousand years. Female condoms are relatively new, first being approved in Europe in 1992 and by the U.S. Food and Drug Administration (FDA) in the United States in 1993. An improved female condom became available in Europe in 2002. As of mid-2004, it

was under review by the FDA but had not been approved for use in the United States.

Male condoms, by far the most popular, consist of a disposable one-time-use tube-shaped piece of thin latex rubber or lambskin. The condom is unrolled over the erect penis before sexual intercourse. The tip of the condom usually has an open space to collect and hold the semen. The condom is a barrier that prevents sperm from entering a woman's uterus. It is also used in anal sex by males with females and other males to prevent transmission of STDs.

Male condoms are available in a wide variety of sizes, styles, textures, colors, and even flavors. Condoms are also recommended for use on a male when oral sex is being performed on him.

Condoms are about 85 percent effective in preventing pregnancies. That means that out of 100 females whose partners use condoms, 15 will still become pregnant during the first year of use, according to the nonprofit advocacy group Planned Parenthood. Unwanted pregnancies usually occur because the condom is not used properly or breaks during intercourse.

More protection against pregnancy is possible if a spermicide is used along with a condom. Spermicide is a pharmaceutical substance used to kill sperm, especially in conjunction with a birth-control device such as a condom or diaphragm. Spermicides come in foam, cream, gel, suppository, or as a thin film. The most common spermicide is called nonoxynol-9, and many condoms come with it already applied as a lubricant.

However, spermicides alone do not kill HIV or other sexually transmitted viruses and do not prevent the spread of HIV and other STDs. Also, nonoxynol-9 can irritate vaginal tissue and thus increase the risk of getting an STD. In anal sex, especially between two males, spermicides also can irritate the rectum, increasing the risk of getting HIV. Spermicides are specifically discouraged for use by gay or bisexual males for anal sex.

Latex condoms are also recommended over condoms made from other materials, especially lambskin, because they are thicker and stronger and have less risk of breakage during sex. Non-latex condoms do not prevent the spread of STDs, including HIV, and should not be used by gay or bisexual men or men who have HIV or other sexually transmitted diseases.

Condoms are available over-the-counter, meaning they do not require a prescription, and there are no age restrictions on purchasing condoms. They are available at a variety of locations, including drug stores, convenience stores, supermarkets, and **family** planning clinics. They are also available for purchase on the Internet.

How to use a male condom

PUTTING IT ON Many people, especially teens, are misinformed or uninformed on how to properly use a condom. In a 2001 study of youths ages 15 to 21, researchers found 33 to 50 percent of youth said it was important for the condom to fit tightly, leaving no air space at the tip, and that petroleum jelly, such as Vaseline, is a good lubricant. Another 20 percent said lambskin condoms offer better protection against HIV than latex condoms. All three beliefs are false.

For pleasure, ease, and effectiveness, both partners should know the correct way to put on and use a condom. Put the condom on before the penis touches the vulva, rectum, or mouth. Men leak fluids from their penises before and after ejaculation that can cause pregnancy and carry STDs. Use a condom only once and use a new one for each erection.

Condoms usually come rolled in a ring shape and are individually sealed in an aluminum foil, cardboard, or plastic pack. Carefully open the package to insure the condom does not tear. Do not use a condom if it is torn, brittle, stiff, or sticky.

To properly put on a male condom, follow these steps:

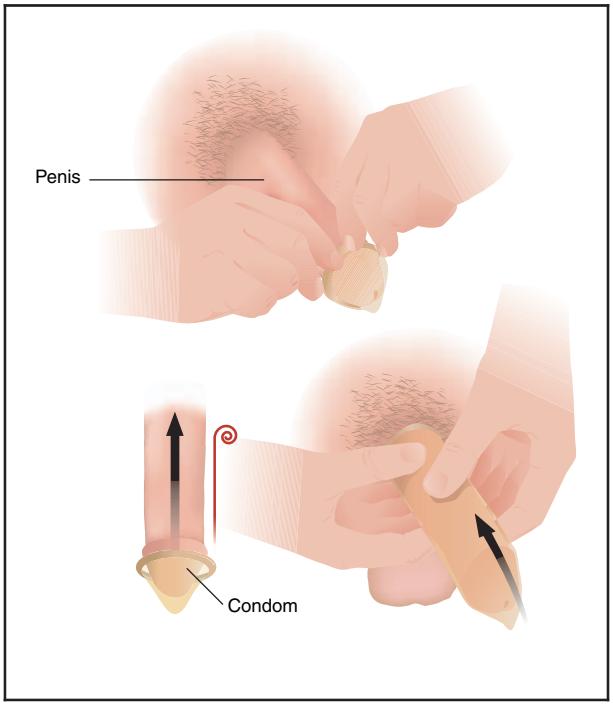
- Put several drops of lubricant inside the condom.
- Pull back the foreskin of an uncircumcised penis before putting on the condom.
- Place the rolled condom over the tip of the erect penis. Leave a half-inch (1 cm) of space at the tip to collect semen. Pinch the air out of the tip with one hand while placing it on the penis.
- Unroll the condom over the penis with the other hand, rolling it all the way down to the base of the penis. Smooth out any air bubbles since they can cause condoms to break.
- Lubricate the outside of the condom.

TAKING IT OFF To properly remove a male condom, follow these steps:

- Remove the penis from the vagina, rectum, or mouth soon after ejaculation and before the penis becomes soft.
- Hold the condom at the base of the penis while pulling out to prevent semen from leaking or spilling.
- Throw the condom away. It is not recommended that it be flushed down a toilet.

Female condom

The female condom is a seven-inch (17-cm) polyurethane pouch that fits into the vagina. It collects semen



A condom is most effective when it is placed on the penis correctly without trapping air between the penis and the condom. *(Illustration by Argosy, Inc.)*

before, during, and after ejaculation, keeping semen from entering the uterus, thus protecting against pregnancy. In one year of use, it is 79 percent effective in preventing pregnancies. It also reduces the risk of many STDs, including HIV.

There is a flexible ring at the closed end of the thin, soft pouch of the female condom. A slightly larger ring is at the open end. The ring at the closed end holds the condom in place in the vagina. The ring at the open end rests outside the vagina. When the condom is in place during sexual intercourse, there is no contact of the vagina and cervix with the skin of the penis or with secretions from the penis. It can be inserted up to eight hours before sex.

To insert the female condom, follow these steps:

- Find a comfortable position, such as standing with one foot on a chair, squatting with knees apart, or lying down with legs bent and knees apart.
- Hold the condom with the open end hanging down. Squeeze the inner ring with a thumb and middle finger.
- With the inner ring squeezed together, insert the ring into the vagina and push the inner ring and pouch into the vagina past the pubic bone.
- When inserted properly, the outer ring will hang down slightly outside the vagina.
- Adding a water-based lubricant to the inside of the condom or to the penis may be helpful.

There are no age restrictions and no prescription is needed to purchase female condoms. They can be used only once, and each costs $2.50 to $5.

School age

In a 2001 study by the Youth Risk Behavior Surveillance System, nearly 46 percent of American high school students reported they had had sexual intercourse at least once. Nearly 7 percent of students surveyed said they had engaged in their first sexual intercourse before age 13.

Of these sexually active students, 42 percent reported they did not use a condom the last time they had sex. Nationwide, male students (65.1%) were significantly more likely than female students (51.3%) to report condom use. This significant sex difference was identified for white and black students and students in grades 10, 11, and 12. Overall, black students (67.1%) were significantly more likely than white and Hispanic students (56.8% and 53.5%, respectively) to report condom use. This significant ethnic difference was identified for both female and male students.

Students in grades 9, 10, and 11 (67.5%, 60.1%, and 58.9%, respectively) were significantly more likely than students in grade 12 (49.3%) to report condom use, and students in grade 9 (67.5%) were significantly more likely than students in grade 11 (58.9%) to report condom use. The 2001 survey was published in the October 2002 issue of the *Journal of School Health*.

Common problems

The most common problems associated with condoms are breakage during use and improper knowledge on how to use condoms. These problems can lead to pregnancy and sexually transmitted diseases, especially HIV.

Parental concerns

Parents of adolescents often are concerned that distribution of condoms leads to increased sexual activity. However, a study of 4,100 high school students published in the June 2003 issue of the *American Journal of Public Health* found that students who had access at school to condoms and instructions on their proper use were no more likely to have sexual intercourse than students at schools without condom distribution programs.

When to call the doctor

It is not well known nor publicized, but having a condom break or leak while having sex is not necessarily

KEY TERMS

Antiretroviral drugs—Several classes of drugs that are used to treat HIV.

Contraceptive—A device or medication designed to prevent pregnancy by either suppressing ovulation, preventing sperm from passing through the cervix to fertilize an egg, or preventing implantation of a fertilized egg.

Diaphragm—The thin layer of muscle that separates the chest cavity containing the lungs and heart from the abdominal cavity containing the intestines and digestive organs. This term is also used for a dome-shaped device used to cover the back of a woman's vagina during intercourse in order to prevent pregnancy.

Ejaculation—The process by which semen (made up in part of prostatic fluid) is ejected by the erect penis.

Polyurethane—A type of synthetic plastic.

Prophylaxis—Protection against or prevention of a disease. Antibiotic prophylaxis is the use of antibiotics to prevent a possible infection.

Rubbers—A slang name for condoms.

Semen—The thick, whitish liquid released from the penis on ejaculation. It contains sperm and other secretions.

Spermicide—A substance that kills sperm. Also called a spermatocide.

Uncircumcised—Not having had the foreskin of the penis removed.

Uterus—The female reproductive organ that contains and nourishes a fetus from implantation until birth. Also called the womb.

Vulva—The external genital organs of a woman, including the outer and inner lips, clitoris, and opening of the vagina.

a health disaster, even if the condom wearer has HIV. The risk of HIV transmission during vaginal sex between a female and a male who has the virus is low, estimated at one-tenth to one-fifth of a percent, according to the Centers for Disease Control and Prevention. The risk for a single exposure through anal sex is estimated at one-tenth of a percent to 3 percent, according to the CDC.

Once exposed to the virus, the person can begin a therapy called post-exposure prophylaxis (PEP). The newly exposed person must begin four weeks of treatment with antiretroviral drugs, which usually prevents the virus from taking hold, according to the CDC. The treatment must begin within 72 hours after exposure but is more effective if begun within 24 hours of exposure. The exposed person should contact a physician immediately or go to the nearest hospital emergency room. The CDC does not have data on the effectiveness of PEP treatment on persons other than healthcare workers.

Pregnancy can also be prevented should a condom break or leak during sex. Emergency contraceptive pills (ECP), also called "morning-after pills," have been available since 1997. The pills have high levels of regular birth control hormones and are effective in preventing pregnancies following unprotected sex 75 to 94 percent of the time. They should be taken within 72 hours of unprotected sex.

As of August 2004, there were two ECPs available: Preven and Plan B. However, 11 brands of regular oral contraceptive pills in varying regimens can be effective in preventing post-sex pregnancies. Prescriptions are required for ECPs except in Washington State, where they can be dispensed without a prescription by selected pharmacies, doctors' offices, and hospital emergency rooms.

There are often financial, legal, and social barriers to persons under 18 getting ECPs. The group Advocates for Youth recommends young women always keep ECPs on hand (in advance) so they can be used as soon as possible following unprotected sex, such as when a condom breaks during sexual intercourse.

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Ken R. Wells

Conduct disorder

Definition

Conduct disorder (CD) is a behavioral and emotional disorder of childhood and **adolescence**. Children with conduct disorder act inappropriately, infringe on the rights of others, and violate the behavioral expectations of others.

Description

Children and adolescents with conduct disorder act out aggressively and express anger inappropriately. They engage in a variety of antisocial and destructive acts, including violence towards people and animals, destruction of property, **lying**, **stealing**, **truancy**, and **running away** from home. They often begin using and abusing drugs and alcohol and having sex at an early age. Irritability, temper **tantrums**, and low **self-esteem** are common personality traits of children with CD.

Demographics

Conduct disorder is present in approximately 6–16 percent of boys and 2–9 percent of girls under the age of 18. The incidence of CD increases with age. Girls tend to develop CD later in life (age 12 or older) than boys. Up to 40 percent of children with conduct disorder grow into adults with **antisocial personality disorder**.

Causes and symptoms

There are two subtypes of CD, one beginning in childhood (childhood onset) and the other in adolescence (adolescent onset). Research suggests that this disease may be caused by one or more of the following factors:

- poor parent-child relationships
- dysfunctional families
- inconsistent or inappropriate parenting habits
- substance abuse
- physical and/or emotional abuse
- poor relationships with other children
- cognitive problems leading to school failures
- brain damage
- biological defects

Difficulty in school is an early sign of potential conduct disorder problems. While the child's IQ may be in the normal range, he or she can have trouble with verbal and abstract reasoning skills and may lag behind classmates, and consequently feel as if he/she does not "fit in." The frustration and loss of self-esteem resulting from this academic and social inadequacy can trigger the development of CD.

A dysfunctional home environment can be another major contributor to CD. An emotionally, physically, or sexually abusive household member; a **family** history of antisocial personality disorder; or parental **alcoholism** or substance abuse can damage a child's self-perception and put him or her on a path toward negative or **aggressive behavior**. Other less obvious environmental factors can also play a part in the development of conduct disorder; several long-term studies have found an association between maternal **smoking** during pregnancy and the development of CD in offspring.

Other conditions that may cause or co-exist with conduct disorder include **head injury**, substance abuse disorder, major depressive disorder, and **attention deficit hyperactivity disorder** (ADHD). Fifty to seventy-five percent of children diagnosed with CD also have ADHD, a disorder characterized by a persistent pattern of inattention and/or hyperactivity.

CD is defined as a repetitive behavioral pattern of violating the rights of others or societal norms. Three of the following criteria or symptoms are required over the previous 12 months for a diagnosis of CD (one of the three must have occurred in the past six months):

- bullies, threatens, or intimidates others
- picks fights
- has used a dangerous weapon
- has been physically cruel to people
- has been physically cruel to animals
- has stolen while confronting a victim (for example, mugging or extortion)
- has forced someone into sexual activity
- has deliberately set a fire with the intention of causing damage
- has deliberately destroyed property of others
- has broken into someone else's house or car
- frequently lies to get something or to avoid obligations
- has stolen without confronting a victim or breaking and entering (e.g., shoplifting or forgery)
- stays out at night; breaks curfew (beginning before 13 years of age)
- has run away from home overnight at least twice (or once for a lengthy period)
- is often truant from school (beginning before 13 years of age)

When to call the doctor

When symptoms of conduct disorder are present, a child should be taken to his or her health care provider as soon as possible for evaluation and possible referral to a mental health care professional. If a child or teen diagnosed with conduct disorder reveals at any time that he/she has had recent thoughts of self-injury or **suicide**, or if he/she demonstrates behavior that compromises personal **safety** or the safety of others, professional assistance from a mental health care provider or care facility should be sought immediately.

Diagnosis

Conduct disorder may be diagnosed by a family physician or pediatrician, social worker, school counselor, psychiatrist, or psychologist. Diagnosis may require psychiatric expertise to rule out such conditions as **oppositional defiant disorder**, **bipolar disorder**, or ADHD. A comprehensive evaluation of the child should ideally include interviews with the child and parents, a full social and medical history, review of educational records, a cognitive evaluation, and a psychiatric exam.

One or more clinical inventories or scales may be used to assess the child for conduct disorder, including the Youth Self-Report, the Overt Aggression Scale (OAS), Behavioral **Assessment** System for Children (BASC), Child Behavior Checklist (CBCL), the Nisonger Child Behavior Rating Form (N-CBRF), Clinical Global Impressions scale (CGI), and Diagnostic Interview Schedule for Children (DISC). The tests are verbal and/or written and are administered in both hospital and outpatient settings.

Treatment

Treating conduct disorder requires an approach that addresses both the child and his/her environment. Behavioral therapy and psychotherapy can help a child with CD to control his/her anger and develop new coping techniques. Social skills training can help a child improve his/her relationship with peers.

Family group therapy may also be effective in some cases. Parents should be counseled on how to set appropriate limits with their child and be consistent and realistic when disciplining. A parental skills training program may be recommended. If an abusive home life is at the root of the conduct problem, every effort should be made to move the child into a more supportive environment.

For children with coexisting ADHD, substance abuse, depression, **anxiety**, or **learning disorders**, treating these conditions first is preferred, and may result in a significant improvement in behavior. In all cases of CD, treatment should begin when symptoms first appear. Several studies have shown **methylphenidate** (Ritalin) to be a useful drug for both ADHD and CD in some children.

When aggressive behavior is severe, mood stabilizing medication, including lithium (Cibalith-S, Eskalith, Lithobid, Lithonate, Lithotabs), and carbamazepine (Tegretol, Carbatrol, Epitol) may be an appropriate option for treating the aggressive symptoms. However, placing the child into a structured setting or treatment program such as a psychiatric hospital may be just as beneficial for easing aggression as medication.

Prognosis

Follow-up studies of conduct-disordered children have shown a high incidence of antisocial personality disorder, affective illnesses, and chronic criminal

KEY TERMS

Attention deficit hyperactivity disorder (ADHD)—A condition in which a person (usually a child) has an unusually high activity level and a short attention span. People with the disorder may act impulsively and may have learning and behavioral problems.

Major depressive disorder—A mood disorder characterized by profound feelings of sadness or despair.

behavior in adulthood. However, proper treatment of coexisting disorders, early identification and intervention, and long-term support may improve the outlook significantly.

Conduct disorder that first occurs in adolescence is thought to have a statistically better prognosis than childhood-onset conduct disorder. Adolescents with CD tend to have better relationships with their peers and are less likely to develop antisocial personality disorder in adulthood than those with childhood-onset CD. There is also less of a gender gap in adolescent-onset conduct disorder, as girls approach boys in CD incidence. Childhood-onset CD is much more common among boys.

Prevention

A supportive, nurturing, and structured home environment is believed to be the best defense against conduct disorder. Children with learning disabilities and/or difficulties in school should get immediate and appropriate academic assistance. Addressing these problems when they first appear helps to prevent the frustration and low self-esteem that may lead to CD later on.

Parental concerns

A child with conduct disorder can have a tremendous impact on the home environment and on the physical and emotional welfare of siblings and others sharing the household. While seeking help for their child with CD, parents must remain sensitive to the needs of their other children and adjust household routines accordingly. This may mean avoiding leaving siblings alone together, getting assistance with childcare, or even seeking residential or hospital treatment for the conduct disordered child if the safety and well-being of other family members is in jeopardy.

See also Aggression; Oppositional defiant disorder.

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Paula Ford-Martin

Congenital adrenal hyperplasia

Definition

Congenital adrenal hyperplasia (CAH) is a genetic disorder characterized by a deficiency in the hormones cortisol and aldosterone and an over-production of the hormone androgen. CAH is present at birth and affects the sexual development of the child.

Description

Congenital adrenal hyperplasia (CAH) is a form of adrenal insufficiency in which 21-hydroxylase, the enzyme that produces two important adrenal steroid hormones, cortisol and aldosterone, is deficient. Because cortisol production is impeded, the adrenal gland overproduces androgens (male steroid hormones). CAH affects both females and males. Females with CAH are born with an enlarged clitoris and normal internal reproductive tract structures. Males have normal genitals at

birth. CAH causes abnormal growth for both sexes; those affected will be tall as children but short as adults because of early bone maturation. Females develop male characteristics, and males experience premature sexual development.

In its most severe form, called salt-wasting (or salt-losing) CAH, where there is a total or near total deficiency of the 21-hydroxylase enzyme, a life-threatening adrenal crisis can occur if the disorder is untreated. Adrenal crisis can cause **dehydration**, shock, and death within 14 days of birth. There is also a milder form of CAH in which children have partial 21-hydroxylase enzyme deficiencies (simple virilizing form). Another type of CAH is characterized by only a slight deficiency in production of the 21-hydroxylase enzyme (nonclassic or late-onset form), in which symptoms occurs later in childhood or during young **adolescence**.

CAH is also called adrenogenital syndrome or 21-hydroxylase deficiency.

Demographics

CAH, a genetic disorder, is the most common adrenal gland disorder in infants and children, occurring in one in 10,000 total births worldwide.

Causes and symptoms

CAH is an inherited recessive disorder, which means that a child must inherit one copy of the defective gene from each parent who is a carrier; when two carriers have children, each pregnancy carries a 25 percent risk of producing an affected child. CAH is related to the deficiency of 21-hydroxylase, an enzyme that is required to transform cholesterol into cortisol. The 21-hydroxylase gene is made by a gene located on the short arm of chromosome 6. This gene is located in an area of the chromosome that contains many other important genes whose products control immune function. Various mutations of the 21-hydroxylase gene result in various degrees of CAH (salt-losing form, simple-virilizing form, and the nonclassic form). When 21-hydroxylase is deficient, this leads to a hyperfunction and increased size (hyperplasia) of the adrenals.

In females, CAH produces an enlarged clitoris at birth, with the urethral opening at the base (ambiguous genitalia, appearing more male than female) and masculinization of features as the child grows, such as deepening of the voice, facial hair, and failure to menstruate or abnormal periods at **puberty**. The internal structures of the reproductive tract, including the ovaries, uterus, and fallopian tubes, are normal. Females with severe CAH may be mistaken for males at birth. In males, the genitals are normal at birth, but the child becomes muscular, the penis enlarges, pubic hair appears, and the voice deepens long before normal puberty, sometimes as early as two to three years of age. At puberty, the testes are small.

In the severe salt-wasting form of CAH, newborns may develop symptoms shortly after birth, including **vomiting**, dehydration, electrolyte (a compound such as sodium or calcium that separates to form ions when dissolved in water) changes, and cardiac arrhythmias. If not treated, this form of CAH can result in death within one to six weeks after birth.

In the mild form of CAH, which occurs in late childhood or early adulthood, symptoms include premature development of pubic hair, irregular menstrual periods, unwanted body hair, or severe **acne**. However, sometimes there are no symptoms, and children affected are diagnosed because of an affected relative.

When to call the doctor

Many cases of CAH will be detected at birth, but in milder cases, symptoms may not develop until later, at which time medical care should be obtained. For children with more severe cases of CAH, regular medical care is necessary to achieve desired treatment results.

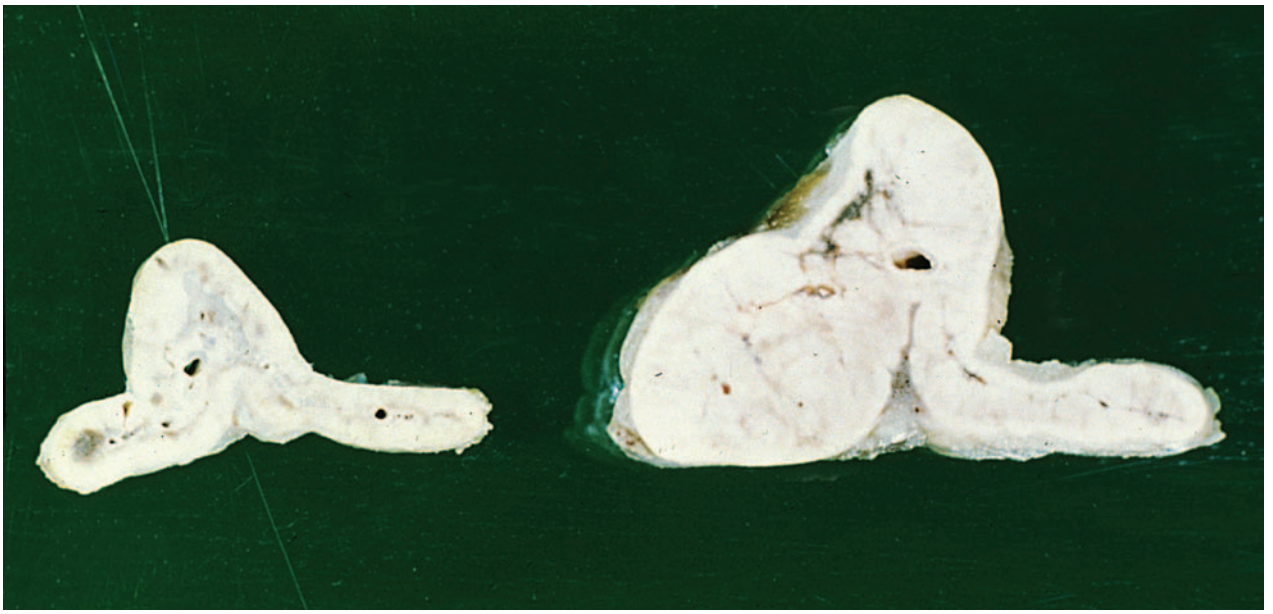
Diagnosis

CAH is diagnosed by a careful examination of the genitals and blood and urine tests that measure the hormones produced by the adrenal gland. A number of states in the United States perform a hormonal test (a heel prick blood test) for CAH and other inherited diseases within a few days of birth. In questionable cases, genetic testing can provide a definitive diagnosis. For some forms of CAH, prenatal diagnosis is possible through chronic villus sampling in the first trimester and by measuring certain hormones in the amniotic fluid during the second trimester.

Treatment

The goal of treatment for CAH is to return the androgen levels to normal. This is usually accomplished through drug therapy, although surgery may be an alternative for children with little or no enzyme activity. Lifelong treatment for CAH is required.

Drug therapy consists of use of a cortisol-like steroid medications called glucocorticoids. Oral hydrocortisone is prescribed for younger children, and prednisone or dexamethasone is prescribed for older children. Side effects of steroids include stunted growth. Steroid



Adrenal cortical hyperplasia. The adrenal on the left is normal, the right shows hyperplasia. *(Photo Researchers, Inc.)*

therapy should not be suddenly stopped, since adrenal insufficiency will result. Treatment results must be monitored carefully, because of large individual variations in enzyme deficiency in children with CAH.

For children with salt-wasting CAH, fludrocortisone (Florinef), which acts like aldosterone (the missing hormone), is also prescribed. Infants and small children may also receive salt tablets, while older children are encouraged to eat salty foods. Serum electrolytes must be checked frequently, especially for children with salt-wasting CAH, to assure that normal levels of sodium and potassium are maintained.

Medical therapy achieves hormonal balance most of the time, but at times appropriate levels can be hard to maintain. CAH patients may have periods of fluctuating hormonal control that lead to increases in the dose of steroids prescribed. Sometimes these doses can become excessive as needs later decrease, leading to growth inhibition.

Increased doses may also be required when the child has a **fever** or a serious injury (a broken bone). If children are vomiting their oral medicine, have severe **diarrhea**, are unconscious, or cannot take anything by mouth before surgery, they may need to receive their medications by injections.

Children with CAH should see a pediatric endocrinologist frequently. The endocrinologist will assess height, weight, and blood pressure, and order an annual x ray of the wrist (to assess bone age), as well as assess blood hormone levels. If they require medical treatment, CAH children with the milder form of the disorder are usually effectively treated with hydrocortisone or prednisone.

Females with CAH who have masculine external genitalia require surgery to reconstruct the clitoris and/or vagina. This is usually performed when the child is an infant. However, some doctors and parents believe that the best time for vaginal surgery is during adolescence.

An experimental type of drug therapy—a three-drug combination, with an androgen blocking agent (flutamide), an aromatase inhibitor (testolactone), and low dose hydrocortisone—was as of 2004 being studied by physicians at the National Institutes of Health. Preliminary results are encouraging, but it will be many years before the **safety** and effectiveness of this therapy is fully known.

Adrenalectomy, a surgical procedure to remove the adrenal glands, is a more radical treatment for CAH. It was widely used before the advent of steroids. In the early 2000s, it is recommended for CAH children with little or no enzyme activity and can be accomplished by laparoscopy. This is a minimally invasive type of surgery done through one or more small one-inch (2.5 cm) incisions and a laparoscope, an instrument with a fiber-optic light containing a tube with openings for surgical instruments. Adrenalectomy is followed by hormone therapy, but in lower doses than CAH patients not treated surgically receive.

KEY TERMS

Adrenal glands—A pair of endocrine glands (glands that secrete hormones directly into the bloodstream) that are located on top of the kidneys. The outer tissue of the glands (cortex) produces several steroid hormones, while the inner tissue (medulla) produces the hormones epinephrine (adrenaline) and norepinephrine.

Aldosterone—A hormone secreted by the adrenal glands that is important for maintaining salt and water balance in the body.

Androgens—Hormones (specifically testosterone) responsible for male sex characteristics.

Congenital—Present at birth.

Cortisol—A steroid hormone secreted by the adrenal cortex that is important for maintenance of body fluids, electrolytes, and blood sugar levels. Also called hydrocortisone.

Hormone—A chemical messenger secreted by a gland or organ and released into the bloodstream. It travels via the bloodstream to distant cells where it exerts an effect.

Hyperplasia—A condition where cells, such as those making up the prostate gland, rapidly divide abnormally and cause the organ to become enlarged.

Steroids—Hormones, including aldosterone, cortisol, and androgens, that are derived from cholesterol and that share a four-ring structural characteristic.

Prognosis

CAH can be controlled and successfully treated in most patients as long as they remain on drug therapy.

Prevention

Prenatal therapy, in which a pregnant woman at risk for a second CAH child is given dexamethasone to decrease secretion of androgens by the adrenal glands of the female fetus, has been in use since 1994. This therapy is started in the first trimester when fetal adrenal production of androgens begins but before prenatal diagnosis is done that would provide definitive information about the sex of the fetus and its disease status. This means that a number of fetuses are exposed to unnecessary steroid treatment in order to prevent the development of male-like genitals in female fetuses with CAH. Several hundred children have undergone this treatment with no major adverse effects, but its long-term risks are unknown. Since there is very little data on the effectiveness and safety of prenatal therapy, it should only be offered to patients who clearly understand the risks and benefits and who are capable of complying with strict monitoring and follow-up throughout pregnancy and after the child is born.

Parental concerns

Parents with a **family** history of CAH or who have a child with CAH should seek genetic counseling. Genetic testing during pregnancy can provide information on the risk of having a child with CAH.

Because children with CAH may not always be able to administer their own treatment (because they are too young or they are unconscious), parents are encouraged to make sure that the child with CAH wears a medical identification bracelet or necklace (Medic-Alert) stating that the child takes glucocorticoids and possibly Florinef. This notifies medical personnel to administer stress doses of medicines if needed.

When taking a child with CAH for emergency care, parents are advised to refer to the condition by its full name rather than CAH. This is because this rare disease could be confused with another condition that shares the same initials: chronic active hepatitis. The parents should inform medical personnel if the child has salt-wasting CAH. It is also recommended that parents have a letter or information prepared concerning CAH and care needed so that this can be given to a new doctors who may treat the child.

Parents should be sensitive to the psychological aspects of the disease and obtain counseling for children with CAH. Topics of concern might include an understanding of the disease, the life-long requirement for medication, genital surgery, and sexuality.

Resources

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ORGANIZATIONS

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Congenital amputation

Definition

Congenital amputation is the absence of a limb or part of a limb at birth.

Description

There are different types of congenital amputation birth defects. An infant with congenital amputation may be missing an entire limb or just a portion of a limb. The complete absence of a limb leaving a stump is called transverse deficiency, or amelia. When a specific part of a limb is missing, for example, when the fibula bone in the lower leg is missing, but the rest of the leg is intact, it is called a longitudinal deficiency. The condition in which only a mid-portion of a limb is missing, as when the hands or feet are attached directly to the trunk, is known as phocomelia.

Congenital amputation may be the result of the constriction of fibrous bands within the membrane that surrounds the developing fetus (amniotic band syndrome), the exposure to substances known to cause birth defects (teratogenic agents), genetic factors, or other, unknown, causes.

Demographics

An estimated one in 2,000 babies is born with all or part of a limb missing. This number includes everything from a missing part of a finger to the absence of both arms and both legs. Congenital amputation is the least common form of amputation. There have been occasional periods in history where the frequency of congenital amputations has increased. For example, in the 1960s many pregnant women were given tranquilizers containing the drug thalidomide. The result was the "thalidomide tragedy" during which there was a drastic increase in the number of babies born with deformations of the limbs. In this case, the birth defect usually presented itself as very small, deformed versions of normal limbs. Subsequently, birth defects as a result of exposure to Agent Orange, the U.S. defoliant used in Vietnam, and radiation exposure near the site of the Chernobyl disaster in Russia have left numerous children with malformed or absent limbs.

Causes and symptoms

Most of the time, the cause of congenital amputations is unknown. According to the March of Dimes, most birth defects have one or more genetic factors and one or more environmental factors, but what the actual factors are in any given case is often difficult, if not impossible, to pinpoint. Most birth defects occur in the first three months of pregnancy when the organs of the fetus are forming.

During the crucial first weeks, frequently before a woman is aware she is pregnant, the developing fetus is most susceptible to substances that can cause birth defects (teratogens). Exposure to teratogens can cause congenital amputation. Congenital amputation can also be caused by genetic factors. In some cases, tight amniotic bands may constrict the developing fetus, preventing a limb from forming properly, if at all. It is estimated that amniotic band syndrome occurs in between one in 12,000 and one in 15,000 live births. It is not known what makes the amniotic bands behave in this way in some instances and not in others.

When to call the doctor

Many congenital amputations are not discovered until the birth of the baby. At that time the doctor overseeing the delivery can give the parents helpful resources and refer them to the appropriate medical professionals to begin to discuss possible treatment paths and to help the parents cope effectively. If the abnormality is discovered before the birth, the obstetrician can help the parents decide what steps should be taken, and parents can begin to plan to meet the special needs their child will have.

Diagnosis

Ultrasound examinations may reveal the absence of a limb in some developing fetuses, but routine ultrasounds may not pick up signs of more subtle defects. However, if a doctor suspects that the fetus is at risk for developing a limb deficiency (for example, if the mother has been exposed to radiation), a more detailed ultrasound examination may be performed.

Treatment

If a problem with amniotic band constriction is detected early enough, it may be possible to correct the bands before there is significant damage to limb development. There have been cases in which physicians have detected amniotic band constriction and performed minimally invasive surgery that freed constricting amniotic bands and preserved the affected limbs. This procedure, however, is not commonly available.

Successful treatment of a child with congenital amputation involves an entire medical team, including a pediatrician, an orthopedist, a psychiatrist or psychologist, a prosthetist (an expert in making artificial limbs, or prosthetics), a social worker, and occupational and physical therapists. There is controversy over whether it is considered sound practice to fit the child early with a functional prosthesis. Some experts believe that this leads to more normal development and less wasting away (atrophy) of the muscles of the limbs. However, some parents and physicians believe that the child should be allowed to learn to **play** and perform tasks without a prosthesis, if possible. This is thought to help build a child's positive self-image because it does not reinforce the idea that the child is missing something that should be replaced. Also, many children reject prosthetic devices and do not want to wear them. When the child is older, he or she can be involved in decisions concerning whether to be fitted for a prosthesis.

Prognosis

A congenital limb deficiency has a profound effect on the life of the child and his or her parents. Children have been found to be extraordinarily good at learning to accomplish tasks using the means they have available and finding ways to compensate for their disability. Parents can help their child by encouraging persistence, allowing the child to do normal activities for him or herself, and not becoming frustrated and doing them for the child. Occupational therapy can help the child learn to accomplish tasks that are more complex if the child encounters difficulties. Prosthetic devices are increasingly sophisticated. Some experts believe that early fitting of a prosthesis enhances acceptance of the prosthesis by the child and parents.

Prevention

There is no known way to prevent congenital amputations, but the prevention of birth defects in general begins with the well being of the mother before and during pregnancy. Prenatal care should be clear and comprehensive, so that the mother understands both her genetic risks and her environmental risks. Several disciplines in alternative therapy also recommend various supplements and **vitamins** that may reduce the chances of birth defects. Studies have suggested that a multivitamin including **folic acid** may reduce birth defects, including congenital abnormalities. **Smoking**, drinking alcohol, using recreational drugs or drugs not prescribed by a physician and having a poor diet while pregnant may increase the risk of congenital abnormalities. Daily, heavy exposure to chemicals is also thought to be especially dangerous for gestating women.

KEY TERMS

Prosthetic—Referring to an artificial part of the body.

Prosthetist—A health care professional who is skilled in making and fitting artificial limbs and other prostheses.

Teratogen—Any drug, chemical, maternal disease, or exposure that can cause physical or functional defects in an exposed embryo or fetus.

Parental concerns

Raising a child with one or more congenital amputations can be challenging. The way in which the child thinks of him or herself is dependent on the way in which the parents treat the child and the ways in which the parents introduce the child to others. Parents should be careful to treat the child as they would any other and not to expect any less from him or her. Children with congenital amputations are remarkably good at compensating for their disability. They are not mentally disabled in any way, and less should not be expected from them in an educational setting because of their disability. Lowered expectations can have a detrimental effect on the child's self-image.

Parents also need to be careful about how they respond to questions and comments about their child's disability when their child is present, always keeping in mind that the child can hear their answers and comments. The way in which the parent feels about the child's disability has a very strong impact on how the child feels about it.

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March of Dimes Birth Defects Foundation. 1275 Mamaroneck Ave., White Plains, NY 10605. Web site: <www.modimes.org>.

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Congenital bladder anomalies

Definition

The two most common congenital bladder abnormalities are exstrophy and congenital diverticula. An exstrophic bladder is one that is open to the outside and turned inside out, so that its inside is visible at birth, protruding from the lower abdomen. A diverticulum is an extension of a hollow organ, usually shaped like a pouch with a narrow opening.

Description

During fetal development, folds enclose tissues and organs and eventually fuse at the edges to form sealed compartments. Both in the front and the back, folds eventually become major body structures. In the back, the entire spinal column folds in like a pipe wrapped in a pillow. In the front, the entire lower urinary system is folded in.

Exstrophy of the bladder represents a failure of this folding process to complete itself, so the organs form with more or less of their front side missing and remain open to the outside. At the same time, the front of the pelvic bone is widely separated. The abdominal wall is open, too. In fact, the defect often extends all the way to the penis in the male or splits the clitoris in the female.

A congenital bladder diverticulum represents an area of weakness in the bladder wall through which extrudes some of the lining of the bladder. (A small balloon squeezed in a fist will create diverticula-like effect between the fingers.) Bladder diverticula may be multiple, and they often occur at the ureterovesical junction (the entrance of the upper urinary system into the bladder). In this location, they may cause urine to reflux into the ureter and kidney, leading to infection and possible kidney damage.

Demographics

Exstrophy is rare, occurring once in approximately 40,000 births.

Causes and symptoms

As with many birth defects, the causes are not well known. Lack of prenatal care and **nutrition** has been linked to many birth defects; however, beyond the avoidance of known teratogens (anything that can cause a birth defect), there is little prevention possible. Diverticula are more common and less serious.

If left untreated, the person with bladder exstrophy will have no control over urination and is more likely than others to develop bladder **cancer**. Diverticula, particularly if it causes urine reflux, may lead to chronic infection and its subsequent consequences.

Diagnosis

A major consideration with congenital abnormalities is that they tend to be multiple. Further, each one is unique in its extent and severity. Exstrophy can involve the rectum and large bowel and coexist with hernias. The obvious bladder exstrophy seen at birth will prompt immediate action and a search for other anomalies.

Diverticula are not visible and will be detected only if they cause trouble. They are usually found in an examination for the cause of recurring urinary infections. **X rays** of the urinary system or a cystoscopy (examination with a telescope-like instrument) will identify them. Often, the two procedures are done together: a urologist performs the cystoscopy, then a radiologist instills a contrast agent into the bladder and takes x rays.

Treatment

Surgery is necessary and can usually produce successful results. If possible, the surgery must be performed within 48 hours of birth. Prior to surgery, the exposed organs must be protected and all related defects identified and managed. Delay in the surgery leads to the frequent need to divert the urine into the bowel because the partially repaired bladder cannot control the flow. After surgery, the likelihood of infection requires monitoring.

After surgery, ongoing precautions to reduce frequency of infection may have to be used. Cranberry juice

has the ability to keep bacteria from adhering to the membranes and can help prevent infection whenever there is increased risk. There are botanical and homeopathic treatments available; however, consultation by a trained practitioner is recommended before treatment.

Prognosis

With immediate surgery, three-fourths of all people can be successfully repaired. They will have control of their urine and no long-term consequences. The rate of infection is greater for those with congenital bladder anomalies, since any abnormality in the urinary system predisposes it to invasion by bacteria.

Prevention

Birth defects often have no precisely identified cause; therefore, prevention is limited to general measures such as early and continuous prenatal care, appropriate nutrition, and a healthy lifestyle.

Parental concerns

Parents must monitor the urinary output of their newborn children for the first few days of life. Parents of children with bladder diverticula should be aware of prevention and treatment practices for urinary tract infections.

See also Cystitis.

KEY TERMS

Congenital—Present at birth.

Cystoscopy—A diagnostic procedure in which a hollow lighted tube (cystoscope) is used to look inside the bladder and the urethra.

Diverticulum—Plural, diverticula; an outpouching in a tubular organ caused when the inner, lining layer bulges out (herniates) through the outer, muscular layer. Diverticula are present most often in the colon (large intestine), but are also found in the bladder.

Exstrophy—A congenital condition in which a hollow organ, such as the bladder, is turned inside out, establishing contact between the organ and the outside of the body.

Radiologist—A medical doctor specially trained in radiology, the branch of medicine concerned with radioactive substances and their use for the diagnosis and treatment of disease.

Teratogen—Any drug, chemical, maternal disease, or exposure that can cause physical or functional defects in an exposed embryo or fetus.

Ureter—The tube that carries urine from the kidney to the bladder; each kidney has one ureter.

Ureterovesical junction—The point where the ureter joins the bladder.

Urologist—A physician who specializes in the anatomy, physiology, diseases, and care of the urinary tract (in men and women) and male reproductive tract.

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American Academy of Pediatrics. 141 Northwest Point Blvd., Elk Grove Village, IL 60007–1098. Web site: <www.aap.org/default.htm>.

American Foundation for Urologic Disease. 1128 North Charles St., Baltimore, MD 21201. Web site: <www.afud.org/>.

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Congenital brain defects

Definition

Congenital brain defects are a group of disorders of brain development that are present at birth.

Description

Brain development begins shortly after conception and continues throughout the growth of a fetus. A complex genetic program coordinates the formation, growth, and migration of billions of neurons, or nerve cells, and their development into discrete, interacting brain regions. Interruption of this program, especially early in development, can cause structural defects in the brain. In addition, normal brain formation requires proper development of the surrounding skull, and skull defects may lead to brain malformation. Congenital brain defects may be caused by inherited genetic defects, spontaneous mutations within the genes of the embryo, or effects on the embryo due to the mother's infection, trauma, or drug use.

Early in fetal development, a flat strip of tissue along the back of the fetus rolls up to form a tube. This so-called neural tube develops into the spinal cord, and at one end, the brain. Closure of the tube is required for subsequent development of the tissue within. Many different types of brain defects are caused by improper closure of this neural tube. One such congenital brain anomaly, anencephaly (literally "without brain") results when the topmost portion of the tube fails to close and the brain does not develop. Anencephaly is the most common severe malformation seen in stillborn births. It is about four times more common in females than males. Anencephaly is sometimes seen to run in families, and for parents who have conceived one anencephalic fetus, the risk of a second is as high as 5 percent. Fewer than half of babies with anencephaly are born alive, and survival beyond the first month is rare.

Another congenital brain defect, encephalocele, is a protrusion of part of the brain through a defect in the skull. The most common site for encephalocele is along the front-to-back midline of the skull, usually at the rear, although frontal encephaloceles are more common among Asians. Pressure within the skull pushes out cranial tissue. The protective layer over the brain, the meninges, grows to cover the protrusion, as does skin in some cases. Defects in skull closure are thought to cause some cases of encephalocele, while defects in neural tube closure may cause others. Encephaloceles may be small and contain little or no brain tissue or may be quite large and contain a significant portion of the brain.

Failure of neural-tube closure below the level of the brain prevents full development of the surrounding vertebral bones and leads to **spina bifida**, or a divided spinal column. Incomplete closure causes protrusion of the spinal cord and meninges, called meningomyelocele. Some cases of spina bifida are accompanied by another defect at the base of the brain, known as the Arnold-Chiari malformation or Chiari II malformation. For reasons that are unclear as of 2004, part of the cerebellum is displaced downward into the spinal column. Symptoms may be present at birth or delayed until early childhood.

The Dandy-Walker malformation is a brain defect marked by incomplete formation or absence of the central section of the cerebellum and the growth of cysts within the lowest of the brain's ventricles. The ventricles are fluid-filled cavities within the brain, through which cerebrospinal fluid (CSF) normally circulates. The cysts may block the exit of the fluid, causing **hydrocephalus**. Symptoms may be present at birth or delayed until early childhood.

Soon after closure of the neural tube, the brain divides into two halves, or hemispheres. Failure of division is termed holoprosencephaly (literally "whole forebrain"). Holoprosencephaly is almost always accompanied by facial and cranial deformities along the midline of the head, including **cleft lip**, **cleft palate**, fused eye sockets and a single eye (cyclopia), and deformities of the limbs, heart, gastrointestinal tract, and other internal organs. Most infants are either stillborn or die soon after birth. Survivors suffer from severe neurological impairments.

The normal ridges and valleys of the mature brain are formed after cells from the inside of the developing brain migrate to the outside and multiply. When these

cells fail to migrate, the surface remains smooth, a condition called lissencephaly (''smooth brain''). Lissencephaly is often associated with facial abnormalities including a small jaw, a high forehead, a short nose, and low-set ears.

If damaged during growth, especially within the first 20 weeks, brain tissue may stop growing, while tissue around it continues to form. This causes an abnormal cleft or groove to appear on the surface of the brain, called schizencephaly (literally ''split brain''). This cleft should not be confused with the normal wrinkled brain surface, nor should the name be mistaken for **schizophrenia**, a mental disorder. Generalized destruction of tissue or lack of brain development may lead to hydranencephaly, in which cerebrospinal fluid fills much of the space normally occupied by the brain. Hydranencephaly is distinct from hydrocephalus, in which CSF accumulates within a normally-formed brain, putting pressure on it and possibly causing skull expansion.

Excessive brain size is termed megalencephaly (literally ''big brain''). Megalencephaly is defined as any brain size above the 98th percentile within the population. Some cases are familial and may be entirely benign. Others are due to metabolic or neurologic disease. The opposite condition, microcephaly, may be caused by failure of the brain to develop or by intrauterine infection, drug toxicity, or brain trauma.

Demographics

Researchers estimate that central nervous system anomalies, congenital brain defects included, occur in approximately 15 of every 10,000 live births. Some congenital brain defects, such as those associated with spina bifida have a higher prevalence, affecting as many as two to three per 1,000 live births. In general, birth defects of the brain are not more prevalent in one gender; however, specific defects such as anencephaly, affect girls four times more often than boys, and in families who have conceived one child with anencephaly, the risk to another pregnancy increases nearly 5 percent.

Causes and symptoms

Causes

Congenital brain defects may have genetic, infectious, toxic, or traumatic causes. In most cases, no certain cause can be identified.

GENETIC CAUSES Some brain defects are caused by trisomy, the inclusion of a third copy of a chromosome normally occurring in pairs. Most trisomies occur because of improper division of the chromosomes during formation of eggs or sperm. Trisomy of chromosome 9 can cause some cases of Dandy-Walker and Chiari II malformation. Some cases of holoprosencephaly are caused by trisomy of chromosome 13, while others are due to abnormalities in chromosomes 7 or 18. Individual gene defects, either inherited or spontaneous, are responsible for other cases of congenital brain malformations.

DRUGS Drugs known to cause congenital brain defects when used by the mother during critical developmental periods include:

- anticonvulsant drugs
- retinoic acid and tretinoin
- warfarin
- alcohol
- cocaine

OTHER Other causes of congenital brain defects include:

- intrauterine infections, including cytomegalovirus, **rubella**, **herpes simplex**, and varicella zoster
- maternal diabetes mellitus
- maternal phenylketonuria
- fetal trauma

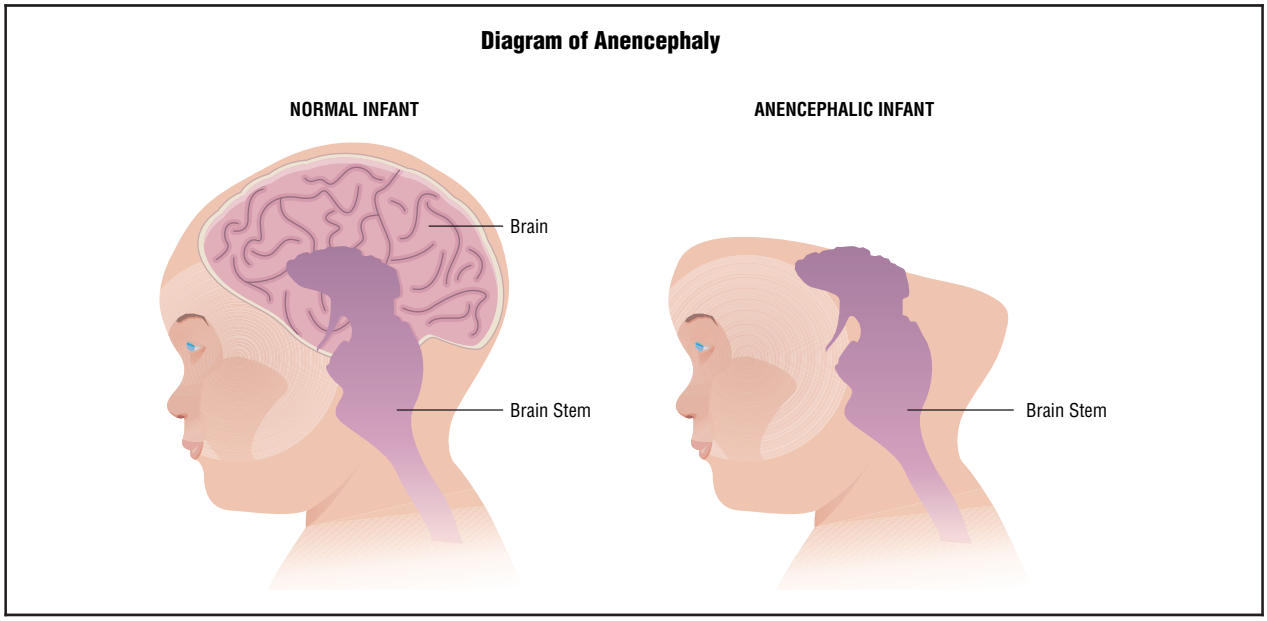
Symptoms

Besides the features listed above, symptoms of congenital brain defects may include:

- Chiari II malformation: Impaired swallowing and gag reflex, loss of the breathing reflex, facial paralysis, uncontrolled eye movements (**nystagmus**), impaired balance and gait.
- Dandy-Walker malformation: Symptoms of hydrocephalus, lack of muscle tone or floppiness, seizures, **spasticity**, deafness, irritability, visual impairment, deterioration of consciousness, paralysis.
- Lissencephaly: Lack of muscle tone, seizures, **developmental delay**, spasticity, cerebral palsy.
- Hydranencephaly: Irritability, spasticity, seizures, temperature oscillations.
- Megalencephaly due to neurological or metabolic disease: **Mental retardation**, seizures.

Diagnosis

Congenital brain defects are diagnosed either from direct physical examination or imaging studies



Comparison of the brain of a normal infant with the brain of an infant with anencephaly. *(Illustration by Argosy, Inc.)*

including **computed tomography** scans (CT) and **magnetic resonance imaging** (MRI). Electroencephalography (EEG) may be used to reveal characteristic abnormalities.

Prenatal diagnosis of neural tube defects causing anencephaly or meningomyelocele is possible through ultrasound examination and maternal blood testing for alpha-fetoprotein, which is almost always elevated. Ultrasound can also be used to diagnose Dandy-Walker and Chiari II malformations. **Amniocentesis** may reveal trisomies or other chromosomal abnormalities.

Treatment

Spina bifida may be treated with surgery to close the open portion of the spinal cord. Surgery for encephalocele is possible only if there is a minimal amount of brain tissue protruding. Malformations associated with hydrocephalus (Dandy-Walker, Chiari II, and some cases of hydranencephaly) may be treated by installation of a drainage shunt for cerebrospinal fluid. Drugs may be used to treat some symptoms of brain defects, including seizures and spasticity or muscle rigidity.

Prognosis

Most congenital brain defects carry a very poor prognosis. Surgical treatment of meningomyelocele and encephalocele may be successful, with lasting neurological deficiencies that vary in severity. Early treatment of hydrocephalus may prevent more severe brain damage.

Prevention

Some cases of congenital brain defects can be prevented with good maternal **nutrition**, including **folic acid** supplements. Folic acid is a vitamin that has been shown to reduce the incidence of neural tube defects. The Centers for Disease Control and Prevention recommends that all women of childbearing age consume 0.4 mg of folic acid daily to prevent neural tube defects. All over-the-counter multivitamins contain this amount of folic acid. Pregnant women should avoid exposure to infection, especially during the first trimester. Abstention from drugs and alcohol during pregnancy may reduce risk. Genetic counseling is advisable for parents who have had one child with anencephaly, since the likelihood of having another is increased.

Parental concerns

Some congenital brain anomalies, such as anencephaly, are not compatible with life, and fetuses affected by them will die. Many more brain anomalies are not lethal. Most congenital brain anomalies, however, will impact mental functioning, development, and, in some cases, physical mobility. When functions of the brain are severely limited, the child may be placed on a life support system. If the essential tasks of the brain are not impaired, the child will most likely face brain dysfunction, including any or all of the following: memory and language problems, neuromotor functioning problems, and behavioral and social problems. It is important that the child's abilities are accurately assessed and that an appropriate plan of treatment is developed. A multidisci-

KEY TERMS

Amniocentesis—A procedure performed at 16-18 weeks of pregnancy in which a needle is inserted through a woman's abdomen into her uterus to draw out a small sample of the amniotic fluid from around the baby for analysis. Either the fluid itself or cells from the fluid can be used for a variety of tests to obtain information about genetic disorders and other medical conditions in the fetus.

Cerebrospinal fluid—The clear, normally colorless fluid that fills the brain cavities (ventricles), the subarachnoid space around the brain, and the spinal cord and acts as a shock absorber.

Congenital—Present at birth.

Fetus—In humans, the developing organism from the end of the eighth week to the moment of birth. Until the end of the eighth week the developing organism is called an embryo.

Spasticity—Increased mucle tone, or stiffness, which leads to uncontrolled, awkward movements.

plinary team approach to the medical, educational, and emotional needs of the child is imperative. The treatment team for these children often includes neurologists, surgeons, nurses, physical and occupational therapists, educators, and social workers, all coordinated by the primary physician.

Many children with congenital brain anomalies, such as Chiari I malformation and nearly 50 percent of those affected by hydrocephalus, have normal intellectual functioning, and some have unimpaired physical mobility. These children, however, still require medical management and benefit from a multidisciplinary healthcare team. Many face multiple surgical procedures and hospital admissions. It is important to prepare children for these medical treatments and surgeries. Children's hospitals, local school systems, and state governments can assist parents in finding the support and resources they need.

See also Chiara malformation; Spina bifida.

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Congenital heart disease

Definition

Congenital heart disease, or congenital heart defect, includes a variety of structural problems of the heart or its major blood vessels, which are present at birth.

Description

The heart, which is completely developed about eight weeks after conception, is one of the earliest organs to completely develop. Congenital heart defects occur when the heart or blood vessels near the heart do not develop properly before birth. Some infants are born with mild types of congenital heart defects, but most need surgery in order to survive. In some cases, the defect may be mild and unnoticed at birth, then diagnosed later in life.

Research is ongoing, and at least 35 congenital heart or cardiovascular defects have been identified. Each defect is defined by its location and severity. Most congenital cardiovascular defects obstruct the flow of blood in the heart or nearby blood vessels, or cause an abnormal flow of blood through the heart. Rarer congenital cardiovascular defects occur when the newborn has only one ventricle (lower chamber), when the pulmonary artery (leading to the lungs) and the aorta (the largest artery that brings blood to the body) come out of the same ventricle, or when one side of the heart is not completely formed.

Patent ductus arteriosus

Ductus arteriosus refers to an open passageway—or temporary blood vessel (ductus)—that carries blood from the heart via the pulmonary artery to the aorta before birth. This passageway allows blood to bypass the lungs, which are not yet functional in the fetus. The ductus should close spontaneously in the first few hours

after birth. When it does not close in the newborn, some of the blood that should flow through the aorta returns to the lungs. **Patent ductus arteriosus** is common in premature babies, but rare in full-term babies. It has been associated with mothers who had German **measles** (**rubella**) while pregnant. Patent ductus arteriosus accounts for 6–11 percent of all cases of congenital cardiovascular defects in the United States.

Hypoplastic left heart syndrome

Although rare, hypoplastic left heart syndrome, a condition in which the left side of the heart is underdeveloped, is the most serious congenital cardiovascular defect. With this syndrome, blood returning from the lungs must flow through an opening or hole in the wall between the atria, called an **atrial septal defect**. The right ventricle pumps blood into the pulmonary artery and blood reaches the aorta through a patent ductus arteriosus (see description in the previous section). In hypoplastic left heart syndrome, the baby seems normal at birth, but as the ductus closes, blood cannot reach the aorta and circulation fails. If left untreated, hypoplastic left heart syndrome is always fatal.

Heart rhythm problems

An arrhythmia is an abnormal heart beat. Normally, the heart beats at 50–150 beats per minute, depending on the child's age. Bradycardia is an irregularly slow heart rhythm, and tachycardia is an irregularly fast heart rhythm. Both conditions reduce the heart's pumping ability.

Obstruction defects

When heart valves, arteries, or veins are narrowed, they partially or completely block the flow of blood. The most common obstruction defects are pulmonary valve stenosis, aortic valve stenosis, and **coarctation of the aorta**. Coarctation of the aorta accounts for 8–11 percent of all cases of congenital cardiovascular defects in the United States.

Stenosis is a narrowing of the valves or arteries. In pulmonary stenosis, the pulmonary valve does not open properly, forcing the right ventricle to work harder. In aortic stenosis, the improperly formed aortic valve is narrowed. As the left ventricle works harder to pump blood through the body, it becomes enlarged. In coarctation of the aorta, the aorta is constricted, reducing the flow of blood to the lower part of the body and increasing blood pressure in the upper body.

Bicuspid aortic valve and subaortic stenosis are less common obstruction defects. A bicuspid aortic valve has only two flaps instead of three, which can lead to stenosis in adulthood. Subaortic stenosis is a narrowing of the left ventricle below the aortic valve that limits the flow of blood from the left ventricle.

Septal defects

When a baby is born with a hole in the septum (the wall separating the right and left sides of the heart), blood leaks from the left side of the heart to the right. A major leakage can lead to enlargement of the heart and failing circulation. The most common types of septal defects are atrial septal defect, an opening between the two upper heart chambers (atria), and ventricular septal defect, an opening between the two lower heart chambers (ventricles). Atrial septal defects account for 4–10 percent of all cases of congenital cardiovascular defects in the United States; ventricular septal defects account for about 14–16 percent.

Two variations of septal defects include atrioventricular canal defect and Eisenmenger's complex. Atrioventricular canal defect (also called endocardial cushion defect or atrioventricular septal defect) is a large hole in the septum, accompanied by abnormal tricuspid and mitral valves that are not formed as individual valves. Instead, a single large valve crosses the defect. The defect allows oxygen-rich blood from the lungs to flow from the left side of the heart to the right side of the heart and back again to the lungs. The heart must work harder to accommodate this extra blood and may become enlarged. Eisenmenger's complex is a ventricular septal defect coupled with pulmonary high blood pressure, an enlarged right ventricle, and sometimes an aorta that is not positioned correctly. With this syndrome, blood flows abnormally from the right side of the heart to the left.

Cyanotic defects

Heart disorders that cause a decreased, inadequate amount of oxygen in blood pumped to the body are called cyanotic defects. Cyanotic defects result in a blue discoloration of the skin due to low oxygen levels. Cyanotic defects include truncus arteriosus, total anomalous pulmonary venous return, **tetralogy of Fallot**, **transposition of the great arteries**, tricuspid atresia, and pulmonary atresia.

Truncus arteriosus is a complex malformation in which only one artery comes from the heart and forms the aorta and pulmonary artery. Total anomalous pulmonary venous return is a condition in which the pulmonary veins that bring oxygen-rich blood from the lungs back to the heart are not connected to the left atrium. Instead, the pulmonary veins drain through abnormal connections to the right atrium.

Nine to fourteen percent of cases of congenital cardiovascular defects in the United States are tetralogy of Fallot, which includes four defects. The major defects are a large hole (ventricular septal defect) between the ventricles, which allows oxygen-poor blood to mix with oxygen-rich blood, and narrowing at or beneath the pulmonary valve. The other defects are an overly muscular right ventricle and an aorta that lies over the ventricular septal defect.

In transposition (reversal of position) of the great arteries, the positions of the pulmonary artery and the aorta are reversed, causing oxygen-rich blood to re-circulate to the lungs while oxygen-poor blood goes to the rest of the body. Transposition of the great arteries comprises 10–14 percent of congenital cardiovascular defect cases in the United States.

In tricuspid atresia, the baby lacks a triscupid valve and blood cannot flow properly from the right atrium to the right ventricle. In pulmonary atresia, the baby lacks a pulmonary valve and blood cannot flow properly from the right ventricle into the pulmonary artery and on to the lungs.

Other defects

Ebstein's anomaly is a rare congenital syndrome that causes malformed tricuspid valve leaflets, which allow blood to leak between the right ventricle and the right atrium. This condition may cause a hole in the wall between the left and right atrium, called an atrial septal defect. Treatment often involves repairing the tricuspid valve. Ebstein's anomaly may be associated with maternal use of the psychiatric drug lithium during pregnancy.

Brugada syndrome is another rare congenital cardiovascular defect that appears in adulthood and may cause sudden death if untreated. Symptoms, which include rapid, uneven heart beat, often appear at night. Scientists believe that Brugada syndrome is caused by mutations in the gene SCN5A, which involves cardiac sodium channels.

Infants born with DiGeorge sequence can have heart defects such as a malformed aortic arch and tetralogy of Fallot. Researchers believe DiGeorge sequence most often is caused by mutations in genes in the region 22q11.

Marfan syndrome is a connective tissue disorder that causes tears in the aorta. Since the disease also causes excessive bone growth, most Marfan syndrome patients are over 6 ft (1.8 m) tall. In athletes and others, it can lead to sudden death. Researchers believe the defect responsible for Marfan's syndrome is found in gene FBN1 on chromosome 15.

Heart muscle abnormalities may lead to congestive heart failure. In heart failure, the heart does not pump blood well enough for the body to get the nourishment it needs for normal function and activity. When the heart does not function properly, fluid can build up in the lungs, causing difficult breathing. Fluid can also build up in the rest of the body, causing swelling.

Demographics

About 44,000 infants (about eight of every 1,000 infants or 1 percent of live births) are born every year with congenital cardiovascular defects, the most common birth defect. It is the number one cause of death from birth defects during the first year of life. Nearly twice as many children die from congenital cardiovascular defects in the United States than from all forms of childhood cancers combined. Most of these children can benefit from surgical treatment, even if the defect is severe. Overall, the mortality rate from congenital cardiovascular defects has significantly declined in the past few decades. About one million adults (over age 20) with cardiovascular defects are currently living in the United States.

Causes and symptoms

Causes

In most cases, the causes of congenital cardiovascular defects are unknown. Genetic and environmental factors, and lifestyle habits can all be involved. However, only a few genes have been discovered that have been linked to the presence of heart defects. The likelihood of having a child with a congenital cardiovascular defect increases if the mother or father, another child, or another relative had congenital cardiovascular defects, or there is a **family** history of sudden death.

Women with diabetes and **phenylketonuria** (an inherited liver condition also called PKU) are at higher risk of having children with congenital heart defects. Many cases of congenital cardiovascular defects result from the mother's excessive use of alcohol or history of taking illegal drugs, such as cocaine, while pregnant. The mother's exposure to certain prescription drugs such as anticonvulsant and dermatologic medications during pregnancy also can cause congenital cardiovascular defects. Her exposure to industrial chemicals, solvents, and ionizing radiation (x ray) also increases the risk of having children with congenital heart defects.

The occurrence of some infections during pregnancy, including viral infections such as rubella (German measles), can cause congenital cardiovascular defects. In

addition, there are many genetic conditions, such as **Down syndrome** or Turner's syndrome, which affect multiple organs and can cause congenital cardiovascular defects. Children with oral clefts are 16 times more likely to have a congenital cardiovascular defect than the normal population, although the reason for this association is unknown.

Symptoms

General symptoms of congenital cardiovascular defects include:

- shortness of breath or rapid breathing
- difficulty feeding in infancy
- sweating
- cyanosis (bluish discoloration of the skin, lips, and fingernails)
- heart murmur
- respiratory infections that recur excessively
- poor weight gain in infants
- stunted growth
- underdeveloped limbs and muscles

Some infants and children have no signs or symptoms of congenital cardiovascular defects.

Symptoms of specific types of congenital cardiovascular defects are as follows:

- Patent ductus arteriosus—quick tiring, slow growth, susceptibility to **pneumonia**, and rapid breathing. If the ductus is small, there are no symptoms.
- Hypoplastic left heart syndrome—ashen color, rapid and difficult breathing, and inability to eat.
- Abnormal heart rhythm—feeling of skipped heart beats, **dizziness**, lightheadedness, fainting, shortness of breath, and fatigue.
- Obstruction defects: cyanosis (skin that is discolored blue)—chest **pain**, unusual fatigue or quick tiring, dizziness or fainting, and high blood pressure.
- Septal defects—difficulty breathing, stunted growth, and high blood pressure. Sometimes there are no symptoms.
- Cyanotic defects: cyanosis—sudden rapid breathing or unconsciousness, and shortness of breath and fainting during **exercise**.
- Congestive heart failure—difficulty breathing, swelling, quick tiring, and feeding problems in infants.

When to call the doctor

The parent or caregiver should call the child's pediatrician or cardiologist when the child has these symptoms or conditions:

- infant who has feeding problems, difficulty sucking, or **vomiting** more than two or three feedings per day
- poor weight gain in young children
- swelling in the ankles or feet
- swollen abdomen
- poor exercise tolerance
- recurrent chest colds and respiratory infections
- abnormal blood pressure
- signs of infection including **sore throat**, general body aches or **fever**

The parent or caregiver should seek emergency treatment by calling 911 in most areas when the child has these symptoms or conditions:

- bluish skin tone
- bluish coloration around the lips, fingernail beds, and tongue
- breathing difficulties or rapid breathing
- dizziness or fainting
- uncontrolled coughing or coughing with blood
- irregular heart beats or palpitations (abnormal heart beats that feel like fluttering in the chest)
- chest pain (rare in children)

Diagnosis

Severe congenital cardiovascular defect is diagnosed in infancy and usually becomes evident shortly after birth. However, significant cardiovascular defects may be found anytime during childhood. In a few cases, a cardiovascular defect is not detected until the child is a teenager or adult.

The medical and family history help the physician determine if the child has any conditions or disorders that might contribute to or cause the cardiovascular defect. A family history of cardiovascular defects might suggest a genetic predisposition to the disease.

During the physical exam, the child's blood pressure is measured, and a stethoscope is used to listen to sounds made by the heart and blood flowing through the arteries. Some **heart murmurs** (abnormal heart sounds) can indi-

cate a congenital heart defect. The child's pulse, reflexes, and height and weight are checked and recorded. The child's blood oxygen level can be measured using a pulse oximeter, a sensor placed on the fingertip or earlobe. Internal organs are palpated, or felt, to determine if they are enlarged.

Blood and urine tests are performed to detect the presence of any abnormal substances that may indicate congenital cardiovascular defects.

Echocardiography and cardiac **magnetic resonance imaging** (MRI) may be used to confirm congenital cardiovascular defects when suggested by the child's symptoms and physical exam results.

An echocardiogram (echo) uses ultrasound, or high-frequency sound waves, to display an image of the heart's internal structures. It can be used to detect valve and other heart problems. A Doppler echo uses sound waves to measure blood flow. Fetal echocardiography is used to diagnose congenital cardiovascular defects in utero, usually after 20 weeks of pregnancy. Between 10 and 14 weeks of pregnancy, physicians may use an ultrasound to look for a thickness at the nuchal translucency, a pocket of fluid in back of the embryo's neck, which may indicate a cardiac defect in 55 percent of cases.

Cardiac MRI, a scanning method that uses magnetic fields and radio waves to create three-dimensional images of the heart, can help physicians evaluate congenital cardiovascular defects, but is not always necessary. MRI reveals how blood flows through the heart and how the heart is working. Physicians also may use a chest x ray to look at the size, shape, and location of the heart and lungs.

An electrocardiogram (ECG, EKG) helps the physician evaluate the electrical activity of the heart. During an EKG, small electrode patches are attached to the skin on the chest and connected to a computer that measures the heart's electrical impulses and records them in a zigzag patter on a moving strip of paper.

Stress tests may be performed to provide information about how the heart responds to stress. The test may involve actual exercise or a medication that simulates exercise. Increasing levels of exercise difficulty are monitored while the electrocardiogram, heart rate, and blood pressure are recorded.

Special monitors may be used to evaluate an abnormal heart rhythm. Ambulatory monitors are small portable electrocardiograph machines that record the heart's rhythm. Each type of monitor has unique features related to length of recording time and ability to send the recordings over the phone.

In some cases, these tests are not conclusive enough to confirm the diagnosis of congenital cardiovascular defects. More invasive diagnostic procedures, such as angiography and cardiac catheterization, may be performed to show the type and severity of heart disease. These procedures should be performed by a specially trained physician and diagnostic team in a well-equipped heart center.

During catheterization, a long, slender tube, called a catheter, is inserted into a vein or artery and slowly directed to the heart using x-ray guidance. To better view the heart and blood vessels, contrast material (dye) is injected through the catheter and viewed and recorded on an x-ray video as it moves through the heart. This imaging technique is called angiography. In some cases, blood vessel blockages or narrowed areas may be treated during the catheterization procedure using a specialized balloon tip or other device at the end of the catheter. When treatment is performed as part of a catheterization procedure, it is referred to as an interventional procedure.

Treatment

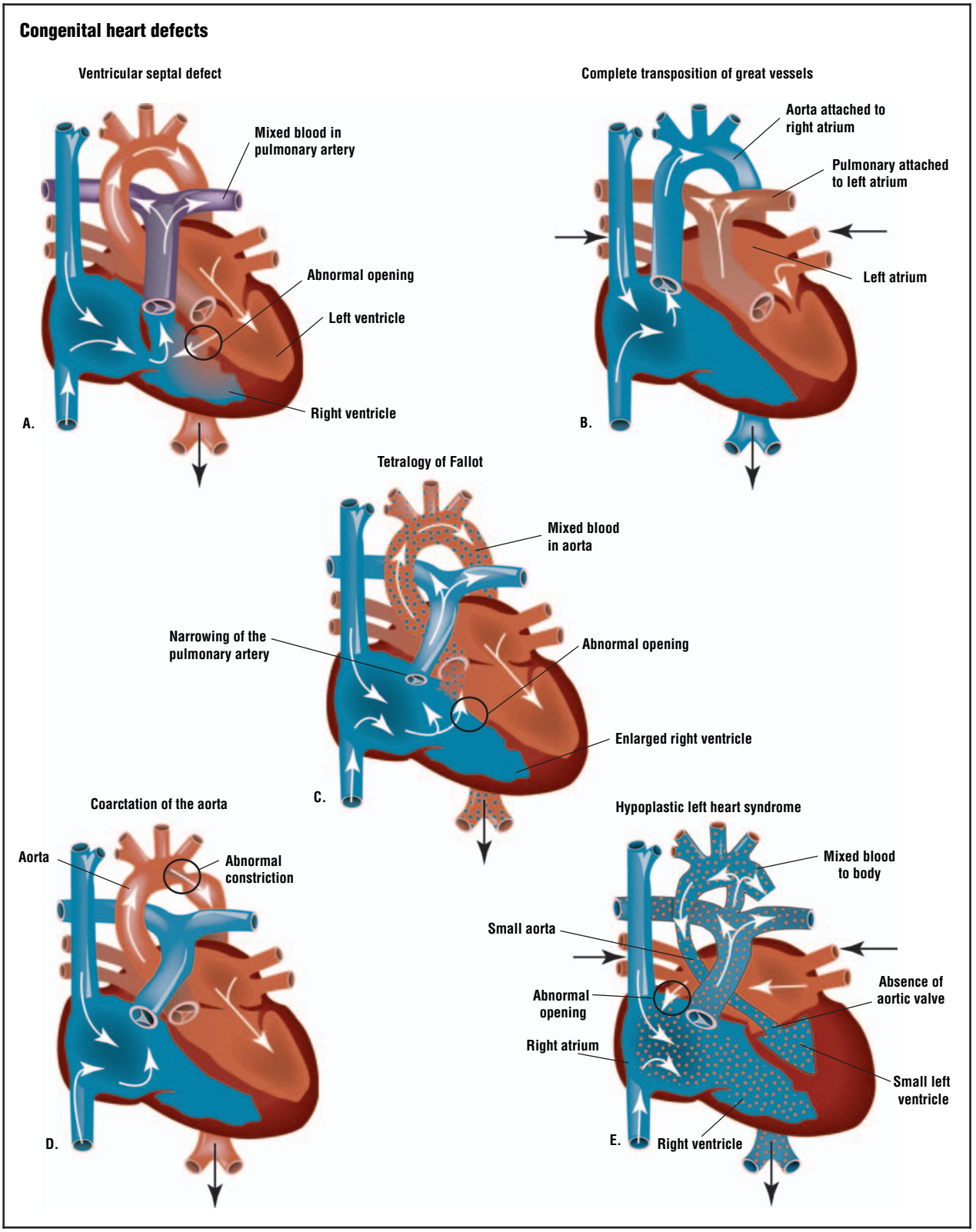
Treatment should be provided by a pediatric cardiologist, a specialist trained to diagnose and treat congenital cardiovascular defects.

Medications

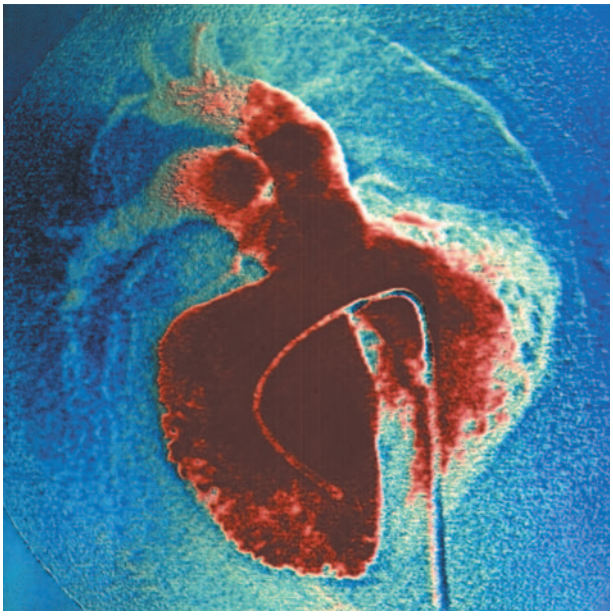
Medications used to treat congenital cardiovascular defects include diuretics, which aid the child in excreting water and salts, and Digoxin, which strengthens the contraction of the heart, slows the heartbeat, and removes fluid from tissues. A potassium supplement may be prescribed along with diuretics, which remove potassium from the body along with excess fluid. Heart rate control drugs and antiarrhythmic drugs may be prescribed to treat irregular heart rhythms. Other medications may include anticoagulants (blood thinners) to reduce the risk of blood clots and **stroke**, ACE inhibitors to decrease artery constriction and improve blood flow, and inotropes to strengthen the heart's contractions.

Electrical therapy and implantable devices

If medications are not effective in controlling a child's heart rate, cardioversion may be required. In this procedure, an electrical shock is delivered to the chest wall to synchronize the heart and allow the normal rhythm to restart. A permanent pacemaker or implantable cardioverter defibrillator (ICD) is sometimes needed to regulate the child's heart rhythm. A pacemaker is a device that sends small electrical impulses to the heart



The most common types of congenital heart defects are ventricular septal defect (A), complete transposition of the great vessels (B), tetralogy of Fallot (C), coarctation of the aorta (D), and hypoplastic left heart syndrome (E). *(Illustration by GGS Information Services.)*



An angiogram of a ventricular septal defect, a congenital heart disease that causes a hole in the center wall of the heart, which normally completely divides the two ventricles, or lower chambers. *(Photograph by Simon Fraser/Science Photo Library/Photo Researchers, Inc.)*

muscle to maintain a suitable heart rate. An ICD is a device used primarily to treat ventricular tachycardia and ventricular fibrillation, two life-threatening heart rhythms. The ICD constantly monitors the heart rhythm. When a very rapid, abnormal heart rhythm is detected, the ICD delivers energy to the heart muscle to cause it to regain a normal rhythm.

Interventional procedures

Catheter-based procedures may be performed to open stenotic valves or vessels, widen the septal opening between the atria, or close abnormal vessels or certain septal defects. During catheterization, a long, slender tube called a catheter is inserted into a vein or artery and slowly directed to the heart, using x-ray guidance. Blood vessel blockages or stenotic valves may be treated during the catheterization procedure using a specialized balloon tip or other device at the end of the catheter. The balloon is rapidly inflated and deflated to open or widen the area. In older patients, a stent (metal mesh tube) can be positioned to act as a scaffold and hold the area open. Several closure devices such as coils, patches, or umbrella-like devices, have been developed that can be inserted through the catheter and are designed to close the defect.

Surgery

The goal of surgery is to repair the defect as much as possible, restore circulation to as close to normal as possible, reduce symptoms, improve survival, and improve quality of life. Sometimes, multiple surgical procedures are necessary. Surgery for most congenital cardiovascular defects has low risk of death (less than 2 percent), compared to 80–100 percent in the 1940s. Surgical procedures used to treat congenital cardiovascular defects include:

- arterial switch
- balloon atrial septostomy
- balloon valvuloplasty
- Damus-Kaye-Stansel procedure
- Fontan procedure
- pulmonary artery banding
- Ross procedure
- shunt procedure
- venous switch or intra-atrial baffle

Arterial switch, to correct transposition of the great arteries, involves connecting the aorta to the left ventricle and connecting the pulmonary artery to the right ventricle. Balloon atrial septostomy, also done to correct transposition of the great arteries, enlarges the atrial opening during heart catheterization. Balloon valvuloplasty uses a balloon-tipped catheter to open a narrowed heart valve, improving the flow of blood in pulmonary stenosis. It is sometimes used to treat aortic stenosis. Transposition of the great arteries also can be corrected by the Damus-Kaye-Stansel procedure, in which the pulmonary artery is cut in two and connected to the ascending aorta and the farthest section of the right ventricle.

For tricuspid atresia and pulmonary atresia, the Fontan procedure connects the right atrium to the pulmonary artery directly or with a conduit, and the atrial defect is closed. Pulmonary artery banding, narrowing the pulmonary artery with a band to reduce blood flow and pressure in the lungs, is used for ventricular septal defect, atrioventricular canal defect, and tricuspid atresia. The band can be removed at a later time, and the defect corrected with open-heart surgery.

To correct aortic stenosis, the Ross procedure grafts the pulmonary artery to the aorta. For tetralogy of Fallot, tricuspid atresia, or pulmonary atresia, the shunt procedure creates a passage between blood vessels, sending blood into parts of the body that need it. For transposition of the great arteries, venous switch creates a tunnel inside the atria to re-direct oxygen-rich blood to the right ventricle and aorta, and venous blood to the left ventricle and pulmonary artery.

When all other options fail, some patients may need a heart transplant. A heart transplant involves replacing the child's heart with a healthy heart from a donor who has died or been declared brain dead. The child's eligibility for a transplant depends on the results of blood tests and other factors related to his or her health and potential for survival.

Nutritional concerns

Infants and children with congenital cardiovascular defects tend to gain weight more slowly. An 8-oz to 1 lb (225–450 g) weight gain in a month may be acceptable. The physician will monitor the child's weight gain and advise the parents of the goal weight gain and any necessary dietary changes. The most common reason for poor growth among children with congenital cardiovascular defects is that they do not consume enough calories or nutrients. Some other factors that may interfere with growth include:

- rapid heart beat and increased breathing rate
- poor appetite
- decreased food intake due to rapid breathing and fatigue
- frequent respiratory infections
- poor absorption of nutrients from the digestive tract
- decreased oxygen in the blood

For infants with congenital cardiovascular defects, **nutrition** supplements may need to be added to regular formula or breast milk. Sometimes additional feedings are required with the aid of a nasogastric tube to provide enough calories and promote weight gain. The nasogastric tube is placed in the baby's nose and passes to the stomach. Formula or breast milk is delivered through the tube. Breastfeeding may not be possible right after birth, depending on the child's condition. A breast pump may be used to maintain the mother's milk supply during times when the baby cannot nurse.

Babies with congenital cardiovascular defects tire quickly during feedings, making frequent feedings necessary. Feedings should be on-demand and may need to be as often as every two hours in the first few months. Some babies have difficulty feeding from a regular bottle nipple; parents may need to try different brands. If medications are prescribed, they should be given before a feeding. Medications should not be mixed in the formula or breast milk unless the doctor advises otherwise.

The pediatrician will advise when solid foods can be started, usually around six months of age. Fat should not be restricted in the diet, especially in the first two years. High-calorie foods and snacks can play an important role in providing good nutrition and helping the child grow at a healthy rate.

Follow-up care

Children with congenital cardiovascular defects require lifelong monitoring, even after successful surgery. Along with routine medical care and standard immunizations, periodic heart check-ups are necessary. Usually, heart check-up appointments are scheduled more frequently just after the diagnosis or after surgery. Additional immunizations, such as the **influenza** vaccine, may be recommended.

Medical identification

A medical identification bracelet or necklace should be worn to alert all health care providers of the child's heart condition in cases of emergency.

Prognosis

The outlook for children with congenital cardiovascular defects has improved markedly since the 1980s. Many types of congenital cardiovascular defects that were once fatal can be treated successfully. Most children with congenital cardiovascular defects grow up to be healthy adults. Children with complex heart disease may continue to need special medical attention throughout **adolescence** and into adulthood for survival and to maintain a good quality of life.

Research on diagnosing cardiovascular defects when the fetus is in the womb may lead to future treatment to correct these conditions before birth. Promising new prevention methods and treatments include genetic screening and the cultivation of cardiac tissue in the laboratory that could be used to repair congenital cardiovascular defects. As scientists continue to advance the study of genetics, they also will better understand the genetic causes of many congenital cardiovascular defects.

Prevention

Congenital cardiovascular defects cannot be prevented. However, to protect patients with congenital cardiovascular defects from heart infections (endocarditis), the American Heart Association recommends regular dental check-ups to prevent infections of the mouth as well as the preventive use of **antibiotics**. Preventive antibiotics should be taken before surgery, invasive tests or procedures, and dental work. A 2003 study reported that preventive antibiotics are underused in people with congenital cardiovascular defects, possibly because they

KEY TERMS

Aneurysm—A weakened area in the wall of a blood vessel which causes an outpouching or bulge. Aneurysms may be fatal if these weak areas burst, resulting in uncontrollable bleeding.

Aorta—The main artery located above the heart that pumps oxygenated blood out into the body. The aorta is the largest artery in the body.

Arteriosclerosis—A chronic condition characterized by thickening, loss of leasticity, and hardening of the arteries and the build-up of plaque on the arterial walls. Arteriosclerosis can slow or impair blood circulation. It includes atherosclerosis, but the two terms are often used synonymously.

Artery—A blood vessel that carries blood away from the heart to the cells, tissues, and organs of the body.

Atrial—Referring to the upper chambers of the heart.

Bacterial endocarditis—An infection caused by bacteria that enter the bloodstream and settle in the heart lining, a heart valve, or a blood vessel. People with congenital cardiovascular defects have an increased risk of developing bacterial endocarditis, so preventive antibiotics are prescribed before surgery, invasive tests or procedures, and dental work to reduce this risk.

Coarctation of the aorta—A congenital defect in which severe narrowing or constriction of the aorta obstructs the flow of blood.

Congenital—Present at birth.

Cyanotic—Marked by a bluish tinge to the skin that occurs when the blood oxygen level drops too low. It is one of the types of congenital heart disease.

Ductus—The blood vessel that joins the pulmonary artery and the aorta. When the ductus does not close at birth, it causes a type of congenital heart disease called patent ductus arteriosus.

Echocardiogram—A record of the internal structures of the heart obtained from beams of ultrasonic waves directed through the wall of the chest.

Electrocardiagram (ECG, EKG)—A record of the electrical activity of the heart, with each wave being labeled as P, Q, R, S, and T waves. It is often used in the diagnosis of cases of abnormal cardiac rhythm and myocardial damage.

Hypertension—Abnormally high arterial blood pressure, which if left untreated can lead to heart disease and stroke.

Hypoplastic—Refers to incomplete or underdeveloped tissues or organs. Hypoplastic left heart syndrome is the most serious type of congenital heart disease.

Nuchal translucency—A pocket of fluid at the back of an embryo's neck, visible via ultrasound. When this pocket of fluid is thickened, it may indicate that the infant will be born with a congenital cardiovascular defect.

Renal artery stenosis—A disorder in which the arteries that supply blood to the kidneys are narrowed or constricted.

Septal—Relating to the septum, the thin muscle wall dividing the right and left sides of the heart. Holes in the septum are called septal defects.

Stenosis—A condition in which an opening or passageway in the body is narrowed or constricted.

Ventricles—The lower pumping chambers of the heart. The ventricles push blood to the lungs and the rest of the body.

do not understand their increased risk of developing bacterial endocarditis.

Parental concerns

If the child needs surgery, it is important for him or her to be as healthy as possible for the operation. If the child has a fever, **cough**, or cold, the parent should inform the surgical team to determine if the procedure should be delayed. The medical team can help parents prepare the child for surgery, and provide information on how to explain the procedure, based on the child's age, ability to understand, and emotions. The child usually stays in the hospital from five to seven days after surgery and returns to normal activities within four to six weeks.

Most children with congenital cardiovascular defects can be fully active and are encouraged to exercise. A scientific statement by the American Heart Association advises children and teens with genetic heart disease to seek advice from their doctors about the types of physical activities that are safe. The statement was intended to help doctors counsel patients who have an

increased risk of sudden cardiac death during physical activity. With some congenital cardiovascular defects, certain athletic activities such as competitive **sports** may be limited, depending on the child's diagnosis and medical condition. Since a child with congenital cardiovascular defects may tire easily, frequent breaks and rest periods should be encouraged, as needed, during activities. Parents should obtain a doctor's note to explain their child's specific exercise limitations. This information should be shared with all teachers and coaches.

Even though most children with congenital cardiovascular defects do not have any mental limitations, some children with congenital cardiovascular defects have developmental delays or other learning difficulties. Therefore, community and school-based resources are important for these children to achieve optimum functioning.

A child with a congenital cardiovascular defect has a greater adulthood risk of having a child with a cardiovascular defect. The frequency of disease increases from less than 1 percent in the general population to 2–20 percent when a parent is affected. Defects such as coarctation of the aorta and aortic valve stenosis have the greatest risk of occurring in the child's offspring. Genetic counseling and further testing, such as chromosome analysis before pregnancy or **amniocentesis** during pregnancy, may be recommended in adults with congenital cardiovascular defects.

Treatment and care for a child with congenital cardiovascular defects can be costly, and some health insurance plans may not cover all the expenses associated with a child's **hospitalization** or surgery. Help is available to cover medical expenses. The parents can discuss financial aid with the hospital. Some organizations, including The Heart of a Child Foundation and Little Hearts on the Mend Fund, provide financial assistance to children in need of heart surgery.

Caring for a child with congenital cardiovascular defects is demanding. Support groups are available to help parents and caregivers cope with these challenges. It is important for parents to take care of themselves, too, by eating properly, exercising regularly, taking care of personal hygiene, keeping in contact with friends and family members for support, and managing stress by practicing relaxation techniques.

See also Atrial septal defect.

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Adult Congenital Heart Association (ACHA). 1500 Sunday Dr., Suite 102, Raleigh NC 27607-5151. (919) 861-4547. Web site: <www.achaheart.org>.

American College of Cardiology. Heart House. 9111 Old Georgetown Rd., Bethesda, MD 20814-1699. (800) 253-4636 ext. 694 or (301) 897-5400. Web site: <www.acc.org>.

American Heart Association. 7320 Greenville Ave., Dallas, TX 75231-4596. (214) 373-6300 or (800) 242-8721. Web site: <www.americanheart.org/children>.

Children's Heart Services. P.O. Box 8275, Bartlett, IL 60108-8275. (630) 415-0282. Web site: <www.childrensheartservices.org>.

The Cleveland Clinic Heart Center. The Cleveland Clinic Foundation, 9500 Euclid Ave., F25, Cleveland, Ohio, 44195. (800) 223-2273 ext. 46697 or (216) 444-6697. Web site: <www.clevelandclinic.org/heartcenter>.

Congenital Heart Disease Information and Resources. 1561 Clark Dr., Yardley, PA 19067. Web site: <www.tchin.org>.

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Verdes, CA 90275. (310) 375-6617. Web sites: <www.heartofachild.org> and <www.littleheartsonthemend.org>.

Heart Support of America. 4873 N. Broadway, Knoxville, TN 37918. Web site: <www.heartsupport.com>.

International Children's Heart Foundation.1750 Madison, Suite 100, Memphis, TN 38104. (877) 869-4243. Web site: <www.babyhearts.com>.

Mended Little Hearts. Support program for parents of children with cardiovascular defects. (888)-HEART99. Web site: <www.mendedhearts.org/MLH/mlh.htm>.

National Heart, Lung and Blood Institute. P.O. Box 30105, Bethesda, MD 20824-0105. (301) 251-1222. Web site: <www.nhlbi.nih.gov>.

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Congenital hip dysplasia

Definition

Congenital hip dysplasia is a condition of abnormal development of the hip, resulting in hip joint instability and potential dislocation of the thigh bone from the socket in the pelvis. This condition has been in the early 2000s been termed developmental hip dysplasia, because it often develops over the first few weeks, months, or years of life.

Description

Congenital hip dysplasia is a disorder in children that is either present at birth or shortly thereafter. During gestation, the infant's hip should be developing with the head of the thigh bone (femur) sitting perfectly centered in its shallow socket (acetabulum). The acetabulum should cover the head of the femur as if it were a ball sitting inside of a cup. In the event of congenital hip dysplasia, the development of the acetabulum in an infant allows the femoral head to ride upward out of the joint socket, especially when the infant begins to walk.

Demographics

In the United States, approximately 1.5 percent of all infants have congenital hip dysplasia. Though the worldwide incidence of congenital hip dysplasia varies, researchers estimate the global incidence to be approximately 1 percent.

Clinical studies show a familial tendency toward hip dysplasia with a greater chance of this hip abnormality in the first born compared to the second or third child. Infants with siblings who have been diagnosed with congenital hip dysplasia or who have parents with the defect are at an increased risk. Females are affected four to eight times more than males, and in children with congenital hip dysplasia, the left leg in more often affected. This disorder is found in many cultures around the world. However, statistics show that infants in colder climates have a higher incidence. It is speculated that this increase may be due to the practice of swaddling which can place the infant's legs in an extreme straightened or adducted position, forcing the hips closer together. The incidence of congenital hip dysplasia is also higher in infants born by cesarean and in breech position births.

Causes and symptoms

Hormonal changes within the mother during pregnancy result in increased ligament looseness or laxity and are thought to possibly cross over the placenta and cause the baby to have lax ligaments while still in the womb. Other symptoms of complete dislocation include a shortening of the leg and limited ability to abduct the leg, or move it outward.

Diagnosis

Because the abnormalities of this hip problem often vary, a thorough physical examination is necessary for an accurate diagnosis of congenital hip dysplasia. The hip disorder can be diagnosed by moving the hip to determine if the head of the femur is moving in and out of the hip joint. One specific method, called the Ortolani test, begins with each of the examiner's hands around the

infant's knees, with the second and third fingers pointing down the child's thigh. With the legs abducted (moved apart), the examiner may be able to hear a distinct clicking sound, called a hip click, with motion. If symptoms are present with a noted increase in abduction, the test is considered positive for hip joint instability. It is important to note this test is only valid a few weeks after birth.

The Barlow method is another test performed with the infant's hip brought together with knees in full bent position. The examiner's middle finger is placed over the outside of the hipbone while the thumb is placed on the inner side of the knee. The hip is abducted to where it can be felt if the hip is sliding out and then back in the joint. In older babies, if there is a lack of range of motion in one hip or even both hips, it is possible that the movement is blocked because the hip has dislocated and the muscles have contracted in that position. Also in older infants, hip dislocation may be present if one leg looks shorter than the other.

X-ray films can be helpful in detecting abnormal findings of the hip joint. **X rays** may also be helpful in finding the proper positioning of the hip joint for treatment. Ultrasound has been noted as a safe and effective tool for the diagnosis of congenital hip dysplasia. Ultrasound has advantages over x rays, as several positions are noted during the ultrasound procedure. This is in contrast to only one position observed during the x ray.

Treatment

The objective of treatment is to replace the head of the femur into the acetabulum and, by applying constant pressure, to enlarge and deepen the socket. In the past, stabilization was achieved by placing rolled cotton diapers or a pillow between the thighs. The child may be dressed in two or three diapers, called double or triple diapering. Both these techniques keep the knees in a frog-like position. In the early 2000s, the Pavlik harness and von Rosen splint are commonly used in infants up to the age of six months to spread the legs apart and force the head of the femur into the acetabulum. A stiff shell cast, called a splint, may be also used to achieve the same purpose. In some cases, older children between six to 18 months old may need surgery to reposition the joint. Also at this age, the use of closed manipulation may be applied successfully, by moving the leg around manually to replace the joint. Operations are performed to reduce the dislocation of the hip and to repair a defect in the acetabulum. A cast is applied after the operation to hold the head of the femur in the correct position. As of 2004 the use of a home traction program was more common. However, after the child is eight years of age, surgical procedures are primarily done for **pain** reduction measures only. Total hip surgeries may be inevitable later in adulthood.

Alternative treatment

Nonsurgical treatments include **exercise** programs, orthosis (a force system, often involving braces), and medications. A physical therapist may develop a program that includes strengthening, range-of-motion exercises, pain control, and functional activities. Chiropractic medicine may be helpful, especially the procedures of closed manipulations, to reduce the dislocated hip joint.

Prognosis

Unless corrected soon after birth, congenital hip dysplasia can cause a characteristic limp or waddling gait in children. If left untreated, the child will have difficulty walking and may experience life-long pain. If diagnosed early, congenital hip dysplasia treatment is highly effective. Children who have received casting, bracing, or surgery, usually go on to have normal hip and leg development. In individuals for whom the diagnosis is made later, the prognosis is not as positive. These children may require more extensive surgery. After surgery, however, the prognosis for normal development of the hip and leg is excellent.

Prevention

Prevention includes proper prenatal care to determine the position of the baby in the womb. This may be helpful in preparing for possible breech births associated with hip problems. Avoiding excessive and prolonged infant hip adduction, or forcing the legs in a straight position close together for periods of time (as in swaddling) may help prevent strain on the hip joints. Early diagnosis remains an important part of prevention of congenital hip dysplasia.

Parental concerns

It is important for infants suspected of having congenital hip dysplasia to receive regular physical examinations. Since this disorder of the hip is progressive and early detection and treatment are essential, the American Academy of Pediatrics has suggested guidelines for examination and treatment of children suspected of having development hip dysplasia. They suggest referral to a pediatric orthopedist if an infant has a positive Ortolani or Barlow test. For infants with mild hip clicks, they suggest the child be seen by the regular pediatrician in two weeks for follow up since most benign hip clicks will resolve within that time period. If signs of hip dysplasia

KEY TERMS

Abduction—Turning away from the body.

Acetabulum—The large cup-shaped cavity at the junction of pelvis and femur (thigh bone).

Adduction—Movement toward the body.

Bracing—Using orthopedic devices to hold joints or limbs in place.

Dislocation—The displacement of bones at a joint or the displacement of any part of the body from its normal position.

Dysplasia—Abnormal changes in cells.

Femur—The thigh bone.

Orthosis—An external device, such as a splint or a brace, that prevents or assists movement.

Placenta—The organ that provides oxygen and nutrition from the mother to the unborn baby during pregnancy. The placenta is attached to the wall of the uterus and leads to the unborn baby via the umbilical cord.

Splint—A thin piece of rigid or flexible material that is used to restrain, support, or immobilize a part of the body while healing takes place.

are still present after two weeks, it is recommended that the child be seen by a pediatric orthopedist. If double or triple diapering is recommended by the pediatrician after the initial newborn exam, it is imperative that parents follow up with their pediatrician for a more extensive examination of the hips soon after the newborn comes home.

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Congenital hypothyroidism *see* **Hypothyroidism**

Congenital megacolon *see* **Hirschsprung's disease**

Congenital thymic hypoplasia *see* **DiGeorge syndrome**

Congenital ureter anomalies

Definition

The ureter drains urine from the kidney into the bladder. Not simply a tube, the ureter is an active organ that propels urine forward by muscular action. It has a valve at its bottom end that prevents urine from flowing backward into the kidney. Normally there is one ureter on each side of the body for each kidney. However, among the many abnormalities of ureteral development, duplication is quite common. Ureters may also be malformed in a variety of ways—some harmful, others not.

Description

There are many different types of ureter anomalies. Ureters can be duplicated completely or partially, they can be in the wrong place, they can be deformed, and they can end in the wrong place. The trouble these abnormalities bring is directly related to their effect on the flow of urine. As long as urine flows normally through them, and only in one direction, no harm is done. A description of ureter anomalies follows.

Duplication of ureters is quite common, either in part or completely. Kidneys are sometimes duplicated as well. Someone may have four kidneys and four ureters or two kidneys, half of each drained by a separate ureter, or a single kidney with two, three, or four ureters attached. As long as urine can flow easily in the correct direction, such malformations may never be detected. If, however, one of the ureters has a dead end, a stricture or stenosis (narrowing), or a leaky ureterovesical valve (between the ureter and bladder), infection is the likely result.

Stricture or stenosis of a ureter prevents urine from flowing freely. Whenever flow is obstructed in the body—urine, bile, mucus, or any other liquid—infection follows. Ureters can be obstructed anywhere along their course, though the ureterovesical valve is the most common place.

A ureter may have an ectopic (out-of-place) orifice (opening): it may enter the bladder, or even another structure, where it does not belong and therefore lack an adequate valve to control reflux.

The primary ureter, or a duplicate, may not even reach the bladder, but rather terminate in a dead end. Urine will stagnate there and eventually cause infection.

A ureter can be perfectly normal but in the wrong place, such as behind the vena cava (retrocaval ureter), the large vein in the middle of the abdomen. In this case the ureter may be pinched by the vena cava so that flow is hindered. Other abnormal locations may also lead to compression and impaired flow.

Besides infection, urine that backs up causes the ureter and the kidney to expand or dilate. Eventually, the kidney stops functioning because of the back pressure. This condition is called hydronephrosis (a kidney swollen with urine).

Demographics

The urogenital system is more likely than any other organ system to have birth defects, and they can occur in endless variety. Congenital ureter anomalies affect as many as one in every 160 individuals.

Causes and symptoms

In general, the causes of birth defects are multiple and often as of 2004 unknown. Furthermore, the precise cause of specific birth defects has only rarely been identified. Such is the case with congenital ureteral anomalies.

Practically the only symptom generated by ureteral abnormalities is urinary tract infection. A lower tract infection, in the bladder, is called **cystitis**. In children it may cause **fever** and systemic symptoms, but in adults it causes only cloudy, burning, and frequent urine. Upper tract infections, by contrast, can be serious for both adults and children, causing high fevers, back **pain**, severe generalized discomfort, and even leading to kidney failure or septicemia (infection spreading throughout the body by way of the blood stream).

In rare cases, urine from an ectopic ureter will bypass the bladder and dribble out of the bottom somewhere, through a natural orifice like the vagina or a completely separate unnatural opening.

KEY TERMS

Congenital—Present at birth.

Contrast agent—Also called a contrast medium, this is usually a barium or iodine dye that is injected into the area under investigation. The dye makes the interior body parts more visible on an x-ray film.

Cystoscopy—A diagnostic procedure in which a hollow lighted tube (cystoscope) is used to look inside the bladder and the urethra.

Ectopic—Out of place or located away from the normal position.

Retrocaval ureter—A ureter that is located behind the vena cava blood vessel.

Septicemia—A systemic infection due to the presence of bacteria and their toxins in the bloodstream. Septicemia is sometimes called blood poisoning.

Ureterovesical valve—A sphincter (an opening controlled by a circular muscle), located where the ureter enters the bladder, that keeps urine from flowing backward toward the kidney.

Urogenital—Refers to both the urinary system and the sexual organs, which form together in the developing embryo.

Diagnosis

For children experiencing serious or recurrent urinary tract infections, the pediatrician will search for underlying abnormalities. Cystoscopy (looking into the bladder with a thin telescope-like instrument) and **x rays** with a contrast agent to illuminate the urinary system will usually identify the defect. **Computed tomography** scans (CT) and **magnetic resonance imaging** (MRI) may provide additional information. Urine cultures to identify the infecting germs will be repeated frequently until the problem is corrected.

Treatment

Sometimes the recurring infections caused by flow abnormalities can be treated with repeated and changing courses of **antibiotics**. Over time, the infecting germs develop resistance to most treatments, especially the safer ones. If it can be done safely, it is better to repair the defect surgically. Urologists have various approaches to urine drainage that range from simply reimplanting a ureter into

the bladder, in such a way that an effective valve is created, to building a new bladder out of a piece of bowel.

Alternative treatment

There are botanical and homeopathic treatments available for urinary tract infection. None can take the place of correcting a problem that is occurring because of a malformed or dysfunctional organ system. Once correction of the cause is addressed and there is unimpeded flow of urine, adequate fluid intake can contribute to prevention of future infections.

Prognosis

As long as damage to the kidneys from infection or back pressure has not become significant, the surgical repair of troublesome ureteral defects produces excellent long-term results in the great majority of cases. Monitoring for recurrent infections is always a good idea, and occasional checking of kidney function will detect hidden ongoing damage.

Prevention

The cause of congenital ureter anomalies is not known. There is no prevention.

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Conjunctivitis

Definition

Conjuctivitis is an inflammation resulting in redness of the lining of the white part of the eye and the underside of the eyelid (conjunctiva) that can be caused by infection, allergic reaction, or physical agents like infrared or ultraviolet light.

Description

Conjunctivitis is the inflammation of the conjunctiva, a thin, delicate membrane that covers the eyeball and lines the eyelid. Conjunctivitis is an extremely common eye disease because the conjunctiva is continually exposed to microorganisms and environmental agents that can cause infections or allergic reactions. Conjunctivitis can be acute or chronic depending on how long the condition lasts, the severity of symptoms, and the type of organism or agent involved. It can affect one or both eyes and, if caused by infection, can be very easily transmitted to others during close physical contact, particularly among children in a school or daycare setting. Other names for conjunctivitis include pink eye and red eye.

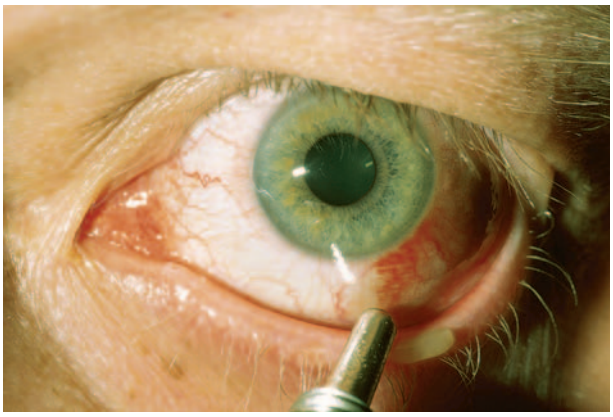
Demographics

Conjunctivitis is the most common eye infection of childhood. It occurs so frequently that records are not kept, so exact demographic information has not been amassed.

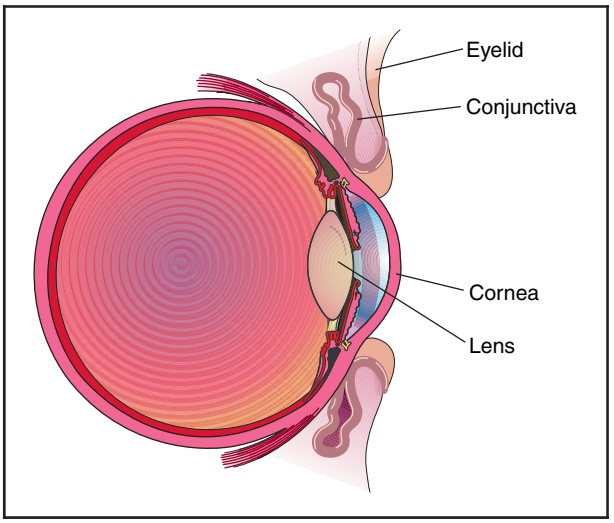
Causes and symptoms

Conjunctivitis may be caused by a viral infection, such as a cold; acute respiratory infection; or other disease such as **measles**, **herpes simplex**, or herpes zoster. Symptoms include mild to severe discomfort in one or both eyes; redness; swelling of the eyelids; and watery, yellow, or green discharge. Symptoms may last anywhere from several days to two weeks. Infection with an adenovirus, however, may also cause a significant amount of pus-like discharge and a scratchy, foreign-body-sensation in the eye. These symptoms may be accompanied by swelling and tenderness of the lymph nodes near the ear.

Bacterial conjunctivitis can occur in adults and children and is caused by organisms such as *Staphylococcus*, *Streptococcus*, and *Hemophilus*. Symptoms of bacterial conjunctivitis include a pus-like discharge and crusty eyelids after awakening. Redness of the conjunctiva can be mild to severe and may be accompanied by swelling.



An antibotic eye ointment is applied to relieve bacterial conjunctivitis. *(© T. Bannor/Custom Stock Medical Photo, Inc.)*



Conjunctivitis is the inflammation of the conjunctiva, a thin, delicate membrane that covers the eyeball and lines the eyelid. It may be caused by a viral infection, such as a cold or acute respiratory infection, or by such diseases as measles, herpes simplex, or herpes zoster. *(Illustration by Electronic Illustrators Group.)*

Persons with symptoms of conjunctivitis who are sexually active may possibly be infected with the bacteria that cause either gonorrhea or chlamydia. There may be large amounts of pus-like discharge, and symptoms may include intolerance to light (photophobia), watery mucus discharge, and tenderness in the lymph nodes near the ear that may persist for up to three months.

Conjunctivitis may also be caused by environmental hazards, such as wind, smoke, dust, and allergic reactions caused by pollen, dust, or grass. Symptoms range from **itching** and redness to a mucus discharge. Persons who wear **contact lenses** may develop allergic conjunctivitis caused by various eye solutions used and the foreign proteins contained in them.

Other less common causes of conjunctivitis include exposure to sun lamps or the electrical arcs used during welding and problems with inadequate drainage of the tear ducts.

Diagnosis

An accurate diagnosis of conjunctivitis centers on taking a patient's history to learn what symptoms are being experienced, when symptoms began, and other predisposing factors, such as upper respiratory complaints, **allergies**, **sexually transmitted diseases**, herpes simplex infections, and exposure to persons with pink eye. It may be helpful to learn whether an aspect of an individual's occupation may be the cause, for example, welding. Diagnostic tests are usually not indicated unless initial treatment fails or an infection with gonorrhea or chlamydia is suspected. In such cases, the discharge may be cultured and tested to determine the organism responsible for causing the condition.

Treatment

The treatment of conjunctivitis depends on what caused the condition. In all cases, warm compresses applied to the affected eye several times a day may help to reduce discomfort.

Conjunctivitis due to a viral infection, particularly those due to adenoviruses, are usually treated by applying warm compresses to the affected area and using topical antibiotic ointments to prevent secondary bacterial infections.

Viral conjunctivitis caused by herpes simplex should be referred to an ophthalmologist. Topical steroids are commonly prescribed in combination with antiviral therapy.

In cases of bacterial conjunctivitis, a physician may prescribe an antibiotic eye ointment or eye drops containing sodium sulfacetamide (Sulamyd) to be applied daily for seven to 14 days. If, after 72 hours, the condition does not improve, a physician or primary care provider should be notified, because the bacteria involved may be resistant to the antibiotic used or the cause may not be bacterial.

For cases of conjunctivitis caused by a gonococcal organism, a physician may prescribe an intramuscular injection of ceftriaxone (Rocephin) and a topical antibiotic ointment containing erythromycin or bactracin to be applied four times daily for two to three weeks. Sexual partners should also be treated.

With accompanying chlamydia infection, a topical antibiotic ointment containing erythromycin (Ilotycin) may be prescribed to be applied one to two times daily. In addition, oral erythromycin or tetracycline therapy

KEY TERMS

Adenovirus—A type of virus that can cause upper respiratory tract infections.

Chlamydia—The most common bacterial sexually transmitted disease in the United States. It often accompanies gonorrhea and is known for its lack of evident symptoms in the majority of women.

Gonococcal—Refers to the bacterium *Neisseria gonorrheae*. This bacterium causes gonorrhea, a sexually transmitted infection of the genitals and urinary tract. The gonococcal organism may occasionally affect the eye, causing blindness if not treated.

Herpes simplex virus—A virus that can cause fever and blistering on the skin and mucous membranes. Herpes simplex 1 infections usually occur on the face (cold sores) and herpes simplex 2 infections usually occur in the genital region.

Herpes zoster virus—Acute inflammatory virus that attacks the nerve cells on the root of each spinal nerve with skin eruptions along a sensory nerve ending. It causes chickenpox and shingles. Also called varicella zoster virus.

Staphylococcus—Any of several species of spherical bacteria that occur in groups of four or in irregular clusters. They can infect various parts of the body, especially the skin and mucous membranes.

Streptococcus—Plural, streptococci. Any of several species of spherical bacteria that form pairs or chains. They cause a wide variety of infections including scarlet fever, tonsillitis, and pneumonia.

may be indicated for three to four weeks. Again, sexual partners should also be treated.

Allergic conjunctivitis can be treated by removing the allergic substance from a person's environment, if possible; by applying cool compresses to the eye; and by administering eye drops four to six times daily for four days. Also, the antihistamine diphenhydramine hydrochloride (Benadryl) may help to relieve itchy eyes.

Prognosis

If treated properly, the prognosis for conjunctivitis is good. Conjunctivitis caused by an allergic reaction should clear up once the allergen is removed. However, allergic conjunctivitis will likely recur if the individual again comes into contact with the particular allergen. Conjunctivitis caused by bacteria or a virus, if treated properly, is usually resolved in ten to 14 days. If there is no relief of symptoms in 48 to 72 hours, or there is moderate to severe eye **pain**, changes in vision, or the conjunctivitis is suspected to be caused by herpes simplex, a physician should be notified immediately. If untreated or if treatment fails and is not corrected, conjunctivitis may cause visual impairment by spreading to other parts of the eye, such as the cornea.

Prevention

Conjunctivitis can, in many cases, be prevented, or at least the course of the disease can be shortened by following these simple practices:

- frequently washing hands with antiseptic soap and using single-use towels while the disease continues
- avoiding chemical irritants and known allergens
- in an area where welding occurs, using the proper protective eye wear and screens to prevent damaging the eyes
- using a clean tissue to remove discharge from eyes and washing hands to prevent the spread of infection
- if medication is prescribed, finishing the course of **antibiotics**, as directed, to make sure that the infection is cleared up and does not recur
- avoiding close contact, such as vigorous physical activities, with other persons until symptoms resolve

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Constipation

Definition

Constipation is an acute or chronic condition in which bowel movements occur less often than usual or consist of hard, dry stools that are painful or difficult to pass. Although constipation is a relative term, with normal patterns of bowel movements varying widely from person to person, generally an adult who has not had a bowel movement in three days or a child who has not had a bowel movement in four days is considered constipated. Infants who are still exclusively breastfed may go seven days without a stool.

Description

The colon (the large intestine) absorbs water while forming waste products (the stool) from digested food. Muscle contractions in the colon (peristalsis) push the stool toward the rectum. By the time the stool reaches the rectum, it is solid because most of the water has been absorbed. However, hard, dry stools and constipation occur when too much water is absorbed by the colon from the stool, which can result from the muscle of the colon contracting too slowly. Constipation is also referred to as irregularity of bowels or lack of regular bowel movements.

Constipation can occur at any age and is more common among individuals who resist the urge to move their bowels at their body's signal. This often happens when children start school or enter daycare. They may feel shy about asking permission to use the bathroom, they may be involved in more enjoyable activities and may not want to stop, or they may be rushed when using the bathroom and not have time to complete the bowel movement. Once constipation has developed and bowel movements become painful or more difficult, the child will attempt to go even less often, and the constipation will worsen.

Although this condition is rarely serious, it can lead to the following:

- tearing of the mucosal membrane of the anus (especially in children), which can cause bleeding and the development of an anal fissure
- bowel obstruction
- chronic constipation
- hemorrhoids (a mass of dilated veins in swollen tissue around the anus)
- hernia (a protrusion of an organ through a tear in the muscle wall)
- spastic colitis (**irritable bowel syndrome**, a condition characterized by alternating periods of **diarrhea** and constipation)
- laxative dependency

Less commonly, chronic constipation may be a symptom of colorectal **cancer**, depression, diabetes, diverticulosis (small pouches in the muscles of the large intestine), **lead poisoning**, or Parkinson's disease (in adults) and should be investigated by a doctor.

Demographics

Constipation is a common complaint in children, occurring in up to 10 percent of youngsters. It accounts for approximately 3 percent of pediatric outpatient visits and 25 percent of visits to a pediatric gastroenterologist.

Causes and symptoms

Constipation usually results from not getting enough **exercise**, not drinking enough fluids (especially water), delays in going to the bathroom when there is the urge to defecate, or from a diet that does not include an adequate amount of fiber-rich foods such as beans, bran cereals, fruits, raw vegetables, rice, and whole-grain breads. Eating too many dairy products such as milk, cheese, yogurt, and ice cream may also result in harder stools. Constipation in children often occurs when they hold back bowel movements for various reasons, such as when they are not ready for **toilet training** or are afraid of toilet training.

Other less common causes of constipation include anal fissure (a tear or crack in the lining of the anus); chronic kidney failure; colon or rectal cancer; depression; hypercalcemia (abnormally high levels of calcium in the blood); **hypothyroidism** (underactive thyroid gland); illness requiring complete bed rest; and irritable bowel syndrome. Stress and travel can also contribute to constipation, as well as other changes in bowel habits.

Constipation can also be a side effect of the use of the following medications, many of which are not commonly used by children:

- aluminum salts in antacids
- antihistamines
- antipsychotic drugs
- aspirin
- belladonna (*Atopa belladonna,* a source of atropine, a medication used to relieve spasms and dilate the pupils of the eye)

- beta blockers (medications used to stabilize irregular heartbeat, lower high blood pressure, and reduce chest **pain**)
- blood pressure medications
- calcium channel blockers (medication prescribed to treat high blood pressure, chest pain, some types of irregular heartbeat and **stroke**, and some non-cardiac diseases)
- diuretics (drugs that promote the formation and secretion of urine)
- iron or calcium supplements
- narcotics (potentially addictive drugs that relieve pain and cause mood changes)
- tricyclic **antidepressants** (medications prescribed to treat chronic pain, depression, headaches, and other illnesses)

A child who is constipated may feel bloated, have a **headache**, swollen abdomen, or pass rock-like feces; or strain, bleed, or feel pain during bowel movements. A constipated baby may strain, cry, draw the legs toward the abdomen, or arch the back when having a bowel movement. Newborns and young infants may also strain, turn red in the face, grunt and draw legs up when passing normal, soft stool. If the stool is not hard (rabbit pellet in consistency), then these infants are not considered constipated.

When to call the doctor

Most people become constipated once in a while, but a doctor should be contacted if significant changes in bowel patterns last for more than a week or if symptoms continue more than three weeks after increasing activity and fiber and fluid intake.

In addition, a doctor should be called if an infant younger than two months is constipated, or if an infant (except those that are exclusively breastfed) goes three days without a stool. If **vomiting** or irritability is also present, then the doctor should be called immediately. A doctor should also be consulted if a child is holding back bowel movements (in order to resist toilet training) or whenever constipation occurs after starting a new prescription, vitamin, or mineral supplement or is accompanied by blood in the stools, changes in bowel patterns, **fever**, and rectal or abdominal pain.

Diagnosis

The child's symptoms and medical history help a primary care physician to diagnose constipation. The doctor uses his fingers to see if there is a hardened mass in the abdomen and may perform a rectal examination. Other diagnostic procedures include a barium enema, which reveals blockage inside the intestine; laboratory analysis of blood and stool samples for internal bleeding or other symptoms of systemic disease; and a sigmoidoscopy (examination of the sigmoid area of the colon with a flexible tube equipped with a magnifying lens).

Treatment

Constipation is usually a temporary problem in children and no cause for concern. A child with constipation should be instructed to drink an adequate amount of water each day (six to eight glasses), exercise on a regular basis, and eat a diet high in soluble and insoluble fibers. Soluble fibers include pectin, flax, and gums; insoluble fibers include psyllium and brans from grains like wheat and oats. Fresh fruits and vegetables contain both soluble and insoluble fibers. Dietary fiber intake should be increased gradually, along with an increase in water consumption, in order to produce soft, bulky stools.

Constipation in infants may be treated by the following:

- if over two months of age, feeding the infant 2–4 ounces (60–120 ml) of fruit juice (grape, pear, apple, cherry, or prune) twice a day
- if over four months of age and the infant has begun solid foods, feeding the baby foods with high fiber content (such as peas, beans, apricots, prunes, peaches, pears, plums, and spinach) twice a day

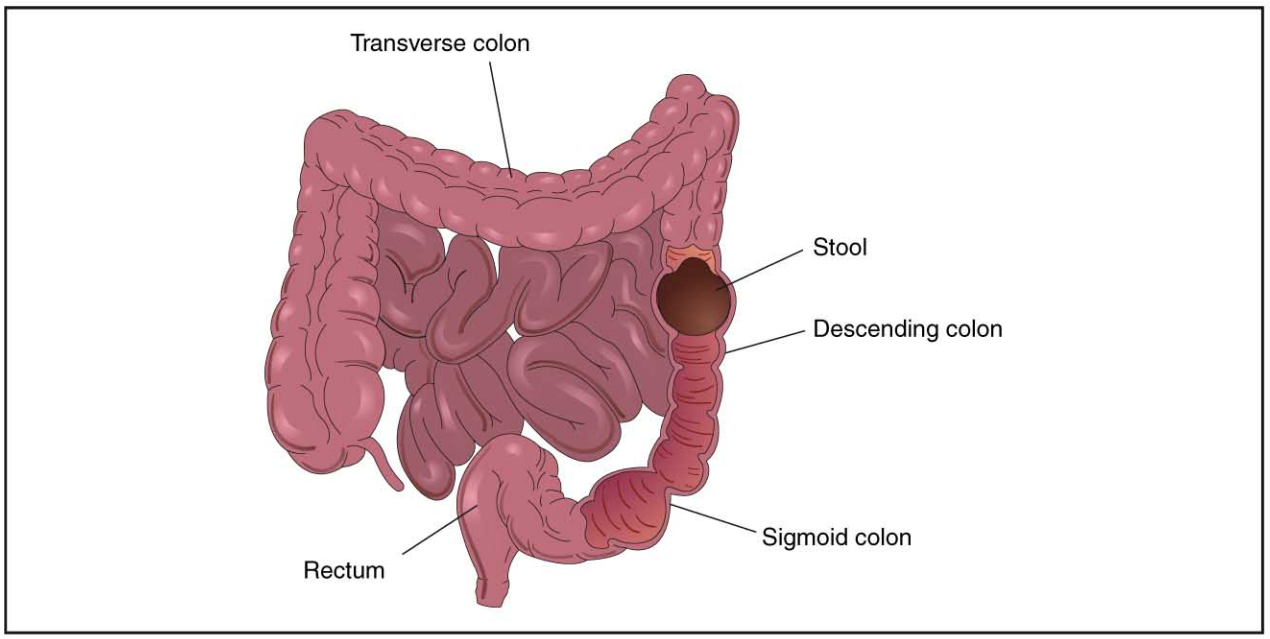
If changes in diet and activity fail to relieve occasional constipation, an over-the-counter laxative may be used for a few days. Preparations that soften stools or add bulk (bran, psyllium) work more slowly but are safer than Epsom salts and other harsh **laxatives** or herbal laxatives containing senna (*Cassia senna*) or buckthorn (*Rhamnus purshianna*), which can harm the nerves and lining of the colon. A child who is experiencing abdominal pain, **nausea**, or vomiting should not use a laxative. Laxatives should not be used for a long period, because the child can become dependent on them.

A warm-water or mineral oil enema can relieve constipation in children with severe or stubborn cases of constipation. However, laxatives or enemas should not be given to children without instruction from a doctor.

If a child has an impacted bowel, the doctor can insert a gloved finger into the rectum and gently dislodge the hardened feces.

Alternative treatment

Castor oil, applied topically to the abdomen and covered by a heat source (a heating pad or hot water



Constipation is an acute or chronic condition in which bowel movements occur less often than usual or consist of hard, dry stools that are painful or difficult to pass. *(Illustration by Electronic Illustrators Group.)*

bottle) can help relieve constipation when used nightly for 20 to 30 minutes.

Acupressure

This needleless form of acupuncture is said to relax the abdomen, ease discomfort, and stimulate regular bowel movements when diet and exercise fail to do so. After lying down, the child closes his or her eyes and takes a deep breath. For two minutes, the child or parent applies gentle fingertip pressure to a point about 2.5 in (14 cm) below the navel.

Acupressure can also be applied to the outer edges of one elbow crease and maintained for 30 seconds before pressing the crease of the other elbow. This should be done three times a day to relieve constipation.

Aromatherapy

Six drops of rosemary (*Rosmarinus officinalis*) and six drops of thyme (*Thymus* spp.) diluted by one ounce of almond oil, olive oil, or another carrier oil can relieve constipation when used to massage the abdomen.

Herbal therapy

A variety of herbal therapies can be useful in the treatment of constipation. Several herbs, including chamomile (*Matricaria recutita*), dandelion (*Taraxacum mongolicum*), and burdock (*Arctium lappa*), act as bitters, stimulating the movement of the digestive and excretory systems.

Homeopathy

Homeopathy also can offer assistance with constipation. There are acute remedies for constipation that can be found in one of the many home remedy books on homeopathic medicine. A constitutional prescription also can help rebalance someone who is struggling with chronic constipation.

Massage

Massaging the leg from knee to hip in the morning, at night, and before trying to move the bowels is said to relieve constipation. There is also a specific Swedish massage technique that can help relieve constipation.

Yoga

The knee-chest position, said to relieve gas and stimulate abdominal organs, involves the following:

- standing straight with arms at the sides
- lifting the right knee toward the chest
- grasping the right ankle with the left hand
- pulling the leg as close to the chest as possible
- holding the position for about eight seconds
- repeating these steps with the left leg

The cobra position, which can be repeated as many as four time a day, involves the following:

- lying on the stomach with legs together
- placing the palms just below the shoulders, holding elbows close to the body
- inhaling, then lifting the head (face forward) and chest off the floor
- keeping the navel in contact with the floor
- looking as far upward as possible
- holding this position for three to six seconds
- exhaling and lowering the chest

Prognosis

Changes in diet and exercise can often eliminate constipation. However, childhood constipation can sometimes be difficult to treat when a child, after having a painful experience, makes a decision to resist and delay going to the bathroom. These cases often require prolonged support, explanation, and medical treatment.

Prevention

Avoiding constipation by making lifestyle changes is easier than treating it. Most American adults only consume between 11 to 18 grams of fiber a day, but to prevent constipation, consumption of 30 to 35 grams of fiber (an amount equal to five servings of fruits and vegetables, and a large bowl of high-fiber cereal) and between six and eight glasses of water each day can generally prevent constipation. A suggested goal for dietary fiber intake during childhood and **adolescence** is consumption in grams equivalent to the age of the child plus 5 grams per day.

Sitting on the toilet for 10 minutes at the same time every day, preferably after a meal, can induce regular bowel movements. This may not become effective for a few months, and it is important to defecate whenever necessary.

Fiber supplements containing psyllium (*Plantago psyllium*) usually become effective within about 48 hours and can be used every day without causing dependency. Powdered flaxseed (*Linium usitatissimum*) works the same way. Insoluble fiber, like wheat or oat bran, is as effective as psyllium but may give the child gas at first.

Parental concerns

Constipation can be a frustrating problem for both children and their parents. Parents need to work closely with the doctor to determine why the child is constipated and to develop an appropriate treatment strategy.

KEY TERMS

Constipation—Difficult bowel movements caused by the infrequent production of hard stools.

Dietary fiber—Mostly indigestible material in food that stimulates the intestine to peristalsis.

Peristalsis—Slow, rhythmic contractions of the muscles in a tubular organ, such as the intestines, that move the contents along.

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Contact dermatitis

Definition

Contact **dermatitis** is the name for any skin inflammation that occurs when the skin's surface comes in contact with a substance originating outside the body. There are two major categories of contact dermatitis, irritant and allergic. Irritant dermatitis is essentially a direct injury to the skin, caused by such compounds as acids, alkalis, phenol, and detergents. The immune system is not involved in irritant dermatitis, and the person's skin is damaged without prior sensitization.

In allergic dermatitis, however, the patient's skin reacts to a substance to which it has become sensitized. A third type of dermatitis, photo contact dermatitis, is triggered by exposure of the skin to light following the application of certain cosmetics or chemicals. Photo contact dermatitis may be either irritant or allergic.

Description

Contact dermatitis may be either an acute or chronic skin disorder. In general, allergic contact dermatitis is more severe and acute in its onset than irritant contact dermatitis. In irritant contact dermatitis, the rash is usually limited to the area that was exposed to the substance, whereas in allergic contact dermatitis, the rash often spreads beyond the area directly exposed to the allergen. Irritant contact dermatitis most commonly affects the hands, while allergic contact dermatitis may be found on almost any part of the body, including the armpits and genitals. Allergic contact dermatitis is more likely to involve swelling of the skin and the development of small fluid-filled blisters than irritant contact dermatitis.

Photo contact dermatitis is usually limited to the area of skin exposed to direct light. If the substance that was applied to the skin was changed to an irritant by light exposure, the primary symptom is a burning sensation resembling **sunburn**. If the substance was changed to an allergen, the primary sensation is **itching**.

Demographics

Contact dermatitis is a common complaint in people of all ages, in part because of the large number of potential irritants and allergens in the contemporary environment. One textbook on contact dermatitis runs to over 1,100 pages of descriptions of the various manufactured products and other substances that can cause these skin reactions.

In the United States, contact dermatitis ranks among the top 10 reasons for visits to primary care doctors and accounts for 7 percent of all visits to dermatologists. Every year between 10 and 50 million Americans in all age groups develop an allergic rash following contact with **poison ivy** or **poison oak**.

About 20 percent of children in the general United States population develop allergic contact dermatitis at some point prior to **adolescence**. Between 20 percent and 35 percent of healthy children react to one or more allergens on standard patch tests. Children of parents with allergic contact dermatitis have a 60 percent greater chance of having a positive reaction on a patch test themselves.

Contact dermatitis is more likely to affect Caucasians than African, Asian, or Native Americans. People with fair skin and red hair are particularly susceptible to contact dermatitis.

With regard to sex, girls are twice as likely as boys to develop both irritant and allergic skin reactions.

Causes and symptoms

Irritant contact dermatitis

Irritant contact dermatitis (ICD) is the more commonly reported of the two kinds of contact dermatitis, and is seen in about 80 percent of cases. It can be caused by soaps, detergents, solvents, adhesives, fiberglass, and other substances that are able to directly injure the skin by breaking or removing the protective layers of the upper epidermis. Irritants remove lipids, which are fatty substances that help to maintain the integrity of skin cells; irritants also damage the skin's ability to hold water. A common form of irritant contact dermatitis in infants is **diaper rash**, which develops when the protective epidermal layer of the baby's skin is damaged by long periods of contact with fecal matter and urine.

Most attacks of ICD are slight and confined to the hands and forearms but can affect any part of the body that comes in contact with an irritating substance. The symptoms can take many forms: redness, itching, crusting, swelling, blistering, oozing, dryness, scaling, thickening of the skin, and a feeling of warmth at the site of contact. In extreme cases, severe blistering can occur and open sores can form. Jobs that require frequent skin exposure to water, such as hairdressing and food preparation, can make the skin more susceptible to ICD.

Thin, moist, or already damaged skin is more susceptible to ICD than thick, dry, or intact skin.

Allergic contact dermatitis

Allergic contact dermatitis (ACD) results when repeated exposure to an allergen (an allergy-causing substance) triggers an immune response that inflames the skin. There are two phases in the development of ACD: an induction phase, in which the allergen penetrates the epidermis and is processed by an antigen-presenting cell; and an elicitation phase, in which the sensitized person has a second exposure to the allergen, which produces an inflammatory response several hours or days after the second exposure. Sensitivity to the specific allergen is often lifelong.

Tens of thousands of drugs, pesticides, cosmetics, food additives, commercial chemicals, and other substances have been identified as potential allergens. Fewer than 30, however, are responsible for the majority of ACD cases. Common culprits include poison ivy, poison oak, and **poison sumac**; fragrances and preservatives in cosmetics and personal care products, such latex items as gloves and condoms; and formaldehyde. Many people find that they are allergic to the nickel in inexpensive jewelry; some adolescents find that they are allergic to the metal alloys used in orthodontic braces. ACD is usually confined to the area of skin that comes in contact with the allergen. Symptoms range from mild to severe and resemble those of ICD.

Photo contact dermatitis

In photo contact dermatitis, certain substances undergo chemical changes as a result of exposure to light that transform them into either irritants or allergens. Aftershave lotions, **sunscreens**, and certain topical sulfa drugs may be changed into allergens, while coal tar and certain oils used in manufacturing may become irritants after light exposure.

When to call the doctor

Contact dermatitis is not a medical emergency. It can often be treated at home once the irritant or allergen has been identified. A visit to the doctor may be necessary, however, in order to identify the cause(s) as well as obtain specific recommendations for treatment.

Diagnosis

Diagnosis of contact dermatitis begins with a physical examination and asking the patient questions about his or her health and daily activities. When contact dermatitis is suspected, the doctor attempts to learn as much as possible about the child or adolescent's school, **sports** participation, hobbies, favorite jewelry, use of medications and cosmetics—anything that might shed light on the source of the problem. The doctor will ask when the symptoms started, whether this is the first time they occurred, whether the rash is spreading, whether the primary sensation is itching or burning, and how severe the itching or burning feels.

In some cases, an examination of the home or school may be undertaken; in one interesting case, the doctors discovered that a rash on the back of the child's thighs was an allergic reaction to nickel in the metal parts of the chairs in the child's school. If the dermatitis is mild, responds well to treatment, and does not recur, ordinarily the investigation is at an end. More difficult cases require patch testing to identify the specific allergen.

Two methods of patch testing are used in the early 2000s. The most widely used method, the Finn chamber method, employs a multiwell aluminum patch. Each well is filled with a small amount of the allergen being tested and the patch is taped to normal skin on the patient's upper back. After 48 hours, the patch is removed and an initial reading is taken. A second reading is made a few days later.

The second method of patch testing involves applying a small amount of the test substance to directly to normal skin and covering it with a dressing that keeps air out and keeps the test substance in (occlusive dressing). After 48 hours, the dressing is taken off to see if a reaction has occurred. Identifying the allergen may require repeated testing, can take weeks or months, and is not always successful. Moreover, patch testing works only with ACD, though it is considered an essential step in ruling out ICD.

In a few cases, the doctor may take a skin biopsy in order to rule out certain infectious skin diseases.

Treatment

The best treatment for contact dermatitis is to identify the allergen or irritating substance and avoid further contact with it. If the culprit is, for instance, a cosmetic, avoidance is a simple matter, but in some situations, avoidance may be impossible or force the sufferer to make drastic changes in his or her life. Barrier creams and such protective clothing as gloves, masks, and long-sleeved shirts are coping devices to reduce the chance of contact dermatitis when avoidance is impossible, though they are not always effective.

For the symptoms themselves, treatments in mild cases include cool compresses and nonprescription lotions and ointments. Diaper rash is often treated by applying various emollient preparations that restore lipids to the child's skin. In older children and adolescents, more severe cases of contact dermatitis are treated with corticosteroids applied to the skin or taken orally. Contact dermatitis that leads to a bacterial skin infection is treated with **antibiotics**. Although **antihistamines** do not cure contact dermatitis, the doctor may prescribe them to relieve severe itching.

Alternative treatment

Herbal remedies have been used for centuries to treat skin disorders including contact dermatitis. An experienced herbalist can recommend the remedies that will be most effective for an individual's condition. Among the herbs often recommended are the following:

- Burdock (*Arctium lappa*) minimizes inflammation and boosts the immune system. It is taken internally as a

tea or tincture (a concentrated herbal extract prepared with alcohol).

- Calendula (*Calendula officinalis*) is a natural antiseptic and anti-inflammatory agent. It is applied topically in a lotion, ointment, or oil to the affected area.
- Aloe (*Aloe barbadensis*) gel soothes skin irritations. The gel is applied topically to the affected area.

A homeopath treating a patient with contact dermatitis will do a thorough investigation of the individual's history and exposures before prescribing a remedy. One homeopathic remedy commonly prescribed to relieve the itching associated with contact dermatitis is *Rhus toxicodendron*, which is taken internally three to four times daily.

Poison ivy, poison oak, and poison sumac are common culprits in cases of allergic contact dermatitis. Within fifteen minutes of exposure to these plants, rash development may be prevented by washing the area with soap and water. The leaves of jewelweed (*Impatiens* spp.), which often grows near poison ivy, may neutralize the poison-ivy allergen if rubbed on the skin right after contact. Several topical remedies may help relieve the itching associated with allergic contact dermatitis, including the juice of plantain leaves (*Plantago major*); a paste made of equal parts of green clay and goldenseal root (*Hydrastis canadensis*); a paste made of salt, water, clay, and peppermint (*Mentha piperita*) oil; and calamine lotion.

Prognosis

If the offending substance is promptly identified and avoided, the chances of a rapid and complete recovery are excellent. Otherwise, symptom management—not cure—is the best doctors can offer. Sensitivity to allergens is typically lifelong. For a few people, contact dermatitis becomes a chronic and disabling condition that can have a profound effect on quality of life.

Prevention

Avoidance or substitution of known or suspected allergens or irritating substances is the best prevention. If avoidance is difficult, barrier creams and protective clothing can be tried. Skin that comes in contact with an offending substance should be thoroughly washed as soon as possible.

Parental concerns

Parents should be concerned primarily with identifying the cause(s) of a child or adolescent's contact dermatitis, as treatment is often ineffective until the offending substance can be removed or avoided. Most cases of contact dermatitis are mild and can be treated without disrupting the child's school routine or severely affecting his or her quality of life. In some cases, parents may find it helpful to consult a dermatologist to identify the specific causes and to suggest products that can be substituted for those that are causing the skin reactions.

See also Diaper rash; Poison ivy, oak, and sumac; Rashes.

KEY TERMS

Antibiotics—Drugs that are designed to kill or inhibit the growth of the bacteria that cause infections.

Corticosteroids—A group of hormones produced naturally by the adrenal gland or manufactured synthetically. They are often used to treat inflammation. Examples include cortisone and prednisone.

Dermatologist—A physician that specializes in diseases and disorders of the skin.

Epidermis—The outermost layer of the human skin.

Immune response—A physiological response of the body controlled by the immune system that involves the production of antibodies to fight off specific foreign substances or agents (antigens).

Lipids—Organic compounds not soluble in water, but soluble in fat solvents such as alcohol. Lipids are stored in the body as energy reserves and are also important components of cell membranes. Commonly known as fats.

Topical—Not ingested; applied to the outside of the body, for example to the skin, eye, or mouth.

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Howard Baker

Contact lenses *see* **Eyeglasses and contact lenses**

Contraception

Definition

Contraception (birth control) prevents pregnancy by interfering with the normal process of ovulation, fertilization, and implantation. There are different kinds of birth control that act at different points in the process.

Purpose

Every month a woman's body begins the process that can potentially lead to pregnancy. An egg (ovum) matures, the mucus that is secreted by the cervix (a cylindrical-shaped organ at the lower end of the uterus) changes to be more inviting to sperm, and the lining of the uterus grows in preparation for receiving a fertilized egg. Any woman who wants to prevent pregnancy must use a reliable form of birth control. Birth control (contraception) is designed to interfere with the normal process and prevent the pregnancy that could result. There are different kinds of birth control that act at different points in the process, from ovulation through fertilization to implantation. Each method has its own side effects and risks. Some methods are more reliable than others.

Although there are many different types of birth control, they can be divided into a few groups based on how they work. These groups include:

- Hormonal methods: These use medications (hormones) to prevent ovulation. Hormonal methods include birth control pills (**oral contraceptives**), Depo Provera injections, and Norplant.
- Barrier methods: These methods work by preventing the sperm from getting to and fertilizing the egg. Barrier methods include male **condom** and female condom, diaphragm, and cervical cap. The condom is the only form of birth control that also protects against **sexually transmitted diseases**, including human **immunodeficiency** virus (HIV) that causes acquired immune deficiency syndrome (AIDS).
- Spermicides: These medications kill sperm on contact. Most spermicides contain nonoxynyl-9. Spermicides come in many different forms such as jelly, foam, tablets, and even a transparent film. All are placed in the vagina. Spermicides work best when they are used at the same time as a barrier method.
- Intrauterine devices (IUDs): These devices are inserted into the uterus, where they stay from one to ten years. An IUD prevents the fertilized egg from implanting in the lining of the uterus and may have other effects as well.
- Tubal ligation: This medical procedure is a permanent form of contraception for women. Each fallopian tube is either tied or burned closed. The sperm cannot reach the egg, and the egg cannot travel to the uterus.
- Vasectomy: This medical procedure is a the male form of sterilization and should be considered permanent. In vasectomy, the vas defrens, the tiny tubes that carry the sperm into the semen, are cut and tied off.

Unfortunately, there is no perfect form of birth control. Only abstinence (not having sexual intercourse) protects against unwanted pregnancy with 100 percent reliability. The failure rates, or the rates at which pregnancy occurs, for most forms of birth control are quite low. However, some forms of birth control are more difficult or inconvenient to use than others. In actual practice, the

birth control methods that are more difficult or inconvenient have much higher failure rates, because they are not used faithfully.

Description

All forms of birth control have one feature in common. They are only effective if used faithfully. Birth control pills work only if taken every day; the diaphragm is effective only if used during every episode of sexual intercourse. The same is true for condoms and the cervical cap. Some methods are automatically working every day, no matter what. These methods include Depo Provera, Norplant, the IUD, and tubal sterilization.

There are many different ways to use birth control. They can be divided into several groups:

- By mouth (oral): Birth control pills must be taken by mouth every day.
- Injected: Depo Provera is a hormonal medication that is given by injection every three months.
- Implanted: Norplant is a long-acting hormonal form of birth control that is implanted under the skin of the upper arm.
- Vaginal: Spermicides and barrier methods work in the vagina.
- Intra-uterine: The IUD is inserted into the uterus.
- Surgical: Tubal sterilization is a form of surgery. A doctor must perform the procedure in a hospital or surgical clinic. Many women need general anesthesia.

The methods of birth control differ from each other regarding when they are used. Some methods of birth control must be used specifically at the time of sexual intercourse (condoms, diaphragm, cervical cap, spermicides). All other methods of birth control must be working all the time to provide protection (hormonal methods, IUDs, tubal sterilization).

Condoms and spermicides

Condoms are about 85 percent effective in preventing pregnancies. That means that out of 100 females whose partners use condoms, 15 will still become pregnant during the first year of use, according to the nonprofit advocacy group Planned Parenthood. Unwanted pregnancies usually occur because the condom is not attached or used properly or breaks during intercourse. More protection against pregnancy is possible if a spermicide is used along with a condom. Spermicide is a pharmaceutical substance used to kill sperm, especially in conjunction with a birth-control device such as a condom or diaphragm. Spermicides come in foam, cream, gel, suppository, or as a thin film. The most common spermicide is called nonoxynol-9, and many condoms come with it already applied as a lubricant. However, spermicides do not kill HIV or other sexually transmitted viruses and do not prevent the spread of HIV and other STDs. Also, nonoxynol-9 can irritate vaginal tissue and thus increase the risk of getting an STD. In anal sex, especially between two males, spermicides also can irritate the rectum, increasing the risk of getting HIV. Spermicides are specifically discouraged for use by gay or bisexual males for anal sex.

Latex condoms are also recommended over condoms made from other materials, especially lambskin, because they are thicker and stronger and have less risk of breakage during sex. Non-latex condoms do not prevent the spread of STDs, including HIV, and should not be used by gay or bisexual men or men who have HIV or other sexually transmitted diseases. Condoms are available over-the-counter, meaning they do not require a prescription, and there are no age restrictions on purchasing condoms. They are available at a variety of locations, including drug stores, convenience stores, supermarkets, and **family** planning clinics. They are also available for purchase on the Internet.

FEMALE CONDOM The female condom is a seven-inch polyurethane pouch that fits into the vagina. It collects semen before, during, and after ejaculation, keeping semen from entering the uterus through the cervix and thus protecting against pregnancy. In one year of use, it is 79 percent effective in preventing pregnancies. It also reduces the risk of many STDs, including HIV. There is a flexible ring at the closed end of the thin, soft pouch of the female condom. A slightly larger ring is at the open end. The ring at the closed end holds the condom in place in the vagina. The ring at the open end rests outside the vagina. When the condom is in place during sexual intercourse, there is no contact of the vagina and cervix with the skin of the penis or with secretions from the penis. It can be inserted up to eight hours before sex.

Precautions

There are risks associated with some forms of birth control. Some of the risks of each method are:

- Birth control pills: The hormone (estrogen) in birth control pills can increase the risk of heart attack in women over forty who smoke.
- IUD: This device can increase the risk of serious pelvic infection. The IUD can also injure the uterus by poking into or through the uterine wall. Surgery might be needed to fix this injury.

Various types of contraception, including birth control pills, condoms, and diaphram. *(Photograph by Michael Keller. The Stock Market.)*

- Tubal sterilization: "Tying the tubes" is a surgical procedure and has all the risks of any other surgery, including the risks of anesthesia, infection, and bleeding.
- Condom: The most common problems associated with condoms are breakage during use and improper technique in using condoms. These can lead to pregnancy and sexually transmitted diseases, especially HIV.

Preparation

No specific preparation is needed before using contraception. However, a woman must be sure that she is not already pregnant before using a hormonal method or having an IUD placed.

Risks

Many methods of birth control have side effects. Knowing the side effects can help a woman to determine which method of birth control is right for her. There is no perfect form of birth control. Every method has a small failure rate and side effects. Some methods carry additional risks. However, every method of birth control has fewer risks than pregnancy. The risks include:

- Hormonal methods: The hormones in birth control pills, Depo Provera, and Norplant can cause changes in menstrual periods, changes in mood, weight gain, **acne**, and headaches. In addition, once a woman stops using Depo Provera or Norplant, she may go many months before she begins ovulating again.
- Barrier methods: A woman must insert the diaphragm in just the right way to be sure that it works properly. Some women get more urinary tract infections if they use a diaphragm because the diaphragm can press against the urethra, the tube that connects the bladder to the outside.

Contraception

Type of contraceptive	Description	Use	Failure rate per 100 women in one year
Abstinence	Refraining from intercourse, anal sex, and oral sex	Universally applicable. Also prevents spread of sexually transmitted diseases	0
Birth control pill	Prescription pill containing estrogen and progestin that suppresses ovulation	Must be taken daily, regardless of the frequency of intercourse	1-2
Cervical cap with spermicide	Soft rubber cup that fits around the cervix, obtained by prescription	Inserted before intercourse. May be difficult to insert	17-23, depending on type
Condom, female	Lubricated sheath that is inserted into the vagina. Similar in shape to the male condom, with a flexible ring	Applied immediately before intercourse, for single use	21
Condom, male	Latex or polyurethane sheath placed over erect penis, widely available in drugstores	Applied immediately before intercourse, for single use. Best protection against sexually transmitted diseases	11
Depo-Provera injection	Injection that inhibits ovulation, obtained by prescription	Injections performed at a doctor's office, once every three months	Less than 1
Diaphragm with spermicide	Dome-shaped rubber disk that covers the cervix, obtained by prescription	Inserted before intercourse and left in place at least six hours after	17
Douching	Use an over-the-counter feminine douche immediately after intercourse in an effort to wash out the sperm	Sperm travel quickly to the cervix, making this an ineffective method of birth control	40
IUD (intrauterine device)	T-shaped device inserted in the uterus during a visit to the doctor	Can remain in place for up to one or 10 years, depending on type	Less than 1
Morning-after pill (emergency contraceptive)	Pills similar to regular birth control pills, obtained by prescription	Must be taken within 72 hours of unprotected intercourse	80% reduction in pregnancy risk
Patch	Adhesive patch worn on the skin that releases hormones preventing ovulation. Obtained by prescription	New patch is applied once a week for three weeks, followed by one week without the patch	1-2
Periodic abstinence	Refraining from intercourse when conception is likely	Requires regular menstrual cycles and close monitoring of body functions pertaining to ovulation	20
Spermicide alone	A foam, cream, jelly, film, or suppository, or tablet containing nonoxynol-9	Depending on product, inserted between five and 90 minutes before intercourse; usually left in place at least six to eight hours after	20-50, depending on product
Withdrawal	Having intercourse, but removing the male penis before ejaculation	Not recommended for teens, and some seminal fluid leaks before ejaculation, making it an ineffective method of birth control	27

SOURCE: Food and Drug Administration, December 2003; Planned Parenthood, March 2004; kidshealth.org, September 2001.

(Table by GGS Information Services.)

- Spermicides: Some women and men are allergic to spermicides or find them irritating to the skin.
- IUD: The device is a foreign object that stays inside the uterus, and the uterus tries to get it out. A woman may have heavier menstrual periods and more menstrual cramping with an IUD in place.
- Tubal ligation: Some women report increased menstrual discomfort after this surgery. It is not known if this side effect is related to the tubal ligation itself.

Parental concerns

Nearly 60 percent of sexually active girls under age 18 would discontinue at least some reproductive health services if their parents were informed that they were seeking contraceptive services, according to a study published in the August 14, 2002 issue of the *Journal of the American Medical Association (JAMA)*. If parental notification would cause the majority of minor girls to stop seeking reproductive health services or to use less effective methods of contraception, the rates of teen pregnancies and STD infections would substantially increase,

KEY TERMS

Fallopian tubes—The pair of narrow tubes leading from a woman's ovaries to the uterus. After an egg is released from the ovary during ovulation, fertilization (the union of sperm and egg) normally occurs in the fallopian tubes.

Fertilization—The joining of the sperm and the egg; conception.

Implantation—The process in which the fertilized egg embeds itself in the wall of the uterus.

Ovulation—The monthly process by which an ovarian follicle ruptures releasing a mature egg cell.

Carol Ford of the Adolescent Medicine Program at the University of North Carolina–Chapel Hill and Abigail English of the Center for Adolescent Health & the Law state in an accompanying *JAMA* editorial. Although there is widespread consensus that communication between adolescents and their parents about sexual decision-making is important, there is no reason that confidential reproductive health care and efforts to improve communication between parents and their adolescent children cannot occur simultaneously, these authors suggest.

Parents of adolescents often are concerned that distribution of contraceptives leads to increased sexual activity. However, a study of 4,100 high school students published in the June 2003 issue of the *American Journal of Public Health* found that students who had access at school to condoms and instructions on their proper use were no more likely to have sexual intercourse than students at schools without condom distribution programs.

See also Condom.

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Amy B. Tuteur
Ken R. Wells

Cooley's anemia *see* **Thalassemia**

Corneal abrasion

Definition

A corneal abrasion occurs when there is a loss of cells from the epithelium or surface of the cornea. It is usually due to trauma but may occur without trauma such as with the overuse of **contact lenses**.

Description

The cornea is the clear curved structure found at the front of the eye. It is comprised of three layers and the membranes that separate these layers. It is very difficult to penetrate past the epithelium or top layer of the cornea. The cornea is normally devoid of blood vessels yet has many sensory nerves. When any trauma to the corneal epithelium occurs, cells are lost or destroyed and **pain** is immediately sensed. When a corneal abrasion occurs, the conjunctiva, or the white of the eye, turns red, as new blood vessels form and those present enlarge,

in an attempt to increase blood flow to the eye as it attempts to bring to the eye those cells needed for the healing of the cornea.

A corneal abrasion heals by the movement of neighboring epithelial cells, which slide over the wounded area, and through a cell division process called mitosis, which fill in the abraded area with new epithelial cells. Within two to three days of trauma to the cornea, these new cells start to adhere to the underlying membrane of the epithelium, called the basement membrane and within seven to eight days the abraded area usually heals completely without scarring. But if a corneal abrasion is deep and penetrates the next layer of the cornea, then scarring is possible and complete healing of the abrasion may be delayed as long as three months.

Demographics

Corneal abrasions account for 10 percent of ocular emergency care. The incidence of non-penetrating injuries to the eye, which includes corneal abrasions, is 1.57 percent per year. More males are treated for corneal abrasions than are females.

Causes and symptoms

Common causes of corneal abrasions include fingernails, make-up implements, paper cuts, plant material, including tree branches, animal scratches, cigarettes, inverted eyelashes, and blunt trauma, such as that with a knife or with scissors. Children are most likely to get a corneal abrasion while playing, while adults are more likely to sustain an abrasion in the workplace. Ultraviolet radiation such as that which occurs with a welder's flash or use of a sunlamp, can also cause an abrasion, as well as misuse and mishandling of contact lenses. When a missile type object causes an abrasion, the object can become embedded in the cornea or penetrate the eye.

Pain, irritation, tearing, red eye, twitching of the eye, decreased vision, and sensitivity to bright lights are common complaints that accompany a corneal abrasion. If there is significant swelling of the cornea, then vision may be decreased. If there is inflammation inside the eye, a dull ache may be felt inside the eye. Very rarely, **nausea** is experienced due to the pain associated with a corneal abrasion. The only symptom in a nonverbal patient, such as an infant, may be that the child is fussier than usual.

When to call the doctor

Any redness or foreign body sensation, especially if only one eye is affected, should be evaluated promptly for a corneal abrasion. If a corneal abrasion is not treated appropriately, scarring and ulceration of the corneal are possible. A corneal abrasion should be treated by a healthcare practitioner capable of evaluating eye conditions. **Herpes simplex**, recurrent corneal erosion (RCE), and acanthamoeba infections are other conditions that can mimic a corneal abrasion but which require very different treatments.

Diagnosis

The individual with a corneal abrasion will usually report a known trauma to the eye area.

To diagnose a corneal abrasion, a topical anesthetic with a yellow dye called flourescein is placed into the eye. Under blue cobalt light, the part of the cornea abraded will be stained by the dye and is easily seen by the examiner. The area and depth of the abrasion can be easily seen under a special microscope called a slit lamp biomicroscope. If a microscope is not available, then a blue light called a Burton lamp may be used.

Usually the anesthetic drop will relieve the ocular pain immediately. If a dull aching sensation persists after this instillation then a co-existing iritis, or inflammation of the iris, also called a uveitis, may also be present. When a biomicroscope is available, the eye is checked for infection or inflammation. The eyelids are everted (turned out) to check for any foreign bodies. These areas may also be rinsed with saline to remove any small foreign body that may be a source of the abrasion.

If any of the instilled dye leaks into the eye, then the cornea has been perforated and a small projectile may be inside the eye. Other testing, such as **x rays**, must be done to rule out foreign bodies inside the eye. Since metal is the most common material seen in penetrating injuries, an MRI is not usually done. If the cornea has been perforated, then the patient is referred to a corneal specialist for surgery.

If an infection is suspected or if an abrasion does not heal, then swabbing of the eye for microscopic culture may be done to definitively establish the organism involved.

Treatment

If the corneal abrasion is very small and superficial, then the application of mild antibiotic drops three to four times a day for a few days to a week is sufficient to prevent an infection. At bedtime, an antibiotic ointment, which will remain on the eye longer, may be necessary for deep abrasions. When an infection is present, then specially compounded fortified **antibiotics**, formulated specifically for the organism that caused the infection, are prescribed topically. For those with very minor abra-

sions, instillation of non-preserved lubricating drops a few times a day for a few days may be all that is required.

The pain of a corneal abrasion can be treated with drugs such as homatropine which keep the eye in a dilated state and stop the spasm of the iris, a major cause of discomfort for the individual with a corneal abrasion. Nonsteroidal drops, called NSAIDs, may also be prescribed for a few days to relieve the pain. In some instances, oral **analgesics** may be prescribed, but children under 12 should not be given aspirin. If the area is large and there is no underlying viral infection, then a mild steroid to decrease future scarring and treat an associated iritis may also be needed. Topical anesthetics are never prescribed because they delay and interfere with the healing process.

For many years the standard treatment for a corneal abrasion included patching. Patching can decrease blinking, which was thought to speed the healing process. But patching is as of 2004 no longer done routinely because it decreases the amount of oxygen that gets to the cornea. Patching is never done for contact lens patients, whose eyes are at a greater risk of a *Pseudomonas* infection, nor when the cause of an abrasion is due to vegetative matter, such as a tree branch. Organisms can thrive in these types of environment. Patching is also not done for patients who are monocular (have one good eye), if the better eye has the abrasion, nor for patients for whom depth perception is important, as this is lost when only one eye is used. Controlled studies have demonstrated that patching a corneal abrasion does not improve healing either in children or adults and that patching of an eye may make walking difficult.

For the contact lens wearer with a corneal abrasion, contact lens wear must be discontinued for at least one week and glasses must be worn. Patients who wear contact lenses are given antibiotics that act on the bacteria that are ubiquitous in the eye of the contact lens wearer. The eye must be evaluated by an eye-care practitioner prior to reinstituting contact lens wear.

For the patient with a large abrasion and without an infection, and who does not wear contact lenses, a bandage contact lens approved for extended overnight wear may be worn. The pressure of the contact lens may comfort the patient, and the antibiotics are absorbed by the contact lens, giving the eye continuous 24 hour protection from a potential bacterial infection.

Treatment of a corneal abrasion with over-the-counter (OTC) drugs advertised to decrease redness should not be used to self treat corneal abrasions. These drugs act by constricting the blood vessels in the eye, decreasing the blood supply to the eye and delaying healing. Also, the preservatives in these drops may irritate the cornea.

Except for a very mild abrasion, the doctor may require daily follow-up examination to ensure that the abrasion is healing. This is especially important if vegetative matter is the cause of the abrasion. Those who have been patched must be re-evaluated within 24 hours to see if the symptoms have improved.

KEY TERMS

Antibiotics—Drugs that are designed to kill or inhibit the growth of the bacteria that cause infections.

Cornea—The clear, dome-shaped outer covering of the eye that lies in front of the iris and pupil. The cornea lets light into the eye.

Epithelium—The layer of cells that covers body surfaces, lines body cavities, and forms glands.

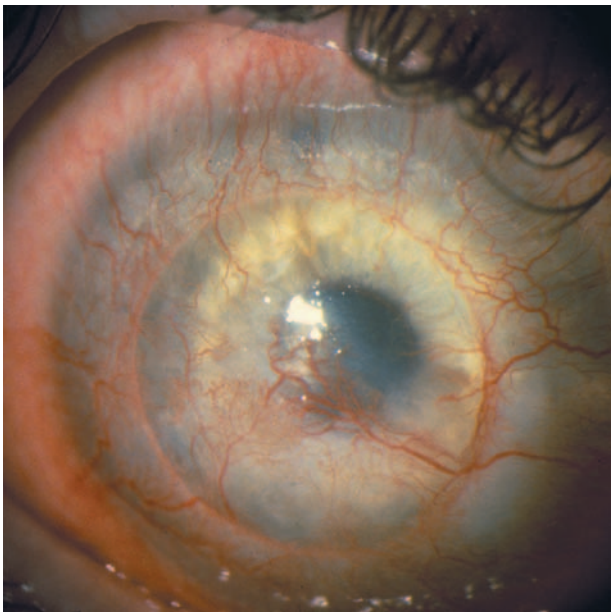
Pseudomonas—A bacterium which can cause ulcers in contact lens wearers.

Uveitis—Inflammation of all or part the uvea. The uvea is a continuous layer of tissue which consists of the iris, the ciliary body, and the choroid. The uvea lies between the retina and sclera.

Prognosis

Corneal abrasions usually heal within a week, but complications can occur. A secondary infection and scarring can result if the abrasion is not treated. Up to 50 percent of those with a corneal abrasion develop a uveitis or inflammation inside the eye.

Approximately 10 to 25 percent of those with corneal abrasions will develop recurrent corneal erosion (RCE) a condition in which the epithelium of the cornea pulls off because it did not heal properly or completely. This can happen weeks, months, or years after the initial trauma. Usually the patient either awakens with sharp pain or is bothered by a foreign body sensation that is worse in the morning. This erosion is usually treated conservatively with lubricating drops and hypertonic saline ointment for a month or more, although some patient need a debridement of the cornea or laser treatment. Oral doxycycline and topical steroids have been shown to help with the restructuring of the cornea with a RCE.



A close-up view of an abrasion on patient's cornea. *(Photograph by Dennis R. Cain. Custom Medical Stock Photo, Inc.)*

The probability of corneal ulcer development from a corneal abrasion increases tenfold in contact lens wearers for those who use extended wear contacts, over those who remove their contacts as night. This risk may be decreased for those wearing the new silicone contact lenses.

Prevention

Prevention of a traumatic injury or an accident is not possible, but for those types of abrasions caused by chronic trauma, such as with contact lenses, the likelihood of abrasions and subsequent ulcers can be reduced by proper contact lens wear and care. It is important that children and adolescents who wear contact lenses have glasses to wear and that glasses are worn every day to give the eyes a chance to breathe. The contact lenses should be replaced and cared for as recommended by an eye care practitioner.

For those who engage in welding, eye protection, including the use of helmets, decreases the incidence a corneal abrasions caused by welding **burns**.

Parental concerns

Rarely do corneal abrasion lead to loss of vision, but any trauma to the eye must be evaluated by a healthcare practitioner for the possibility of an abrasion. When an abrasion is present, then it is imperative that prescribed treatment, including cessation of contact lenses and use of prescribed drops be followed. If the corneal abrasion is due to contact lens abuse, then the consequences of further misuse of contact lenses must be thoroughly discussed with the eye care practitioner.

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Cough

Definition

A cough is a forceful release of air from the lungs that can be heard. Coughing protects the respiratory system by clearing it of irritants and secretions.

Description

While people can generally cough voluntarily, a true cough is usually a reflex triggered when an irritant stimulates one or more of the cough receptors found at different points in the respiratory system. These receptors then send a message to the cough center in the brain, which in turn tells the body to cough. A cough begins with a deep

breath in, at which point the opening between the vocal cords at the upper part of the larynx (glottis) shuts, trapping the air in the lungs. As the diaphragm and other muscles involved in breathing press against the lungs, the glottis suddenly opens, producing an explosive outflow of air at speeds greater than 100 miles (160 km) per hour.

In normal situations, most people cough once or twice an hour during the day to clear the airway of irritants. However, when the level of irritants in the air is high or when the respiratory system becomes infected, coughing may become frequent and prolonged. It may interfere with **exercise** or **sleep**, and it may also cause distress if accompanied by **dizziness**, chest **pain**, or breathlessness. In the majority of cases, frequent coughing lasts one to two weeks and tapers off as the irritant or infection subsides. If a cough lasts more than three weeks it is considered a chronic cough, and physicians try to determine a cause beyond an acute infection or irritant.

Coughs are generally described as either dry or productive. A dry cough does not bring up a mixture of mucus, irritants, and other substances from the lungs (sputum), while a productive cough does. In the case of a bacterial infection, the sputum brought up in a productive cough may be greenish, gray, or brown. In the case of an allergy or viral infection it may be clear or white. In the most serious conditions, the sputum may contain blood.

KEY TERMS

Antitussive—A drug used to suppress coughing.

Expectorant—A drug that promotes the discharge of mucus from respiratory system.

Gastroesophageal reflux—The backflow of stomach contents into the esophagus.

Glottis—The opening between the vocal cords at the upper part of the larynx.

Larynx—Also known as the voice box, the larynx is the part of the airway that lies between the pharynx and the trachea. It is composed of cartilage that contains the apparatus for voice production–the vocal cords and the muscles and ligaments that move the cords.

Sputum—The substance that is coughed up from the lungs and spit out through the mouth. It is usually a mixture of saliva and mucus, but may contain blood or pus in patients with lung abscess or other diseases of the lungs.

Demographics

Formal statistics on coughs are not maintained. Virtually all persons will experience coughs several times each year throughout their lives.

Causes and symptoms

In the majority of cases, coughs are caused by respiratory infections, including the following:

- colds or **influenza**, the most common causes of coughs
- **bronchitis**, an inflammation of the mucous membranes of the bronchial tubes
- croup, a viral inflammation of the larynx, windpipe, and bronchial passages that produces a bark-like cough in children
- whooping cough, a bacterial infection accompanied by the high-pitched cough for which it is named
- **pneumonia**, a potentially serious bacterial infection that produces discolored or bloody mucus
- **tuberculosis**, another serious bacterial infection that produces bloody sputum
- fungal infections, such as aspergillosis, histoplasmosis, and cryptococcosis

Environmental pollutants, such as cigarette smoke, dust, or smog, can also cause a cough. In the case of cigarette smokers, the nicotine present in the smoke paralyzes the hairs (cilia) that regularly flush mucus from the respiratory system. The mucus then builds up, forcing the body to remove it by coughing. Post-nasal drip, the irritating trickle of mucus from the nasal passages into the throat caused by **allergies** or **sinusitis**, can also result in a cough. Some chronic conditions, such as **asthma**, chronic bronchitis, emphysema, and **cystic fibrosis**, are characterized in part by a cough. A condition in which stomach acid backs up into the esophagus (gastroesophageal reflux) can cause coughing, especially when a person is lying down. A cough can also be a side-effect of medications that are administered via an inhaler. It can be a side-effect of beta-blockers and ACE inhibitors, which are drugs used for treating high blood pressure.

When to call the doctor

A physician or other healthcare provider should be called when a cough does not subside after three or four

days. Individuals such as smokers, who have chronic coughs, should consult a doctor if the nature of their cough changes or they produce blood when they cough.

Diagnosis

To determine the cause of a cough, a physician should take an exact medical history and perform an exam. Information regarding the duration of the cough, what other symptoms may accompany it, and what environmental factors may influence it aid the doctor in his or her diagnosis. The appearance of the sputum also helps determine what type of infection, if any, may be involved. The doctor may even observe the sputum microscopically for the presence of bacteria and white blood cells. Chest **x rays** may help indicate the presence and extent of such infections as pneumonia or tuberculosis. If these actions are not enough to determine the cause of the cough, a bronchoscopy or laryngoscopy may be ordered. These tests use slender tubular instruments to inspect the interior of the bronchi and larynx.

Treatment

Treatment of a cough generally involves addressing the condition causing it. An acute infection such as pneumonia may require **antibiotics**, an asthma-induced cough may be treated with the use of bronchodilators, or an antihistamine may be administered in the case of an allergy. Physicians prefer not to suppress a productive cough, since it aids the body in clearing respiratory system of infective agents and irritants. However, cough medicines may be given if the person cannot rest because of the cough or if the cough is not productive, as is the case with most coughs associated with colds or flu. The two types of drugs used to treat coughs are antitussives and **expectorants**.

Antitussives

Antitussives are drugs that suppress a cough. Narcotics—primarily codeine—are used as antitussives and work by depressing the cough center in the brain. However, they can cause such side effects as drowsiness, **nausea**, and **constipation**. Dextromethorphan, the primary ingredient in many over-the-counter cough remedies, also depresses the brain's cough center but without the side effects associated with narcotics. Demulcents relieve coughing by coating irritated passageways.

Expectorants

Expectorants are drugs that thin mucus in order to make it easier to cough up. Guaifenesin and terpin hydrate are the primary ingredients in most over-the-counter expectorants. However, some studies have shown that in acute infections, simply increasing fluid intake has the same thinning effect as taking expectorants.

Coughs due to bacterial or viral upper respiratory infections may be effectively treated with botanical and homeopathic therapies. The choice of remedy will vary and be specific to the type of cough the person has. Some combination over-the-counter herbal and homeopathic cough formulas can be very effective for cough relief. Lingering coughs or coughing up blood should be treated by a trained practitioner.

Many health practitioners advise increasing fluids and breathing in warm, humidified air as ways of loosening chest congestion. Others recommend hot tea flavored with honey as a temporary home remedy for coughs caused by colds or flu. Various **vitamins**, such as vitamin C, or **minerals**, such as zinc, may be helpful in preventing or treating conditions (including colds and flu) that lead to coughs. Avoiding of mucus-producing foods can be effective in healing a cough condition. These mucus-producing foods can vary, based on individual intolerance, but dairy products are a major mucus-producing food for most people.

Prognosis

Because the majority of coughs are related to the **common cold** or influenza, most will end in seven to 21 days. The outcome of coughs due to a more serious underlying disease depends on the pathology of that disease.

Prevention

It is important to identify and treat the underlying disease and origin of the cough. It is helpful to avoid cigarette smoke and coming in direct contact with people experiencing cold or flu symptoms. Hands should be washed frequently during episodes of upper-respiratory illnesses.

Nutritional concerns

Persons with coughs should be sure to maintain balanced and healthy diets.

Parental concerns

Parents of children under the age of five should closely monitor their children when they have a cough. Parents of children over five years of age must accept the fact that their children are likely to acquire coughs and

related illnesses from schoolmates. They should remain vigilant and consider having their children seen by a physician if the cough does not resolve after five to seven days.

See also Common cold.

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American Academy of Family Physicians. 11400 Tomahawk Creek Parkway, Leawood, KS 66211–2672. Web site: <www.aafp.org/>.

American Academy of Pediatrics. 141 Northwest Point Blvd., Elk Grove Village, IL 60007–1098. Web site: <www.aap.org/default.htm>.

American College of Physicians. 190 N Independence Mall West, Philadelphia, PA 19106–1572. Web site: <www.acponline.org/>.

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Cough suppressants

Definition

Cough suppressants are medicines that prevent or stop a person from coughing.

Description

Cough suppressants act on the center in the brain that controls the cough reflex. They are meant to be used only to relieve dry, hacking coughs associated with colds and flu. They should not be used to treat coughs that bring up mucus or the chronic coughs associated with **smoking**, **asthma**, emphysema, or other lung problems.

The most effective cough suppressants are narcotics. Heroin, which is not approved for medicinal use in the United States, and codeine have been widely used to stop coughs. These compounds, in addition to relieving coughs, also relieve **pain**, cause sedation, and are addictive. The most popular drug in this class is dextromethorphan, which is quite safe and is available without prescription. Dextromethorphan is an ingredient in most over-the-counter cough preparations:

- Vicks Formula 44
- Drixoral Cough Liquid Caps
- Sucrets Cough Control
- Benylin DM

The letters DM in a product's name normally indicates the presence of dextromethorphan, but it is always best to read the ingredients. Dextromethorphan works best in liquid formulations but is also available in capsules, lozenges, and tablets.

General use

Dextromethorphan is used for the temporary relief of coughs caused by minor throat and bronchial irritation such as may occur with common colds or with inhaled

irritants. Dextromethorphan is most effective in the treatment of chronic, nonproductive cough.

Dextromethorphan has been reported to be effective in reversing some of the adverse effects of methotrexate, a drug that has found use in many conditions including **cancer**, **psoriasis**, and some types of arthritis.

Precautions

Lozenges containing dextromethorphan hydrobromide should not be used in children younger than six years of age. Liquid-filled capsules containing the drug should not be used in children younger than 12 years of age.

Dextromethorphan is not meant to be used for coughs associated with asthma, chronic **bronchitis**, or other lung conditions. It should not be used for coughs that produce mucus.

A lingering cough could be a sign of a serious medical condition. Patients with a cough that lasts more than seven days or is associated with **fever**, rash, **sore throat**, or lasting **headache** should have medical attention. Parents should call a physician as soon as possible if their child has these symptoms.

Side effects

Dextromethorphan rarely causes side effects but has been reported to cause **dizziness**, drowsiness, and stomach upset. There have been rare reports of **vomiting** caused by dextromethorphan.

Although dextromethorphan is very safe, it can cause problems when taken in too large a dose. In overdose, dextromethorphan can cause extreme dizziness, shallow breathing, and coma.

Interactions

Dextromethorphan has no clinically significant interactions with medications that are likely to be given to children. However, dextromethorphan should not be used in combination with narcotic **analgesics** such as meperidine or codeine, since dextromethorphan will increase the side effects of the analgesic.

Parental concerns

Lozenges containing dextromethorphan hydrobromide should not be used in children younger than six years of age. Liquid-filled capsules containing the drug should not be used in children younger than 12 years of age. Doses must be measured carefully. Measuring teaspoons should be used in place of household teaspoons.

Adolescent behavior must be observed, since some multi-ingredient over-the-counter cough remedies have become drugs of abuse. While these products are not addictive, they are toxic when misused.

See also Expectorants.

KEY TERMS

Chronic—Refers to a disease or condition that progresses slowly but persists or recurs over time.

Narcotic—A drug derived from opium or compounds similar to opium. Such drugs are potent pain relievers and can affect mood and behavior. Long-term use of narcotics can lead to dependence and tolerance. Also known as a narcotic analgesic.

Nonproductive—A cough in which no mucus is coughed up, also called dry cough.

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American Academy of Emergency Medicine. 555 East Wells Street, Suite 1100, Milwaukee, WI 53202–3823. Web site: <www.aaem.org>.

American Academy of Pediatrics. 141 Northwest Point Boulevard, Elk Grove Village, IL 60007–1098. Web site: <www.aap.org>.

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CPR *see* **Cardiopulmonary resuscitation**

Cradle cap *see* **Seborrheic dermatitis**

Cramps, menstrual *see* **Dysmenorrhea**

Craniosynostosis

Definition

Craniosynostosis is one of a diverse group of deformities in the head and facial bones called craniofacial anomalies. An infant or child with craniosynostosis has improperly fused or joined bones (sutures) in the skull. ("Cranio" means skull; "synostosis" means fused bones.) When children with craniosynostosis also show other body deformities, their condition is called syndromic craniosynostosis. Primary craniosynostosis occurs when one or more of an infant's sutures (where skull bones meet) fuse prematurely. Secondary craniosynostosis results when one or more of an infant's sutures fuse prematurely as a result of lack of proper brain growth.

Description

A baby's skull is often thought of as a single piece of bone. However, it is actually made up of several bones that fit together like a jigsaw puzzle. These areas meet at what are called sutures. Sutures allow a growing baby's brain to expand. The four sutures come together at the fontanel, or "soft spot" in a baby's head. Eventually the sutures stop growing, and the cranial bones fuse.

Sometimes a suture is fused too early, however, preventing a growing child's brain from expanding. This condition can cause the brain to grow more rapidly in another area of the skull. The result is an abnormally shaped skull. Sometimes this happens before birth (congenital), or sometimes it occurs as the baby develops after birth.

There are four sutures of the skull that may be affected by craniosynostosis:

- Metopic: This suture extends from the top of the head down the middle of the forehead to the nose.
- Coronal: This suture extends from each ear to the fontanelle.
- Sagittal: This suture extends from the front of the head to the back, down the middle of the top to the head.
- Lambdoidal: This suture extends across the back of the head.

Types

The form of craniosynostosis depends on the suture or sutures that are affected.

Plagiocephaly (unicoronal synostosis)

Plagiocephaly is the most common form of craniosynostosis. It occurs in approximately one out of every 2,500 births. Plagiocephaly involves early fusion of either the right or left side of the coronal suture, the suture that extends from each ear over the top of the head to the fontanelle. The forehead and brow of a child with plagiocephaly look as if they have been pushed back or flattened because the forehead and brow have stopped their normal growth.

Brachycephaly (bicoronal synostosis)

Brachycephaly, which means "short headed," occurs when the right and left coronal sutures close prematurely. Brachycephaly results in an abnormally broad head with a high forehead. It is often associated with other craniofacial abnormalities, including Crouzon syndrome, Apert syndrome, Pfeiffer syndrome, and Saethre-Chotzen syndrome. It also is associated with **Down syndrome** (trisomy 21).

Trigonocephaly

This type of craniosynostosis involves fusion of the metopic suture that runs from the top of the head toward the nose, which can create a ridge running down the

forehead and gives the front of the head a wedge-shaped effect. The eyes also may be close together.

Scaphocephaly (sagittal craniosynostosis)

This early fusion involves the sagittal suture that runs from front to back on the top of the skull. The result can be a long, narrow skull.

Positional nonsyndromic plagiocephaly (positional molding)

Positional nonsyndromic plagiocephaly is a form of craniosynostosis. In 1992, the American Academy of Pediatrics recommended that infants **sleep** on their backs to reduce the risk of **sudden infant death syndrome** (SIDS). This successfully reduced the number of infants with SIDS, but also increased the number of infants suffering from positional plagiocephaly due to back sleeping. An infant with positional nonsyndromic plagiocephaly has a flattened skull at the back of the head. This condition is also commonly called positional molding or deformational plagiocephaly.

Demographics

Craniosynostosis occurs in one out of 2,000 live births in the United States. It affects males twice as often as females. Of those affected, 2–8 percent have primary craniosynostosis and the remaining cases are secondary craniosynostosis. Plagiocephaly is the most common form of craniosynostosis. It occurs in approximately one out of every 2,500 live births. Sagittal craniosynostosis is the most common type of single suture craniosynostosis. It is estimated to occur in one in 4,000 to 8,500 live births. Although sagittal craniosynostosis mostly occurs by chance, about 2–6 percent of cases are considered to be inherited.

Frequencies of the types of craniosynostosis based on suture classification include: sagittal (50–58%); coronal (20–29%); metopic (4–10%); and lambdoid (2–4%).

Causes and symptoms

As of 2004 the exact cause of craniosynostosis is not understood. Many scientists believe it is the result of a defect in the ossification (bone formation) in the bones of the skull. Craniosynostosis usually occurs by chance (sporadic). In some families, however, it is inherited.

Genetic abnormalities such as craniosynostosis are described by the type of chromosome that carries the abnormal gene and whether the gene is recessive or dominant. The autosomal chromosomes are the nonsex chromosomes.

Autosomal recessive

In order for a child to inherit an autosomal recessive abnormality, both parents have to be carriers of the abnormal gene. When both parents are carriers, there is a 25 percent chance that each child born will inherit the abnormal gene and develop craniosynostosis. The child also has a 50 percent chance of inheriting the abnormal gene and becoming a carrier. Males and females are affected equally.

Autosomal dominant

When one parent has the abnormal gene and the other parent has normal genes, craniosynostosis can still result. That is because the abnormal gene dominates the normal gene. For an autosomal dominant disorder when one parent is a carrier of the abnormal gene, there is a 50 percent chance each child born will inherit the abnormal gene. Males and females are affected equally.

Other genetic syndromes, such as Crouzon syndrome, Apert syndrome, and Pfeiffer syndrome, are associated with craniosynostosis. All have different patterns of inheritance and chances of recurrence.

The physical symptoms of craniosynostosis depend on the sutures affected. Since other syndromes are associated with craniosynostosis, other symptoms may also be present, including the following:

- Endocrine disorders: **Hyperthyroidism** (overactivity of the thyroid gland), **vitamin D deficiency**, renal osteodystrophy (defective bone development), hypercalcemia (high levels of calcium in the blood), and rickets (weakened bones resulting from vitamin D deficiency).
- Hematologic disorders: Bone marrow diseases including sickle cell disease, and thalassemia.
- Inadequate growth of brain: Microcephaly (abnormal smallness of head), and **hydrocephalus** (abnormal buildup of cerebrospinal fluid in the head).

When to call the doctor

Craniosynostosis is a progressive condition that must be treated. The skull and facial asymmetry associated with craniosynostosis are frequently observed by an infant's pediatrician or **family** physician. Parents may also observe the condition.

Diagnosis

Craniosynostosis may be present at birth or may be observed later when a child has delays in neurological development. It often may be diagnosed by physical examination alone. Craniosynostosis may be suspected

KEY TERMS

Anomaly—Something that is different from what is normal or expected. Also an unusual or irregular structure.

Apert syndrome—A craniofacial abnormality characterized by abnormal head shape, small upper jaw, and fusion of fingers and toes.

Asymmetrical—Unbalanced, disproportionate, or unequal.

Brachycephaly—An abnormal thickening and widening of the skull.

Chromosome—A microscopic thread-like structure found within each cell of the human body and consisting of a complex of proteins and DNA. Humans have 46 chromosomes arranged into 23 pairs. Chromosomes contain the genetic information necessary to direct the development and functioning of all cells and systems in the body. They pass on hereditary traits from parents to child (like eye color) and determine whether the child will be male or female.

Computed tomography (CT)—An imaging technique in which cross-sectional x rays of the body are compiled to create a three-dimensional image of the body's internal structures; also called computed axial tomography.

Coronal suture—Skull suture that lies behind the forehead area, across the head from left side to the right side.

Craniosynostosis—A premature closure of one or more of the joints (fissures) between the bones of the skull, which causes an abnormally shaped skull.

Crouzon syndrome—A disorder characterized by malformations of the skull and face.

Deformational plagiocephaly (positional molding)—A form of craniosynostosis in which the head is misshapen, the result of constant pressure to the same area of the head.

Fontanelle—One of several "soft spots" on the skull where the developing bones of the skull have yet to fuse.

Lambdoidal suture—The suture between the two parietal bones and the occipital bone in the skull.

Metopic suture—Suture extending from the top of the head down the middle of the forehead to the nose.

Pfeiffer syndrome—This condition includes craniosynostosis, shallow eye sockets, underdevelopment of the midface, short thumbs and big toes, and possible webbing of hands and feet.

Plagiocephaly—A form of craniosynostosis that involves fusion of the right or left side of coronal suture.

Sagittal suture—The suture between the two parietal bones in the top of the skull.

Scaphocephaly—An abnormally long and narrow skull.

Suture—A "seam" that joins two surfaces together, such as is found between the bones of the skull. Also refers to stitching together the torn or cut edges of tissue.

Trigonocephaly—An abnormal development of the skull characterized by a triangular shaped forehead.

when an infant has an abnormally shaped head or a small bony ridge along the skull in various locations. The condition may also be suspected if a baby's fontanel (soft spot) closes off earlier than expected. The physician will do a complete physical exam and take a complete prenatal and birth history of the child, including position in the uterus. Family history of craniosynostosis or other craniofacial abnormalities also will be discussed.

Since craniosynostosis may be associated with other neurological and muscular disorders, such as Crouzon syndrome, Apert syndrome, or Pfeiffer syndrome, the physician will also discuss whether there have been any delays in the child's developmental progression.

If craniosynostosis is suspected, an x ray or a CT scan of the child's skull will be taken. In addition, DNA testing can help identify the gene mutations that can cause the condition. Mutations in what are called fibroblast growth factor receptors (FGR1, 2, and 3) and the transcription factor TWIST are responsible for several types of craniosynostosis.

Treatment

For most children with craniosynostosis, facial and skull deformity will be obvious and may be expected to worsen as the child grows. Surgical management by experienced neurosurgeons (brain surgeons) and orthopedic (bone) surgeons will be necessary in most cases. It usually is performed between the ages of six and ten months.

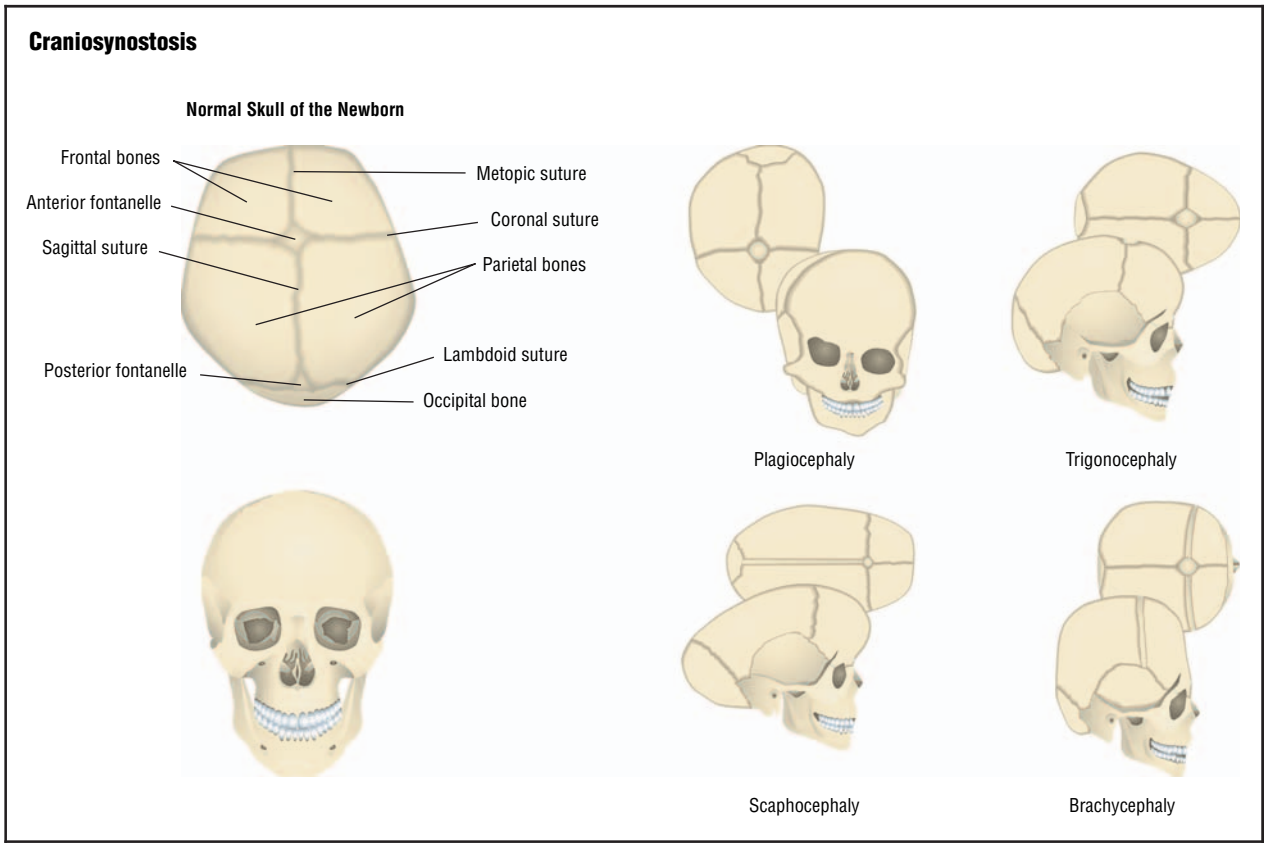

Illustration of a normal skull (left) and those with the four types of of craniosynostosis. Plagiocephaly, in which one side of the coronal suture closes prematurely, is the most common type. *(Illustration by GGS Information Services.)*

Surgery involves releasing the fused sutures and reshaping the bone of the skull and eye orbit. The fused sutures are excised (removed), and the skull reshaped and replaced with a variety of materials. Surgery usually lasts for three to seven hours, and several days of **hospitalization** are necessary to monitor and treat any complications of the surgery. When more severe deformities are present, repeat surgery of the skull and eye sockets may be necessary. This will help release and advance the child's mid-face.

In the early 2000s, newer, less invasive techniques are being developed using endoscopes. Endoscopes are narrow tubes that are inserted into the brain through narrow incisions (cuts). They allow surgeons to visualize the brain and pass tiny surgical instruments through the tube to perform surgery to re-open the sutures and allow the brain to grow normally.

Children born with co-existing craniofacial conditions, such as Apert syndrome, which affects the growth of the jaw, may benefit from a technique called mandibular bone lengthening or distraction osteogenesis. This technique was first used in Russia to treat bone problems in the leg and has now been used successfully to correct deformities of the jaw.

Positional plagiocephaly (positional molding, deformational plagiocephaly)

Positional plagiocephaly, the result of an infant's back sleeping position, can be treated by varying the infant's sleeping position from back to side sleeping, and providing tummy **play** time. A wedge-shaped foam pillow sold at many baby stores may help position the infant for side sleeping. The child may also be repositioned in the crib or the crib's location changed. When positional molding is identified at less than three months of age, repositioning is usually successful in stopping the plagiocephaly and reversing the flattening. If the flattening is severe or the condition is not caught until an infant is older, a helmet may be necessary. The helmet is specifically made for the infant by an orthotic specialist, a person who creates devices that provide more normal functioning for impaired people. The infant may wear the helmet for up to 23 hours daily, removing it only during baths. Average length of treatment is three to six months. The helmet gently redirects

the skull's growth and is most successful when the skull is most pliable, from about three months to about six months. The plagiocephaly will be monitored throughout this time, and the helmet adjusted as the child grows.

Prognosis

It is important to detect and treat craniosynostosis early. Untreated craniosynostosis will remain the same or worsen as a child grows and can affect a child's mental and physical development. Associated neuromuscular conditions also may affect the child's development. A child with craniosynostosis will require ongoing medical evaluations to ensure that the brain, skull, and facial bones are developing properly.

Prevention

As of 2004 there was no known prevention for craniosynostosis. Nothing that parents did or did not do causes the condition. The exception is positional plagiocephaly, which results from an infant being put to sleep on the back. This can be prevented by varying the infant's sleeping position from back to side sleeping, and providing tummy play time. A wedge-shaped foam pillow sold at many baby stores may help position the infant for side sleeping.

Parents who have an increased likelihood of carrying the genes that result in craniosynostosis may seek genetic counseling to better understand inheritance patterns and chances for reoccurrence.

Parental concerns

The physical symptoms of an asymmetrical face and head seen when a child has craniosynostosis are readily apparent and may cause the child embarrassment. The pressure a growing brain exerts on a fused suture also can cause a delay in development or, rarely, permanent brain damage. In addition, several conditions exist along with craniosynostosis and need to be evaluated.

A child needs to be carefully evaluated when craniosynostosis is present to distinguish between positional plagiocephaly, caused by back sleeping, and other forms of craniosynostosis. Positional plagiocephaly can be treated by repositioning the infant and perhaps by having the infant wear a helmet. Craniosynostosis must be treated by surgery. The differential diagnosis and treatment of posterior plagiocephaly is a challenging aspect of craniofacial surgery.

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Christine Kuehn Kelly

Crawling

Definition

Crawling is a slow creeping mode of locomotion, consisting of forward motion with weight supported by the infant's hands (or forearms) and knees. It is the primary means of mobility in infants.

Description

Crawling is the primary form of mobility achieved by infants before they learn to walk. It is the baby's first method of getting around efficiently on his or her own. In the traditional crawl, babies start by learning to balance on their hands and knees. Then they figure out how to move forward and backward by pushing off with their knees. At the same time they are strengthening the muscles that will soon enable them to walk.

Most babies learn to crawl between six and ten months. Some babies opt for another method of locomotion around this time, like bottom shuffling (scooting around on their bottom, using a hand behind and a foot in front to propel them), slithering on their stomach, or rolling across the room. Five percent of babies skip crawling altogether and move directly to pulling, standing, and walking. Parents should not worry about the infant's style; getting mobile is more important than how the baby does it.

Babies have a primitive crawling reflex at birth, which is instinctively activated when they are on their abdomens. Their legs flex, and they move forward, raising their heads to free them for motion. However, this reflex disappears during the early weeks of life, and true crawling does occur until six months, normally around the same time that an infant is able to sit up alone for extended periods of time. Learning to crawl occurs gradually and is usually complete by the time the baby is nine to ten months old.

Infancy

For most babies, creeping, wriggling, or slithering forward on the stomach comes before crawling, typically by the age of seven months. Infants also find that they can cover a distance simply by rolling from place to place. Especially on smooth floors, it is easy for them to move forward using only their arms or elbows and pulling their legs along, which are held out straight behind them. Infants can also get around while remaining in a seated position and pulling themselves with one or both arms, a form of mobility sometimes called hitching or bottom shuffling. From the infant's perspective, it has several advantages over crawling: it can leave one arm free, it allows better visibility, and the baby is already in a sitting position when she reaches her destination. Often, these alternate means of mobility are so convenient the child never learns to crawl, advancing directly to pulling herself upright and learning to walk. This is normal and not a cause for concern.

In creeping the infant is prone, with the abdomen touching the floor, and the head and shoulders supported with the weight borne on the elbows. The body is pulled along by movements of the arms, and the legs drag. The leg movements may resemble swimming or kicking.

Crawling is a more advanced locomotion than creeping. The trunk is above the floor, but parallel to it. The infant uses both his hands and knees in propelling himself forward. Not all infants follow this pattern of hitching, creeping, and crawling. Different children use different means of locomotion and may even skip a stage. (Skipping is especially likely if an infant is sick or for some other reason is unable to practice moving about).

Learning to crawl involves gradual trial-and-error attempts. When infants first get up on their hands and knees, they make modest attempts at movement, rocking or swaying in the direction they want to go. When they try to move, their balance is unstable, and they have trouble coordinating their movements, often moving an arm or leg and toppling over. One source of difficulty comes from the fact that neurological control over the arms and shoulders develops faster than control of the legs. For this reason once the infant is finally able to make real progress, he or she often moves backward, because it is easier to push harder with the hands and arms than with the feet. Although parents can provide temporary support by firmly placing their hands against the baby's feet, propelling them into forward motion despite themselves, backwards crawling typically persists for several weeks until the infant's coordination develops. Infants with greater strength in their hands, arms, and shoulders than in their legs and feet may learn to grasp, pull-up, and stand before crawling.

After crawling, the next stage is learning to walk. To that end, an infant soon begins pulling up on everything within reach. Once he or she gets the feel of balancing on the legs, an infant is ready to stand on his own and walk while holding onto furniture.

Common problems

Infant safety

Accidents are a leading cause of death in children from one to 24 months of age. They are second only to

KEY TERMS

Coordination—The ability to do activities with precision and proficiency.

Creeping—A form of locomotion in infants, in which the baby pulls the body forward with the arms while the belly and legs drag behind.

Developmental milestone—The age at which an infant or toddler normally develops a particular skill. For example, by nine months, a child should be able to grasp and toss a bottle.

acute infections as a cause of acute morbidity and doctor visits. Most accidents in infancy occur because parents either underestimate or overestimate the child's ability. Parents need to learn about their infant's developmental progress to use appropriate **childproofing** measures.

Parental concerns

To encourage crawling, parents can place **toys** and other desirable objects just beyond the baby's reach. They can also use billows, boxes, and sofa cushions to create obstacles courses for the baby to negotiate. This kind of **play** improves the baby's agility and speed.

Once an infant can crawl, the parent needs to provide a safe, roomy area for exploration. The baby is at the beginning of one of the most intense periods of educational development of her life and needs to satisfy her natural curiosity and her enormous capacity to learn by exploring. Rather than restrict her to a small area, it is recommended that parents childproof the home and keep it that way for the next two to three years. The greatest dangers to an inquisitive infant include uncovered electrical outlets; ungated stairways; and household cleaners, medications, and other potentially toxic substances. Other childproofing precautions include removing or securely anchoring lightweight furniture; hiding or securing electrical cords that could be pulled on; keeping valuable items or small objects that could be swallowed out of the baby's reach; keeping crib bars raised high; and strapping the infant securely into high chairs, car seats, and strollers.

When to call the doctor

If a child has not shown an interest in getting mobile by some means, figured out how move his harms and legs together in a coordinated motion, or learned to use both arms and both legs equally by one year of age, parents may want to discuss the matter with their pediatrician.

Infant crawling on the hands and knees. Most infants learn to crawl between six and eight months of age. *(© Jim Craigmyle/Corbis.)*

See also Gross motor skills.

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Creativity

Definition

Creativity is the ability to think up and design new inventions, produce works of art, solve problems in new ways, or develop an idea based on an original, novel, or unconventional approach.

Description

Creativity is the ability to see something in a new way, to see and solve problems no one else may know exists, and to engage in mental and physical experiences that are new, unique, or different. Creativity is a critical aspect of a person's life, starting from inside the womb onward through adulthood.

Although many people equate creativity with **intelligence**, the two terms are not synonymous, and it is not necessary to have a genius-level IQ in order to be creative. While creative people do tend to have average or above-average scores on IQ tests, beyond an IQ of about 120 there is little correlation between intelligence and creativity. Researchers have found environment to be more important than heredity in influencing creativity, and a child's creativity can be either strongly encouraged or discouraged by early experiences at home and in school.

Standard intelligence tests measure convergent thinking, which is the ability to come up with a single correct answer. However, creativity involves divergent thinking, which is the ability to come up with new and unusual answers.

Creative individuals tend to share certain characteristics, including a tendency to be more impulsive or spontaneous than others. Nonconformity (not going along with the majority) can also be a sign of creativity. Many creative individuals are naturally unafraid of experimenting with new things; furthermore, creative people are often less susceptible to **peer pressure**, perhaps because they also tend to be self-reliant and unafraid to voice their true feelings even if those go against conventional wisdom.

Creativity in childhood is typically assessed through paper-and-pencil measures such as the Torrance Test of Creative Thinking. These tests are designed to measure divergent thinking, such as fluency, flexibility, originality, and elaboration. Signification criticisms have been raised about these tests as measures of creativity. First is the general problem that there are no universally accepted definitions of creativity. Second, critics of creativity tests argue that these tests do not measure creativity per se but instead reflect the specific abilities that are assessed by the tests. Third, the scores on these tests often depend partly on speed, which is not necessarily a criterion for creativity. A final consistent concern relates to the scoring of creativity tests, which by definition are somewhat subjective. Thus, the reliability of such tests is commonly questioned.

Infancy

Scientific research in the late twentieth century revealed how the quality of interaction with unborn infants affects their later development of creative abilities. From birth to 18 months, infants can be encouraged to engage in creativity by playing with a variety of safe household materials, such as margarine tubs, empty boxes, and large empty spools. Parents and caregivers can encourage experimentation by showing excitement and interest in what babies do.

Parents can encouraged infants to develop creativity by singing to the infant and playing music, moving the infant's hands to music, hanging a colorful mobile over the crib, placing pictures and photos where the baby can focus on them, and playing sound games with infants, such making up nonsense words or using rhyming words when talking to them.

Toddlerhood

From ages 18 months to four years, toddlers have progressively better hand and eye coordination. Caregivers should give them opportunities to develop this coordination by allowing them to draw with water-based paints, with chalk, and with crayons. Toddlers also can develop their creativity by pasting, tearing, cutting, printing, modeling with clay or play dough, or working with various materials to create collage, and for the older child, experimenting with fabric, tie dye, batik, printing, and simple woodwork.

From around 12 months, children may begin to imitate things that adults do. Real fantasy **play** begins at around ages 18 to 21 months. This should not prevent caregivers from playing imaginatively from a younger age, since fantasy play is linked to creativity. Studies have shown that children with very active fantasies tend to have personality traits that contribute to creativity—originality, spontaneity, verbal fluency, and a higher degree of flexibility in adapting to new situations.

Children who fantasize a lot have unusually good inner resources for amusing themselves. Parents can provide materials that lend themselves to fantasy play (dressing-up clothes, dolls, housecleaning sets, and stuffed animals), play pretending games with their children, and

make suggestions and encourage new ideas when toddlers play alone.

Adults should start involving toddlers with creative activities as soon as they feel the child will enjoy them. Adults need to remember that young toddlers are not skillful enough to consciously produce works of art. At 18 months they may be more ready for creative play and even at this age, they may spend no more than five minutes of concentration on any one activity.

Preschool

Preschoolers can use the same materials as toddlers but can use them in more complex ways. By age five, many children start drawing recognizable objects. By age six, they are usually interested in explaining their art works. They also like to tell stories and can make books of their stories, including drawing pictures to accompany the writing.

At this age fantasy play becomes more complex. Preschoolers often direct each other on what to do or say as they play "Let's pretend." Play is a critical part of developing creativity, according to Mary Mindess, a child psychology professor at Lesley University in Cambridge, Massachusetts. "Play allows children to construct meaning for themselves," Mindess stated in an article in the August 2001 newsletter *The Brown University Child and Adolescent Behavior Letter*. "Two children may share an experience, but each will process the experience differently. Very often during play, children take things they see in real life, or things they imagine they experience—like something they read in a book or saw on television—and make meaning of it," she wrote. As an example, she cites Mark Twain's stories about Tom Sawyer and Huckleberry Finn as good role-playing examples. "They include many examples of play," she wrote. "If, as in a scene in *The Adventures of Tom Sawyer*, a child pretends to be a riverboat captain, there's a lot more to that role-playing than simply knowing what a captain does and some basic boat terminology. There are feelings that accompany the role-playing: mainly, the power of being captain and the satisfaction in the ability to make decisions."

School age

Early school-age children, six to nine years, incorporate lots of fantasy into their play, including action games with superheroes. Children of this age group spend much of their time daydreaming. Some daydreams become "real" as children begin to act them out in stories and plays.

Many researchers believe that in order to foster creativity in schools, education should be based on the discovery of knowledge and the development of critical attitudes, rather than on the passive absorption of knowledge. They believe this applies whether the class is in art, history, science, or humanities. However, most school teaching in the United States is based on the child's ability to memorize. The highest marks are often given to those who merely studied their lessons well. The pupil whose creative side is more developed may be considered a disruptive member of the class.

For this reason some educators decided to encourage creativity outside the school system. Science clubs are open to the young, in different countries, in which students can unleash their ideas and imagination. Student science fairs are also useful in developing creativity.

In the United States, children who participate in the nationwide invention contest organized by the *Weekly Reader* do not have to submit a model. A drawing or a photograph is sufficient to enter the contest, the purpose of which is to stimulate creative thinking among all the students in a class, all becoming involved in the process of invention either individually or in small groups. The class then chooses the best invention that will be presented later at the level of the national contest.

At ages nine to 12, children's creativity is greatly affected by peer influence. They increase the amount of detail and use of symbols in **drawings**. They also have expanded their individual creative differences and begin to develop their own set of creative values.

Teenagers are highly critical of the products they make and ideas they have. They try to express themselves creatively in a more adult-like way. Their creativity is influenced by their individual differences, physically, mentally, emotionally, and socially. In most high schools, classes that stress creativity, such as art, music, writing, and drama are electives and many may not be required. For many adolescents, high school is their last opportunity to take these creative classes.

Also, teens become more self-aware and self-conscious. This focus often causes them to conform to their peers, which stifles their creativity and makes their thoughts less flexible. Flexibility refers to the ability to consider various alternatives at the same time.

Common problems

Rewards or incentives appear to interfere with creativity and reduce children's flexibility of thought. Studies show that any constraints such as structured instructions reduce creative flexibility in children. Many

parents and teachers do not understand that children who are creative are often involved in imaginary play and are motivated by internal rather than external factors.

Parental concerns

Environment appears to play a greater role than heredity in the development of creativity: identical **twins** reared apart show greater differences in creativity than in intellectual ability. **Family** environments with certain characteristics have been found to be more conducive to creativity than others. One of these characteristics is a relaxed parental attitude rather than one that is overly anxious or authoritarian.

On the whole, the families of creative children **discipline** them without rigid restrictions, teaching them respect for values above rules. Similarly, they emphasize achievement rather than grades. The parents in such homes generally lead active, fulfilling lives themselves and have many interests. Finally, they reinforce creativity in their children by a general attitude of respect and confidence toward them and by actively encouraging creative pursuits and praising the results. It has been found that creativity in both children and adults is affected by positive reinforcement.

Positive reinforcement has also been shown to boost fifth graders' scores on creativity tests, help sixth graders write more original stories, and lead college students to produce novel word associations. Studies have also found that positively reinforcing one kind of creative activity encourages original thinking in other areas as well.

Just as certain actions and attitudes on the part of parents can encourage creativity, others have been found to discourage it. Devising restrictive guidelines or instructions for an activity reduces its potential as a creative experience. Unrestricted, imaginative play is central to creativity in children—exposure to new objects and activities stimulates the senses, reinforces exploratory impulses, and results in the openness to new experiences and ideas that foster creative thinking. In addition, anything that takes the focus away from the creative act itself and toward something external to it can be damaging. For example, knowing that one's efforts are going to be evaluated tends to restrict the creative impulse, as does knowing of the possibility of a prize or other reward.

Schools as well as families can encourage creativity by offering children activities that give them an active role in their own learning, allow them freedom to explore within a loosely structured framework, and encourage them to participate in creative activities for the sheer enjoyment of it rather than for external rewards.

When to call the doctor

Several studies have shown relationships sometimes exist between creativity and mental illness, including depression, **schizophrenia**, and **attention-deficit hyperactivity disorder** (ADHD).

For decades, scientists have known that eminently creative individuals have a much higher rate of manic depression or **bipolar disorder** than does the general population. But few controlled studies have been done to build the link between mental illness and creativity. One study that does support such a link was presented at the 2002 annual meeting of the American Psychiatric Association by Stanford University researchers Connie Strong and Terence Ketter. Using personality and **temperament** tests, they found healthy artists to be more similar in personality to individuals with manic depression than to healthy people in the general population.

While creativity itself is not a sign of mental illness, parents should be aware that there is a much higher degree of mental illness, especially depression and bipolar disorder, in creative children than in their less creative peers.

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KEY TERMS

Attention deficit disorder (ADD)—Disorder characterized by a short attention span, impulsivity, and in some cases hyperactivity.

Batik—A method of hand-printing a fabric by covering with removable wax the parts that will not be dyed.

Bipolar disorder—A severe mental illness, also known as manic depression, in which a person has extreme mood swings, ranging from a highly excited state, sometimes with a false sense of well being, to depression.

Convergent thinking—The ability to come up with a single correct answer.

Divergent thinking—The ability to come up with new and unusual answers.

Hereditary—Something which is inherited, that is passed down from parents to offspring. In biology and medicine, the word pertains to inherited genetic characteristics.

Manic depression—A psychiatric disorder characterized by extreme mood swings, ranging between episodes of acute euphoria (mania) and severe depression; also called bipolar depression.

Schizophrenia—A severe mental illness in which a person has difficulty distinguishing what is real from what is not real. It is often characterized by hallucinations, delusions, and withdrawal from people and social activities.

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Ken R. Wells

Cretinism *see* **Hypothyroidism**

Cri du chat syndrome

Definition

Cri du chat (a French phrase that means "cry of the cat") syndrome is a group of symptoms that result when a piece of chromosomal material is missing (deleted) from a particular region on chromosome 5. Children born with this chromosomal deletion have a characteristic mewing cat-like cry as infants that is thought to be caused by abnormal development of the larynx (organ in the throat responsible for voice production). They also have unusual facial features, poor muscle tone (**hypotonia**), small head size (microcephaly), and **mental retardation**.

Description

Jerome Lejeune first described cri du chat syndrome in 1963. Cri du chat syndrome is also called 5p minus syndrome or chromosome 5p deletion syndrome because it is caused by a deletion, or removal, of genetic material from chromosome 5. The deletion that causes cri du chat syndrome occurs on the short or p arm of chromosome 5. This deleted genetic material is vital for normal development. Absence of this material results in the features associated with cri du chat syndrome.

A high-pitched mewing cry during infancy is a classic feature of cri du chat. Infants with cri du chat also typically have low birth weight, slow growth, a small head (microcephaly), and poor muscle tone (hypotonia). Infants with cri du chat may also have congenital heart defects, language difficulties, delayed motor skill development, **scoliosis**, and varying degrees of mental retardation. Behavioral problems such as hyperactivity may also develop as the child matures.

Demographics

It has been estimated that cri du chat syndrome occurs in one of every 50,000 live births. According to the 5p minus Society, approximately 50 to 60 children are born with cri du chat syndrome in the United States each year. The syndrome can occur in all races and in both sexes.

Causes and symptoms

Cri du chat is the result of a chromosome abnormality—a deleted piece of chromosomal material on chromosome 5. In 90 percent of children with cri du chat syndrome, the deletion is sporadic. This means that it happens randomly and is not hereditary. If a child has cri du chat due to a sporadic deletion, the chance the parents could have another child with cri du chat is 1 percent. In approximately 10 percent of children with cri du chat, there is a hereditary chromosomal rearrangement that causes the deletion. If a parent has this rearrangement, the risk for their having a child with cri du chat is greater than 1 percent.

An abnormal larynx causes the unusual cat-like cry made by infants that is a hallmark feature of the syndrome. As children with cri du chat get older, the cat-like cry becomes less noticeable. This can make the diagnosis more difficult in older children. In addition to the cat-like cry, individuals with cri du chat also have unusual facial features. These facial differences can be very subtle or more obvious. Microcephaly (small head size) is common. During infancy many children with cri du chat do not gain weight or grow normally. Approximately 30 percent of infants with cri du chat have a congenital heart defect. Hypotonia (poor muscle tone) is also common, leading to problems with eating and slow, but normal development. Mental retardation is present in all children with cri du chat, but the degree of mental retardation varies between children.

When to call the doctor

A doctor should be consulted if a child exhibits symptoms typical of cri du chat syndrome.

Diagnosis

During infancy, the diagnosis of cri du chat syndrome is strongly suspected if the characteristic cat-like cry is heard. If a child has this unusual cry or other features seen in cri du chat syndrome, chromosome testing should be performed. Chromosome analysis provides the definitive diagnosis of cri du chat syndrome and can be performed from a blood test. Chromosome analysis, also called karyotyping, involves staining the chromosomes and examining them under a microscope. In some cases the deletion of material from chromosome 5 can be easily seen. In other cases, further testing must be performed. Fluorescence in-situ hybridization (FISH) is a special technique that detects very small deletions. The majority of the deletions that cause cri du chat syndrome can be identified using the FISH technique.

Treatment

As of 2004, there is no cure for cri du chat syndrome. Treatment consists of supportive care and developmental therapy.

Prognosis

Individuals with cri du chat have a 10 percent mortality during infancy due to complications associated with congenital heart defects, hypotonia, and feeding difficulties. Once these problems are controlled, most individuals with cri du chat syndrome have a normal lifespan. The extent of mental retardation and other symptoms depends on the site of the chromosomal deletions, with larger deletions resulting in more serious symptoms. With extensive early intervention and special schooling, many cri du chat children can develop adequate social, motor, and language skills.

Prevention

As of 2004, cri du chat syndrome had no known prevention.

Parental concerns

Cri du chat syndrome can be detected before birth if the mother undergoes **amniocentesis** testing or chorionic villus sampling (CVS). This testing would only be recommended if the mother or father is known to have a chromosome rearrangement, or if they already have a child with cri du chat syndrome.

Families may wish to seek counseling regarding the effects of the syndrome on relationships within the **family**. Many people respond with guilt, **fear**, or blame when a genetic disorder is diagnosed in the family, or they may overprotect the affected member. Support groups are often good sources of information about cri du chat syndrome; they can offer helpful suggestions about living with it as well as emotional support.

KEY TERMS

Amniocentesis—A procedure performed at 16–18 weeks of pregnancy in which a needle is inserted through a woman's abdomen into her uterus to draw out a small sample of the amniotic fluid from around the baby for analysis. Either the fluid itself or cells from the fluid can be used for a variety of tests to obtain information about genetic disorders and other medical conditions in the fetus.

Chorionic villus sampling—A procedure performed at 10 to 12 weeks of pregnancy in which a needle is inserted either through the mother's vagina or abdominal wall into the placenta to withdraw a small amount of chorionic membrane from around the early embryo. The amniotic fluid can be examined for signs of chromosome abnormalities or other genetic diseases.

Chromosome—A microscopic thread-like structure found within each cell of the human body and consisting of a complex of proteins and DNA. Humans have 46 chromosomes arranged into 23 pairs. Chromosomes contain the genetic information necessary to direct the development and functioning of all cells and systems in the body. They pass on hereditary traits from parents to child (like eye color) and determine whether the child will be male or female.

Congenital—Present at birth.

Deletion—The absence of genetic material that is normally found in a chromosome. Often, the genetic material is missing due to an error in replication of an egg or sperm cell.

Hypotonia—Having reduced or diminished muscle tone or strength.

Karyotyping—A laboratory test used to study an individual's chromosome make-up. Chromosomes are separated from cells, stained, and arranged in order from largest to smallest so that their number and structure can be studied under a microscope.

Microcephaly—An abnormally small head.

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ORGANIZATIONS

5p- Society. 7108 Katella Ave. #502, Stanton, CA 90680. Web Site: <www.fivepminus.org>.

Alliance of Genetic Support Groups. 4301 Connecticut Ave. NW, Suite 404, Washington, DC 20008. Web site: <www.geneticalliance.org>.

Cri du Chat Society. Department of Human Genetics, Box 33, MCV Station, Richmond VA 23298. Telephone: 804/786-9632.

Cri du Chat Syndrome Support Group. Web site: <www.cridchat.u-net.com>.

National Organization for Rare Disorders (NORD). PO Box 8923, New Fairfield, CT 06812-8923. Web site: <www.rarediseases.org>.

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Holly Ann Ishmael, M.S.

Crib death *see* **Sudden infant death syndrome**

Cromolyn *see* **Antiasthmatic drugs**

Crossed eyes *see* **Strabismus**

Croup

Definition

Croup is one of the most common respiratory illnesses in children. It is an inflammation of the larynx and the trachea. When a child has croup, that portion of the airway just below the vocal cords narrows and becomes swollen, making breathing both noisy and labored.

Description

Croup is a broad term describing a group of illnesses that affect the larynx, trachea, and bronchi. The key symptom is a harsh, barking **cough**. One of the most common respiratory illnesses in children, croup is frequently noted in infants and children and can have a variety of causes. Before the days of **antibiotics** and immunizations, croup was a dreaded and often deadly disease usually caused by the **diphtheria** bacteria. Though in the

early 2000s cases of croup are normally mild, it can still be dangerous. Croup affects the vocal cords and the area just below, the voice box, or larynx, and the windpipe, or trachea. The lower breathing passages (bronchi) may also be affected. Swelling of these areas causes the airway to narrow, which makes breathing difficult. It is also sometimes called laryngotracheitis, a medical term describing the inflammation of the trachea and larynx.

The characteristic symptoms of croup can be better understood by knowing the anatomic makeup of a child's larynx. Small children typically have quite a narrow larynx, so even a slight decrease in the airway's radius may lead to a large decrease in the air flow, leading to the symptoms of croup.

There are two primary types of croup: viral and spasmodic. Viral croup is caused by a viral infection in the trachea and larynx. It often starts with a cold that over time develops into a barking cough. When the child's airway becomes increasingly swollen and more mucus is secreted, it becomes more challenging to breathe. Breathing gets increasingly noisy, and a condition known as **stridor** may occur. (Stridor is a sign of respiratory obstruction that presents as a high-pitched, coarse, musical sound that occurs during breathing.) Children with viral croup usually have a low-grade temperature, but a few may have fevers up to 104°F (40°C). As breathing requires more effort, the child may stop eating and drinking. The child may also become too fatigued to even cough. If the airway continues to swell, it may approach a point at which the child can no longer breathe. Stridor is fairly common with a mild case of croup, especially if the child is active or crying. However, if a child has stridor at rest, the child may have severe croup. Symptoms are usually worse at night. The symptoms peak between 24 and 48 hours and usually resolve within one week.

Spasmodic croup is usually precipitated by an allergy or mild upper respiratory infection. It can be quite alarming, both because of the noise of the cough and because it usually comes on suddenly in the middle of the night. A child may go to **sleep** with a mild cold and wake up a few hours later, gasping for air. In addition, the child may have a cough that sounds like a seal barking, and will have a hoarse voice. Children with spasmodic croup normally do not have a **fever**.

Spasmodic croup can sometimes be difficult to differentiate from viral croup. Although spasmodic croup is associated with the same viruses that cause viral croup, spasmodic croup tends to recur and may be an indication of some type of allergic reaction instead of a direct infection.

Transmission

The viruses causing croup are highly contagious and easily transmitted between individuals through sneezing and coughing. It is usually transmitted via the respiratory route, entering through the nose and nasopharynx.

Demographics

Croup accounts for about 15 percent of all respiratory tract infections in children seen by physicians. It typically is seen in late fall and winter, and primarily occurs in children aged six months to three years. It has an annual peak incidence of 50 new cases per 1,000 children during the second year of life. Males are twice as likely as females to get the disease. The incidence decreases significantly after age six.

Causes and symptoms

Croup is most commonly brought on by a viral infection. The parainfluenza viruses (types 1, 2, and 3) are the most frequent causes of croup, accounting for approximately 75 percent of all cases diagnosed. Human parainfluenza virus 1 (HPIV-1) is the most common cause. Croup may also be caused by **influenza** A and B, adenovirus, **measles**, and respiratory syncytial virus (RSV). Other possible causes of croup are bacteria, inhaled irritants, **allergies**, and acid reflux.

The following are usually true of viral croup:

- It commonly occurs in individuals between the ages six months to six years.
- Stridor, and the classic barking cough are usually present.
- The child may have a fever.
- Wheezing may be present.
- It usually lasts two to seven days.

The following items are characteristic of spasmodic croup:

- The symptoms come on suddenly, often in the middle of the night.
- Stridor occurs along with the barking cough.
- It typically lasts two to four hours.

When to call the doctor

Most cases of croup can be safely managed at home, but parents should call their child's doctor for advice,

even if it is in the middle of the night. Call 911 for emergency help if any of the following is true:

- The croup is possibly caused by an inhaled object or by an insect sting.
- The child is drooling.
- The child has blue lips or skin.
- The child has a very high fever.
- The child is very anxious, has rapid breathing, and/or is struggling to get a breath.
- The child insists on sitting up or complains of a **sore throat** and is drooling. This is a possible indication that he or she may have a disease called **epiglottitis**, which is potentially life-threatening.
- The child makes a whistling sound that gets louder with each breath.
- The child has stridor when resting.

Diagnosis

The diagnosis of croup is usually made based on the description of symptoms by the parent, as well as a physical examination. Sometimes other studies, such as **x rays**, may be required. The doctor may note chest retractions with breathing and may hear wheezing and decreased breath sounds when listening to the chest with a stethoscope. Sometimes a foreign object or narrowing of the trachea is seen on a neck x ray.

Treatment

The most important part of treating patients with croup is maintaining an open airway. If a child wakes up in the middle of the night with croup, he or she should be taken to the bathroom. The door should be closed and the shower turned on to allow the bathroom to steam up. The parent should then sit in the steamy bathroom with the child. The moist, warm air should assist the child in breathing within 15 to 20 minutes, though the child will still have the barking cough. For the rest of that night and for two to three nights following, a humidifier or cold-water vaporizer should be placed in the child's room. If another attack of croup recurs that night or the next, the steam treatment should be repeated. If the steam does not work, sometimes taking the child outside, where he or she can inhale the cool, moist night air will be enough to improve breathing. Though a study in the early 2000s cast some doubt on the efficacy of using steam or mist, it does seem to be helpful for most children with croup. Parents may also give **acetaminophen** to reduce fevers and increase the child's comfort level. Cough medicines should usually be avoided.

Several other treatments are possible if the croup is severe enough to warrant the child's being seen by a physician. Aerosolized racemic epinephrine as well as oral dexamethasone (a steroid) may be used to help shrink the upper airway swelling. A bacterial infection will require antibiotics. If the airway becomes increasingly obstructed, the child may require intubation (the placing of a tube through the nose or mouth through the larynx into the main air passage to the lungs.) If the patient is dehydrated, intravenous fluids will be administered.

Prognosis

Croup is normally a self-limiting disease with an excellent prognosis. Only a few who are diagnosed require **hospitalization**, and less than 5 percent require intubation. If proper airway management is maintained, death is rare. There is some speculation that children with a history of croup may be at a higher risk for developing **asthma**, but the evidence was not clear as of 2004.

Prevention

The best way to prevent croup is to prevent the causative infections. Parents should practice excellent hand washing, especially during the cold and flu season, and avoid close contact with anyone who has a respiratory infection.

Parental concerns

The onset of croup can be frightening, especially when it comes on suddenly. Parents can help their child by not panicking or appearing anxious, as this may increase anxiety in the child, which can worsen symptoms. If they are at all unsure about how their child is responding to home treatment, parents should not hesitate to seek medical advice or treatment, no matter the time of day or night.

See also Influenza.

Resources

PERIODICALS

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KEY TERMS

Epiglottitis—Inflammation of the epiglottis, most often caused by a bacterial infection. The epiglottis is a piece of cartilage behind the tongue that closes the opening to the windpipe when a person swallows. An inflamed epiglottis can swell and close off the windpipe, thus causing the patient to suffocate. Also called supraglottitis.

Larynx—Also known as the voice box, the larynx is the part of the airway that lies between the pharynx and the trachea. It is composed of cartilage that contains the apparatus for voice production–the vocal cords and the muscles and ligaments that move the cords.

Retractions—Tugging-in between the ribs when breathing in.

Stridor—A term used to describe noisy breathing in general and to refer specifically to a high-pitched crowing sound associated with croup, respiratory infection, and airway obstruction.

Trachea—The windpipe. A tube composed of cartilage and membrane that extends from below the voice box into the chest where it splits into two branches, the bronchi, that lead to each lung.

"Patient Education Guide: What to Do When Your Child Has Croup." *Journal of Respiratory Diseases* 23 (March 2002): 23, 192–5.

ORGANIZATIONS

American Academy of Pediatrics. 141 Northwest Point Boulevard, Elk Grove Village, IL 60007–1098. Web site: <www.aap.org>.

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Crying and fussing in an infant

Definition

All babies cry and fuss. Many infants spend a considerable amount of time being fussy. Young infants cry between one and five hours out of 24. Crying is important for babies; it is the baby's first way of communicating and an important way to release tension. Constant crying, though, can be a symptom of **colic** or a sign that something else is wrong.

Description

The baby's cry is a perfect signal of life. It has three features:

- The newborn's cry is automatic and reflexive. The infant senses a need, which triggers a sudden inspiration of air followed by a forceful expelling of that air through vocal cords, which vibrate to produce the sound called a cry.
- The baby's cry is disturbing, even ear-piercing, loud enough to catch the caregiver's attention but not so disturbing as to make the listener want to avoid the sound altogether.
- Third, the cry can be personalized as both the sender and the listener learn ways to make the signal more precise. Each baby's cry is as unique as his or her fingerprints.

Infancy

Crying in infants is a normal, healthy means of expression and communication. The average six-week-old baby cries for two-and-a-half hours every day. Infants cry because they are hungry, uncomfortable, in **pain**, overstimulated, tired, or just bored. A new mother can distinguish her infant's crying from that of other babies within three days, and some fathers can make this distinction as well. A hungry cry begins softly and then becomes loud and rhythmic; an angry cry is similar to a hungry cry but louder. A cry of pain has a distinctive pattern, beginning with a single shriek followed by a short silence and then continuous loud wailing. Neglected or abused infants have a high-pitched cry that is difficult for adults to tolerate. This cry is characteristic of babies born to crack-addicted mothers and has been linked to abnormalities in the central nervous system. An infant's crying patterns and ability to be comforted are important indicators of **temperament**, both in infancy and even in later years.

The most common way to comfort a crying infant is to hold him or her close to the chest. Some infants are soothed by the motion of a cradle, rocking chair, stroller, swing, or automobile. Sucking on a pacifier is another comfort. Other methods include a warm bath, a massage, music, or some background noise, such as the sound of a

hair dryer, a washing machine, or fan. There are also special recordings that reproduce sounds similar to those the infant heard while in the mother's womb. Some infants are hypersensitive to stimuli, and their crying will get worse if they receive any more than a minimum of comforting, such as parental holding or cuddling. A sign of healthy emotional development is the degree to which an infant learns to comfort him- or herself, either with the aid of an object such as a stuffed toy or blanket, or by certain patterns of behavior, such as sucking on a thumb.

Another cause of excessive crying is hypersensitivity. Hypersensitive infants cry in response to new experiences that do not normally upset other babies; ordinary comforting measures, such as holding, rocking, feeding, or swaddling do not work and may even make the crying worse. Hypersensitivity can be a matter of temperament, and it may be influenced by the behavior and attitude of the parents. Some children get into the habit of excessive crying as a way of demanding parental attention. The parents of such children may be overprotective, not giving them the chance to develop independence and resourcefulness by solving problems on their own.

Common problem

Many new parents are not prepared for the amount of time a newborn spends crying. Infants typically cry an average of two hours of every 24 for the first seven weeks of life. The duration peaks at about six or seven weeks. Almost all infants have a period during the day when they are fussy. New parents need to recognize this as normal and not worry. Parents might use this fussy time for bathing or playing with the infant. The most typical time for fussy times is between 6 p.m. and 11 p.m., often when parents are tired and less able to tolerate crying.

A common cause of persistent crying in infants is colic, which is caused by gastrointestinal distress. Colicky infants may have a hard abdomen, get red in the face, and curl their legs up. Often times the colic begins in the evening after the baby's last meal before bedtime. Rocking or walking around with the baby held up against the shoulder can sometimes soothe the infant. Holding the infant face-down across the lap puts pressure on the belly that can sometimes ease the distress.

Among the most common physical reasons for excessive crying are earaches, viral illnesses, and other causes of low-grade **fever**. Teething also causes increased crying. Medical attention may be necessary if an infant is crying more than usual or if the cries themselves sound different, for example, the cries are weaker or more high-pitched than usual.

Different kinds of cries

As parents get to know their baby, they become experts in understanding the baby's cries. Cries are the baby's form of communication. Following are several common reasons babies cry:

- Hunger: If three or four hours have passed since the last feeding, if the infant has just awakened, or if he or she had just had a full diaper and begins to cry, the baby is probably hungry. Most babies eat six to ten times in a 24-hour period. For a least the first three months, babies usually wake for night feedings.
- Tiredness: The baby has decreased activity, loses interest in people and **toys**, rubs eyes, looks glazed, and yawns. If the infant cries, he or she may just need to take a nap.
- Discomfort: If babies are uncomfortable, too wet, too hot, or too cold, they will squirm or arch their back when crying, as they try to get away from the source of discomfort. The distress of gas or indigestion can cause the baby to cry, as can wet or soiled diapers and uncomfortable positions. Parents should try to find the source of the child's distress and solve the problem.
- Pain: A sudden shrill cry followed by a brief silence and then more crying communicates pain. The parents should examine the baby carefully to locate the source of pain and remove it.
- Overstimulation: When overtired or overstimulated, babies cry to release tension. If the room is noisy and people are trying to get the baby's attention, the baby may close his eyes, turn his head away, and cry. The parent should find a quiet, dark room and hold the baby until he or she is calmer.
- Illness: When babies are sick, they may cry in a weak, moaning way. If the baby seems ill, parents should take their temperature and call the healthcare provider.
- Frustration: Some babies cry out of frustration because they cannot do what they want to do. Sometimes they want a toy but cannot control their arms and hands enough to reach it. Taking note of the baby's attention to an object and putting it within reach may solve the problem.
- Loneliness: Babies who fall asleep feeding and are placed in a crib may wake soon afterwards crying. These babies are signaling that they miss the warmth of their parent's embrace and do not like being alone. A baby seeking such comfort may calm down simply with the assuring sight of mother, hearing her voice, feeling her touch, cuddling, or being offered something to suck.

- Worry or **fear**: When the baby suddenly finds himself in the arms of a stranger and cannot see the parent, the baby may begin to cry. Some babies need more time than others to warm up to someone new.
- Boredom: If the baby has been left to **play** while a parent is busy with another task, boredom may set in. The child is not tired, hungry, or uncomfortable, but starts a whiny, fussy cry. A new position or different toy may help.

Parental concerns

Responding to baby's cry is biologically correct. Mothers are biologically programmed to give a nurturing response to their newborn's cries and not to restrain themselves. Biological changes take place in a mother's body in response to their infant's cry. On hearing the baby cry, the blood flow to the mother's breast increases, with a biological urge to nurse. The act of breastfeeding itself causes a surge of prolactin. Hormones that cause a mother's milk to let down brings feelings of relaxation and pleasure; it is a pleasant release from the tension built up by the baby's cry. These biological responses help mothers to connect with and meet their babies' needs.

New parents sometimes feel guilty when their babies cry. It is not the parent's fault that babies cry. Parents should be responsive to their babies and try to help them feel secure in their world. The parent's job is to create a sympathetic environment that lessens the baby's need to cry, to offer a set of caring and relaxed arms so the baby does not need to cry, and to do as much detective work to figure out why the baby is crying as possible.

There are many times when parents cannot tell why their baby is crying. Some things to try include:

- Hold the baby. No matter the reason for the baby's crying, holding offers security and may calm the baby.
- Breastfeed. Nursing the baby is as much for comfort as food. Most babies calm easily when offered the breast.
- Provide motion. Babies enjoy repetitive, rhythmic motion such as rocking, swinging, swaying, and jiggling. Many parents sway when they are holding a fussy baby because it soothes the baby.
- Turn on some white noise. The womb was a noisy place. Sometimes the baby can be calmed by white noise that is continuous and uniform, such as that of a heartbeat, the rain, static between radio stations, and the vacuum cleaner. Some alarm clocks even have a white noise.

KEY TERMS

Diagnosis—The art or act of identifying a disease from its signs and symptoms.

Hormone—A chemical messenger secreted by a gland or organ and released into the bloodstream. It travels via the bloodstream to distant cells where it exerts an effect.

Hypersensitivity—A condition characterized by an excessive response by the body to a foreign substance. In hypersensitive individuals even a tiny amount of allergen can cause a severe allergic reaction.

Swaddling—To wrap the infant securely in clothing or blankets; to provide comfort and control.

- Let music soothe the baby. Soft, peaceful music calms some babies.
- Give the baby a massage. Babies love to be touched and stroked, so a massage is one way to calm a fussy baby. A variation of massage is the baby pat; many babies love a gentle rhythmic pat on their backs or bottoms.
- Swaddle the baby. During the first three or four months of life, many babies feel comforted if they are held tightly in a way the reminds them of the womb.
- Distract the baby. Sometimes a new activity or change of scenery is calming. Maybe a walk outside, a dance with a song, or a splashy bath can be helpful in turning a fussy baby into a happy one.
- Let the baby have something to suck on. The most natural pacifier is mother's breast, but pacifiers and teething rings also may work. It is important for everything to be clean that goes into the baby's mouth.

When to call the doctor

Parents should call the healthcare provider if there are concerns about why the baby continues to cry. It is important not to misdiagnose a serious condition and call it colic. If the baby's behavior or crying pattern changes suddenly or if the crying is associated with fever, forceful **vomiting**, **diarrhea**, bloody stools, or other abnormal spasms or symptoms, call the doctor immediately. Parents should not hesitate to seek help immediately if they feel overwhelmed and are afraid that they will hurt or neglect their baby.

See also Colic.

Resources

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Cryptochidism *see* **Undescended testes**

CT scan *see* **Computed tomography**

Custody laws *see* **Child custody laws**

Cuts *see* **Wounds**

Cyclic vomiting syndrome

Definition

First described in 1882, cyclic **vomiting** syndrome (CVS) is a rare idiopathic disorder characterized by recurring periods of vomiting in an otherwise normal child. The word, idiopathic, means that the origin of the disorder is unknown. The syndrome is sometimes called abdominal migraine because it may be caused by some of the same mechanisms in the central nervous system that cause migraine headaches.

Description

Children with cyclic vomiting syndrome have bouts of severe **nausea and vomiting** that may last for hours or days. In some cases the vomiting is so severe that the child is unable to go to school for several days. The episodes alternate with periods of normal digestive functioning.

The bouts of vomiting that characterize CVS usually begin at the same time of day as previous episodes, last about the same length of time, and have the same symptoms. The most common pattern is severe **nausea** and vomiting that begins late at night or early in the morning. The child may vomit as often as six to 12 times an hour over a period of one to five days, although cases have been reported in which the episode lasts for ten days. The vomited material may contain blood or bile as well as mucus or watery fluid.

In addition to the vomiting, the child may have a **headache**, low-grade **fever**, **dizziness**, **pain** in the abdomen, heavy drooling, and **diarrhea**. Some children also become unusually sensitive to light, while others may be unable to walk or talk.

Demographics

Children between the ages of three and seven years are most susceptible to CVS, although it can appear at any time from infancy to adulthood. The average age of patients at onset is 5.2 years, but CVS has been diagnosed in patients as old as 73.

The frequency of cyclic vomiting syndrome in the general population is not known for certain as of the early 2000s, but it is thought that the disorder is probably underdiagnosed because other diseases and disorders can also cause periods of acute nausea and vomiting. Some researchers think that as many as one child in 50 may have CVS.

CVS appears to affect all races and ethnic groups equally. The female-to-male ratio has been reported as 11 to nine.

Causes and symptoms

Causes

The cause of CVS is as of 2004 a mystery. Similarities to migraine suggest a common cause, but no firm evidence has been found. It is known, however, that 82 percent of patients with CVS have a **family** history of migraine compared to 14 percent of control subjects. Patients can usually identify some factor that precedes an attack. Common triggers of CVS episodes include the following:

- stress and excitement
- certain foods, particularly chocolate and cheese
- bacterial or viral infections, particularly colds, sinus infections, and influenza
- hot and humid weather

- **motion sickness**
- lack of sleep
- menstruation

In the summer of 2003, two teams of researchers in Italy and the United States reported that some cases of CVS appear to be caused by a DNA mutation that affects the proper functioning of the mitochondria (energy generators) in human cells and that this mutation is inherited from the mother. Further research is needed, however, in order to determine whether other genetic factors are involved in CVS.

Symptoms

Vomiting associated with CVS can be protracted and lead to such complications as **dehydration**; erosion of tooth enamel leading to **tooth decay**; unbalanced blood electrolyte levels; and tearing, burning, or bleeding of the esophagus (swallowing tube). Between attacks, however, the child has no sign of any illness.

CVS has four distinct stages or phases:

- Prodrome: A warning symptom (or group of symptoms) appears just before an acute attack of an illness. Patients with CVS often feel pain in the abdomen a few minutes or hours before the vomiting starts. Adults with CVS often have **anxiety** or panic attacks as a prodrome.
- Episode phase: The patient is actively nauseated and vomiting and cannot keep down any food or medications given by mouth. He or she may also feel drowsy, dizzy, or exhausted.
- Recovery phase: The vomiting stops and the child's normal appetite and level of energy return.
- Symptom-free interval.

When to call the doctor

The vomiting and other symptoms associated with CVS are so severe that parents will usually call the doctor during the first episode, before a pattern has been identified. It may take several episodes of the disorder before the parents or the doctor notice a pattern.

Diagnosis

The most important and difficult aspect of diagnosing CVS is to make sure there is not an acute and life-threatening event in progress. So many different diseases can cause vomiting—from bowel obstruction to epilepsy—that an accurate and timely diagnosis is critical. Because there is no way to prove the diagnosis of CVS, the physician must instead disprove every other diagnosis. This process, which is known as a diagnosis of exclusion, can be tedious, expensive, exhausting, and involve almost every system in the body. The first episode of cyclic vomiting syndrome may be diagnosed as stomach flu when nothing more serious turns up. Only after several episodes and several fruitless searches for a cause will a physician normally consider the diagnosis of CVS.

A careful history-taking is critical to making the correct diagnosis of CVS. A family history of migraine, particularly on the mother's side of the family, should alert the doctor to the possibility that the patient has CVS. The doctor may also order blood tests for metabolic screening or imaging studies of the kidneys, gall bladder, small bowel, or sinuses in order to rule out endocrine disorders, gastrointestinal disorders, kidney disease, and chronic **sinusitis**.

In some cases, the doctor may refer the patient to a psychiatrist for evaluation in order to rule out depression, anxiety disorders, or an eating disorder.

Treatment

There is no permanent cure for cyclic vomiting syndrome as of the early 2000s. Doctors as of 2004 recommend a combination of several strategies for managing the disorder:

- Avoidance of known dietary or stress-related triggers: Such triggers as hot weather or automobile transportation, however, may be difficult or impossible to avoid.
- Prophylactic treatment with medications: Prophylactic treatment refers to therapy that is given to prevent a disease. This approach is recommended for children with CVS who have 10 to 12 episodes per year or have episodes of vomiting lasting longer than three days. Several different medications have given good results in small trials. The antimigraine drugs amitriptyline (Elavil) and cyproheptadine (Periactin) performed well for children in one study group. Propranolol (Inderal) is sometimes effective for children with CVS, and erythromycin helped several patients in one study—not because it is an antibiotic but because it irritates the stomach and encourages it to move its contents forward instead of in reverse.
- Abortive treatment: Abortive treatment is therapy given to stop an attack of CVS after it has begun. Drugs that have been found to work well as abortive agents are ondansetron (Zofran, an antinausea drug) and sumatriptan (Imitrex, an antimigraine drug). These drugs can be given intravenously, and sumatriptan is also available as a nasal solution.

- Supportive care: Supportive care for episodes of CVS includes such antinausea drugs as diphenhydramine (Benadryl) or chlorpromazine (Largactil), and intravenous fluids when necessary.

Another medication that has been reported to be successful in treating children with CVS is dexmedetomidine (Precedex), which was originally developed to sedate patients on respirators in intensive care settings. The researchers found that dexmedetomidine relieved the anxiety as well as the nausea associated with CVS.

Alternative treatment

Constitutional homeopathic medicine can work well in treating CVS because it addresses the person's overall health, not just the treatment of acute symptoms.

Stress management techniques may be helpful for older children or teenagers in preventing episodes of CVS triggered by emotional or psychological stress. These techniques may include the relaxation response developed by Herbert Benson, meditation, and biofeedback.

Weekly outpatient acupuncture treatments are also helpful to some children with CVS.

Nutritional concerns

Avoiding dehydration is the primary nutritional concern during episodes of cyclic vomiting syndrome. In most cases the child will bring up water that is offered during the acute phase of an attack even though he or she may be very thirsty. About 50 percent of children require an intravenous infusion of glucose and water to prevent dehydration.

Some children have a normal appetite for food soon after the vomiting stops, while others may take several days to return to a full diet. Parents should offer the child clear liquids first to prevent dehydration and gradually reintroduce solid foods as the child's appetite improves.

Prognosis

The average duration of cyclic vomiting syndrome is 2.5 to 5.5 years. Some children, however, continue to have episodes of the disorder into adulthood. About 60 percent of children diagnosed with CVS eventually develop migraine headaches in **adolescence** or early adulthood. If the more severe complications of prolonged vomiting can be successfully prevented or managed, however, most patients can lead normal lives between acute attacks.

Prevention

Some episodes of vomiting may be prevented by avoiding specific triggers or by taking prophylactic medications. As the cause of the disorder is as of 2004 not yet fully understood, however, there is no way to prevent CVS as a whole.

Parental concerns

Cyclic vomiting syndrome can be a heavy emotional and financial burden on the families of affected children. Episodes of CVS are often upsetting or downright frightening to other family members, in addition to the fact that they often spoil family outings or vacations when they are triggered by excitement or motion sickness. Moreover, CVS can interfere with a child's schooling; most children diagnosed with the disorder miss an average of 20 school days per year and may require tutoring or **home schooling**.

See also Dehydration; Motion sickness; Nausea and vomiting.

Resources

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ORGANIZATIONS

Cyclic Vomiting Syndrome Association in the United States and Canada (CVSA—USA/Canada). 3585 Cedar Hill Road, NW, Canal Winchester, OH 43110. Web site: <www.cvsaonline.org>.

National Organization for Rare Disorders Inc. (NORD). 55 Kenosia Avenue, PO Box 1968, Danbury, CT 06813. Web site: <www.rarediseases.org>.

KEY TERMS

Abdominal migraine—Another term that is sometimes used for cyclic vomiting syndrome (CVS).

Idiopathic—Refers to a disease or condition of unknown origin.

Mitochondria—Spherical or rod-shaped structures of the cell. Mitochondria contain genetic material (DNA and RNA) and are responsible for converting food to energy.

Prodrome—Early symptoms that warn of the beginning of disease. For example, the herpes prodrome consists of pain, burning, tingling, or itching at a site before blisters are visible while the migraine prodrome consists of visual disturbances.

Prophylactic—Preventing the spread or occurrence of disease or infection.

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Cystic fibrosis

Definition

Cystic fibrosis (CF) is an inherited disease that affects the lungs, digestive system, sweat glands, and male fertility. Its name derives from the fibrous scar tissue that develops in the pancreas, one of the principal organs affected by the disease.

Description

Cystic fibrosis affects the body's ability to move salt and water in and out of cells. This defect causes the lungs and pancreas to secrete thick mucus, blocking passageways and preventing proper function.

Many of the symptoms of CF can be treated with drugs or nutritional supplements. Close attention to and prompt treatment of respiratory and digestive complications have dramatically increased the expected life span of a person with CF. While in the 1970s, most children with CF died by age two, in the early 2000s about half of all people with CF live past age 31. That median age is expected to grow as new treatments are developed, and it is estimated that a person born in 1998 with CF has a median expected life span of 40 years.

Demographics

CF affects approximately 30,000 children and young adults in the United States, and about 3,000 babies are born with CF every year. CF primarily affects people of white northern-European descent; rates are much lower in non-white populations.

Causes and symptoms

Causes

Cystic fibrosis is a genetic disease, meaning it is caused by a defect in the person's genes. Genes, found in the nucleus of all the body's cells, control cell function by serving as the blueprint for the production of proteins. Proteins carry out a wide variety of functions within cells. The gene that, when defective, causes CF is called the CFTR gene, which stands for cystic fibrosis transmembrane conductance regulator. A simple defect in this gene leads to all the consequences of CF. There are over 500 known defects in the CFTR gene that can cause CF. However, 70 percent of all people with a defective CFTR gene have the same defect, known as delta-F508.

Much as sentences are composed of long strings of words, each made of letters; genes can be thought of as long strings of chemical words, each made of chemical letters, called nucleotides. Just as a sentence can be changed by rearranging its letters, genes can be mutated, or changed, by changes in the sequence of their nucleotide letters. The gene defects in CF are called point mutations, meaning that the gene is mutated only at one small spot along its length. In other words, the delta-F508 mutation is a loss of one "letter" out of thousands within the CFTR gene. As a result, the CFTR protein made from its blueprint is made incorrectly and cannot perform its function properly.

The CFTR protein helps to produce mucus. Mucus is a complex mixture of salts, water, sugars, and proteins that cleanses, lubricates, and protects many passageways in the body, including those in the lungs and pancreas. The role of the CFTR protein is to allow chloride ions to

exit the mucus-producing cells. When the chloride ions leave these cells, water follows, thinning the mucus. In this way, the CFTR protein helps to keep mucus from becoming thick and sluggish, thus allowing the mucus to be moved steadily along the passageways to aid in cleansing.

In CF, the CFTR protein cannot allow chloride ions out of the mucus-producing cells. With less chloride leaving, less water leaves, and the mucus becomes thick and sticky. It can no longer move freely through the passageways, so they become clogged. In the pancreas, clogged passageways prevent secretion of digestive enzymes into the intestine, causing serious impairment of digestion—especially of fat—which may lead to **malnutrition**. Mucus in the lungs may plug the airways, preventing good air exchange and, ultimately, leading to emphysema. The mucus is also a rich source of nutrients for bacteria, leading to frequent infections.

INHERITANCE OF CYSTIC FIBROSIS Each person actually has two copies of each gene, including the CFTR gene, in each of their body cells. During sperm and egg production, however, these two copies separate, so that each sperm or egg contains only one copy of each gene. When sperm and egg unite, the newly created cell once again has two copies of each gene.

The two gene copies may be the same or they may be slightly different. For the CFTR gene, for instance, a person may have two normal copies, or one normal and one mutated copy, or two mutated copies. A person with two mutated copies will develop cystic fibrosis. A person with one mutated copy is said to be a carrier. A carrier will not have symptoms of CF but can pass on the mutated CFTR gene to his/her children.

When two carriers have children, they have a one-in-four chance of having a child with CF each time they conceive. They have a two-in-four chance of having a child who is a carrier, and a one-in-four chance of having a child with two normal CFTR genes.

Approximately one in every 25 Americans of northern-European descent is a carrier of the mutated CF gene, while only one in 17,000 African Americans and one in 30,000 Asian Americans are carriers. Since carriers are symptom-free, very few people know if they are carriers unless there is a **family** history of the disease. Two white Americans with no family history of CF have a one in 2,500 chance of having a child with CF.

It may seem puzzling that a mutated gene with such harmful consequences would remain so common; one might guess that the high mortality of CF would quickly lead to loss of the mutated gene from the population. Some researchers in the early 2000s believe the reason for the persistence of the CF gene is that carriers, those with only one copy of the gene, are protected from the full effects of cholera, a microorganism that infects the intestine, causing intense **diarrhea** and eventual death by **dehydration**. It is believed that having one copy of the CF gene is enough to prevent the full effects of cholera infection, while not enough to cause the symptoms of CF. This so-called ''heterozygote advantage'' is seen in some other genetic disorders, including sickle-cell anemia.

Symptoms

The most severe effects of cystic fibrosis are seen in two body systems: the gastrointestinal (digestive) system and the respiratory tract from the nose to the lungs. CF also affects the sweat glands and male fertility. Symptoms develop gradually, with gastrointestinal symptoms often the first to appear.

GASTROINTESTINAL SYSTEM Approximately 10 to 15 percent of babies who inherit CF have meconium **ileus** at birth. Meconium is the first dark stool that a baby passes after birth; ileus is an obstruction of the digestive tract. The meconium of a newborn with meconium ileus is thickened and sticky, due to the presence of thickened mucus from the intestinal glands. Meconium ileus causes abdominal swelling and **vomiting** and often requires surgery immediately after birth. Presence of meconium ileus is considered highly indicative of CF. Borderline cases may be misdiagnosed, however, and attributed instead to milk allergy.

Other abdominal symptoms are caused by the inability of the pancreas to supply digestive enzymes to the intestine. During normal digestion, as food passes from the stomach into the small intestine, it is mixed with pancreatic secretions that help to break down the nutrients for absorption. While the intestines themselves also provide some digestive enzymes, the pancreas is the major source of enzymes for the digestion of all types of foods, especially fats and proteins.

In CF, thick mucus blocks the pancreatic duct, which is eventually closed off completely by scar tissue formation, leading to a condition known as pancreatic insufficiency. Without pancreatic enzymes, large amounts of undigested food pass into the large intestine. Bacterial action on this rich food source can cause gas and abdominal swelling. The large amount of fat remaining in the feces makes it bulky, oily, and foul-smelling.

Because nutrients are only poorly digested and absorbed, the person with CF is often ravenously hungry, underweight, and shorter than expected for his age. When CF is not treated for a longer period, a child may

develop symptoms of malnutrition, including anemia, bloating, and, paradoxically, appetite loss.

Diabetes becomes increasingly likely as a person with CF ages. Scarring of the pancreas slowly destroys those pancreatic cells which produce insulin, producing type I, or insulin-dependent diabetes.

Gallstones affect approximately 10 percent of adults with CF. Liver problems are less common but can be caused by the buildup of fat within the liver. Complications of liver enlargement may include internal hemorrhaging, accumulation of abdominal fluid (ascites), spleen enlargement, and liver failure.

Other gastrointestinal symptoms can include a prolapsed rectum, in which part of the rectal lining protrudes through the anus; intestinal obstruction; and rarely, intussusception, in which part of the intestinal tube slips over an adjoining part, cutting off blood supply.

Somewhat less than 10 percent of people with CF do not have gastrointestinal symptoms. Most of these people do not have the delta-F508 mutation but a different one, which presumably allows at least some of their CFTR proteins to function normally in the pancreas.

RESPIRATORY TRACT The respiratory tract includes the nose, the throat, the trachea (or windpipe), the bronchi (which branch off from the trachea within each lung), the smaller bronchioles, and the blind sacs called alveoli, in which gas exchange takes place between air and blood.

Swelling of the sinuses within the nose is common in people with CF. This usually shows up on an x ray and may aid the diagnosis of CF. However, this swelling, called pansinusitis, rarely causes problems and does not usually require treatment.

Nasal polyps, or growths, affect about one in five people with CF. These growths are not cancerous and do not require removal unless they become annoying. While nasal polyps appear in older people without CF, especially those with **allergies**, they are rare in children without CF.

The lungs are the site of the most life-threatening effects of CF. The production of a thick, sticky mucus increases the likelihood of infection, decreases the ability to protect against infection, causes inflammation and swelling, decreases the functional capacity of the lungs, and may lead to emphysema. People with CF live with chronic populations of bacteria in their lungs, and lung infection is the major cause of death for those with CF.

The bronchioles and bronchi normally produce a thin, clear mucus that traps foreign particles including bacteria and viruses. Tiny hair-like projections on the surface of these passageways slowly sweep the mucus along, out of the lungs and up the trachea to the back of the throat, where it may be swallowed or coughed up. This "mucociliary escalator" is one of the principal defenses against lung infection.

The thickened mucus of CF prevents easy movement out of the lungs and increases the irritation and inflammation of lung tissue. This inflammation swells the passageways, partially closing them down, further hampering the movement of mucus. A person with CF is likely to **cough** more frequently and more vigorously as the lungs attempt to clean themselves out.

At the same time, infection becomes more likely since the mucus is a rich source of nutrients. **Bronchitis**, **bronchiolitis**, and **pneumonia** are frequent in CF. The most common infecting organisms are the bacteria *Staphylococcus aureus, Haemophilus influenzae*, and *Pseudomonas aeruginosa*. A small percentage of people with CF have infections caused by *Burkholderia cepacia*, a bacterium which was resistant to most **antibiotics** as of 2004. (*Burkholderia cepacia* was formerly known as *Pseudomonas cepacia*.) The fungus *Aspergillus fumigatus* may infect older children and adults.

The body's response to infection is to increase mucus production; white blood cells fighting the infection thicken the mucus even further as they break down and release their cell contents. These white blood cells also provoke more inflammation, continuing the downward spiral that marks untreated CF.

As mucus accumulates, it can plug up the smaller passageways in the lungs, decreasing functional lung volume. Getting enough air can become difficult; tiredness, shortness of breath, and intolerance of **exercise** become more common. Because air passes obstructions more easily during inhalation than during exhalation, over time, air becomes trapped in the smallest chambers of the lungs, the alveoli. As millions of alveoli gradually expand, the chest takes on the enlarged, barrel-shaped appearance typical of emphysema.

For unknown reasons, recurrent respiratory infections lead to digital clubbing, in which the last joint of the fingers and toes becomes slightly enlarged.

SWEAT GLANDS The CFTR protein helps to regulate the amount of salt in sweat. People with CF have sweat that is much saltier than normal, and measuring the saltiness of a person's sweat is the most important diagnostic test for CF. Parents may notice that their infants taste salty when they kiss them. Excess salt loss is not usually a problem except during prolonged exercise or heat. While most older children and adults with

CF compensate for this extra salt loss by eating more salty foods, infants and young children are in danger of suffering its effects (such as heat prostration), especially during summer. Heat prostration is marked by lethargy, weakness, and loss of appetite and should be treated as an emergency condition.

FERTILITY Some 98 percent of men with CF are sterile, due to complete obstruction or absence of the vas deferens (the tube carrying sperm out of the testes). While boys and men with CF form normal sperm and have normal levels of sex hormones, sperm are unable to leave the testes, and fertilization is not possible. Most women with CF are fertile, though they often have more trouble getting pregnant than women without CF. In both boys and girls, **puberty** is often delayed, most likely due to the effects of poor **nutrition** or chronic lung infection. Women with good lung health usually have no problems with pregnancy, while those with ongoing lung infection often do poorly.

Diagnosis

The decision to test a child for cystic fibrosis may be triggered by concerns about recurring gastrointestinal or respiratory symptoms or salty sweat. A child born with meconium ileus will be tested before leaving the hospital. Families with a history of CF may wish to have all children tested, especially if there is a child who already has the disease. Some hospitals require routine screening of newborns for CF.

Sweat test

The **sweat test** is both the easiest and most accurate test for CF. In this test, a small amount of the drug pilocarpine is placed on the skin. A very small electrical current is then applied to the area, which drives the pilocarpine into the skin. The drug stimulates sweating in the treated area. The sweat is absorbed onto a piece of filter paper and is then analyzed for its salt content. A person with CF will have salt concentrations that are 1.5 to 2 times greater than normal. The test can be done on persons of any age, including newborns, and its results can be determined within an hour. Virtually every person who has CF will test positively on it, and virtually everyone who does not will test negatively.

Genetic testing

The discovery of the CFTR gene in 1989 allowed the development of an accurate genetic test for CF. Genes from a small blood or tissue sample are analyzed for specific mutations; presence of two copies of the mutated gene confirms the diagnosis of CF in all but a very few cases. However, since there are so many different possible mutations and since testing for all of them would be too expensive and time-consuming, a negative gene test cannot rule out the possibility of CF.

Couples planning a family may decide to have themselves tested if one or both have a family history of CF. Prenatal genetic testing is possible through **amniocentesis**. Many couples who already have one child with CF decide to undergo prenatal screening in subsequent pregnancies and use the results to determine whether to terminate the pregnancy. Siblings in these families are also usually tested, to determine if they will develop CF and to determine if they are carriers, to aid in their own family planning. If the sibling has no symptoms, determining his carrier status is often delayed until his teen years or later, when he is closer to needing the information to make decisions.

Newborn screening

Some states in the early 2000s require screening of newborns for CF, using a test known as the IRT test. This blood test measures the level of immunoreactive trypsinogen, which is generally higher in babies with CF than those without it. This test gives many false positive results immediately after birth and so requires a second test several weeks later. A second positive result is usually followed by a sweat test.

Treatment

As of 2004 there was no cure for CF. However, treatment advanced during the last quarter of the twentieth century, increasing both the life span and the quality of life for most people affected by CF. Early diagnosis is important to prevent malnutrition and infection from weakening the young child. With proper management, many people with CF engage in the full range of school and **sports** activities.

Nutrition

People with CF usually require high-calorie diets and vitamin supplements. Height, weight, and growth of a person with CF are monitored regularly. Most people with CF need to take pancreatic enzymes to supplement or replace the inadequate secretions of the pancreas. Tablets containing pancreatic enzymes are taken with every meal; depending on the size of the tablet and the meal, as many as 20 tablets may be needed. Because of incomplete absorption even with pancreatic enzymes, a person with CF needs to take in about 30 percent more food than a person without CF. Low-fat diets are not recommended except in special circumstances, since fat

is a source of both essential fatty acids and abundant calories.

Some people with CF cannot absorb enough nutrients from the foods they eat, even with specialized diets and enzymes. For these people, tube feeding is an option. Nutrients can be introduced directly into the stomach through a tube inserted either through the nose (a nasogastric tube) or through the abdominal wall (a gastrostomy tube). A jejunostomy tube, inserted into the small intestine, is also an option. Tube feeding can provide nutrition at any time, including at night while the person is sleeping, allowing constant intake of high-quality nutrients. The feeding tube may be removed during the day, allowing normal meals to be taken.

Respiratory health

The key to maintaining respiratory health in a person with CF is regular monitoring and early treatment. Lung function tests are done frequently to track changes in functional lung volume and respiratory effort. Sputum samples are analyzed to determine the types of bacteria present in the lungs. Chest x-rays are usually taken at least once a year. Lung scans, using a radioactive gas, can show closed off areas not seen on the x ray. Circulation in the lungs may be monitored by injection of a radioactive substance into the bloodstream.

People with CF live with chronic bacterial colonization; that is, their lungs are constantly host to several species of bacteria. Good general health, especially good nutrition, can keep the immune system healthy, which decreases the frequency with which these colonies begin an infection or attack on the lung tissue. Exercise is another important way to maintain health, and people with CF are encouraged to maintain a program of regular exercise.

In addition, clearing mucus from the lungs helps to prevent infection, and mucus control is an important aspect of CF management. Postural drainage is used to allow gravity to aid the mucociliary escalator. For this technique, the person with CF lies on a tilted surface with head downward, alternately on the stomach, back, or side, depending on the section of lung to be drained. An assistant thumps the rib cage to help loosen the secretions. A device called a flutter offers another way to loosen secretions: it consists of a stainless steel ball in a tube. When a person exhales through it, the ball vibrates, sending vibrations back through the air in the lungs. Some special breathing techniques may also help clear the lungs.

Several drugs are available to prevent the airways from becoming clogged with mucus. Bronchodilators and theophyllines open up the airways; steroids reduce inflammation; and mucolytics loosen secretions. Acetylcysteine (Mucomyst) has been used as a mucolytic during the 1980s and 1990s but is not prescribe frequently in the early 2000s, while DNase (Pulmozyme) is a newer product gaining in popularity. DNase breaks down the DNA from dead white blood cells and bacteria found in thick mucus.

People with CF may pick up bacteria from other CF patients. This is especially true of *Burkholderia cepacia*, which is not usually found in people without CF. While the ideal recommendation from a health standpoint might be to avoid contact with others who have CF, this is not usually practical (since CF clinics are a major site of care), nor does it meet the psychological and social needs of many people with CF. At a minimum, CF centers recommend avoiding prolonged close contact between people with CF and scrupulous hygiene, including frequent hand washing. Some CF clinics schedule appointments on different days for those with and without *B. cepacia* colonies.

Some doctors choose to prescribe antibiotics only during infection, while others prefer long-term antibiotic treatment against *S. aureus*. The choice of antibiotic depends on the particular organism or organisms found. Some antibiotics are given as aerosols directly into the lungs. Antibiotic treatment may be prolonged and aggressive.

Supplemental oxygen may be needed as lung disease progresses. Respiratory failure may develop, requiring temporary use of a ventilator to perform the work of breathing.

Lung transplantation has become increasingly common for people with CF, although the number of people who receive lungs was as of 2004 much lower than those who want them. Transplantation is not a cure, however, and has been likened to trading one disease for another. Long-term immunosuppression is required, increasing the likelihood of other types of infection. About 50 percent of adults and more than 80 percent of children who receive lung transplants live longer than two years. Liver transplants are also done for CF patients whose livers have been damaged by fibrosis.

Long-term use of ibuprofen has been shown to help some people with CF, presumably by reducing inflammation in the lungs. Close medical supervision is necessary, however, since the effective dose is high and not everyone benefits. Ibuprofen at the required doses interferes with kidney function and, together with aminoglycoside antibiotics, may cause kidney failure.

A number of experimental treatments were as of 2004 the subject of much research. Some evidence indi-

cates that aminoglycoside antibiotics may help overcome the genetic defect in some CF mutations, allowing the protein to be made normally. While promising, these results would apply to only about 5 percent of those with CF.

Gene therapy is the most ambitious approach to curing CF. In this set of techniques, non-defective copies of the CFTR gene are delivered to affected cells, where they are taken up and used to create the CFTR protein. While elegant and simple in theory, gene therapy has met with a large number of difficulties in trials, including immune resistance, very short duration of the introduced gene, and inadequately widespread delivery.

Prognosis

People with CF may lead relatively normal lives, with the control of symptoms. The possible effect of pregnancy on the health of a woman with CF requires careful consideration before she and her partner begin a family; as do issues of longevity, and the children's status as carriers. Although most men with CF are functionally sterile, new procedures for removing sperm from the testes are being tried and may offer more men the chance to become fathers.

Approximately half of people with CF live past the age of 30. Because of better and earlier treatment, a person born in 2004 with CF is expected, on average, to live to age 40.

Prevention

Adults with a family history of cystic fibrosis may obtain a genetic test of their carrier status for purposes of family planning. Prenatal testing is also available. There is no known way to prevent development of CF in a person with two defective gene copies.

KEY TERMS

Carrier—A person who possesses a gene for an abnormal trait without showing signs of the disorder. The person may pass the abnormal gene on to offspring. Also refers to a person who has a particular disease agent present within his/her body, and can pass this agent on to others, but who displays no symptoms of infection.

Cystic fibrosis transmembrane conductance regulator (CFTR)—The protein responsible for regulating chloride movement across cells in some tissues. Cystic fibrosis results when a person has two defective copies of the CFTR gene.

Emphysema—A chronic respiratory disease that involves the destruction of air sac walls to form abnormally large air sacs that have reduced gas exchange ability and that tend to retain air within the lungs. Symptoms include labored breathing, the inability to forcefully blow air out of the lungs, and an increased susceptibility to respiratory tract infections. Emphysema is usually caused by smoking.

Mucociliary escalator—The coordinated action of tiny projections on the surfaces of cells lining the respiratory tract, which moves mucus up and out of the lungs.

Mucolytic—An agent that dissolves or destroys mucin, the chief component of mucus.

Pancreatic insufficiency—Reduction or absence of pancreatic secretions into the digestive system due to scarring and blockage of the pancreatic duct.

Resources

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ORGANIZATIONS

Cystic Fibrosis Foundation. 6931 Arlington Road, Bethesda, MD 20814. Web site: <www.cff.org>.

WEB SITES

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Cystitis

Definition

Cystitis is inflammation of the urinary bladder. Urethritis is an inflammation of the urethra, which is the

tube that connects the bladder with the exterior of the body. Sometimes cystitis and urethritis are referred to collectively as a lower urinary tract infection (UTI). Infection of the upper urinary tract involves the spread of bacteria to the kidney and is called pyelonephritis.

Description

Cystitis in women

Cystitis is a common female problem. It is estimated that 50 percent of adult women experience at least one episode of dysuria (painful urination); half of these people have a bacterial UTI. Between 2 percent and 5 percent of women's visits to primary care physicians are for UTI symptoms. About 90 percent of UTIs in women are uncomplicated but recurrent.

Cystitis in men

UTIs are uncommon in younger and middle-aged men but may occur as complications of bacterial infections of the kidney or prostate gland.

Cystitis in children

In children, cystitis often is caused by congenital abnormalities (present at birth) of the urinary tract. **Vesicoureteral reflux** is a condition in which the child cannot completely empty the bladder. The condition allows urine to remain in or flow backward (reflux) into the partially empty bladder. In addition, cystitis can also be caused by wiping forward instead of backward after a bowel movement, especially in girls that are newly toilet trained.

Demographics

The frequency of bladder infections in humans varies significantly according to age and sex. The male/female ratio of UTIs in children younger than 12 months is four to one because of the high rate of birth defects in the urinary tract of male infants. Urinary tract infections are fairly common in young girls. In adult life, the male/female ratio of UTIs is one to 50. After age 50, however, the incidence among males increases due to prostate disorders.

Causes and symptoms

The causes of cystitis vary according to gender because of the differences in anatomical structure of the urinary tract.

Females

Most bladder infections in women are so-called ascending infections, which means they are caused by disease agents traveling upward through the urethra to the bladder. The relative shortness of the female urethra (1.2 to 2 inches [3-5 cm] in length for adults) facilitates bacteria gaining entry to the bladder and multiplying there. The most common bacteria associated with UTIs in women (including teens) are: *Escherichia coli* (approximately 80% of cases), *Staphylococcus saprophyticus*, *Klebsiella*, *Enterobacter*, and *Proteus* species. Risk factors for UTIs in women include:

- sexual intercourse (The risk of infection increases if the woman has multiple partners.)
- use of a diaphragm for contraception
- an abnormally short urethra
- diabetes or chronic **dehydration**
- the absence of a specific enzyme (fucosyltransferase) in vaginal secretions (The lack of this enzyme makes it easier for the vagina to harbor bacteria that cause UTIs.)
- inadequate personal hygiene (Bacteria from fecal matter or vaginal discharges can enter the female urethra because its opening is very close to the vagina and anus.)
- history of previous UTIs (About 80 percent of women with cystitis develop recurrences within two years.)

The early symptoms of cystitis in women are dysuria (**pain** on urination); urgency (a sudden strong desire to urinate); and increased frequency of urination. About 50 percent of females experience **fever**, pain in the lower back or flanks, **nausea and vomiting**, or shaking chills. These symptoms indicate pyelonephritis (spread of the infection to the upper urinary tract).

Males

Most UTIs in adult males are complications of kidney or prostate infections. They usually are associated with a tumor or kidney stones that block the flow of urine and often are persistent infections caused by drug-resistant organisms. UTIs in men are most likely to be caused by *E. coli* or another gram-negative bacterium. Risk factors for UTIs in men include lack of **circumcision** and urinary catheterization. The longer the period of catheterization, the higher the risk of contracting a UTI.

The symptoms of cystitis and pyelonephritis in men are the same as in women.

Children

In children, cystitis causes pain and tenderness in the lower abdomen, frequent urination, blood in the urine, and fever. However, some foods, including citrus juices, **caffeine**, and carbonated beverages, can irritate the lower urinary tract and mimic the symptoms of an infection.

Hemorrhagic cystitis

Hemorrhagic cystitis, which is marked by large quantities of blood in the urine, is caused by an acute bacterial infection of the bladder. In some cases, hemorrhagic cystitis is a side effect of radiation therapy or treatment with cyclophosphamide. Hemorrhagic cystitis in children is associated with adenovirus type 11.

When to call the doctor

A doctor or other healthcare provider should be contacted whenever urination becomes painful or the voided urine is cloudy or bloody, or when a child complains of pain when voiding urine.

Diagnosis

When cystitis is suspected, the doctor first examines a person's abdomen and lower back, to evaluate unusual enlargements of the kidneys or swelling of the bladder. In small children, the doctor checks for fever, abdominal masses, and a swollen bladder.

The next step in diagnosis is collection of a urine sample. The procedure involves voiding into a cup, so small children may be catheterized to collect a sample. Laboratory testing of urine samples as of the early 2000s can be performed with dipsticks that indicate immune system responses to infection, as well as with microscopic analysis of samples. Normal human urine is sterile. The presence of bacteria or pus in the urine usually indicates infection. The presence of hematuria (blood in the urine) may indicate acute UTIs, kidney disease, kidney stones, inflammation of the prostate (in men), endometriosis (in women), or **cancer** of the urinary tract. In some cases, blood in the urine results from athletic training, particularly in runners.

Other tests

Women and children with recurrent UTIs can be given ultrasound exams of the kidneys and bladder together with a voiding cystourethrogram to test for structural abnormalities. (A cystourethrogram is an x-ray test in which an iodine dye is used to better view the urinary bladder and urethra.) In some cases, **computed tomography** scans (CT scans) can be used to evaluate people for possible cancers in the urinary tract.

Treatment

Medications

Uncomplicated cystitis is treated with **antibiotics**. These include penicillin, ampicillin, and amoxicillin; sulfisoxazole or sulfamethoxazole; trimethoprim; nitrofurantoin; cephalosporins; or fluoroquinolones. (Fluoroquinolones generally are not used in children under 18 years of age.) A 2003 study showed that fluoroquinolone was preferred over amoxicillin, however, for uncomplicated cystitis in young women. Treatment for women is short-term; most women respond within three days. Men and children do not respond as well to short-term treatment and require seven to 10 days of oral antibiotics for uncomplicated UTIs.

Persons of either gender may be given phenazopyridine or flavoxate to relieve painful urination.

Trimethoprim and nitrofurantoin are preferred for treating recurrent UTIs in women.

Individuals with pyelonephritis can be treated with oral antibiotics or intramuscular doses of cephalosporins. Medications are given for ten to 14 days and sometimes longer. If the person requires **hospitalization** because of high fever and dehydration caused by **vomiting**, antibiotics can be given intravenously.

Surgery

A minority of women with complicated UTIs may require surgical treatment to prevent recurrent infections. Surgery also is used to treat reflux problems (movement of the urine backward) or other structural abnormalities in children and anatomical abnormalities in adult males.

Alternative treatment

Alternative treatment for cystitis may emphasize eliminating all sugar from the diet and drinking lots of water. Drinking unsweetened cranberry juice not only adds fluid but also is thought to help prevent cystitis by making it more difficult for bacteria to cling to the bladder wall. A variety of herbal therapies also are recommended. Generally, the recommended herbs are antimicrobials, such as garlic (*Allium sativum*), goldenseal (*Hydrastis canadensis*), and bearberry (*Arctostaphylos uva-ursi*); and/or demulcents that soothe and coat the urinary tract, including corn silk and marsh mallow (*Althaea officinalis*).

Homeopathic medicine also can be effective in treating cystitis. Choosing the correct remedy based on the individual's symptoms is always key to the success of this type of treatment. Acupuncture and Chinese traditional herbal medicine can also be helpful in treating acute and chronic cases of cystitis.

Prognosis

The prognosis for recovery from uncomplicated cystitis is excellent.

Prevention

Females

Women and teens with two or more UTIs within a six-month period sometimes are given prophylactic treatment, usually nitrofurantoin or trimethoprim for three to six months. In some cases the woman is advised to take an antibiotic tablet following sexual intercourse.

Other preventive measures for women include drinking large amounts of fluid; voiding frequently, particularly after intercourse; and proper cleansing of the area around the urethra. Children with UTIs should be encouraged to drink plenty of fluids and wipe themselves properly after a bowel movement.

In 2003, clinical trials in humans tested a possible vaccine for recurrent urinary tract infections. The vaccine was administered via a vaginal suppository.

Nutritional concerns

Many experts recommend that people with a UTI should drink cranberry juice, which contains hippuric acid that tends to lower the pH (acidify) of urine. This change reduces the ability of bacteria to thrive, thus helping to cure a UTI.

Parental concerns

Parents should monitor the urine of their young children. Older children should be encouraged to discuss episodes of painful urination with their parents or other knowledgeable persons.

KEY TERMS

Bacteriuria—The presence of bacteria in the urine.

Dysuria—Painful or difficult urination.

Hematuria—The presence of blood in the urine.

Pyelonephritis—An inflammation of the kidney and upper urinary tract, usually caused by a bacterial infection. In its most serious form, complications can include high blood pressure (hypertension) and renal failure.

Urethritis—Inflammation of the urethra, the tube through which the urine moves from the bladder to the outside of the body.

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L. Fleming Fallon, Jr., MD, DrPH

Cytomegalovirus infection

Definition

Cytomegalovirus (CMV) is a virus related to the group of herpes viruses. Infection with CMV can cause

no symptoms or can be the source of serious illness in people with weak immune systems. CMV infection is also an important cause of birth defects.

Description

CMV is an extremely common organism worldwide. It is believed that about 85 percent of the adults in the United States have been infected by CMV at some point in their lives. CMV is found in almost all of the body's organs. It is also found in body fluids, including semen, saliva, urine, feces, breast milk, blood, and secretions of the cervix (the narrow, lower section of the uterus).

CMV is also able to cross the placenta (the organ that provides oxygen and nutrients to the unborn baby in the uterus). Because CMV can cross the placental barrier, initial infection in a pregnant woman can lead to infection of the developing baby.

Demographics

In the United States, about 40 to 60 percent of all adults in the middle- and upper-socioeconomic classes show antibody proof of prior infection with CMV; antibody proof is as high as 80 percent in adults in the lower socioeconomic class. Worldwide, about 0.2 to 2.2 percent of all babies are born with congenital CMV infection. Of those babies born with congenital CMV infection, about 10 percent to 20 percent ultimately suffer form **hearing impairment**, eye damage, or problems with intellectual or motor function.

Causes and symptoms

CMV is passed between people through contact with body fluids. CMV also can be passed through sexual contact. Babies can be born infected with CMV, either becoming infected in the uterus (congenital infection) or during birth (from infected cervical secretions).

Like other herpes viruses, CMV remains inactive (dormant) within the body for life after the initial infection. Some of the more serious types of CMV infections occur in people who have been harboring the dormant virus, only to have it reactivate when their immune system is stressed. Immune systems may be weakened because of **cancer chemotherapy**, medications given after organ transplantation, or diseases that significantly lower immune resistance like acquired **immunodeficiency** syndrome (**AIDS**).

In a healthy person, initial CMV infection often occurs without symptoms and is rarely noticed. Occasionally, a first-time infection with CMV may cause a mild illness called mononucleosis. Symptoms include swollen glands, liver, and spleen; **fever**; increased white blood cells; **headache**; fatigue; and **sore throat**. About 8 percent of all mononucleosis cases are due to CMV infection. A similar infection, though slightly more serious, may occur two to four weeks after receiving a blood transfusion containing CMV.

In people with weakened immune systems, CMV infection can cause more serious and potentially life-threatening illnesses. These illnesses include **pneumonia** and inflammations of the liver (hepatitis), brain (**encephalitis**), esophagus (esophagitis), large intestine (colitis), and retina of the eye (retinitis).

Babies who contract CMV from their mothers during birth rarely develop any illness from these infections. Infants born prematurely who become CMV infected during birth have a greater chance of complications, including pneumonia, hepatitis, decreased blood platelets.

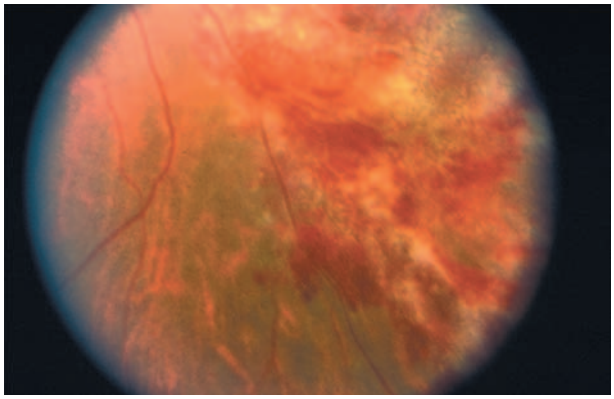
However, an unborn baby is at great risk for serious problems when the mother becomes infected with CMV for the first time while pregnant. About 10 percent of these babies will be born with obvious problems, including **prematurity**, lung problems, an enlarged liver and spleen, **jaundice**, anemia, low birth weight, small head size, and inflammation of the retina. About 90 percent of these babies may appear perfectly normal at birth. Unfortunately, about 20 percent later develop severe hearing impairments and **mental retardation**. A 2003 report found that pregnant women 25 years of age and older who are immune to CMV are much less likely to pass the virus to their babies than younger women who have never been exposed to CMV.

Diagnosis

Body fluids or tissues can be tested to reveal CMV infection. However, this information is not always particularly helpful because CMV stays dormant in the cells for life. Tests to look for special immune cells (antibodies) that are directed specifically against CMV are useful in proving that a person has been infected with CMV. However, these tests do not give any information regarding when the CMV infection first occurred.

Treatment

Ganciclovir and foscarnet are antiviral medications that have been used to treat patients with weak immune systems who develop a serious illness from CMV (including retinitis). As of 1998, research was still being done to try to find useful drugs to treat newborn babies



An infected retina caused by a cytomegalovirus. *(Photograph by Paula Ihnat. Custom Medical Stock Photo, Inc.)*

KEY TERMS

Cervix—A small, cylindrical structure about an inch or so long and less than an inch around that makes up the lower part and neck of the uterus. The cervix separates the body and cavity of the uterus from the vagina.

Congenital—Present at birth.

Placenta—The organ that provides oxygen and nutrition from the mother to the unborn baby during pregnancy. The placenta is attached to the wall of the uterus and leads to the unborn baby via the umbilical cord.

suffering from congenital infection with CMV. **Antiviral drugs** are not used to treat CMV infection in otherwise healthy patients because the drugs have significant side effects that outweigh their benefits. In 2003, researchers in Europe announced a new compound that appeared to be highly effective against CMV infections. The new drug acted earlier in the viral replication of the infection and showed promise; however, clinical trials were continuing.

Prognosis

Prognosis in healthy people with CMV infection is excellent. About 0.1 percent of all newborn babies have serious damage from CMV infection occurring while they were developing in the uterus. About 50 percent of all transplant patients develop severe illnesses due to reactivation of dormant CMV infection. These illnesses have a high rate of serious complications and death.

Prevention

Prevention of CMV infection in the normal, healthy person involves good hand washing. Blood products can be screened or treated to insure that they do not contain CMV. In 2003, a new high-dose prophylactic (preventive) treatment was being tested to reduce CMV risk in stem cell transplant recipients.

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The GALE ENCYCLOPEDIA *of* CHILDREN'S HEALTH

INFANCY THROUGH ADOLESCENCE

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Dandruff *see* **Seborrheic dermatitis**

Day care

Definition

Day care refers to the care provided for infants and toddlers, preschoolers, and school-aged children, either in their own homes, in the home of a relative or other caregiver, or in a center-based facility.

Description

The last half of the twentieth century saw a dramatic rise in the numbers of women with young children who worked outside of the home. In 2000, 55 percent of mothers with infants were in the labor force. In 2001, 64 percent of mothers with children under the age of six, and 78 percent of mothers with children ages six to seventeen were in the labor force. These developments led to an increased demand for childcare providers by parents while they are at work.

In 2001, 61 percent of all children participated in some sort of nonparental care. As children grow older, the likelihood they will receive care from someone other than a parent increases.

Types of day care

Center-based care

Center-based care may also be labeled child or daycare centers, nursery schools, or preschools. These facilities care for children in groups. They may have different sponsors, including universities, schools, churches, social service agencies, independent owners or chains, and employers. Many parents choose center-based care because they believe the presence of multiple caregivers, larger groups of children, and state inspections make them both safer and more dependable. Some parents also consider these types of centers a better learning environment for their children.

The National Association for the Education of Young Children (NAEYC) issues recommendations relating to the organization and structure of daycare centers, particularly those that provide care for infants and toddlers. These recommendations are considered to be the minimum standards a daycare center should observe. Their recommendations concerning staff to child ratios are as follows:

- There should be no more than four infants per caregiver, and no more than eight infants per one group of children in center-based care.
- There should be no more than four young toddlers (12–24 months) per caregiver, with a maximum of 12 young toddlers and three caregivers per group. They recommend there be no more than six older toddlers (24–36 months) per caregiver, and a maximum of 12 older toddlers and two caregivers per group.

Some of the advantages of center-based care are:

- The staff are trained and supervised.
- There are more resources and equipment available.
- Care is still available when a staff member is absent.
- The centers are more likely to be licensed and subject to state regulation.
- Children in center-based care demonstrate slightly better **cognitive development** than those cared for in homes, possibly because they have more opportunities to interact with other children and are exposed to more learning materials.

Some of the disadvantages of center-based care are:

- The costs are higher than for other types of care.
- The background of staff can vary greatly, and there is often greater staff turnover.

- Larger groups of children may mean less individual attention for the child.
- There is a greater likelihood of exposure to communicable illnesses.

Family childcare providers

Family childcare providers offer care for children in the provider's home. Requirements differ from state to state. However, the majority of states require that providers be regulated if they are watching more than four children. Many states may have a voluntary regulation process in place for those providers caring for four or fewer children. Regulations usually require providers to meet minimum health, **safety**, and **nutrition** standards. In addition, they are usually required to have a criminal background check. Some states yearly inspect the homes of family childcare providers, and many require ongoing training. Parents often make this childcare choice because they prefer their children to stay in a more home-like environment. This arrangement may be less expensive and more flexible than center-based care. Parents may also believe that their children are better off in smaller groups with a single caregiver.

The American Academy of Pediatrics recommends that family childcare providers should have six children or fewer per one adult caregiver, including the caregiver's own children. The total number should be fewer if infants and toddlers are involved. No caregiver who works alone should be caring for more than two children younger than two years of age.

Some of the advantages of family child care are:

- There are usually fewer children than in center-based care.
- There may be children of different ages.
- The child gets to stay in a home-like environment.

Some of the disadvantages of family child care are:

- Many family childcare providers are not licensed or regulated.
- Resources and equipment may vary widely.
- Family childcare providers normally work alone, which may make it more difficult to judge their work.

In-home caregivers

In-home care occurs in the child's own home. This care includes both live-in and live-out nannies and **babysitters**. Most in-home caregivers are not state-regulated, though many nanny-placement agencies are subject to state regulation. If in-home caregivers receive childcare subsidy payments, they may be required by many states to have a criminal background check done, and a very few states have minimal health and safety training requirements.

The advantages of in-home caregivers are:

- Children receive one-on-one care.
- Children may be safer and feel more secure in their own home.
- Parents may feel they have more control over the type of care their children receive.
- There is the possibility of more flexible scheduling.
- Care will usually be available even if the child is ill.

There are also disadvantages to in-home care. These include:

- It is often the most expensive type of care.
- The parent may bear the burden of obtaining background checks and providing ongoing supervision.

Care provided by relatives, friends, and neighbors

This type of care is often referred to as kith and kin care and may take place either in the child's or the caregiver's home. Some of the advantages of this type of care are:

- Parents may believe their children are receiving more loving, affectionate care and that the child is more secure.
- A relative, friend, or neighbor may be more likely to share the parents' values.
- The child receives one-on-one care.
- There may be a great deal of flexibility in this option.
- Care may be low- or no-cost.

Some of the disadvantages of having friends and relatives caring for children include:

- There is minimal regulation required by most states (though some parents may view this as an advantage).
- There may be a lack of care if the friend or relative is sick or on vacation.

The type of care chosen is related to the child's age. Twenty-three percent of newborn to two-year-olds and 22 percent of three- to six-year-olds are cared for in a home by a relative. Eighteen percent and 14 percent of these same respective ages were cared for by a nonrelative in a home environment. Higher percentages of three- to six-year-olds (56%) participated in center-based programs while only 17 percent of newborn to two-year-olds did. Some children may participate in more than one type of arrangement.

Chidren during play time at a day-care center. *(AP/Wide World Photos.)*

Center care is more common for black and white children with **working mothers** (30% and 24%, respectively) than for Hispanic children (10%). However, relative care is more common for Hispanic children (39%) versus black and white children (27% for blacks; 25% for whites). Use of parent care does not differ depending on racial and ethnic background. Black children with working mothers are more likely to be in care full-time than are white and Hispanic children (58% for black children, 36% for white children, and 34% for Hispanic children).

Choosing and finding high-quality child care is important and may play a key role in a child's health and development. Parents need to consider a variety of factors when deciding who should care for their child. There are several positive factors parents should look for when evaluating child-care options. These include:

- There is adequate supervision and attention to each child. Parents can find out their state's specific daycare regulations by contact their state's department of health and human services.
- Caregivers are well-trained and professional.
- Close attention is paid to health and safety issues. There are proper hand-washing routines, and the facility (or home) is clean. Caregivers have training in first aid and **cardiopulmonary resuscitation**. Safety precautions and accident prevention measures are in place.
- Children are encouraged to explore and are exposed to games, songs, and conversation in order to foster language development.
- There is appropriate and sufficient equipment and play materials.
- Parents are welcomed to make unscheduled visits and are encouraged to voice concerns and suggestions.
- Stability of caregivers is recognized as an important component of quality care. There is a low rate of staff turnover.

The following signs may indicate that there are problems with the child care provider or facility:

- The caregiver or center staff do not answer questions or address parent concerns.
- There is no written copy of center day-care policies.
- There is a high turnover of staff.
- The child indicates he or she is not happy with the day-care experience.
- There are recurring unexplained accidents.
- Parents are discouraged from participating in activities or voicing opinions about policies or practices.
- Other parents report concerns or problems.

Common problems

Parents who are deciding to place their children in some sort of day care should be aware that some problems may occur. A comprehensive study of early child care was started in 1991 by the National Institute of Child Health and Human Development (NICHD), part of the National Institutes of Health. More than 1,300 children were followed from birth to find out how the amount, type, and quality of day care they encounter affected their development. The study found that, at 15 months, child care neither promoted nor negatively affected infants' attachment to their mothers. However, a low-quality child-care environment combined with less sensitive mothering did leave infants less securely attached. The findings from this study also suggest that toddlers who spend long hours in day care display a slightly weaker bond with their mothers.

Child care can influence the behavior of the child, as well. More time spent in day care during the first two years led to more caregiver-reported behavioral problems at age two, although the effect was negated by age three. Higher quality care led to better child compliance and self-control, and children in larger groups (over three) appeared to be more cooperative than those in smaller groups. The biggest indicator of a child's behavior, however, was the family environment, particularly the sensitivity of mothering practices. This also carried over into the area of cognitive development, in which researchers found no benefit for children being raised exclusively by the mother. Those in high-quality care were at an advantage compared to those with exclusive maternal care, while low-quality child care presented a disadvantage. In general, fewer problem behaviors, higher cognitive performance, and better mother-child attachments were noted when children received higher quality care.

An additional concern for parents is that children who attend day care, especially in center-based environments, are more frequently exposed to communicable diseases and more frequently experience respiratory illnesses, ear infections, and **diarrhea** than children who are cared for primarily at home. The size of the group the child is in seems to play a role. Larger groups have higher incidences than smaller groups. In contrast, however, children who spend more time in day care miss fewer days of school than their peers who were cared for at home. Finally, parents need to consider who will care for their child if the child is ill, since most daycare providers will not accept ill children for **fear** of infecting other children.

KEY TERMS

Center-based care—Also called childcare centers or daycare centers, these facilities care for children in groups.

Parental concerns

Most parents are concerned about how their child will cope and adapt to being cared for by someone else. Parents can help their child adjust to a new childcare arrangement in several ways. They can arrange a visit to the center or home where they will receive care. Introducing them to the caregiver(s) may make the first days away from their parents easier. Some children like to bring a reminder of home with them when they attend day care. Parents may also choose one of the several books for children about day care and read it to their child.

See also Attachment between infant and caregiver.

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Deanna M. Swartout-Corbeil, RN

Death and mourning

Definition

Mourning is the grieving process an individual experiences in response to the loss (often through death) of someone.

Description

Almost every child or adolescent faces the death of someone close (a relative, friend, or even a pet) at some point in his or her life. In fact, it is estimated that about 6 percent of children under age 15, or about three of every 50 children, will lose a parent. Though this figure is low compared to 25 percent at the turn of the twentieth century, children in the twenty-first century are more likely to experience more types of violent and catastrophic death, such as in wars, natural disasters, and homicides. Children experience deaths of their peers through **suicide**, acts of violence in school or on the street, and in terminal illness.

In 1900, children experienced death firsthand, seeing a loved one die on the farm or in the home. Then, two world wars came and children experienced death in the remote events of far off places. By the 1950s, though some children did experience the death of a loved one in the Korean War, these were few. Death became an abstraction, something children only read about or experienced in a movie or television. The Vietnam War and racial unrest during the civil rights era connected death as reported by newspapers and television to events occurring U.S. city streets. By the last two decades of the twentieth century, death came knocking on the schoolhouse door as children brought weapons and shot other children. Drive-by shootings became more frequent in some neighborhoods. With the September 11, 2001, terrorist attacks on the United States, children began to see widespread death in a very personal way.

Children experience grief or mourning in very different ways than adults do. That is why over 160 bereavement centers have opened across the United States to help children mourn in ways that are appropriate to their age and developmental stage. These centers also allow children to participate in their own mourning rituals, which can be a comfort to children and a healthy way to move on. Unlike in some religious organizations, children are allowed to express anger as well as sadness about the death of their loved ones. They are encouraged to remember and create tangibles, such as paintings, stories, and even quilts, to remind them of the deceased.

Parents, caregivers, and teachers can provide support and minimize **fear** by answering honestly a child's questions about death. Encouraging communication helps the child through the essential grieving period. At one time, well-meaning adults felt that it was in the child's best interests to avoid discussing death. However, research has shown that children cope more successfully with a death if they feel included in the group that has experienced the loss and share in grieving and mourning.

When listening to a child's observations about death, adults must keep an open mind. A child may respond to the death of a grandmother, who used to make cupcakes for her, by observing that there will be no more cupcakes for dessert. This response could be interpreted as selfish, but it is in fact an expression of the child's loss in her own, very personal, terms. When a child learns of the accidental death of a playmate, he may ask to go out to **play**. This too may be an expression of the loss, as the child might want to remember his friend by engaging in the activity the two of them shared. The child's response to loss can be misunderstood by adults, especially by those who are also grieving. By passing judgment on the child's reactions, adults undermine the child's feelings and make the loss even more difficult for the child to handle.

For most people, deeply felt grief and loss are felt for about a week, followed by sorrow which can last two years or longer. In the days, weeks, and months that follow a

death, adults should refrain from criticizing or reacting negatively to the child's feelings. When the child seems to repeat the same questions over and over, the same answers, as open and honest as possible, must be repeated patiently. Young children may express concern, either directly or through behavior, about being abandoned or neglected, or they may fear they have in some way caused the death. Changes in appetite, complaints of feeling sick, and changes in activity patterns can be indications that the child is worried or anxious. Adults can help a child deal with these feelings by acknowledging them and by reassuring the child that he will still be cared for and that no one can cause a death by thoughts and feelings.

When a death is unanticipated, as in a case of accident or violence, children may grieve longer and more intensely. Sad feelings may resurface over the years when the child experiences the loss anew, such as on holidays or other occasions. When a parent is deeply affected by the death of a loved person, the child may need the steady support of another adult. Books about illness and death can also be helpful. Adults should review the books in advance or ask a librarian, teacher, or counselor for advice. Issues of concern include age-appropriateness, situation-appropriateness, and religious point of view.

Children mourn a bit at a time, returning to their grief anew at different stages. Understanding a child's developmental stages helps parents, teachers, and caregivers provide appropriate responses and support for the child.

Toddlerhood and preschool age

By the time a child is about two and a half or three, the child is able to acknowledge that a death has occurred but will not really understand the reality of death. The child may echo the parents' words but may also express their lack of understanding. This is less denial than it is the inability to understand the concept of death. If a child has experienced the death of a **family** pet and understands that the dog or cat will not come back, the child is more likely to understand that the person who dies will not come back either.

Children between two and five years of age will often be restless, have **sleep** disturbances, or frightening dreams. They will sometimes revert to behaviors they had at a younger age, such as wetting the bed or **thumb sucking**. Because children of this developmental stage do not fully understand the concept of death, they may blame themselves, thinking that if only they had been good enough, their loved one would not have died.

Parents and other adults need to offer comfort in simple but honest words and in physical contact. They need to make sure the child's routine remains intact to preserve a sense of security in a loving environment. Caregivers can draw pictures of the loved one or read books about mourning so the child can feel that they are not alone. Even children of this age can participate in funerals or other death rituals.

School age

By the age of six, children begin to understand that death is the cessation of bodily functions and that it is permanent. They show a morbid curiosity about death. They may talk about the details of death repeatedly. They want to know what happens physically and spiritually. Children of this age may also begin to acknowledge the universality of death, that it happens to everyone. But they may find it difficult to believe that it could ever happen to them.

Around this age, children are capable of taking part in rituals of death, such as visits with the deceased's family, the wake, and the funeral or memorial service. Prior to participating in a visit or funeral, it is helpful to prepare the child for the experience by explaining the purpose of the visit, how long it will last, and what the child will experience there. If a child expresses reluctance to participate in any aspect of the rituals of death, adults should accept the child's feelings and not exert pressure. Children look to adults and their peers for role models of how to conduct themselves at these events.

School-aged children can understand what death means, but they may be so overwhelmed that they act as if nothing has happened. Unexpressed feelings may surface as physical symptoms, such as stomachache, **headache**, and unusual complaint of tiredness. Behavior may also change, demonstrated by reluctance to go to school, daydreaming in class, or a decline in academic performance. Children of this age usually need extra time for physical activity. Even a short walk or a turn at the playground can help them work off some of the physical tensions connected with mourning.

Children both grieve alone and share their grief with others. Families can take a number of actions to support emotional healing, such as openly acknowledging the death, letting children participate in the rituals, and maintaining familiar routines such as school and bedtime activities. Parents should also let children witness their own adult grief. Rather than avoiding any mention of the deceased, it may help to display a photograph in a prominent place as a way of letting family members maintain memories. The visual reminder provides a way to help the child understand that it is okay to talk about the person who died.

For some children, the threat of being alone is central. One of their first questions is usually, "Who is going to take care of me?" Caregivers need to reassure the child that he or she is still loved and will be cared for.

Some older children begin to understand that death can happen to anyone, especially if a child their own age dies. The child may also worry that he or she may die at any time. This is especially a problem when the child's peer dies suddenly in an accident or after a long illness. Questions like "Will I die, too?" need reassuring answers.

Adolescence

Teenagers understand more as an adult does, but they may find it even more difficult than younger children to deal with their sorrow. Behavior problems, dropping out of school, physical complaints such as headache or chest **pain**, sexual promiscuity, and even suicide attempts may result from their feelings of pain and loss. Often, teenagers are reluctant to talk to adults who could help them through their grief.

The death of a peer, even someone they hardly knew, affects adolescents differently than the death of an older person. They must cope not only with the shock of life's unpredictability, but their own mortality.

Some adolescents may feel anger and want to rage at the world for letting death take a loved one. Research has shown that in the majority of cases adolescents in juvenile facilities have lost someone close to them.

Adolescents can also feel very vulnerable. Some express the need to feel like a child again, to be taken care of and protected, to feel secure in a safe world.

Common problems

Nighttime is particularly problematic since the child is now able to remember his or her loss without other distractions intervening. The child may also feel abandoned and alone. Many children will want to sleep with a parent or a sibling. Others request that a light be left on, even if they have long since stopped needing a nightlight.

Many children will also revert to a previous developmental stage. Some children will wet the bed. Others will request permission to sleep with a stuffed animal or return to sucking their thumbs. Some of these behaviors are comfort actions. They seek the security of the earlier developmental stage.

Older children and adolescents may express their bottled up anger through aggression toward other children. They may also turn that unresolved anger inward, becoming depressed or adopting a dark, gloomy lifestyle. Some adolescents may participate in risk-taking behaviors or want to do everything because they are worried that they may not have time to do everything before some random act kills them. Other children and adolescents may mutilate themselves or get body piercings or tattoos. (It should be noted that **piercing and tattoos** are not necessarily a cry for help; they may simply be acts of self-expression.)

Students at Columbine High School in Littleton, Colorado comfort each other after the death of classmates. *(Photograph by David Zalubowski. AP/Wide World Photos.)*

Parental concerns

Parents need to help their children work through their grief, not get around it or avoid it. Parents should never minimize their child's grief. It is as individual as any adult's. Parents and other caregivers need to be available to listen as children express their grief in their own ways. This means that parents should seek counseling themselves if their own grief gets in the way of helping their children move through the grieving process.

Grieving children need their friends' support as much as adults do. Parents should make sure that children are allowed to phone their friends or play with them during the grieving process. Children will often express their sadness to friends much more easily than to their parents or other adults.

In addition, funerals and other death rituals are important to children as well as adults. These gatherings allow

KEY TERMS

Bereavement—The emotional experience of loss after the death of a friend or relative.

Grief reaction—The normal depression felt after a traumatic major life occurrence such as the loss of a loved one.

Mourn—To express grief or sorrow, usually for a death.

the bereaved to express their sorrow, remember those they have lost, and receive and offer support from a community of friends and relatives. Death rituals often are the moments when the death of a loved one becomes real and not just tragic news. The bereaved often need to see the body of the deceased or participate in some activity that expresses the letting go of their dead loved one's presence in this life. Some families release balloons after a funeral; others light candles and let them burn down; both activities express the release of the deceased from life to death. Many of these activities coincide with religious beliefs and customs. Children who participate in a funeral by reading a poem or offering a gift are often better able to cope with their loss.

When to call the doctor

About one third of grieving children may need to seek professional counseling or therapy of some kind. This can be in individual sessions or in a group. It can also come from religious sources. Behaviors that warrant grief counseling or therapy are unresolved anger and hostility, not expressing grief at all or minimally, or depression or **anxiety** that interferes with daily activities that lasts for weeks or months.

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Decongestants

Definition

Decongestants are medicines used to relieve nasal congestion (stuffy nose).

Description

Decongestant drugs are chemically similar to epinephrine and norepinethrine, which are hormones that cause excitation in the body. These hormones cause constriction of blood vessels. A stuffy nose is caused by dilated blood vessels, which swell the nasal passages and make it harder to breathe.

Because these drugs have actions similar to the natural hormones, they have been used for purposes other than the treatment of nasal congestion. Some of these uses, such as treatment of hypotension, can be very important. Some of these drugs have also been abused because of their stimulant effects.

General use

A congested or stuffy nose is a common symptom of colds and **allergies**. This congestion results when membranes lining the nose become swollen. Decongestants relieve the swelling by narrowing the blood vessels that supply the nose. This narrowing reduces the blood supply to the swollen membranes, causing them to shrink.

These medicines do not cure colds or reverse the effects of histamines, chemicals released as part of the allergic reaction. They will not relieve all of the symptoms associated with colds and allergies, only the stuffiness.

Nasal decongestants may be used in many forms, including tablets, nose drops, and nasal sprays.

Precautions

Because decongestants have the potential for many side effects and adverse effects, they must be dosed carefully.

Side effects

When decongestants are used in the form of nose drops or nasal spray, the following adverse effects are common:

- stinging
- burning
- sneezing
- increased nasal discharge
- altered sense of taste

The following adverse effects are very rare when decongestants are given by drops or spray and also quite rare but possible when given by mouth:

- restlessness
- anxiety
- nervousness
- weakness
- difficulty breathing

Even more severe adverse effects are possible when decongestants are taken in large overdose. These include heart problems and tremors.

Some people complain of rebound congestion, which occurs when, after the decongestant has worn off, the congestion returns even worse than before.

Interactions

Decongestants do not have any interactions with drugs that would be taken by a generally healthy child. Even so, people using decongestants should review their drug therapy with a physician or pharmacist before starting treatment.

Although decongestants have the potential for serious side effects and adverse effects, they are very safe when used properly. However, nasal decongestants should only be used for three days at a time to avoid significant rebound effect. The most severe adverse effects can be avoided by using nose drops and nasal sprays in place of tablets or capsules.

Parental concerns

Parents administering these drugs to their children should use nose drops or nasal spray and avoid tablets or capsules, which are more likely to cause adverse effects. They should also review the proper administration of nose drops and nasal spray with a physician or nurse.

Decongestants are subject to abuse. Parents should observe the behavior of adolescents and teens who may be purposely overdosing on these drugs.

In the event of severe adverse effects, parents should get medical care immediately for their child.

See also Cough suppressants; Expectorants.

KEY TERMS

Constricted—Made smaller or narrower.
Dilate—To expand in diameter and size.
Hypotension—Low blood pressure.
Tremor—Involuntary shakiness or trembling.

Resources

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Dehydration

Definition

Dehydration is the loss of water and salts that are essential for normal body function.

Description

Dehydration occurs when the body loses more fluid than it takes in. Dehydration can upset the delicate fluid-salt balance needed to maintain healthy cells and tissues. The human body is generally over 60 percent water. The body works to maintain water balance through mechanisms such as the thirst sensation. When the body requires more water, the brain stimulates nerve centers to encourage a person to drink in order to replenish the water stores. Water intake can vary widely on a daily basis, influenced by such factors as access to water, thirst, habit, and cultural factors.

The kidneys are responsible for maintaining water balance through the elimination of waste products and excess water. Water is primarily absorbed through the gastrointestinal tract and excreted by the kidneys as urine. The variation in water volume ingested is dependent on the ability of kidneys to dilute and concentrate the urine as needed.

Children need more water than adults because they expend more energy, and most children who drink when they are thirsty get as much water as their systems require. Dehydration in children usually results from losing large amounts of fluid and not drinking enough water to replace the loss. This condition generally occurs in children who have a stomach flu characterized by **vomiting** and **diarrhea** or who cannot or will not take enough fluids to compensate for excessive losses associated with **fever** and sweating of acute illness. Dehydration can result from illness; a hot, dry climate; prolonged exposure to sun or high temperatures; not drinking enough water; and overuse of diuretics or other medications that increase urination.

Types of dehydration

Dehydration is classified as mild, moderate, or severe based on how much of the body fluid is lost, estimated by loss in bodyweight. Mild dehydration is the loss of no more than 5 to 6 percent loss of body weight. Loss of 7 to 10 percent is considered moderate dehydration. Severe dehydration (loss of over 10 percent of body weight) is a life-threatening condition that requires immediate medical care.

Complications of dehydration

When the body's fluid supply is severely depleted, hypovolemic shock is likely to occur. This condition, which is also called physical collapse, is characterized by pale, cool, clammy skin; rapid heartbeat; and shallow breathing.

Blood pressure sometimes drops so low it cannot be measured, and skin at the knees and elbows may become blotchy. **Anxiety**, restlessness, and thirst increase. After a child's temperature reaches 107°F (41.7°C) damage to the brain and other vital organs occurs quickly.

Demographics

Dehydration is a major cause of infant illness and death throughout the world. Dehydration is often a result of gastrointestinal disease and diarrhea in children. Among children in the United States, short-term diarrhea results in approximately 200,000 hospitalizations and 300 deaths per year. In developing countries, dehydration from illness is a common cause of death in children under five years of age, accounting for about 2 million deaths per year.

Causes and symptoms

Dehydration is a deficit of body water that results when the output of water exceeds intake. Dehydration stimulates a child's thirst mechanism. Causes of dehydration may include the following:

- decreased water or fluid intake
- diarrhea
- vomiting
- excessive heat
- excessive sweating
- fever
- excessive urination (polyuria)
- diuretics or other medication that increase fluid loss
- caffeine or alcohol consumption

Sweating and the output of urine both decrease during dehydration. If water intake continues to fall short of

water loss, dehydration worsens and a child may become critically ill.

Reduced fluid intake may be a result of the following:

- appetite loss associated with acute illness
- nausea
- bacterial or viral infection or inflammation of the pharynx (pharyngitis)
- inflammation of the mouth caused by illness, infection, irritation, **canker sores**, or vitamin deficiency

Other conditions that can lead to dehydration include the following:

- disease of the adrenal glands, which regulate the body's water and salt balance and the function of many organ systems
- diabetes mellitus
- eating disorders
- kidney disease
- chronic lung disease

With mild dehydration, increased thirst and restlessness are usually the only apparent symptoms. In moderately dehydrated children, eyes are somewhat sunken, and the mouth and tongue are dry. Thirst is increased: an older child asks for water, and a younger child drinks eagerly when offered a cup or spoon of water. The skin is less elastic than it should be and is slow to return to its normal position after being pinched. The radial pulse (wrist area) is detectable, but rapid. The soft spot on a baby's head (fontanelle) is somewhat sunken. Two of the following symptoms usually indicate some degree of dehydration: drinks eagerly, thirsty, restless, irritable, sunken eyes, or skin pinch goes back slowly.

Children with severe dehydration are usually lethargic, in a stupor, or even in a coma. Symptoms are even more apparent (deeply sunken eyes without tears, very dry mouth and tongue, rapid and deep breathing). A skin pinch retracts very slowly (over two seconds). Children who are awake are very thirsty, although a child may drink poorly if in a stupor. A child may not have urinated for six hours or longer. When in hypovolemic shock, systolic blood pressure taken in the arm is low or not detectable, the arms and legs are cool, and the nail beds may have a bluish or purplish discoloration. Two of the following symptoms indicate severe dehydration: lethargic or unconscious, very slow skin pinch, sunken eyes, and not able to drink or drinking poorly.

Dehydration can cause confusion, **constipation**, discomfort, drowsiness, and fever. The skin turns pale and cold, the mucous membranes lining the mouth and nose lose their natural moisture. The pulse sometimes races and breathing becomes rapid. Significant fluid loss can cause serious neurological problems or death.

When to call the doctor

A doctor should be notified whenever an infant or child exhibits signs of dehydration or a parent is concerned that a stomach virus or other acute illness may lead to dehydration.

A doctor should also be notified if any of the following is the case:

- Symptoms of dehydration worsen.
- A breast-fed or bottle-fed infant is unable to feed or feeds poorly.
- An infant or child urinates very sparingly or does not urinate at all during a eight-hour period.
- An infant younger than two months of age has diarrhea or is vomiting.
- Dizziness, listlessness, or excessive thirst occurs.
- The child's heart is beating fast.
- The child has dry eyes, sunken eyes, a dry mouth, or is not producing tears.
- There is blood in the stool or vomit.

An infant can become dehydrated within hours after the onset of illness. In general, the smaller the child, the lower the threshold should be for healthcare intervention if dehydration is suspected.

Diagnosis

A child's symptoms and medical history alone usually suggest dehydration. Physical symptoms are usually all that is necessary for diagnosing dehydration, although laboratory tests may be ordered by the physician. Physical examination may reveal shock, rapid heart rate, and/or low blood pressure. Laboratory tests, including blood tests (to check electrolyte levels) and urine tests (e.g. urine specific gravity and creatinine), may be used to evaluate the severity of the problem.

Treatment

Increased fluid intake and replacement of lost electrolytes are extremely important for restoring fluid balances in infants and children who are dehydrated. Treatment is given based on severity of dehydration. Treatment should include two phases: a rehydration phase and a maintenance phase. In the rehydration phase, fluid losses are replaced quickly, within three to four hours until normal hydration is achieved. In the maintenance phase, calories and fluids

are given. Rapid refeeding should follow rapid rehydration with the goal of returning the child to an unrestricted, age-appropriate diet including solids. Withholding foods to rest the gut is not recommended. Breastfeeding should be continued at all times through both stages of treatment. Full-strength formula is usually tolerated. Changing formula or diluting to half strength are common practices but are usually unnecessary and may even prolong symptoms and delay nutritional recovery.

To replace calories quickly during acute illness, food should be given as soon as the child will tolerate it. During both rehydration and maintenance phases, fluid losses from vomiting and diarrhea should be replaced continuously. Restricting lactose (milk and milk products) is usually not necessary but may be helpful in a child with a severe intestinal disease or diarrhea in a malnourished child.

Children with minimal dehydration weighing less than 10 kilograms (22 pounds) should be given 60 to 120 mL (2–4 ounces) of an oral rehydration solution (ORS) for each episode of vomiting or diarrheal stool. Those weighing more than 10 kg (22 lbs) should be given 120 to 240 mL (4–8 ounces). Food should not be restricted. Children with mild to moderate dehydration should be given 50 to 100 mL (roughly 2–3.5 ounces) of an ORS per kilogram body weight during two to four hours to replace fluid losses. Additional ORS should be administered to replace ongoing losses from vomiting and diarrhea. In a sick child, a teaspoon, syringe, or medicine dropper can be used to offer a small amount at first with amounts increasing as tolerated. If the child appears to want more, more can be given. Severe dehydration is a medical emergency requiring intravenous fluids immediately.

For moderate or severe dehydration, a child should be treated in a medical facility. Moderate dehydration can be treated orally, but severe dehydration requires the child to take fluids intravenously (IV). When treating dehydration, the underlying cause must also be addressed. For example, if dehydration is caused by vomiting, medications may be prescribed to resolve these symptoms. However, anti-diarrheal medications are not recommended in children. A child who is dehydrated due to diabetes, kidney disease, or adrenal gland disorders must receive treatment for these conditions as well as for the resulting dehydration.

For older children who are mildly dehydrated, just drinking plain water may be all the treatment that is needed. For infants and younger children, especially when ill, drinking a commercial ORS should be encouraged. Parents should follow label instructions when giving children Pedialyte or other commercial products recommended for relieving dehydration. Sports drinks are not recommended as they contain a lot of sugar and may worsen diarrhea.

In order to accurately calculate fluid loss, it is important to chart weight changes every day and keep a record of how many times a child vomits or has diarrhea. Parents should note how many times a baby's diaper must be changed.

Alternative treatment

Gelatin water may be substituted for electrolyte-replacement solutions if an ORS is unavailable. It is made by diluting a 3-oz package in a quart of water or by adding one-fourth teaspoon of salt and a tablespoon of sugar to a pint of water. Receiving the right amount of electrolytes is very important, and thus homemade remedies such as gelatin (or adding salt or sugar to water) are not recommended because of the potential for quantity errors when mixing. However, these may be useful if ORS cannot be obtained in an emergency. Parents should keep a can of ORS on hand for emergencies.

Formulas containing soy fiber have been reported to reduce liquid stools.

Prognosis

Mild dehydration rarely results in complications. If the cause is eliminated and lost fluid is replaced, mild dehydration can usually be cured quickly.

Vomiting and diarrhea that continue for several days without adequate fluid replacement can be fatal. However, dehydration that is rapidly recognized and treated has a good outcome.

Prevention

Ensuring that children always drink adequate fluids during an illness helps to prevent dehydration. Parents can prevent dehydration in infants and children who are vomiting or who have diarrhea by increasing fluids to compensate for losses. Infants and children with diarrhea and vomiting should be given ORS such as Pedialyte immediately to help prevent dehydration.

Children who are not ill can maintain proper fluid balance by drinking water or fluids even before they are thirsty. Children should drink fluids before going outside to **exercise** or **play** (especially on a hot day). Dehydration can usually be prevented by drinking enough fluid for urine to remain the color of pale straw. Water in foods, especially fruits and vegetables, is a great source of fluid. Fruits and vegetables can contain up to 95 percent water, so a well-balanced diet is a good way to stay hydrated.

Parents should know whether any medication their child is taking can cause dehydration and should get

KEY TERMS

Diuretic—A group of drugs that helps remove excess water from the body by increasing the amount lost by urination.

Electrolytes—Salts and minerals that produce electrically charged particles (ions) in body fluids. Common human electrolytes are sodium chloride, potassium, calcium, and sodium bicarbonate. Electrolytes control the fluid balance of the body and are important in muscle contraction, energy generation, and almost all major biochemical reactions in the body.

prompt medical care to correct any underlying condition that increases the risk of dehydration.

Other methods of preventing dehydration and ensuring adequate fluid intake are as follows:

- eating more soup at mealtime
- drinking plenty of water and juice at mealtime and between meals
- keeping a glass of water nearby

Children should not be given coffee or tea, because they increase body temperature and water loss. Avoiding caffeinated soft drinks may also reduce the risk of dehydration. These beverages are all diuretics (substances that increase fluid loss).

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Dental development

Definition

Dental development is the process by which children develop their first and second (permanent) teeth.

Description

A child's first set of 20 teeth are called baby, primary, deciduous, or milk teeth. As these teeth fall out, they are replaced by 32 permanent, adult, or secondary teeth. The entire process of dental development may take more than two decades. Both primary and permanent teeth usually erupt (break through the gum) in a specific order on each side of the upper and lower jaws. However, the timing of both primary and permanent tooth eruption can vary by two or more years.

Both the timing of dental development and tooth size are determined primarily by heredity. Individuals differ greatly in the size of the crown (the part of the tooth above the gum line). Except for the earliest stages of **prenatal development**, and possibly the third permanent molars or wisdom teeth, dental development in girls proceeds ahead of that in boys, often by as much as 6 percent. Girls also have slightly smaller crowns and slightly shorter tooth roots than boys.

Prenatal

Dental development begins at about three weeks of gestation. By six weeks of gestation the tips or cusps of

the primary teeth appear. By the fourth month the hard tissues (the enamel and dentin) of the primary teeth have begun to form. The enamel crowns of most primary teeth are fully formed by eight months of gestation. Permanent teeth begin to form shortly before or at birth.

Baby teeth

At birth the developing teeth usually are still embedded in the gums. Occasionally a baby is born with some erupted teeth or teeth that erupt shortly after birth. These natal or neonatal teeth usually are poorly formed and mobile. However in most infants the front teeth begin to peek through the gums between four and eight months. Generally from about six months on, children get four new teeth every four months. By 12 to 15 months all of the baby teeth within the gums have formed crowns. Most children have all 20 baby teeth by the age of two-and-a-half to three years. The permanent teeth continue to develop within the jaw.

Baby teeth erupt in pairs on the right and left of the mouth, alternating between the lower and upper jaws, and proceeding from front to back. The 20 primary teeth usually erupt in the following order:

- four front teeth or central incisors, first in the lower jaw and then in the upper jaw
- four lateral incisors, on each side of the front teeth, uppers before lowers
- four first molars, uppers first
- four canines or cuspids, between the lateral incisors and the first molars, usually uppers before lowers
- four second molars behind the first molars, lowers first

Baby teeth may come in straight or at an angle, appearing crooked, although they eventually straighten out. Once all of the baby teeth have erupted, the tongue adapts to their shape and the child's pre-teeth swallowing pattern switches to an adult pattern.

Children start losing their baby teeth at about age six, after the permanent front teeth are almost formed beneath the gums. The pressure of the developing permanent teeth causes the roots of the baby teeth to dissolve. Without their anchor in the jaw, the baby teeth loosen and eventually fall out. Most children lose their lower front baby teeth first. The earlier that the baby teeth come in, the earlier they will fall out. Most children have lost all of their baby teeth by age 13.

Permanent teeth

Between the ages of two-and-a-half and six, the permanent teeth continue to develop within the jaw. The first permanent teeth, the six-year-molars that become the first permanent molars, erupt behind each of the four second baby molars, usually between the ages of five and six. If the baby teeth are properly positioned and aligned, the six-year-molars usually erupt properly. If the baby teeth are pushed too close together, the six-year-molars will be too far forward, crowding the permanent teeth that erupt in front of them. However if the six-year-molars erupt properly and if the jaw is large enough, the permanent teeth have a good chance of coming in correctly.

By about age eight, enamel has formed on all of the permanent teeth except the wisdom teeth. A permanent tooth comes in completely about two months after the corresponding baby tooth is lost. Between the ages of about six and 12 to 14, as the jaw grows, 28 permanent teeth erupt, replacing the primary teeth, incisor for incisor, canine for canine, premolar or bicuspid for molar. The 32 permanent teeth generally erupt in the following order:

- four six-year or first molars
- four central incisors or front teeth, first in the lower jaw and then in the upper jaw
- four lateral incisors, lowers usually first
- four canine teeth, lowers first
- four first premolars or bicuspids, between the canines and the six-year molars, uppers usually first
- four second premolars or bicuspids, between the first premolars and the six-year or first molars, uppers usually first
- four second molars, behind the first molars, lowers usually first
- four third molars or wisdom teeth at the back, usually between ages 17 and 21

Common problems

Teething

Teething (the eruption of the primary teeth through the gums) may cause discomfort or **pain**, particularly with the large molars. Teething babies may:

- be restless and irritable
- lose their appetites
- sleep poorly
- cry excessively
- have flushed cheeks
- have a slight **fever**
- have congestion
- dribble or drool
- have red, swollen gums at the new teeth sites

- rub their gums
- suck their thumbs
- want something to chew on

For teething symptoms, parents may massage the gums to relieve discomfort and offer teething **toys** to help speed tooth eruption. A frozen teething toy numbs the gums and reduces swelling, although it should not be left on the gum for more than one minute without a break. They may also relieve symptoms by the following:

- massaging the gums, with or without ice
- giving the baby a cold teething ring
- encouraging the baby to chew on cold, wet washcloth or frozen bagel
- administering **acetaminophen** (Tylenol)

Eruptions of the permanent teeth are usually much less distressing, although the eruption of the first four broad permanent molars may cause discomfort. As the permanent molars push through the gums, they often leave a flap of tissue over the tooth. If food becomes trapped under the flap, the gums may become sore, swollen, and painful, infected, or abscessed.

Developmental delay

While dental development may be slightly advanced in obese children, development delay can occur with the emergence of some permanent teeth. Delay can be caused by the following:

- hereditary factors
- chronic **malnutrition**
- developmental disorders
- hyperdontia (extra or supernumerary teeth)
- **Down syndrome**
- radiation or chemotherapy
- cysts or tumors
- the absence of lateral incisors or wisdom teeth

Decay

The enamel on baby teeth is thinner and softer than on permanent teeth and decay can move through it very rapidly. Some children develop decay as soon as a tooth erupts and about 50 percent of two-year-olds have at least one cavity.

Baby bottle **tooth decay** occurs when sugary liquids, including milk, juice, or formula, cling to the baby's teeth, particularly when the child is put to bed with a bottle. The decay occurs most often in the upper front teeth but other teeth also can be affected. If a decayed baby tooth is lost too early, the adjacent teeth may move into the space, causing crooked and overcrowded permanent teeth.

Malocclusion

In a perfect jaw, all of the teeth fit exactly without crowding or spacing. The teeth are not rotated, twisted, or leaning forward or backward. With a perfect bite (occlusion), the teeth of the upper jaw slightly overlap those of the lower jaw. The points of the molars fit into the grooves of the opposing molars.

Few children develop perfect teeth and occlusion. More than 90 percent of children have some degree of **malocclusion** or poor bite. Skeletal malocclusions occur when the upper and lower jaws are not properly aligned with each other and with the skull. Dental malocclusions occur when the teeth are crowded or the upper and lower teeth are not properly aligned with each other. Malocclusions can be caused by the following:

- heredity
- crowded or misaligned baby teeth
- premature loss of baby teeth due to decay or injury, so that the permanent first molars move forward, causing crowding and misalignment of the new front teeth
- loss of permanent tooth structure due to untreated decay in the baby teeth
- a tooth emerging at an angle such that it pushes on and damages an adjacent tooth
- sucking that continues after the permanent teeth erupt
- wisdom teeth that erupt crookedly
- accidental injury

Some children experience mild, temporary symptoms of malocclusion resulting from a growth spurt. Severe malocclusion may require orthodontic intervention to improve appearance or to prevent problems with eating and speaking. Most alignment problems develop gradually, although they may be apparent at eruption. Symptoms of alignment problems include the following:

- lack of space between teeth
- teeth that are out-of-line or abnormally spaced
- front teeth that do not meet
- protruding of the upper or lower jaw
- protruding upper front teeth (overbite)
- protruding lower front teeth (underbite)
- an open bite wherein the upper and lower front teeth do not touch during biting

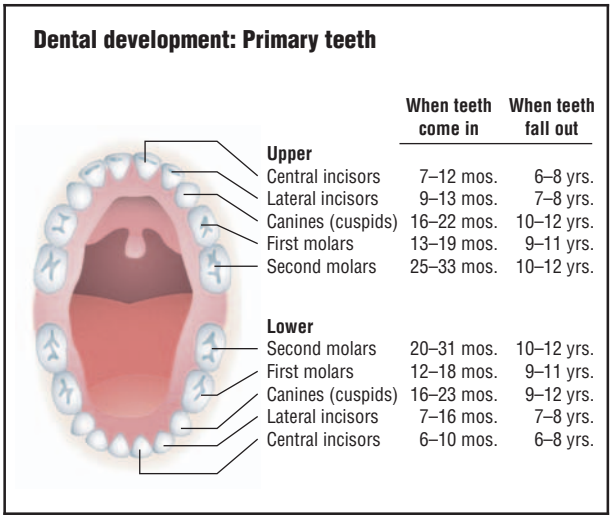

Illustration of the eruption of primary, or baby teeth. *(Illustration by GGS Information Services.)*

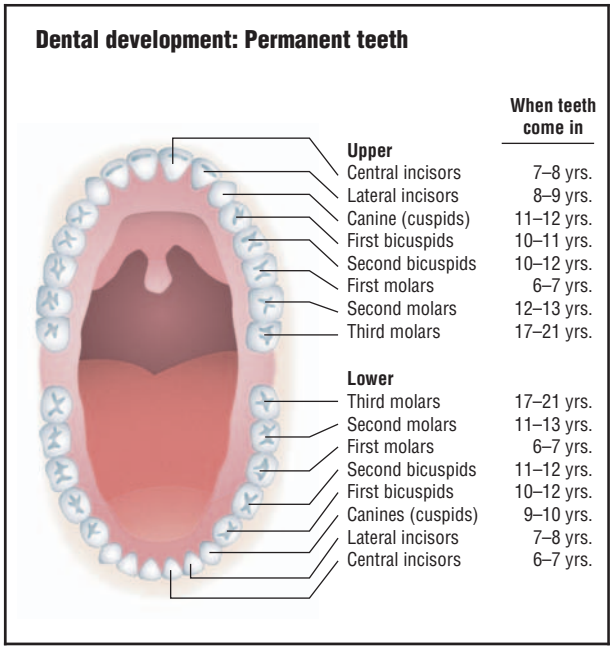

Illustration of the eruption of permanent teeth. *(Illustration by GGS Information Services.)*

Developmental disorders

Dental development disorders may occur as a result of the following:

- numerous inherited syndromes
- improper prenatal development
- endocrine disorders
- environmental factors

Developmental disorders include the following:

- hypodontia, in which one or more permanent teeth, usually the wisdom teeth or the lateral incisors, fail to form and the remaining teeth tend to be smaller
- anodontia, a very rare condition in which many or all of the permanent teeth fail to form
- supernumerary or extra teeth (These are more common in permanent teeth than in baby teeth; extra teeth usually are somewhat shapeless pegs, although occasionally an extra molar develops fully.)

Discolored and misshapen teeth

Disorders that may cause discoloration of the teeth include:

- hypoplasia, in which insufficient or irregular enameling of the teeth caused by the administration of tetracycline to a pregnant or nursing mother or to the infant or young child
- enamel and dentin hypoplasia, in which the enamel and dentin are not calcified (hardened by the depositing of **minerals**) due to **vitamin D deficiency** during tooth development
- hypocalcification, in which the enamel is of poor quality due to genetic factors, extensive plaque deposits, excessive sucking on citrus fruits, or high consumption of very acidic carbonated beverages
- amelogenesis imperfecta, an inherited defect that causes thin and discolored enamel
- dentinogenesis imperfecta, a defect of the dentin that causes discoloration and loss of enamel
- extrinsic enamel coloration due to liquid iron supplements administered for anemia or due to plaque or stains adhering to calculus, hard mineral deposits on the crowns and roots
- intrinsic enamel coloration due to pigments carried in the blood from tetracycline or other drugs or from excessive fluoride

Illness or trauma during infancy or early childhood, including infections, high fever, malnutrition, or disorders such as congenital syphilis or Down syndrome, can cause misshapen or discolored teeth. Both the baby teeth and the permanent teeth are usually affected, particularly the eight front teeth and the six-year or first molars. Crowns may be pitted, grooved, and discolored.

Wisdom teeth

Wisdom teeth frequently have difficulty erupting because the jaw is too small. A wisdom tooth may rotate,

KEY TERMS

Abscess—A localized collection of pus in the skin or other body tissue caused by infection.

Bicuspid—Premolar; the two-cupped tooth between the first molar and the cuspid.

Calcification—A process in which tissue becomes hardened due to calcium deposits.

Calculus—Plural, calculi. Any type of hard concretion (stone) in the body, but usually found in the gallbladder, pancreas, and kidneys. They are formed by the accumulation of excess mineral salts and other organic material such as blood or mucus. Calculi (pl.) can cause problems by lodging in and obstructing the proper flow of fluids, such as bile to the intestines or urine to the bladder. In dentistry, calculus refers to a hardened yellow or brown mineral deposit from unremoved plaque, also called tartar.

Canines—The two sharp teeth located next to the front incisor teeth in mammals that are used to grip and tear. Also called cuspids.

Crown—The natural part of the tooth covered by enamel. A restorative crown is a protective shell that fits over a tooth.

Dentin—The middle layer of a tooth, which makes up most of the tooth's mass.

Enamel—The hard, outermost surface of a tooth.

Eruption—The process of a tooth breaking through the gum tissue to grow into place in the mouth.

Impacted tooth—Any tooth that is prevented from reaching its normal position in the mouth by another tooth, bone, or soft tissue.

Incisor—One of the eight front teeth.

Malocclusion—The misalignment of opposing teeth in the upper and lower jaws.

Molars—The teeth behind the primary canines or the permanent premolars, with large crowns and broad chewing surfaces for grinding food.

Occlusion—The way upper and lower teeth fit together during biting and chewing. Also refers to the blockage of some area or channel of the body.

Plaque—A deposit, usually of fatty material, on the inside wall of a blood vessel. Also refers to a small, round demyelinated area that develops in the brain and spinal cord of an individual with multiple sclerosis.

Premolar—Bicuspid; the two-cupped teeth between the first molars and the cuspids.

Wisdom teeth—The third molars at that back of the mouth.

tilt, or be displaced as it attempts to emerge, and it can become impacted (partially buried) in the gums. Impacted wisdom teeth do not always cause problems. However wisdom teeth are always difficult to clean and susceptible to decay and gum disease; thus an impacted wisdom tooth is usually extracted. An impacted tooth can cause:

- bad breath
- an unpleasant taste in the mouth
- gum pain
- recurrent infection of the tooth and surrounding gums

Parental concerns

Some evidence indicates that infants' jaws and teeth develop better and more completely if babies are fed breast milk rather than infant formula. To promote their baby's dental development, pregnant and nursing mothers should do the following:

- practice good nutrition
- refrain from prescription and nonprescription drugs, particularly during the first trimester of pregnancy
- refrain from alcohol, tobacco, and excessive sugar

Parents must clean and care for their children's teeth until children are able to do it themselves. Furthermore, parents are the first to teach their children good dental hygiene. After feeding, a baby's gums should be wiped with clean gauze. Brushing should begin with the first tooth eruption and the remaining gums should be cleaned and massaged. Flossing should begin as soon as all of the baby teeth have erupted.

When to call the dentist

The rapidity with which decay can advance in baby teeth necessitates periodic dental examinations and cleanings. Pediatric dentists often recommend a first dental appointment at 12 to 18 months of age. Some recommend a first appointment at six months. At the very least, a child should see a dentist by age two-and-a-half or when all of the baby teeth have erupted.

Between two-and-a-half and six years of age is a critical period for dental development. Parents should

regularly examine a child's teeth for signs of decay, crowdedness, or misalignment. A dentist should be consulted if any of the following occurs:

- A child has an inherited disorder that affects the teeth or jaws, such as a protruding or recessed lower jaw.
- Tooth eruption occurs at least a year sooner or later than normal.
- The baby teeth do not erupt properly or seem crowded.
- A baby tooth becomes loose before the age of four or five years.
- A permanent tooth begins to erupt before the baby tooth that it is replacing has been lost.
- Swelling or infection occurs during eruption of the molars.
- A child has difficulty chewing or closing their jaw.
- A child continues to thumb-suck or suck on a pacifier after all of the baby teeth have erupted.
- A child's diet contains excessive sugar.
- A child has a serious fall or blow to the head.
- A permanent tooth is loosened in an accident.

Parents should pay close attention to the child-dentist relationship and voice any concerns, since it is very important to prevent a child from having an unpleasant dental experience. Many parents choose to take their child to a pediatric dentist. A pediatric dentist has undergone additional training and may be more experienced with dental development in children. Their offices are usually designed specifically to make children feel comfortable.

See also Fluoridation; Orthodontics.

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American Academy of Pediatric Dentistry. 211 East Chicago Avenue, Suite 700, Chicago, IL 60611–2663. Web site: <www.aapd.org>.

American Dental Association. 211 East Chicago Avenue, Chicago, IL 60611–2678. Web site: <www.ada.org>.

National Maternal and Child Oral Health Resource Center (OHRC). Georgetown University, Box 571272, Washington, DC 20057–1272. Web site: <www.mchoralhealth.org>.

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Dental hygiene *see* **Oral hygiene**

Dental trauma

Definition

Dental trauma is injury to the teeth, gums, and jawbones. The most common dental trauma is a broken or displaced tooth.

Description

Dental trauma may be inflicted in a number of ways: contact **sports**, motor vehicle accidents, fights, falls, eating hard foods, drinking hot liquids, and other such mishaps. Dental trauma includes teeth that are knocked out (dental avulsion), cracked (fractured), forced out of position (dental luxation, lateral displacement, or extrusion), pushed up into the jawbone (dental intrusion), or loosened by impact (subluxation or dental **concussion**). Oral tissues are sensitive, and injuries to the mouth are typically very painful. Dental trauma should receive prompt treatment from a dentist and in some cases is considered a dental emergency.

Demographics

Children between the ages of 1.5 and 3.5 years are most likely to experience dental trauma to their primary (baby) teeth, because this is the age at which they are learning to run. According to the International Association of Dental Traumatology, half of children experience dental injury, with injury occurring most often in children ages eight to 12. Fracture of the tooth crown (the part that is above the gum line) is the most common injury. School-age boys are twice as likely to experience dental trauma as girls.

Causes and symptoms

The cause of dental trauma varies depending on the age of the child. Toddlers are more likely to injure a tooth by falling, while older children are more likely to suffer dental trauma from a sports injury. Teenagers often present with dental trauma as the result of fights. The incisors in the upper jaw are the most commonly injured teeth.

Pain characterizes all dental trauma. The tooth may be knocked out and the socket bleeding, or it may be loose. There may be additional damage to the bones of the jaw and to the soft tissues of the mouth.

When to call the dentist

A permanent tooth that has been knocked out is a dental emergency. The dentist should also be called whenever dental trauma results in pain, dislocation of the tooth, or tooth sensitivity to hot or cold.

Diagnosis

Dental trauma is readily apparent upon examination. Dental **x rays** may be taken to determine the extent of the damage to fractured teeth. More comprehensive x rays are needed to diagnose a broken jaw.

Treatment

There is a possibility that a permanent tooth that has been knocked out can be re-implanted if handled promptly and correctly. If possible, the tooth should be reinserted in the socket and held there until the child sees a dentist or visits the emergency room. If it is not possible to replace the tooth in the socket, the tooth should immediately be placed in milk, saliva, or cool water with a pinch of saline solution (not contact lens solution or plain water). The tooth should be handled only by the crown and never be allowed to dry out. If a dentist can see the child within half an hour and the tooth has been preserved correctly, there is a possibility that it may be successfully re-implanted. Primary teeth are usually not re-implanted.

For lesser dental trauma, soft tissue injuries may require only cold compresses or ice to reduce swelling. Bleeding may be controlled with direct pressure applied with clean gauze. Deep lacerations and punctures may require stitches. Pain may be managed with aspirin or **acetaminophen** (Tylenol, Aspirin Free Excedrin), or ibuprofen (Motrin, Advil).

Treatment of a broken tooth will vary depending on the severity of the fracture. For immediate first aid, the injured tooth and surrounding area should be rinsed gently with warm water to remove dirt, then covered with a cold compress to reduce swelling and ease pain. A dentist should examine the injury as soon as possible. Any pieces from the broken tooth should be saved and taken to the dentist with the child.

If a piece of the outer tooth has chipped off, but the inner core (pulp) is undisturbed, the dentist may simply smooth the rough edges or replace the missing section with a small composite filling. In some cases, a fragment of broken tooth may be bonded back into place. If enough tooth is missing to compromise the entire tooth structure, but the pulp is not permanently damaged, the tooth will require a protective coverage with a gold or porcelain crown. If the pulp has been seriously damaged, the tooth will require root canal treatment before it receives a crown. A tooth that is vertically fractured or fractured below the gum line will require root canal treatment and protective restoration. A tooth that no longer has enough remaining structure to retain a crown may have to be extracted (surgically removed).

A broken jaw must be set back into its proper position and stabilized with wires while it heals. This is usually done by an oral surgeon. Healing may take six weeks or longer, depending on the patient's age and the severity of the fracture.

Alternative treatment

There is no substitute for treatment by a dentist or other medical professional. There are, however, homeopathic remedies and herbs that can be used simultaneously with dental care and throughout the healing process. Homeopathic arnica (*Arnica montana*) should be taken as soon as possible after the injury to help the body deal with the trauma. Repeating a dose several times daily for the duration of healing is also useful. Homeopathic hypericum (*Hypericum perforatum*) can be taken if nerve pain is involved, especially with a tooth extraction or root canal. Homeopathic comfrey (*Symphytum officinale*) may be helpful in treating pain due to broken jaw bones but should only be used after the bones have been reset. Calendula (*Calendula officinalis*) and plantain (*Plantago major*) can be used as a

KEY TERMS

Avulsion—The forcible separation of a piece from the entire structure.

Crown—The natural part of the tooth covered by enamel. A restorative crown is a protective shell that fits over a tooth.

Extraction—The removal of a tooth from its socket in the bone.

Pulp—The soft, innermost layer of a tooth that contains its blood vessels and nerves.

Root canal treatment—The process of removing diseased or damaged pulp from a tooth, then filling and sealing the pulp chamber and root canals.

mouth rinse to enhance tissue healing. These herbs should not be used with deep lacerations that need to heal from the inside first.

Prognosis

When dental trauma receives timely attention and proper treatment, the prognosis for healing is good. As with other types of trauma, infection may be a complication, but treatment with **antibiotics** is generally effective.

Prevention

Most dental trauma is preventable. Car seat belts should always be worn, and young children should be secured in appropriate car seats. Homes should be monitored for potential tripping and slipping hazards. **Childproofing** measures should be taken, especially for toddlers. Parents can place gates across stairs and pad sharp table edges.

Everyone who participates in contact sports should wear a mouth guard to avoid dental trauma. Athletes in football, ice hockey, wrestling, and boxing commonly wear mouth guards. The mandatory use of mouth guards in football prevents about 200,000 oral injuries annually. Mouth guards should also be worn along with helmets in noncontact sports such as skateboarding, in-line skating, and bicycling. An athlete who does not wear a mouth guard is 60 times more likely to sustain dental trauma than one who does. Any activity involving speed, an increased chance of falling, and potential contact with a hard piece of equipment has the likelihood of dental trauma that may be prevented or substantially reduced in severity with the use of mouth guards.

Parental concerns

Parents are sometimes concerned about the appearance of their child after he or she loses a permanent tooth. Cosmetic dentistry and orthodonture can with time and patience correct almost any problems arising from dental trauma.

Resources

ORGANIZATIONS

American Academy of Pediatric Dentistry. 211 East Chicago Ave., Ste. 700, Chicago, IL 60611–2616. Web site: <www.aapd.org>.

American Association of Endodontists. 211 East Chicago Ave., Ste. 1100, Chicago, IL 60611–2691. Web site: <www.aae.org>.

American Association of Oral and Maxillofacial Surgeons. 9700 West Bryn Mawr Ave., Rosemont, IL 60018–5701. Web site: <www.aaoms.org>.

American Dental Association. 211 E. Chicago Ave., Chicago, IL 60611. Web site: <www.ada.org>.

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Dependent personality disorder

Definition

Dependent personality disorder is a lack of self-confidence coupled with excessive dependence on others.

Description

Persons affected by dependent personality disorder have a disproportionately low level of confidence in their own **intelligence** and abilities and have difficulty making decisions and undertaking projects on their own. Their pervasive reliance on others, even for minor tasks

or decisions, makes them exaggeratedly cooperative out of **fear** of alienating those whose help their need. They are reluctant to express disagreement with others and are often willing to go to abnormal lengths to win the approval of those on whom they rely. Another common feature of the disorder is an exaggerated fear of being left to fend for oneself. Adolescents with dependent personality disorder rely on their parents to make even minor decisions for them, such as what they should wear or how they should spend their free time, as well as major ones, such as what college they should attend or which career they should choose.

It is important to note that in other societies where cultural norms are different, dependent and/or passive traits may be valued, particularly in women. The criteria outlined here for dependent personality disorder is applicable to Americans only, and even then may not apply to all cultural groups within the United States.

Demographics

Dependent personality disorder occurs equally in males and females and usually begins by early adulthood. Overall prevalence is approximately one to two percent of the general population. Because children and adolescents are dependent on adults by necessity, dependent personality disorder is very rarely diagnosed in these age groups.

Causes and symptoms

In the *Diagnostic and Statistical Manual of Mental Disorders, 4th Edition Text Revision (DSM-IV-TR)*, the American Psychiatric Association states that five of the following criteria should be present for a diagnosis of dependent personality disorder:

- difficulty making decisions, even minor ones, without guidance and reassurance from others
- requiring others to take responsibility for major decisions and responsibilities beyond what would be age-appropriate (e.g., letting a parent choose a college without offering any input on the decision)
- difficulty disagreeing with others due to an unreasonable fear of alienation
- unable to initiate or complete projects or tasks due to a belief that he or she is either inept or that the appearance of success would lead a support person(s) to abandon him or her
- takes on unreasonably unpleasant tasks or sacrifices things in order to win the approval of others
- unable to spend time alone due to a lack of self-reliance and an unreasonable fear of being unable to care for oneself
- inability to remain independent of a close relationship as manifested by seeking a substitute support relationship immediately after one ends (e.g., a teenager who feels she must have a boyfriend constantly to validate her self-worth)
- unrealistic preoccupation with the thought of being left to care for oneself

Dependent personality disorder is more common in those who have suffered from chronic illness in childhood. A child may also exhibit dependent behavior in response to a specific stressful life event (such as the death of a caregiver or a **divorce**). However, it should not be considered a potential symptom of dependent personality disorder unless the behavior becomes chronic and significantly interferes with day-to-day functioning and/or causes the child significant distress.

When to call the doctor

It is developmentally suitable for young children to go through "clingy" stages where overt dependent behavior on a parent or caregiver is commonplace. However, if dependency in a child or adolescent starts to interfere with school work, daily living, and the child's sense of **self-esteem** and well-being, parents should seek professional help from their child's doctor. If a child or teen indicates at any time that he/she has had recent thoughts of self-injury, **suicide**, or of inflicting harm on others, professional assistance from a mental health care provider or care facility should be sought immediately.

Diagnosis

Older teens or young adults who have demonstrated at least five of the *DSM-IV-TR* criteria (or symptoms) outlined above are eligible for a diagnosis of dependent personality disorder. In the *DSM-IV-TR*, the APA warns that a diagnosis of dependent personality disorder "should be used with great caution, if at all, in children and adolescents, for whom dependent behavior may be developmentally appropriate." Children are dependent on parents and other adults in their lives for support and physical and emotional **safety** by necessity; it is only when the behaviors are excessive and age inappropriate that a diagnosis of dependent personality disorder can be contemplated.

KEY TERMS

Borderline personality disorder—A mental disorder characterized by mood swings, unstable interpersonal relationships, poor self-image, and self-destructive and impulsive behaviors.

Histrionic personality disorder—A mental disorder characterized by inappropriate attention-seeking behavior, rapid emotional shifts, and exaggerated expression of emotion.

Separation anxiety—Childhood fear of leaving parents for any reason.

Treatment

The primary treatment for dependent personality disorder is psychotherapy, with an emphasis on learning to cope with **anxiety**, developing assertiveness, and improving decision-making skills. Group therapy can also be helpful. In cases where parents or other adult caregivers seem to be facilitating the behavior, therapy for them is also appropriate.

Prognosis

Dependent personality disorder frequently occurs in tandem with other personality-based mental illness, such as borderline, histrionic, and avoidant **personality disorders**. It is also believed that those diagnosed with dependent personality disorder are at an increased risk of mood and anxiety disorders.

Prevention

There is no known prevention strategy for dependent personality disorder. However, some tactics that can promote healthy socialization and positive self-esteem from an early age include encouraging healthy peer relationships, investing a child with increasing levels of age-appropriate responsibilities and independence, and offering choices to even the smallest children.

Parental concerns

Dependent personality disorder is an extremely rare diagnosis in children. Parents of children who have been diagnosed with dependent personality disorder may wish to seek a second opinion from a trained psychologist or psychiatrist specializing in pediatric care. **Separation anxiety** shares some common features with dependent personality disorder, and should be considered as a differential diagnosis.

Resources

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ORGANIZATIONS

The American Academy of Child and Adolescent Psychiatry. 3615 Wisconsin Ave., N.W., Washington, D.C. 20016–3007. (202) 966–7300. Web site: <www.aacap.org>.

National Institute of Mental Health. 6001 Executive Boulevard, Rm. 8184, MSC 9663, Bethesda, MD 20892–9663. (301) 443–4513.

Paula Ford-Martin

Depersonalization disorder *see* **Dissociative disorders**

Depression *see* **Depressive disorders**

Depressive disorders

Definition

Depression and depressive disorders (unipolar depression) are mental illnesses characterized by a profound and persistent feeling of sadness or despair and/or a loss of interest in things that once were pleasurable. Disturbance in **sleep**, appetite, and mental processes are common symptoms of depression.

Description

Everyone experiences feelings of unhappiness and sadness occasionally. However, when these depressed feelings start to dominate everyday life and cause physical and mental deterioration, they become what are known as depressive disorders.

There are two main categories of depressive disorders: major depressive disorder and dysthymic disorder.

Major depressive disorder is a moderate to severe episode of depression lasting two or more weeks. Individuals experiencing this major depressive episode may have trouble sleeping, lose interest in activities they once took pleasure in, experience a change in weight, have difficulty concentrating, feel worthless and hopeless, or have a preoccupation with death or **suicide**. In children, the major depression may often appear as irritability.

While major depressive episodes may be acute (intense but short-lived), dysthymic disorder is an ongoing, chronic depression that lasts two or more years (one or more years in children) and has an average duration of 16 years. The mild to moderate depression of dysthymic disorder may rise and fall in intensity, and those afflicted with the disorder may experience some periods of normal, non-depressed mood of up to two months in length. Its onset is gradual, and dysthymic patients may not be able to pinpoint exactly when they started feeling depressed. Individuals with dysthymic disorder may experience a change in sleeping and eating patterns, low **self-esteem**, fatigue, trouble concentrating, and feelings of hopelessness. Parents of children suffering from dysthymic disorder may notice their child experience a fall in grades and a lack of interest in **extracurricular activities** that were once enjoyable.

Depression also can occur in **bipolar disorder**, an affective mental illness that causes radical emotional changes and mood swings, from manic highs to depressive lows. The majority of bipolar individuals experience alternating episodes of mania and depression.

Demographics

The Substance Abuse and Mental Health Services Administration (SAMSHA) estimates that one out of every 33 children may suffer from depression. Among adults 18 and older, depressive disorders affect an estimated 18.8 million Americans each year. Women are twice as likely to suffer from a depressive disorder than men; approximately 12.4 million American women and 6.4 million men deal with depression. The average age a first depressive episode occurs is in the mid-20s, although the disorder strikes all age groups indiscriminately, from children to the elderly.

According to the U.S. Surgeon General, major depression occurs in about 5 percent of children between age nine and 17, and at any one point in time, 10 to 15 percent of U.S. children and adolescents experience some symptoms of depression.

Causes and symptoms

The causes behind depression are complex and as of 2004 not fully understood. While an imbalance of certain neurotransmitters—the chemicals in the brain that transmit messages between nerve cells—is believed to be key to depression, external factors such as upbringing may be as important. For example, it is speculated that, if an individual is abused and neglected throughout childhood and **adolescence**, a pattern of low self-esteem and negative thinking may emerge. From that, a lifelong pattern of depression may follow.

Heredity seems to play a role in who develops depressive disorders. Individuals with major depression in their immediate **family** are up to three times more likely to have the disorder themselves. It would seem that biological and genetic factors may make certain individuals pre-disposed to depressive disorders, but environmental circumstances often may trigger the disorder as well.

External stressors and significant life changes such as chronic medical problems, death of a loved one, or **divorce** or estrangement of parents also can result in a form of depression known as adjustment disorder. Although periods of adjustment disorder usually resolve themselves, occasionally they may evolve into a major depressive disorder.

Common red flags that children may be experiencing a depressive disorder include a sudden decline in grades and/or disinterest in schoolwork, avoidance of friends, loss of interest in extracurricular activities, and withdrawal from family.

Major depressive episode

Individuals experiencing a major depressive episode have a depressed mood and/or a diminished interest or pleasure in activities. Children experiencing a major depressive episode may appear or feel irritable rather than depressed. In addition, five or more of the following symptoms will occur on an almost daily basis for a period of at least two weeks:

- significant change in weight
- insomnia or hypersomnia (excessive sleep)
- psychomotor agitation or retardation
- fatigue or loss of energy
- feelings of worthlessness or inappropriate guilt
- diminished ability to think or to concentrate, or indecisiveness
- recurrent thoughts of death or suicide and/or suicide attempts

Dysthymic disorder

Dysthymia commonly occurs in tandem with other psychiatric and physical conditions. Up to 70 percent of dysthymic patients have both dysthymic disorder and major depressive disorder, known as double depression. Substance abuse, panic disorders, **personality disorders**, social **phobias**, and other psychiatric conditions also are found in many dysthymic patients. Dysthymia is prevalent in patients with certain medical conditions, including multiple sclerosis, **AIDS**, **hypothyroidism**, chronic fatigue syndrome, diabetes, and post-cardiac transplantation. The connection between dysthymic disorder and these medical conditions is unclear, but it may be related to the way the medical condition and/or its pharmacological treatment affects neurotransmitters. Dysthymic disorder can lengthen or complicate the recovery of patients also suffering from medical conditions.

Along with an underlying feeling of depression, people with dysthymic disorder experience two or more of the following symptoms on an almost daily basis for a period for two or more years (most suffer for five years) or one year or more for children:

- under-eating or overeating
- insomnia or hypersomnia
- low energy or fatigue
- low self-esteem
- poor concentration or trouble making decisions
- feelings of hopelessness

When to call the doctor

Just like adults, children have days when they are feeling down. But if those blue or bad moods begin to interfere with schoolwork and daily living and start to increase in frequency, parents or caregivers need to seek help from their child's doctor. If a child or teen reveals at any time that they have had recent thoughts of self-injury or suicide, professional assistance from a mental healthcare provider or care facility should be sought immediately.

Diagnosis

In addition to an interview, a clinical inventory or scale such as the Child Depression Inventory (CDI) or the Child Depression Rating Scale (CDRS) may be used to assess a child's mental status and determine the presence of depressive symptoms. Tests may be administered in an outpatient or hospital setting by a pediatrician, general practitioner, social worker, psychiatrist, or psychologist.

Treatment

Major depressive and dysthymic disorders are typically treated with **antidepressants** or psychosocial therapy. Psychosocial therapy focuses on the personal and interpersonal issues behind depression, while antidepressant medication is prescribed to provide more immediate relief for the symptoms of the disorder. When used together correctly, therapy and antidepressants are a powerful treatment plan for the depressed child or adolescent.

Antidepressants

Selective serotonin reuptake inhibitors (SSRIs) such as fluoxetine (Prozac) and sertraline (Zoloft) reduce depression by increasing levels of serotonin, a neurotransmitter. Some clinicians prefer SSRIs for treatment of dysthymic disorder. **Anxiety**, **diarrhea**, drowsiness, **headache**, sweating, **nausea**, and insomnia all are possible side effects of SSRIs. As of 2004, fluoxetine was the only SSRI (and the only antidepressant drug) approved by the U.S. Food and Drug Administration for use in children and adolescents with major depressive disorder. However, physicians may prescribe other SSRIs in younger patients in an off-label use of these drugs.

In 2004, fluoxetine and nine other antidepressant drugs came under scrutiny when the FDA issued a public health advisory and announced it was requesting the addition of a warning statement in drug labeling that outlined the possibility of worsening depression and increased suicide risk. These developments were the result of several clinical studies that found that some children taking these antidepressants had an increased risk of suicidal thoughts and actions. The FDA announced at the time that the agency would embark on a more extensive analysis of the data from these clinical trials and decide if further regulatory action was necessary.

Older classes of antidepressant drugs—(tricyclic antidepressants (TCAs), heterocyclics, and monoamine oxidase inhibitors (MAOIs)—do not have any substantial demonstrated effectiveness in pediatric populations and have potentially serious side effects that make them undesirable for child and adolescent use.

For severe depression that does not respond well to antidepressant, mood stabilizer drugs (e.g., lithium, carbamazepine, valproic acid) may be recommended.

Depressive disorders

Diagnosis	Symptoms	Treatment
Sadness	Transient, normal depressive response or mood change due to stress.	Emotional support
Bereavement	Sadness related to a major loss that persists for less than two months after the loss. Thoughts of death and morbid preoccupation with worthlessness are also present.	Emotional support; counseling
Sadness problem	Sadness or irritability that begins to resemble major depressive disorder, but lower in severity and more transient.	Support; counseling; medication possible
Adjustment disorder with depressed mood	Symptoms include depressed mood, tearfulness, and hopelessness, and occur within three months of an identifiable stressor. Symptoms resolve in six months.	Psychotherapy; medication
Major depressive disorder	A depressed or irritable mood or diminished pleasure as well as three to seven of the following criteria almost daily for two weeks. The criteria include: recurrent thoughts of death and suicidal ideation; weight loss or gain; fatigue or energy loss; feelings of worthlessness; diminished ability to concentrate; insomnia or hypersomnia; feeling hyper and jittery, or abnormally slow.	Psychotherapy; medication
Dysthymic disorder	Depressed mood for most of the day, for more days than not, for one year, including the presence of two of the following symptoms: poor appetite or overeating; insomnia/hypersomnia; low energy/fatigue; poor concentration; feelings of hopelessness. Symptoms are less severe than those of a major depressive episode but are more persistent.	Psychotherapy; medication
Bipolar I disorder, most recent episode depressed	Current major depressive episode with a history of one manic or mixed episode. (Manic episode is longer than four days and causes significant impairment in normal functioning.) Moods are not accounted for by another psychiatric disorder.	Psychotherapy; medication
Bipolar II disorder, recurrent major depressive episodes with hypomanic episodes	Presence or history of one major depressive episode and one hypomanic episode (similar to manic episode but shorter and less severe). Symptoms are not accounted for by another psychiatric disorder and cause clinically significant impairment in functioning.	Psychotherapy; medication

SOURCE: Academy of American Family Physicians. 2000. http://www.aafp.org.

(Table by GGS Information Services.)

Psychotherapy

Psychotherapy, or talk therapy, involves analyzing a child's life to bring to light possible contributing causes of the present depression. During treatment, the therapist helps the patient to become aware of his or her thinking patterns and how they came to be, and works with them to develop healthy problem solving and copying skills. In very young patients, a therapist may use **toys**, games, and dolls as a vehicle for helping a child express her emotions. This type of therapy, sometimes referred to as play therapy, is useful in children who may not have the developmental capacity or language skills to express the thoughts and feelings behind their depression.

Cognitive-behavioral therapy assumes that the patient's faulty thinking is causing the current depression and focuses on changing the depressed patient's thought patterns and perceptions. The therapist helps the patient identify negative or distorted thought patterns and the emotions and behavior that accompany them and then retrains the depressed individual to recognize the thinking and react differently to it.

Electroconvulsant therapy

Electroconvulsive therapy (ECT) is only considered after all therapy and pharmaceutical treatment options have been unsuccessful, and even then it is a treatment of last resort, typically employed when a patient has become catatonic, suicidal, or psychotic as well as depressed.

The treatment consists of a series of electrical pulses that move into the brain through electrodes on the patient's head. ECT is given under anesthesia, and patients are administered a muscle relaxant to prevent convulsions. Although the exact mechanisms behind the success of ECT therapy are not known, it is believed that the electrical current modifies the electrochemical processes of the brain, consequently relieving depression. Headaches, muscle soreness, nausea, and confusion are possible side effects immediately following an ECT pro-

cedure. Memory loss, typically transient, also has been reported in ECT patients.

Alternative treatment

St. John's wort (*Hypericum perforatum*) is used throughout Europe to treat mild depressive symptoms. Unlike traditional prescription antidepressants, this herbal antidepressant has few reported side effects. Despite uncertainty concerning its effectiveness, a 2003 report said acceptance of the treatment continues to increase. A poll showed that about 41 percent of 15,000 science professionals in 62 countries said they would use St. John's wort for mild to moderate depression. The usual adult dose is 300 mg three times daily and may be lowered for children and adolescents.

In several small studies, S-adenosyl-methionine (SAM, SAMe) was shown to be more effective than placebo and equally effective as tricyclic antidepressants in treating depression. In 2003, a U.S. Department of Health and Human Services team reviewed 100 clinical trials on SAMe and concluded that it worked as well as many prescription medications without side effects of stomach upset.

Parents and caregivers of children who suffer from depression should consult their child's physician before administering any herb or dietary supplement. Some supplements can interfere with the action of other prescription and over-the-counter medications. In addition, some supplements may not be appropriate for use in children with certain medical conditions.

A report from Great Britain published in 2003 emphasized that more physicians should encourage alternative treatments such as behavioral and self- help programs, supervised **exercise** programs, and watchful waiting before subscribing antidepressant medications for mild depression.

Nutritional concerns

Poor **nutrition**, especially eating habits that lead to overweight or **obesity** in children, can also contribute to depression. A 2003 study in the journal Pediatrics found that children who are substantially overweight for long periods of time are more likely to suffer from depression. Whether the depression causes the weight problem or the weight issue triggers the depression was not completely clear.

Prognosis

Untreated or improperly treated depression is the number one cause of suicide in the United States. Proper treatment relieves symptoms in 80 to 90 percent of depressed patients. After each major depressive episode, the risk of recurrence climbs significantly: 50 percent after one episode, 70 percent after two episodes, and 90 percent after three episodes. For this reason, patients need to be aware of the symptoms of recurring depression and may require long-term maintenance treatment of antidepressants and/or therapy.

KEY TERMS

Hypersomnia—An abnormal increase of 25% or more in time spent sleeping. Individuals with hypersomnia usually have excessive daytime sleepiness.

Neurotransmitter—A chemical messenger that transmits an impulse from one nerve cell to the next.

Psychomotor agitation—Disturbed physical and mental processes (e.g., fidgeting, wringing of hands, racing thoughts); a symptom of major depressive disorder.

Psychomotor retardation—Slowed mental and physical processes characteristic of a bipolar depressive episode.

Prevention

Good nutrition, proper sleep, exercise, and family support are all important to a healthy mental state, particularly in children. Extended maintenance treatment with antidepressants may be required in some patients to prevent relapse. Early intervention for children with depression can be effective in avoiding the development of more severe psychological problems later in life.

Parental concerns

Children who are diagnosed with depression should be reassured that the condition is quite common and that it is due to factors beyond their control (i.e., genetics, neurochemical imbalance) rather than any fault of the child. For those children and teens who feel stigmatized or self-conscious about their depression, arranging psychotherapy sessions outside school hours may lessen their burden. Any child prescribed antidepressants should be carefully monitored for any sign of side effects, and these should be reported to their physician when they do occur. A dosage adjustment or medication change may be warranted if side effects are disruptive or potentially dangerous.

See also Bipolar disorder.

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Dermatitis

Definition

Dermatitis is a general term used to describe inflammation of the skin.

Description

Most types of dermatitis are characterized by an itchy pink or red rash.

Contact dermatitis is an allergic reaction to something that irritates the skin and is manifested by one or more lines of red, swollen, blistered skin that may itch or seep. It usually appears within 48 hours after touching or brushing against a substance to which the skin is sensitive. The condition is more common in adults than in children.

Contact dermatitis can occur on any part of the body, but it usually affects the hands, feet, and groin. Contact dermatitis usually does not spread from one person to another, nor does it spread beyond the area exposed to the irritant unless affected skin comes into contact with another part of the body. However, in the case of some irritants, such as **poison ivy**, contact dermatitis can be passed to another person or to another part of the body.

Atopic dermatitis is characterized by **itching**, scaling, swelling, and sometimes blistering. In early childhood it is called infantile eczema and is characterized by redness, oozing, and crusting. It is usually found on the face, inside the elbows, and behind the knees.

Seborrheic dermatitis may be dry or moist and is characterized by greasy scales and yellowish crusts on the scalp, eyelids, face, external surfaces of the ears, underarms, breasts, and groin. In infants it is called cradle cap.

Demographics

Allergic reactions are common. No formal statistics are kept on such attacks.

Causes and symptoms

Allergic reactions are genetically determined, and different substances cause contact dermatitis to develop in different people. A reaction to resin produced by poison ivy, **poison oak**, or **poison sumac** is the most common source of symptoms. It is, in fact, the most common

allergy in the United States, affecting one of every two people in the country.

Flowers, herbs, and vegetables can also affect the skin of some people. **Burns** and **sunburn** increase the risk of dermatitis developing. Chemical irritants that can cause the condition include:

- chlorine
- cleansers
- detergents and soaps
- fabric softeners
- glues used on artificial nails
- perfumes
- topical medications

Contact dermatitis can develop when the first contact occurs or after years of use or exposure.

Atopic dermatitis can be caused by **allergies**, **asthma**, or stress, and there seems to be a genetic predisposition for atopic conditions. It is sometimes caused by an allergy to nickel in jewelry.

Seborrheic dermatitis (for which there may also be a genetic predisposition) is usually caused by overproduction of the oil glands. In adults it can be associated with **diabetes mellitus** or gold allergy. In infants and adults it may be caused by a biotin deficiency.

When to call the doctor

A doctor or other healthcare provider should be consulted when **rashes** appear. With some experience, common rashes can be accurately identified by parents. Rashes that cannot be accurately identified should be referred to competent healthcare professional for identification and possible treatment.

Diagnosis

The diagnosis of dermatitis is made on the basis of how the rash looks and its location. The doctor may scrape off a small piece of affected skin for microscopic examination or direct the person to discontinue use of any potential irritant that has recently come into contact with the affected area. Two weeks after the rash disappears, the person may resume use of the substances, one at a time, until the condition recurs. Eliminating the substance most recently added should eliminate the irritation.

If the origin of the irritation has still not been identified, a dermatologist may perform one or more patch tests, which involves dabbing a small amount of a suspected irritant onto skin on the person's back. If no irritation develops within a few days, another patch test is performed. The process continues until the person experiences an allergic reaction at the spot where the irritant was applied.

Treatment

Treating contact dermatitis begins with eliminating or avoiding the source of irritation. Prescription or over-the-counter corticosteroid creams can lessen inflammation and relieve irritation. Creams, lotions, or ointments not specifically formulated for dermatitis can intensify the irritation. Oral **antihistamines** are sometimes recommended to alleviate itching, and **antibiotics** are prescribed if the rash becomes infected. Medications taken by mouth to relieve symptoms of dermatitis can make skin red and scaly and cause hair loss.

People who have a history of dermatitis should remove their rings before washing their hands. They should use bath oils or glycerin-based soaps and bathe in lukewarm saltwater.

Patting rather than rubbing the skin after bathing and thoroughly massaging lubricating lotion or nonprescription cortisone creams into still-damp skin can soothe red, irritated nummular dermatitis. Highly concentrated cortisone preparations should not be applied to the face, armpits, groin, or rectal area. Periodic medical monitoring is necessary to detect side effects in people who use such preparations on rashes covering large areas of the body.

Coal-tar salves can help relieve symptoms of nummular dermatitis that have not responded to other treatments, but these ointments have an unpleasant odor and stain clothing.

Coal-tar shampoos may be used for seborrheic dermatitis that occurs on the scalp. Sun exposure after the use of these shampoos should be avoided because the risk of sunburn of the scalp is increased.

Alternative treatment

Some herbal therapies can be useful for skin conditions. Among the herbs most often recommended are:

- burdock root (*Arctium lappa*)
- calendula (*Calendula officinalis*) ointment
- chamomile (*Matricaria recutita*) ointment
- cleavers (*Galium* ssp.)
- evening primrose oil (*Oenothera biennis*)
- nettles (*Urtica dioica*)

Diprobase cream, an emollient, used to treat contact dermatitis. *(© Jim Selby/Photo Researchers, Inc.)*

Contact dermatitis can be treated botanically and homeopathically. Grindelia (*Grindelia* spp.) and sassafras (*Sassafras albidum*) can help when applied topically. Determining the source of the problem and eliminating it is essential. Oatmeal baths are very helpful in relieving the itch. Bentonite clay packs or any mud-pack draws the fluid out and helps dry up the lesions. Cortisone creams are not recommended.

Selenium-based shampoos, topical applications of flax oil and/or olive oil, and biotin supplementation are among the therapies recommended for seborrheic dermatitis.

Prognosis

Dermatitis is often chronic, but symptoms can generally be controlled.

Prevention

Contact dermatitis can be prevented by avoiding the source of irritation. If the irritant cannot be avoided completely, the person should wear gloves and other protective clothing whenever exposure is likely to occur.

Immediately washing the exposed area with soap and water can stem allergic reactions to poison ivy, poison oak, or poison sumac, but because soaps can dry the skin, people susceptible to dermatitis should use them only on the face, feet, genitals, and underarms.

KEY TERMS

Allergic reaction—An immune system reaction to a substance in the environment; symptoms include rash, inflammation, sneezing, itchy watery eyes, and runny nose.

Corticosteroids—A group of hormones produced naturally by the adrenal gland or manufactured synthetically. They are often used to treat inflammation. Examples include cortisone and prednisone.

Patch test—A skin test in which different antigens (substances that cause an allergic reaction) are introduced into a patient's skin via a needle prick or scratch and then observed for evidence of an allergic reaction to one or more of them. Also known as a scratch test.

Rash—A spotted, pink or red skin eruption that may be accompanied by itching and is caused by disease, contact with an allergen, food ingestion, or drug reaction.

Ulcer—A site of damage to the skin or mucous membrane that is characterized by the formation of pus, death of tissue, and is frequently accompanied by an inflammatory reaction.

Clothing should be loose fitting and 100 percent cotton. New clothing should be washed in dye-free, unscented detergent before being worn.

Yoga and other relaxation techniques may help prevent atopic dermatitis caused by stress.

Avoidance of sweating may aid in preventing seborrheic dermatitis.

A person who has dermatitis should also notify a doctor if any of the following occurs:

- fever develops
- skin oozes or other signs of infection appear
- symptoms do not begin to subside after seven days of treatment
- he/she comes into contact with someone who has a wart, **cold sore**, or other viral skin infection

Nutritional concerns

Eating a balanced and adequate diet is generally helpful. People who are susceptible to dermatitis that is linked to particular foods obviously should avoid consuming them.

Parental concerns

Parents should try to monitor new substances and foods when their children encounter them for the first time.

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Dermatomyositis

Definition

Dermatomyositis is one of a group of relatively uncommon diseases known as inflammatory **myopathies**, or inflammatory disorders of the muscles. Dermatomyositis is distinguished from other diseases in this category by the fact that it causes a characteristic skin rash as well as affecting the strength and functioning of the muscles. Dermatomyositis in children and adolescents is called juvenile dermatomyositis (abbreviated JDMS or simply JD) because it is different from the adult form of the disorder in several respects. The most significant differences between JDMS and adult dermatomyositis are as follows:

- Children are more likely than adults to develop calcinosis (calcium deposits in the skin) and gastrointestinal symptoms.
- Children are more likely to develop pains in the joints.
- Adults with dermatomyositis over the age of 50 have a 15 percent risk of developing **cancer**, whereas juvenile dermatomyositis is rarely associated with malignancy.

Description

JDMS is sometimes called childhood idiopathic dermatomyositis. The word *idiopathic* means that the cause of the disease is unknown and that it appears to begin spontaneously. The disorder is also occasionally defined as a systemic vasculopathy, *systemic* meaning that it affects the body as a whole rather than just one part, and *vasculopathy* meaning that it affects the blood vessels.

JDMS usually begins with a reddish or reddish-purple rash, called a heliotrope rash because of its color. In most children the rash first appears on the eyelids or cheekbone area and is often mistaken for an allergy symptom. It may also appear as dry patches of reddened skin on the child's knuckles, knees, elbows, or ankles that are often misdiagnosed as eczema. In a few children, the rash may spread over the entire body. In some children, the rash is made worse by exposure to sunlight.

The heliotrope rash is either accompanied or followed by weakness of the body's central muscles; that is, the muscles on or close to the trunk of the body. These muscles are also called proximal muscles. The child may complain of tiredness and have trouble sitting up, standing, or moving the neck, shoulders, abdomen, back, or hips. The muscular weakness varies in severity; while some children may simply have less energy than usual, others may be literally unable to get out of bed or may have trouble swallowing or breathing. In some cases the child's voice may sound as if he or she is talking through the nose.

The third major symptom of juvenile dermatomyositis is a low-grade **fever** (one or two degrees Fahrenheit above normal).

Transmission

Although fever is one of the most common symptoms of JDMS, the disease cannot be transmitted from one child to another.

Demographics

Juvenile dermatomyositis most commonly affects children between the ages of five and 15 years of age. The ratio of girls to boys is 2:1. The disease is thought to be equally common around the world, affecting about three children per million. It is estimated that 3,000 to 5,000 children in the United States have JDMS as of the early 2000s. The incidence appears to be increasing, however. Although Caucasian children are affected more often than African-American children, the rate is rising faster among African Americans. The reasons for this increase are not known as of 2004.

JDMS has a seasonal pattern in North America, occurring more frequently in the spring and summer months.

Causes and symptoms

Causes

The precise causes of JDMS are not yet fully understood. One theory holds that the disease is an autoimmune reaction caused by the body's abnormal response to a virus. In an autoimmune reaction, the body begins to attack its own tissues after it has successfully eliminated the virus. Some researchers have identified virus-like structures in the muscle cells of patients known to have dermatomyositis, while others have noted that children newly diagnosed with JDMS often have a history of infection with a Coxsackie virus within three months of the first JDMS symptoms.

Genetic factors are also thought to be involved in juvenile dermatomyositis. In 2002 a group of researchers at Northwestern University reported that susceptibility to JDMS is related to a genetic marker known as DQA1*0501. Another team of doctors at the Children's National Medical Center in Washington, DC, has suggested that children with this particular genetic marker develop JDMS when a viral infection triggers an abnormal interaction among the body's immune system, the muscles, and the vascular system. There may also be other genes that increase children's susceptibility to the disease that have not yet been identified.

Symptoms

The major symptoms of juvenile dermatomyositis include a characteristic reddish or purplish rash called a heliotrope rash; weakness or **pain** in the proximal muscles; and a low-grade fever.

Other symptoms that may occur in children with juvenile dermatomyositis include:

- Contractures: A contracture is an abnormal shortening of the muscles near a joint that causes the joint to remain in a bent position. JDMS may lead to contractures for two reasons. The first is that the muscle may form scar tissue during the healing process. The second is that the child may avoid exercising his or her muscles because he or she feels weak. The muscles then gradually lose their ability to hold the joint in its proper position.
- Stunted or slowed growth: The child may grow more slowly than normal during an acute attack of JDMS because some of the medications used to treat the disease slow down the growth of bones. In addition, some

of the body's energy that is ordinarily used for growth is used instead to fight off the disease.

- Sore or swollen joints: About half of all children diagnosed with JDMS have sore or swollen joints, caused by the inflammation of the muscles around the joints. The joint may feel warm to the touch and look reddish as well as swollen.
- Vasculitic ulcers: Vasculitis refers to inflammation of a blood vessel. A vasculitic ulcer is a hole or tear that develops in the tissues around an inflamed blood vessel. In children with JDMS, vasculitic ulcers usually appear either in the skin rash or in the digestive tract. Vasculitic ulcers in the skin look like open sores within the reddish-purple rash; they vary in size from small spots to sores as much as an inch across. Vasculitic ulcers in the digestive tract may lead to perforation of the intestines, which is a medical emergency.
- Calcinosis: Calcinosis is a condition in which small lumps of calcium compounds develop beneath the skin or in the muscles of children with JDMS. They affect between 50 percent and 60 percent of children with the disorder and range in size from less than a millimeter across to lumps the size of small pebbles. Large lumps may interfere with the movement of the muscles, cause pain if they are located close to a joint, or even break through the skin. In most cases, however, the pieces of calcium are reabsorbed by the body during the recovery process.
- Dysphagia: Dysphagia refers to difficulty or discomfort when swallowing. Children whose throat muscles are affected by the disorder may experience difficulty in swallowing food; some lose weight because the dysphagia affects their appetite.
- Abnormal heart rhythms and myocarditis: Myocarditis refers to inflammation of the muscles of the walls of the heart. About 50 percent of children with JDMS develop an abnormal heart rhythm.

When to call the doctor

It is not always easy to tell when a child might have juvenile dermatomyositis. The skin rash associated with JDMS is often mistaken for eczema. In addition, some children may develop a mild form of muscle weakness before the telltale rash appears. While about 50 percent of children diagnosed with JDMS have an acute onset of symptoms, the other 50 percent have what is called a subacute onset, which means that the symptoms are milder and come on more slowly. While most children with acute symptoms are diagnosed within three months, the correct diagnosis of children with subacute symptoms may take a year or even longer.

Children who have developed sudden weakness of the muscles that control breathing or swallowing, or those who have developed vasculitic ulcers in the digestive tract may need to be hospitalized. Parents should call the doctor *at once* if they notice any of the following symptoms:

- choking on food or being unable to swallow
- weak voice or total loss of voice
- severe pain in the abdomen
- coal-black or tarry-looking stools
- change in bowel habits
- passing red blood with stools

Diagnosis

History and physical examination

The first step in diagnosing juvenile dermatomyositis is the taking of a complete history and giving the child a thorough physical examination. The doctor will ask the child and the parents when the symptoms began, what parts of the body are affected, whether the child can keep up his or her normal activities, and (in some cases) whether other **family** members have arthritis or muscle diseases.

During the physical examination, the doctor looks for several specific signs and symptoms, including heliotrope rash on the child's face, knuckles, knees, elbows, or the cuticles of the fingers; swelling around the eyes; a nasal quality to the child's voice; sore or weak muscles; and sore or swollen joints. The doctor will test the strength of the muscles by asking the child to lift his or her head, arms, or legs while the doctor gently pushes or presses downward.

The child's doctor may use a set of criteria first established in 1975 as part of the process of diagnosis. These so-called Bohan-Peter criteria are interpreted as follows: If the child meets the first criterion (the characteristic rash), three of the remaining four criteria must be met to make the diagnosis of juvenile dermatomyositis. The last three criteria listed below require special laboratory tests. If two out of four are met, the child is considered to have "probable" JDMS:

- heliotrope rash
- weakness of the central muscles on both sides of the body
- a higher than normal level of muscle enzymes in the blood
- a specific pattern of changes in the muscle tissue caused by inflammation

- an abnormal pattern of electrical activity in the muscles

Laboratory tests

The doctor will have a sample of the child's blood tested for certain muscle enzymes known as aldolase and CPK. These enzymes are leaked into the blood stream when muscles become inflamed. Abnormally high levels of aldolase and CPK indicate muscle damage. In addition to testing for muscle enzymes, the doctor may also have the blood sample tested for antinuclear antibodies (ANA), which are produced when a person's immune system is producing antibodies against the body's own tissues. Between 60 percent and 80 percent of children with JDMS have elevated levels of ANA.

A muscle biopsy may also be performed. In this test, the doctor removes a small piece of muscle tissue and has it examined under a microscope to see whether the muscle fibers and nearby blood vessels have undergone certain changes that indicate JDMS. Many doctors, however, skip this test if the child has the typical heliotrope rash, shows signs of muscle weakness during the physical examination, and has high muscle enzyme levels in the blood test. A muscle biopsy is necessary, however, if the child has the heliotrope rash but normal enzyme levels.

An electromyogram (EMG) measures electrical activity in the muscles. The doctor pierces the child's skin with a thin needle connected to a wire running to a machine that records the pattern of electrical activity in the muscle tissue. EMGs are not always performed, however, because the test is somewhat painful.

Imaging studies

Some doctors may order a **magnetic resonance imaging** (MRI) test in order to evaluate the presence of inflammation in the muscles of children with normal muscle enzyme levels in their blood. It may also be done in order to identify appropriate muscles for testing when a biopsy is necessary.

Treatment

The treatment of juvenile dermatomyositis involves a combination of approaches. The treatment plan may also have to be changed periodically as the child's symptoms change.

Medications

Medications are the mainstay of treatment for juvenile dermatomyositis. The most common drug used is prednisone, a steroid that is given to reduce pain, control the fever and skin rash, and improve the strength of the child's muscles. Prednisone may be given in pill form or as a weekly intravenous infusion. Unfortunately, prednisone has a number of side effects, ranging from high blood pressure, weight gain, and stretch marks to mood changes and weak or damaged bones. If taken for a long period of time, prednisone may also lead to the development of eye cataracts and slow down the child's growth.

Another drug that is used to treat JDMS is methotrexate, an immunosuppressive drug that may be taken as pills or given as an injection. Methotrexate works by slowing down the immune system. It is usually given together with prednisone. It also has some potentially serious side effects, including **nausea and vomiting**, **diarrhea**, increased susceptibility to infections, skin **rashes**, a decrease in the number of blood cells, and potential liver damage. In some cases, the doctor may recommend another immunosuppressive drug known as cyclosporine to be taken together with methotrexate. If the child's skin rash is unusually severe, the doctor may prescribe hydroxychlorethotrexate (Plaquenil). Methotrexate, cyclosporine, and hydroxychlorethotrexate all belong to a category of medications known as disease-modifying anti-rheumatic drugs, or DMARDs.

Exercise and physical therapy

Exercise and physical therapy are an important part of treatment for JDMS because they help to prevent contractures, keep the child's joints flexible, and strengthen muscles. In most cases, the child will be referred to a physical therapist who can design an exercise program for the specific sets of muscles affected by the disease. The exercise program is modified as the child's strength gradually returns.

Psychotherapy

Because JDMS usually requires two years or even longer of drug treatments, exercise programs, limitations on some activities, and special attention to diet, children often become angry, depressed, or self-pitying. In some cases they may express resentment toward healthy siblings or classmates. The child's doctor may recommend either individual psychotherapy for the affected child or **family therapy** for the family as a whole.

Alternative treatment

Very little has been published regarding complementary and alternative (CAM) treatments for juvenile dermatomyositis, although some practitioners of traditional Chinese medicine (TCM) have reported success in

treating the fever and heliotrope rash of JDMS with various Chinese herbal formulae.

Nutritional concerns

Children diagnosed with JDMS must eat a regular balanced diet with generous amounts of protein and calcium. The protein is necessary to repair damaged muscle tissue, and the calcium is needed to keep the child's bones strong. Because of the possible side effects of prednisone, however, the diet must be low in salt and sugar. More specifically, the child should not be allowed to eat take-out or fast foods except on rare occasions as a special treat. Parents should consult a professional dietitian or nutritionist to help plan a diet that is appealing to the child as well as healthful.

Prognosis

The prognosis of juvenile dermatomyositis varies but is usually related to the child's age and the severity of the vasculitis associated with the disease. Younger children generally recover more rapidly than adolescents. Most children with JDMS have active symptoms for about two years, although some may be able to do without medications after the first year and others may need drug treatment and physical therapy for many years. Some children recover without any relapses; however, most children with the disease have periods of remission alternating with recurrences of the symptoms.

Most children eventually recover completely from juvenile dermatomyositis; however, some have lifelong stiffness or muscle weakness from the disease. In a very few cases the child may die from complications related to myocarditis or from bowel perforation or lung disease caused by vasculitic ulcers.

Prevention

Because the causes are still unknown, there is no way to prevent juvenile dermatomyositis as of 2004.

Parental concerns

Although JDMS is rarely fatal, it can lead to medical emergencies if the child develops vasculitic ulcers. More commonly, however, the disease has a severe impact on families because of the length and complexity of treatment as well as the possibility of complications from the disease itself and side effects from the medications. Although two years is the average length of acute symptoms, some children are affected for years, even into adulthood. In addition, the unpredictable nature of

KEY TERMS

Calcinosis—A condition in which calcium salts are deposited in various body tissues. In juvenile dermatomyositis, calcinosis usually takes the form of small lumps of calcium compounds deposited in muscles or under the skin.

Contracture—A tightening or shortening of muscles that prevents normal movement of the associated limb or other body part.

Coxsackie virus—A type of enterovirus that may produce a variety of illnesses, including upper respiratory infections, myocarditis, and pericarditis. Coxsackieviruses resemble the virus that causes polio.

Cutaneous—Pertaining to the skin.

Disease-modifying anti-rheumatic drugs (DMARDs)—A group of medications given to treat severe cases of arthritis, JDMS, and other diseases that affect the joints. All DMARDs work by modifying the immune system.

Dysphagia—Difficulty in swallowing.

Heliotrope—A plant (*Heliotropium arborescens*) related to borage that has lavender or deep violet flowers. The characteristic skin rash of JDMS is sometimes called a heliotrope rash because of its reddish-purple color.

Idiopathic—Refers to a disease or condition of unknown origin.

Myocarditis—Inflammation of the heart muscle (myocardium).

Myopathy—Any abnormal condition or disese of muscle tissue, characterized by muscle weakness and wasting.

Myositis—Inflammation of the muscle.

Proximal muscles—The muscles closest to the center of the body.

Remission—A disappearance of a disease and its symptoms. Complete remission means that all disease is gone. Partial remission means that the disease is significantly improved, but residual traces of the disease are still present. A remission may be due to treatment or may be spontaneous.

Rheumatologist—A doctor who specializes in the diagnosis and treatment of disorders affecting the joints and connective tissues of the body.

Vasculopathy—Any disease or disorder that affects the blood vessels.

remissions and recurrences can be difficult to handle. If the child with JDMS has siblings, the parents must balance the needs of the affected child with the needs of healthy siblings. It is not unusual for either the affected child or other children in the family to develop emotional or behavioral problems. The Myositis Association and the **Muscular Dystrophy** Association, which are listed below under Resources, can help parents find support groups in their area for dealing with family members' emotional reactions to the disease as well as other problems of daily living.

With regard to education, most children with JDMS can continue to attend their regular school although they may need special transportation. They should not be isolated from other children.

See also Juvenile arthritis; Myopathies; Vasculitides.

Resources

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Zipitis, C. S., et al. "Treatment Approaches to Juvenile Dermatomyositis." *Expert Opinion on Pharmacotherapy* 5 (July 2004): 1509–15.

ORGANIZATIONS

American Academy of Dermatology (AAD). PO Box 4014, Schaumburg, IL 60168–4014. Web site: <www.aad.org>.

Arthritis Foundation. PO Box 7669, Atlanta, GA 30357–0669. Web site: <www.arthritis.org>.

Muscular Dystrophy Association (MDA). 3300 East Sunrise Drive, Tucson, AZ 85718–3208. Web site: <www.mdausa.org>.

Myositis Association. 1233 20th Street NW, Suite 402, Washington, DC 20036. Web site: <www.myositis.org>.

National Institute of Arthritis and Musculoskeletal and Skin Diseases (NIAMS). 1 AMS Circle, Bethesda, MD 20892–3675. Web site: <www.niams.nih.gov>.

National Institute of Neurological Disorders and Stroke (NINDS). National Institutes of Health. 9000 Rockville Pike, Bethesda, MD 20892. Web site: <www.ninds.nih.gov>.

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Rebecca Frey, PhD

Development tests

Definition

Development tests are tools that are used to help measure a child's developmental progress from infancy through **adolescence**.

Purpose

Every child develops at an individual pace. However, development tests may help to discriminate between normal variations in development among children and early signs of a developmental problem. About 16 percent of children have some form of developmental difficulty or delay, and more than 500,000 American children are assessed for early-intervention programs every year.

Development tests have different purposes depending on the age of the child and may be administered under a variety of circumstances. They are designed according to the expected skills of children at a specific age. The tests range from the passive evaluation of an infant to the complex testing of adolescents.

Development testing begins at birth in order to identify any problems as early as possible and try to correct them. The testing of a newborn can be used to detect neurological problems such as **cerebral palsy**. Testing continues with well-baby visits to the pediatrician. Although there are various schedules for routine well-child visits, the American Academy of Pediatrics recommends visits at the following ages:

- two to four days after birth or discharge from the hospital
- one month
- two months
- four months
- six months
- nine months
- 12 months
- 15 months
- 18 months
- yearly between the ages of two and six
- eight years
- ten years
- yearly until age 21

Many daycare centers and preschools use development tests. Most schools administer school readiness tests before admission. Many states and metropolitan school districts have devised or adapted their own development tests to be administered by schools. Development tests are also used to identify specific social or academic problems.

Developmental assessments usually combine standardized tests and observations to cover all aspects of a child's development, including the following:

- motor skills
- **language development**
- mental development
- social/emotional development
- self-help skills, including dressing and toileting

The types of developmental **assessment** include:

- developmental screening to identify children with special needs or who may be at risk for developmental delays or school difficulty
- diagnostic evaluation, if indicated by the screening, to confirm the presence and extent of a disability
- readiness tests to assess a child's specific skills and information
- observational and performance assessments to provide ongoing information about a child's development

Developmental screening tests usually are brief, general, play-based tests of skills. Screenings include tests administered to the child by an educator or health-care professional and questionnaires for parents or child-care providers that inquire about developmental milestones. Screening tests only try to identify children who may have one or more problems. A screening test is not a diagnosis. Rather it may indicate that a child should be referred for developmental assessment or evaluation.

Developmental evaluations are lengthy, in-depth assessments of a child's skills. They are administered by trained professionals. They provide a profile of a child's strengths and weaknesses in all developmental areas and may be used to determine if the child is in need of an early-intervention and/or treatment program.

Readiness tests measure the extent to which a child has acquired certain skills for successfully undertaking some new learning activity. Although school-readiness tests may concentrate on academic skills, most of them also evaluate other aspects of development.

Description

A multitude of different development tests address every aspect of development at every age. They vary greatly in their reliability or validity (how consistently the test measures what it purports to measure). Many widely used tests have been administered, analyzed, and revised by professionals over a period of years. These, as well as new development tests, undergo frequent examination and review for reliability and validity. Other tests are devised by individuals and sold to parents over the Internet. Parents administer these tests to their child and return them for evaluation.

Some tests use developmental ages to describe a child's physical, perceptual, social, and emotional maturity. Development tests do not necessarily correlate in any way with **intelligence** tests. A standardized development has the following features or functions:

- attempts to obtain a systematic sample of a child's performance under prescribed conditions
- is scored by defined rules
- enables professionals to compare a child's performance to the performance of every other child who takes the same test
- has defined norms (averages, means, or patterns) that are regarded as typical for the specific population being tested

Some development specialists use a standard battery of tests. Others customize tests for the individual child, choosing the most appropriate. A written evaluation of the child is based on the child's test results. Standardized tests for infants and toddlers may be used to assess social, emotional, and intellectual development. Such tests usually consist of presenting a variety of tasks to the child, from very simple to challenging, in order to assess the child's full range of skills.

Test types

Types of development tests include the following:

- infant development scales
- sensory-motor tests
- speech and hearing tests
- neuropsychological tests that measure neurological functioning
- **preschool** psychoeducational batteries
- early screeners
- developmental surveys or profiles
- early learning profiles
- kindergarten or school readiness tests
- tests of **play** behavior
- social skills and social acceptance tests

Sensory-motor tests include general or specific measures of each of the five senses and **gross motor skills** (large muscle movement and control), **fine motor skills** (hand and finger skills), and **hand-eye coordination**. The development of motor skills generally progresses from head to toe. Thus babies usually gain control of their body parts in the following order:

- head and neck at about two months of age
- arms and hands, with grasping at about three months
- trunk, with sitting well by about eight months
- legs and feet, with most children walking by 14 or 15 months

Sensory-motor tests may include:

- visual acuity
- visual perception and organization
- color discrimination or color blindness
- visual-motor skills
- sensory integration
- perceptual skills
- perceptual-motor skills
- motor development
- movement and posture
- manual dexterity

An audiologist may test an infant or young child for signs of **hearing impairment** or loss, usually by transmitting sounds through earphones. Speech and hearing tests measure the mechanics of speaking and hearing the spoken word. These include tests for the following:

- auditory discrimination and comprehension
- speech perception and discrimination
- articulation
- voice fluency
- hearing loss or impairment
- stuttering

Infant/toddler tests

There are countless development tests designed for children from birth to about seven years of age, as well as tests for assessing development in disabled school-age children. The tests measure various skills, including the following:

- gross motor skills
- fine motor skills
- communication
- memory
- number concepts
- letter recognition
- social competence

Development tests are performed at each well-baby visit to the healthcare provider. Children are weighed and measured for height and head circumference. The

results may be plotted on a growth chart that is specific for the child's age and gender (EpicCare Growth Chart) and compares a child's growth to other children of the same age and gender. The pediatrician may check the child's eyesight and hearing and ask various questions about the child's development, including the following areas:

- eating and sleeping patterns
- head control
- alertness
- response to voices
- voice recognition
- eye focusing
- strength and coordination
- posture
- noises
- smiling

THE BRAZELTON TEST In 1973 Harvard University pediatrician T. Berry Brazelton and his colleagues developed the Neonatal Behavioral Assessment Scale (NBAS), commonly known as "the Brazelton." It is based on the assumption that babies are highly capable at birth and communicate through their behavior. Test examiners are trained to support the infant in achieving the best possible scores. The test assumes that infants are born with four major developmental tasks:

- regulation of their autonomic nervous systems, including breathing and temperature regulation
- controlling their motor systems
- controlling their states or levels of consciousness
- social interaction

The NBAS examines a wide range of behaviors in newborns up to two months of age, creating a portrait of the infant's strengths, adaptive responses, possible vulnerabilities, and individuality. It tests 28 behavioral and 18 reflex items, including the following:

- signals that may overtax the infant, such as looking at the mother's face, noise, startles, or color changes
- muscle tone, reflexes, and activity levels
- ability to follow a red ball, a face, or a voice
- ability to control his or her state of consciousness and transitions between states

The examiner tests the infant's response to light, sound, or touch during **sleep** and the infant's ability to block out stimulation during sleep. The states examined are as follows:

- quiet sleep
- active sleep
- drowsy waking
- quiet alert
- fussing
- active crying

CLINICAL NEWBORN BEHAVIORAL ASSESSMENT SCALE (CLNBAS) The Clinical Newborn Behavioral Assessment Scale (CLNBAS), based on the NBAS, was developed in 2004 for use by clinicians. It is conducted in the presence of the parents. It focuses on the baby's individuality and unique adaptive or temperamental style and is designed to develop parent-infant and clinician-family relationships.

Ages and stages questionnaires are used to identify infants and young children who may need further evaluation. These questionnaires are completed by the parent or primary caregiver and are administered at the following ages:

- two-month intervals between the ages of four and 24 months
- three month intervals between the ages of 24 and 36 months
- six-month intervals between the ages of 36 and 60 months

Development tests for infants and toddlers usually include testing for the typical sequence of development that most children go through; for example, most children crawl before they walk and eat with their fingers before using utensils. These tests often are administered by a development assessment specialist, a developmental pediatrician, or an early-childhood special educator. The tests may measure developmental domains that include the following:

- gross motor development
- fine motor development
- language/communication and speech
- relationships to **toys** and other objects, to people, and to the larger world
- emotions
- coping behavior
- self-help skills

ARNOLD L. GESELL TESTS One of the earliest development tests was designed by Arnold L. Gesell, who founded the Clinic of Child Development at Yale University in 1911. By observing and filming infants and

young children, and analyzing their functioning by studying the films frame by frame, Gesell delineated 10 normal stages of early childhood development. Gesell's tests include:

- Gesell Developmental Schedules
- Gesell Child Developmental Age Scale (GCDAS)
- Gesell Preschool Test to measure relative maturity in four basic fields of behavior
- Gesell Developmental Observation to assess a child's developmental age for grade placement and the development of instructional programs

NANCY BAYLEY SCALES OF INFANT DEVELOPMENT Developmental psychologist Nancy Bayley authored the **Bayley Scales of Infant Development** in the mid-twentieth century. Her mental scale evaluates various abilities, yielding a normalized standard score called the Mental Development Index. The Bayley Scales of Mental and Motor Development are used worldwide as standardized measures of infant development at eight months of age. The motor scale assesses:

- the degree of body control
- large-muscle coordination
- fine-motor manipulatory skills
- postural imitation
- motor quality

The behavior rating scale consists of 30 items, which measure the following:

- attention
- arousal
- orientation
- engagement
- emotional regulation
- test-taking behaviors

OTHER TESTS Other infant/toddler development tests include:

- Peabody Developmental Gross Motor Scale for infants
- Bury Infant Check to help identify children with special needs
- Infant Monitoring System for children aged four months to 36 months
- Early Coping Inventory of 48 items on sensory-motor organization, reactive behavior, and self-initiated behavior that are used to assess everyday coping strategies in children between the ages of four and 36 months

Preschool tests

Developmental milestones are widely used tests for development in infants and children of all ages. Milestones in preschoolers evaluate the development of the following skills:

- gross motor skills
- fine motor skills
- language and communication
- emotional and social competency
- thinking, reasoning, and problem solving
- reading and writing
- creativity

The milestone test uses collection forms that are completed by a parent, caregiver, or educator on a monthly basis. The forms cover **family**, friends, and milestones corresponding to a specific area of development. They include a photo or anecdote that illustrates the activity corresponding to the milestone. The forms request that the activities be categorized according to the following descriptors:

- child-initiated
- teacher-initiated
- a new task
- a familiar task
- performed independently
- performed with adult guidance
- performed with peers
- one to five minutes in duration
- five to 15 minutes in duration
- more than 15 minutes in duration

Children who are slow to reach developmental milestones in one area may be ahead of their age in other areas. Sometimes developmental milestones are used as part of an assessment method known as minimum adequate surveillance that combines simple testing with the collection of relevant data.

TYPES OF PRESCHOOL TESTS The Assessment, Evaluation, and Programming System (AEPS) for infants and children often is used to test three to six-year-olds. Using activity stations in a school setting, the test measures skills such as balance, mobility, standing, and walking as well as play skills. The AEPS consists of the following:

- a 98-item family report on functional and social skills used in the child's everyday environment
- 87 cognitive items
- 49 social/communicative items
- 33 social items
- 39 adaptive items
- eight gross motor items
- four fine motor items

Other development tests that frequently are administered to preschoolers include the following:

- Griffith's mental development scales, which measure gross motor skills, personal-social development, hand and eye coordination, and performance, providing a general developmental quotient (GDQ) and separate subquotients (DQs) for each area of development
- Mullen Scales of Early Learning (MSEL), a comprehensive assessment of language, motor, and perceptual abilities in children from birth to five years, eight months of age
- Vineland Social-Emotional Early Childhood Scales

Screening tests

The Denver Developmental Screening Test (DDST) is a widely-used test of motor, language, speech, and interpersonal skills for children from birth to six years of age. It is used by physicians in well-baby visits and may include parental questionnaires.

Other screening tests include the following:

- Infant Developmental Screening Scale
- ages and stages questionnaires
- Battelle Developmental Inventory (BDI) screening test for screening, preliminary assessment, and/or initial identification of possible developmental strengths and weaknesses
- Brigance Early Preschool Screen for two-year-old and two-and-a-half-year-old children
- Brigance K and 1 Screen for kindergarten and first-grade children
- Miller FirstSTEP Screening Test for evaluating preschoolers
- Preschool Development Inventory, a brief screening inventory to help identify children with developmental, behavioral, or health problems
- Children at Risk Screener: preschool and kindergarten
- Early Screening Inventory for children aged three to six
- Howell Prekindergarten Screening Test
- Humanics National Child Assessment Form
- Kindergarten Screening Inventory
- Milani-Comparetti Motor Development Screening Test

Readiness tests

Readiness tests include the following:

- ABC Inventory to Determine Kindergarten and School Readiness
- Developmental Tasks for Kindergarten Readiness
- Kindergarten Readiness Test
- Phelps Kindergarten Readiness Scale
- Pediatric Examination of Educational Readiness at Middle Childhood

Communication tests

Communication development tests include the following:

- Sequenced Inventory of Communication Development for testing various early **communication skills** and assigning a communication age
- Assessing Semantic Skills Through Everyday Themes for preschool and early-elementary-aged children
- Expressive One-Word Picture Vocabulary Test to measure a child's ability to verbally label objects and people; a standardized test that yields age equivalents, standard scores, scaled scores, and percentile ranks
- Receptive One-Word Picture Vocabulary Test to provide information about a child's ability to understand language; a standardized test that yields age equivalents, standard scores, scaled scores, and percentile ranks
- Early Language Milestone (ELM) Scale to measure language development in children from birth to three years of age
- Peabody Picture Vocabulary Test, called "the Peabody," a short test that measures vocabulary in two-and-a-half to four-year-olds; sometimes used for screening

Speech and hearing tests include the following:

- Early Speech Perception Test
- Joliet 3-Minute Preschool Speech and Language Screen

- Fluharty Preschool Speech and Language Screening Test to identify preschool children who may warrant a comprehensive communication evaluation
- Assessing Linguistic Behaviors, Assessing Prelinguistic and Early Linguistic Behaviors in Developmentally Young Children
- Assessment of Fluency in School-Age Children
- Children's Articulation Test
- Clinical Evaluation of Language Fundamentals
- Phonological Assessment of Child Speech

Intelligence tests

Intelligence tests attempt to measure a child's ability to learn. Many such tests generally measure what a child has already learned. Results of these tests may be represented on a scale, as a mental age, or as an intelligence quotient (IQ). Some research indicates that IQ tests for children aged 18 to 20 months are not good indicators of future or even current abilities and that they usually measure motor rather than mental skills. However, other research has indicated that IQ tests in children as young as six months are good predictors of school IQ test results years later.

Frequently used intelligence tests include the following:

- Wechsler Preschool and Primary Scale of Intelligence for children aged three to seven years with norms for 17 age groups at three-month intervals
- Wechsler Intelligence Scale for Children
- Differential Ability Scales (DAS), for measuring overall cognitive ability and specific abilities in children aged two years and six months to seventeen years and eleven months
- Stanford-Binet Intelligence Scale for ages two years to adult, for scoring verbal reasoning, abstract and visual reasoning, quantitative reasoning, and short-term memory and yielding a composite score with a mean of 100

The Columbia Mental Maturity Scale (CMMS) does not depend on reading skills. The child makes perceptual discriminations involving color, shape, size, use, number, missing parts, and symbolic material. It appears to measure general reasoning ability, although there is some evidence that it is more a test of the ability to form and use concepts than a test of general intelligence. It provides standard age deviation scores for chronological ages between three years and six months and nine years and eleven months. The Maturity Index indicates the age group of the child in terms of test performance.

The Test of Nonverbal Intelligence is a quick, language-free measure of cognitive ability in children aged five years and older. It contains 55 problem-solving tasks of progressively increasing difficulty. It often is used for assessing children with speech, language, or hearing impairments, academic handicaps, or brain impairments, and for children who do not speak English. The Merrill-Palmer Scale of Mental Tests (MPSMT) is widely used as a nonverbal test for assessing visual-spatial skills in children aged one year and six months to six years.

Precautions

Designing child development tests presents numerous difficulties that may affect the results, including the following:

- motivating the child
- keeping the child's attention
- not discriminating among children

Some healthcare and education professionals are concerned that children are being over-tested, over-screened, and overanalyzed. Some experts question the use of conventional early childhood development tests for the following reasons:

- Young children are inexperienced at test-taking.
- High-stakes tests, such as those that influence major educational decisions, can have long-term negative consequences for children.
- Tests may be too narrow or one-dimensional.
- Teaching and learning may be negatively affected because of a focus on test results.
- There are major differences in the learning opportunities available to children.
- Sometimes tests are used for purposes other than those for which they were designed.

Preparation

The parent, the person who knows the child best, should participate in development tests as much as possible. Tests should be explained to parents who can then explain them to their children in terms that they will understand. The parents' feelings and personal observations should be considered when evaluating a child's development.

Risks

The major risk of development tests is that some children may be labeled in inappropriate ways because

KEY TERMS

Developmental assessment—The ongoing process of testing, observing, and analyzing a child's skills.

Developmental domains—Areas of a child's development.

Diagnostic testing—Testing performed to determine if a person has a particular disease.

Intelligence test—A questionnaire or series of exercises designed to attempt to measure intelligence.

Readiness test—A test that measures the extent of a child's acquired skills for successfully undertaking a new learning activity such as kindergarten.

Screening—A process through which carriers of a trait may be identified within a population.

Standardized test—A test that follows a regimented structure, and each individual's scores may be compared with those of groups of people. In the case of the Cognistat, test taker's scores can be compared to groups of young adults, middle-aged adults, the geriatric, and people who have undergone neurosurgery.

of their test results. Development tests are not infallible. Developmental screenings may over- or under-identify children with developmental delays. For example, some children who under-perform on a school readiness test go on to perform very well in school. Development tests may become outdated because of new, improved methods. Other tests may be found to be unreliable. Development tests must keep pace with demographic changes in the United States, in order to meet the needs, for example, of large numbers of young children who speak languages other than English.

Normal results

The range of normal development is very large. No two children develop at exactly the same rate. Children reach developmental milestones on their individual schedules and at their own pace. Some children completely skip developmental milestones such as rolling over or **crawling**. Although some children begin walking at eight months, others do not walk until 18 months; both are within the normal range. Children also may regress periodically. For example, children who sleep through the night may begin waking up often as they learn to talk.

Parental concerns

Although development tests often are used to reassure parents that their child is normal, many parents may feel anxious or defensive when told that their child should be tested. Parents should try to become active participants in their child's testing and understand the testing process and terminology. Increasingly parents and other family members are joining healthcare professionals and educators in administering development tests.

ZERO TO THREE: National Center for Infants, Toddlers, and Families recommends the following:

- Young children should not be separated from their parent or caregiver during testing since separation can cause anxiety.
- Children should not be assessed by a person they have just met for the first time.
- Tests should not be limited to easily measured factors such as motor or cognitive skills.
- Normative tests or milestone scales should not be the major basis for the developmental assessment of infants and young children.

Development tests can provide parents with a better understanding of their child's development and any possible need for intervention. Parents should feel free to disagree with test results and participate in further discussions concerning their child's development.

When to call the doctor

A pediatrician should be consulted if any of the following occurs:

- an infant's growth or development seems abnormal
- an infant or child seems to be losing developmental milestones

Resources

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ORGANIZATIONS

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The Brazelton Institute. Children's Hospital Boston, 1295 Boylston Street, Suite 320, Boston, MA 02215. Web site: <www.brazelton-institute.com>.

Buros Center for Testing. University of Nebraska-Lincoln, Lincoln, NE 68588. Web site: <www.unl.edu/buros/index.html>.

Child Development Institute. 3528 E. Ridgeway Road, Orange, CA 92867. Web site: <ww.cdipage.com/index.htm>.

Children's Institute. 27 N. Goodman St., Suite D103, Rochester, NY 14607. Web site: <www.childrensinstitute.net>.

ZERO TO THREE: National Center for Infants, Toddlers and Families. 2000 M Street NW, Suite 200, Washington, DC 20036. Web site: <www.zerotothree.org>.

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Developmental delay

Definition

A developmental delay is any significant lag in a child's physical, cognitive, behavioral, emotional, or social development, in comparison with norms.

Description

Developmental delay refers to when a child's development lags behind established normal ranges for his or her age. Sometimes the term is used for **mental retardation**, which is not a delay in development but rather a permanent limitation. If most children crawl by eight months of age and walk by the middle of the second year, then a child five or six months behind schedule in reaching these milestones may be classified as developmentally delayed regarding mobility.

At least 8 percent of all children from birth to six years have developmental problems and delays in one or more areas of development. Some have global delays, which means they lag in all developmental areas.

Doctors try to locate the source of the delay and then design a treatment plan. When the cause of a child's delay is identified, the pediatrician and **family** know better what to expect, and the child can begin to receive appropriate treatment and support. If the problem is a genetic disorder, then parents may seek genetic counseling regarding their decision on having additional children.

The doctor's **assessment** has various components. The following are some of them:

- Developmental assessment: The physician's review of a child's current competencies (including knowledge, skills, and personality), and consideration of the best ways to help the child develop further.
- Family assessment: Interpretation of a child's development from family members, as well as their ideas about priorities and concerns about the child's future development.
- Multidisciplinary assessment: The assessment by a group of professionals who work with the child and family, directly or indirectly. The assessment interprets different phases of a child's development and types of behavior and skills.
- Play-based assessment: This assessment involves observation of the child playing alone, with peers, or with parents or other familiar caregivers, in free **play** or in special games. Play provides a diagnostic framework within which children show abilities, feelings, learning style, and social skills in groups.

Infancy

Infants who have medical problems at birth have an increased chance of developmental difficulties. High-risk infants should be in a follow-up program to track their progress because of an increased likelihood of

developmental problems that may appear gradually in the first years of life.

Most children begin to speak their first words before they are 18 months old, and by age three the vast majority of children speak short sentences. Therefore, any child who is not speaking words or sentences by the third birthday may be developmental delayed.

Toddlerhood

Between the ages of 12 and 30 months, a child begins to strike out independently from a secure base of trust set up with the primary caregiver during the first year. As toddlers learn to walk, there is access to new territory. Boundless energy and insatiable curiosity drives the child to explore the environment and master new skills. Increased motor skills, immaturity, and lack of experience also place the toddler at risk for accidental injury. Children with developmental delays may tend to be more reserved and less adventuresome. They may tend not to explore their environment or take risks in it.

The healthy toddler years are characterized by the struggle for autonomy as the child develops a sense of personhood separate from the parent. Toddlers' egocentric and demanding behavior, often marked by temper **tantrums** and negativism, has given this period a negative reputation. However, toddlers who do not evince this challenging behavior may be delayed. Dramatic growth of language and cognitive skills during the second year enables the healthy toddler to think and solve problems for the first time. For the child who is not progressing in language skills, developmental delays are readily identifiable.

Preschool

The **preschool** period, from age three to five years, is a time of relative tranquility after the tumultuous toddler period. The healthy preschooler becomes increasingly independent, mastering many motor skills and developing greater social and emotional maturity. The preschooler is imaginative, creative, and curious. The developmentally delayed preschooler may act more egocentrically and show more signs of demanding behavior.

School age

Children from six to 12 years of age experience slow, steady physical growth and rapid cognitive and social development. The school-age child develops a sense of industry and learns the basic skills needed to be comfortable in society. The child develops appreciation of rules and a conscience that influences compliance and affects disobedience. Cognitively, the child grows from egocentrism in early childhood to more mature thinking. This maturity supports the ability to solve problems and make reasonably independent decisions. Competence and **self-esteem** increase with each academic, social, and athletic achievement. The relative stability and security of the school-age period prepare the child to manage the challenges of **adolescence**. However, the developmentally delayed child might not evince this growing competence.

Common problems

Although there are several areas of developmental areas, this article is restricted to global delay, delay in speech and language, motor and fine motor delays, and personal and social developmental delays.

Global developmental delay

Children with a diagnosis of mental retardation often have mixed or global development delays. However, low IQ may or may not be causally related to the delays. Two to three children out of every 100 have a mental handicap, and those with IQs of 55 or lower may have a physiological reason for their delay. Some children experience global developmental delay due to chromosomal abnormalities such as **Down syndrome** or **fragile X syndrome**. Global delays also are common in children with **fetal alcohol syndrome**.

Speech and language delay

Speech and language developmental delays are often prevalent in children with developmental disabilities. Eleven percent of toddlers have a speech and language problem. Expressive **language delay** is the most common developmental presentation. The social and educational development of children with delayed speech and language may be significantly disruptive (even in mild delays), so early identification and intervention is essential. Clinical diagnosis of delayed speech and language in children also considers hearing loss and **autism**, among other possible causes.

All children with delayed speech and language should have an audiometric assessment. Congenital sensorineural hearing loss (most common birth deficiency, affecting roughly two to four per 1,000 children) may cause delayed speech and language. Hearing problems occur often in newborns and in a higher number of babies who are in a neonatal intensive care unit. Universal newborn hearing screening programs should help in determining hearing acuity.

Hearing loss

Approximately half of all preschool children have varying hearing loss from **otitis media**. Language skills are affected by hearing loss, and more than one third of children with unilateral deafness fail one or more school grades. In general, children with the greatest hearing loss have the greatest language deficits. The earlier the hearing loss is identified, the better the outcome.

Autism

Children with delayed speech and language should be evaluated for cognitive disabilities. There is a close association among social and affective abilities and cognitive, sensory, and **language development**. Children who are unable to communicate effectively may have problems interacting verbally with their peers. Because social and pragmatic deficits are core characteristics of autism, it is important to look for dissociation among language, social adaptive skills, and motor behavior. Autism is a common disorder, occurring once in 500 children. It is one of the most complex neurodevelopment disorders. Children with autism have significant communication impairment.

In order to be eligible for programs and services for autism, a student must have delayed or abnormal functioning in at least one of three areas with onset before the age of three. These children have difficulty communicating and lack the ability to connect with peers. There may be a delay or total lack of language or the use of repetitive and idiosyncratic language. Other behaviors can include preoccupation with parts of objects, hand or finger flapping, and rocking.

Pervasive developmental disorder is two to three times more common than autism but less severe. When behaviors resembling autistic disorders are present with abnormalities of speech and language development, other syndromes and disorders are considered, for example, Asperger disorder, childhood disintegrative disorder, and Rett syndrome.

Musculature dysfunction

Oral motor dysfunction of the speech-producing musculature (in which children have dysarthria, or mechanical difficulties in speaking) is present in children with **cerebral palsy** and other conditions. The dysfunction leads to uncoordinated oral musculature.

Verbal difficulties

Verbal learning disability is often associated with speech and language problems in preschool children. Children with a specific learning disability, like children with severe mental retardation or autism, may present with dissociation in developmental skills. For example, language may be more delayed than motor skills. Also, lack of academic success at school can reflect dissociation between academic achievement and general intellectual abilities. Delays in language and cognitive areas may suggest a neurodevelopment diagnosis that presents as a nonverbal learning disability. In such cases, a child may have impaired visual-spatial perceptual abilities.

It is helpful to consider a child's expressive and receptive language skills. Children with an ability to understand are more likely to improve than children with expressive and receptive delays. Children whose primary difficulty involves receptive language are more likely to have developmental cognitive disability or autism spectrum disorder. Neurological problems may also be present when a child's head circumference is increasing either too fast or too slowly. Although physical and cognitive delays may occur together, one is not necessarily a sign of the other.

Neurological or medical conditions

In developmental language disorder, impaired language cannot be attributed to a neurological or general medical condition. There is a slow rate of language development, in which speech begins late and advances slowly. Children with developmental language disorder have an inconsistency between their cognitive functioning (nonverbal or performance measures) and their language skills. Different patterns of language impairment in developmental **language disorders** have distinct profiles of linguistic strengths and weaknesses. Developmental dysphasia may be the problem with some of these children. There are many reasons for a developmental language disorder, which occurs in about 10 percent of the population.

An unusual cause of acquired language disorder is an epileptic syndrome called Landau-Kleffner syndrome. Children with Landau-Kleffner develop typical language skills, which then deteriorate. The characteristics of this condition may be confused with autism.

Motor delay

Physician referrals of motor delay are most common during the first six to 18 months of a child's life. By evaluating a child's developmental profile, a doctor may develop a differential diagnosis.

Early motor delays are often a sign of neurological dysfunction. When a child has primarily motor delays, conditions such as cerebral palsy, ataxia, **spina bifida**, **spinal muscular atrophy** (withering) and myopathy may be present. If there is no motor delay, a child does

not have cerebral palsy. When a motor delay exists with delays in other developmental areas, the child should be examined for visual impairment or mental handicap.

Older children with poor motor skills may have a developmental coordination disorder in which their motor skills are substantially below their cognitive abilities. Their clumsiness may link with a learning disability or attention-deficit hyperactivity disorder. Children with Asperger disorder are often clumsy; their neuropsychological profiles display significantly stronger verbal skills than nonverbal abilities.

HYPOTONIA **Hypotonia** is the most common symptom of motor dysfunction in newborns and infants. The child's developmental assessment should include the quality of the pregnancy, including the onset and vitality of fetal movements and problems during labor and delivery. The child's presentation in the neonatal period should be described, with special attention to the family history to document the potential for genetic disorder.

The key to diagnosing a hypotonic infant is a neurodevelopment examination. The challenge in correctly diagnosing a "floppy" child lies in distinguishing between central and neuromuscular hypotonia. A hypotonic infant who is not weak has low tone because of a central nervous disorder. Weakness strongly implies neuromuscular involvement. Normal or increased deep tendon reflexes suggest central hypotonia.

FINE-MOTOR ADAPTIVE DELAY If there is a delay in fine-motor adaptive development combined with delays in other developmental domains, the doctor will consider whether the child is visually impaired or mentally handicapped. It is important to assess the eyes and visual acuity of a child presenting with delayed fine-motor adaptive development.

If the delay occurs mainly in one developmental area, the child may have hemiplegia, a brachial plexus injury, such as Erb's or Klumple's palsy, or a broken clavicle. All symmetries of movement in the first two or three years should be watched.

In older preschool or elementary school children with fine-motor delays, developmental coordination disorder or a disorder of written expression may be causal. Developmental coordination disorder presents in about 6 percent of all children. It is often associated with attention deficit hyperactivity disorder or a learning disability.

PERSONAL AND SOCIAL DELAY When a child presents with personal and social delays, the doctor will consider whether the child has developmental cognitive disability, has autism, or is living in an environment of abuse, neglect, or deprivation.

KEY TERMS

Accommodation—The ability of the lens to change its focus from distant to near objects and vice versa. It is achieved through the action of the ciliary muscles that change the shape of the lens.

Assessment—In the context of psychological assessment (a structured interview), assessment is information-gathering to diagnose a mental disorder.

Child development specialist—A professional who is trained in infant and toddler development and in the tools used to identify developmental delays and disabilities.

Development—The process whereby undifferentiated embryotic cells replicate and differentiate into limbs, organ systems, and other body components of the fetus.

Developmental milestone—The age at which an infant or toddler normally develops a particular skill. For example, by nine months, a child should be able to grasp and toss a bottle.

Disability—An inability to do something others can do; sometimes referred to as handicap or impairment.

Hypotonia—Having reduced or diminished muscle tone or strength.

Motor skills—Controlled movements of muscle groups. Fine motor skills involve tasks that require dexterity of small muscles, such as buttoning a shirt. Tasks such as walking or throwing a ball involve the use of gross motor skills.

Parental concerns

Many doctors routinely include developmental screening in physical examinations. Parents concerned about any of their child's development should seek the opinion of their pediatrician.

See also Cognitive development; Emotional development; Fine motor skills.

Resources

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Developmental reading disorder *see* **Dyslexia**

Dextromethorphan *see* **Cough suppressants**

Diabetes mellitus

Definition

Diabetes mellitus is a chronic disease in which the body is not able to correctly process glucose for cell energy due to either an insufficient amount of the hormone insulin or a physical resistance to the insulin the body does produce. Without proper treatment through medication and/or lifestyle changes, the high blood glucose (or blood sugar) levels caused by diabetes can cause long-term damage to organ systems throughout the body.

Description

There are three types of diabetes mellitus: type 1 (also called juvenile diabetes or insulin-dependent diabetes), type 2 (also called adult-onset diabetes), and gestational diabetes. While type 2 is the most prevalent, consisting of 90 to 95 percent of diabetes patients in the United States, type 1 diabetes is more common in children. Gestational diabetes occurs in pregnancy and resolves at birth.

Every cell in the human body needs energy in order to function. The body's primary energy source is glucose, a simple sugar resulting from the digestion of foods containing carbohydrates (primarily sugars and starches). Glucose from the digested food circulates in the blood as a ready energy source for any cells that need it. However, glucose requires insulin in order to be processed for cellular energy.

Insulin is a hormone or chemical produced by cells in the pancreas, an organ located behind the stomach. Insulin bonds to a receptor site on the outside of a cell. It acts like a key to open a doorway into the cell through which glucose can enter. When there is not enough insulin produced (as is the case with type 1 diabetes) or when the doorway no longer recognizes the insulin key (which happens in type 2 and gestational diabetes), glucose stays in the bloodstream rather entering the cells. The high blood glucose, or blood sugar, levels that result are known as **hyperglycemia**.

Type 1 diabetes

Type 1 diabetes occurs when the beta cells of the pancreas are damaged and stop producing the hormone insulin. While the exact cause of this cell damage is not completely understood, it is thought to be a combination of environmental and autoimmune factors. Despite the name juvenile diabetes, type 1 diabetes can be diagnosed at any stage of life, although diagnosis in childhood through young adulthood is most common.

Children who develop type 1 diabetes must eventually take regular insulin injections to keep blood glucose levels under control and do the job of the pancreas. Regular home testing of blood sugar levels is also important to make sure that the treatment is working effectively and to avoid a diabetic emergency such as **hypoglycemia** (low blood sugar) or hyperglycemia (high blood sugar).

Type 2 diabetes

The hallmark characteristic of type 2 diabetes is insulin resistance. The pancreas typically produces enough insulin (often too much insulin); however, cells are resistant to the insulin and it may not work as effectively. Type 2 is the most common form of diabetes, and most individuals with the disease are adults. However, children and adolescents can develop type 2 diabetes too,

particularly if they are overweight and have a history of type 2 diabetes in their **family**.

Type 2 diabetes is treated with diet, **exercise**, and in some cases, oral medication and/or insulin. Self-monitoring of blood glucose levels is also important to assess how well treatment is working.

Demographics

An estimated 18.2 million Americans live with diabetes, and over 5 million of those remain undiagnosed. Up to 95 percent of diabetes patients in the United States have type 2 diabetes; the vast majority of Americans with diabetes are over 20 years of age. Those under 20 represent only 206,000 of the total cases of diabetes in the United States.

While type 2 diabetes is a growing problem among American youth due to climbing **obesity** rates and more sedentary lifestyles, type 1 diabetes is more prevalent in children and adolescents. An estimated one in 400 to 500 children have type 1 diabetes.

The American Diabetes Association reports that in 2002, diabetes cost Americans an estimated $132 billion in direct medical costs and indirect expenses such as lost productivity and disability payments.

Causes and symptoms

The causes of diabetes are not completely understood; however, there seem to be both genetic and environmental factors involved in the development of both type 1 and type 2 diabetes, meaning that a person may have a genetic predisposition to developing diabetes, but it takes an environmental factor such as a viral infection or excessive weight gain to actually make the disease surface.

Research has shown that some people who develop diabetes have common genetic markers. In type 1 diabetes, the immune system, the body's defense system against infection, is believed to be triggered by a virus or another microorganism that causes an autoimmune reaction that eventually destroys the insulin-producing cells (i.e., beta cells) in the pancreas. Up to 90 percent of cases of type 1 diabetes are the autoimmune subtype, sometimes called type 1A or immune-mediated diabetes.

The other subtype of type 1 diabetes is called idiopathic, or type 1B diabetes. People who have idiopathic type 1 diabetes also experience beta cell destruction, but it is due to a chromosomal abnormality or an unknown cause rather than any autoimmune process. Only tests for islet cell antibodies and other autoimmune markers can differentiate between the two subtypes, and because testing can be costly and treatment for both is the same (i.e., insulin), a physician may not necessarily order tests for autoimmunity.

Finally, damage caused by diseases of the pancreas (such as pancreatitis), endocrine disorders (e.g., endocrine tumors), and drugs or toxins can also destroy beta cell function.

In type 2 diabetes, family history, age, weight, activity level, and ethnic background can all play a role in the genesis of the disease. Individuals who are at high risk of developing type 2 diabetes mellitus include the following groups:

- people who are overweight or obese (more than 20 percent above their ideal body weight)
- people who have a parent or sibling with type 2 diabetes
- those who belong to a high-risk ethnic population (African-American, Native American, Asian-American, Hispanic, or Pacific Islander)
- people who live a sedentary lifestyle (i.e., exercise less than three times a week)
- women who have been diagnosed with gestational diabetes or have delivered a baby weighing more than 9 lbs (4 kg)
- people with high blood pressure (140/90 mmHg or above)
- people with high density lipoprotein cholesterol (HDL, or "good" cholesterol) level less than or equal to 35 mg/dl and/or a triglyceride level greater than or equal to 250 mg/dl

Several common medications can cause chronic high blood sugar levels and/or promote insulin resistance. These include atypical antipsychotics, beta blockers, corticosteroids, diuretics, estrogens, lithium, protease inhibitors, niacin, and some thyroid preparations.

Both type 1 and type 2 diabetes share similar symptoms caused by chronically high blood glucose levels.

Symptoms of both type 1 and type 2 diabetes include:

- excessive thirst
- frequent urination
- weight loss
- increased appetite
- unexplained fatigue
- slow healing cuts, **bruises**, and **wounds**

- frequent or lingering infections (e.g., urinary tract infection)
- mood swings and irritability
- blurred vision
- headache
- high blood pressure
- dry and itchy skin
- tingling, **numbness**, or burning in hands or feet

Symptoms of diabetes can develop suddenly (over days or weeks) in previously healthy children or adolescents, or can develop gradually, particularly in the case of type 2 diabetes.

Children and adolescents sometimes develop a condition known as diabetic ketoacidosis (DKA) at the time of their diagnosis. Ketones are acid compounds that form in the blood when the body breaks down fats and proteins for energy. When blood sugars are high (i.e., over 249 mg/dl, or 13.8 mmol/L) for prolonged periods of time, ketones build up in the bloodstream to dangerous levels. Symptoms of DKA include abdominal **pain**, excessive thirst, **nausea and vomiting**, rapid breathing, extreme lethargy, and drowsiness. Patients with ketoacidosis will also have a fruity or sweet breath odor. Left untreated, this condition can lead to coma and has the potential to be fatal. DKA is more common in people with type 1 diabetes, although it can occur in type 2 diabetes as well.

Symptoms of type 2 diabetes can begin so gradually that a person may not know that he or she has it. It is not unusual for type 2 diabetes to be detected while a patient is seeing a doctor about another health concern that is actually being caused by the yet undiagnosed diabetes, such as heart disease, chronic infections (e.g., urinary tract infections, yeast infections), blurred vision, numbness in the feet and legs, or slow-healing wounds.

When to call the doctor

If left untreated, diabetes is a life-threatening condition. Any child displaying symptoms of diabetes should be taken to a doctor or emergency care facility for evaluation immediately.

Diagnosis

Diagnosis of diabetes is suspected based on symptoms and confirmed by blood tests that measure the level of glucose in blood plasma. Dipstick or reagent test strips that measure glucose in the urine can only detect glucose levels above 180 mg/dl and are non-specific, so they are not useful in the diagnosis of diabetes. However, they are a non-invasive way to obtain a fast and simple reading that a physician might use as a basis for ordering further diagnostic blood tests for diabetes, particularly in children.

Blood tests are the gold standard for the diagnosis of both type 1 and type 2 diabetes in children and adults. The American Diabetes Association recommends that a random plasma glucose, fasting plasma glucose, or oral glucose tolerance test (OGTT) be used for diagnosis of diabetes. The OGTT is commonly used as a screening measure for gestational diabetes. Fasting plasma glucose is the test of choice unless a child is exhibiting classic symptoms of diabetes, in which case a random (or casual) plasma glucose test is acceptable.

Unless hyperglycemia is obvious (e.g., blood glucose levels are extremely high or the child experiences DKA), the fasting or random plasma glucose test should be confirmed on a subsequent day with a repeat test.

Fasting plasma glucose test

Blood is drawn from a vein in the child's arm following an eight-hour fast (i.e., no food or drink), usually in the morning before breakfast. The red blood cells are separated from the sample and the amount of glucose is measured in the remaining plasma. A fasting plasma glucose level of 126 mg/dl (7.0 mmol/l) or higher indicates diabetes (with a confirming retest on a subsequent day).

Random plasma glucose test

Blood is drawn at any time of day, regardless of whether the patient has eaten. A random plasma glucose concentration of 200 mg/dl (11.1 mmol/l) or higher in the presence of symptoms indicates diabetes.

Oral glucose tolerance test

Blood samples are taken both before and several times after a patient drinks 75 grams of a glucose-based beverage. If plasma glucose levels taken two hours after the glucose drink is consumed are 200 mg/dl (11.1 mmol/L) or higher, the test is diagnostic of diabetes (and should be confirmed on a subsequent day if possible).

Although the same diagnostic blood tests are used for both types of diabetes, whether a child is diagnosed as type 1 or type 2 can typically be determined based on her personal and medical history. The majority of children diagnosed in childhood are type 1, but if blood test results indicate prediabetes and a child is significantly overweight and has a history of type 2 diabetes in her family, type 2 is a possibility.

Further blood tests can help to differentiate between type 1 and type 2 when the diagnosis is unclear. One of

these is an assessment of c-peptide levels, a protein released along with insulin that can help a physician determine whether or not a patient is producing sufficient amounts of insulin. The other is a GAD (Glutamic Acid Decarboxylase) autoantibody test. The presence of GAD autoantibodies may indicate the beginning of the autoimmune process that destroys pancreatic beta cells.

Treatment

Children with type 1 diabetes must take insulin injections or infusions. Their dosage needs may change over time. Sometimes children will experience a decreased need for insulin once blood sugars are brought under control following diagnosis. Their insulin needs may go down, and in some cases, they can stop taking injections for a time. This phenomenon, known as the honeymoon period, can last anywhere from a few days to months.

Children with diabetes and their parents should learn to operate a home blood glucose monitor. Home testing can prevent dangerous highs and lows and help parents and children understand how food and exercise impact blood sugar levels. Blood glucose levels taken before meals are also used to calculate dose size of insulin. A small needle or lancet is used to prick the finger or alternate site and a drop of blood is collected on a test strip that is inserted into a monitor. The monitor then calculates and displays the blood glucose reading on a screen. Although individual blood glucose targets should be determined by a medical professional in light of a child's medical history, the general goal is to keep them as close to normal (i.e., 90 to 130 mg/dl or 5 to 7.2 mmol/L before meals) as possible.

Insulin

Children with type 1 diabetes need daily injections of insulin to help their bodies use glucose. The amount and type of insulin required depends on the height, weight, age, food intake, and activity level of the individual diabetic patient. Some patients with type 2 diabetes may also need to use insulin injections if their diabetes cannot be controlled with diet, exercise, and oral medication. Injections are given subcutaneously, that is, just under the skin, using a small needle and syringe, an insulin pen injector, an insulin infusion pump, or a jet injector device. Injection sites can be anywhere on the body where there is a layer of fat available, including the upper arm, abdomen, or upper thigh.

Insulin may be given as an injection of a single dose of one type of insulin once a day, or different types of insulin can be mixed and given in one dose or split into two or more doses during a day. Patients who require multiple injections over the course of a day may be able to use an insulin pump that administers small doses of insulin on demand. The small battery-operated pump is worn outside the body and is connected to a cannula (a thin, flexible plastic tube) that is inserted into the abdomen called an insertion set. Pumps are programmed to infuse a small, steady infusion of insulin (called a basal dose) throughout the day, and larger doses (called boluses) before meals. Because of the basal infusion, pumps can offer many children much tighter control over their blood glucose levels and more flexibility with their diet than insulin shots afford them.

Regular insulin is fast-acting and starts to work within 15 to 30 minutes, with its peak glucose-lowering effect about two hours after it is injected. Its effects last for about four to six hours. NPH (neutral protamine Hagedorn) and Lente insulin are intermediate-acting, starting to work within one to three hours and lasting up to 18 to 26 hours. Ultra-lente is a long-acting form of insulin that starts to work within four to eight hours and lasts 28 to 36 hours. Peakless, or basal-action insulin (insulin glargine, or Lantus) starts working in 15 minutes and has a duration of between 18 and 26 hours.

Nutritional concerns

Because dietary carbohydrates are the primary source of glucose for the body (the other source being the liver), it is very important that children with diabetes learn to read labels and be aware of the amount of carbohydrates in the foods they eat. Children and their parents are usually advised to consult a registered dietitian (RD) to create an individualized, easy to manage food plan that fits their family's health and lifestyle needs. A well-balanced, nutritious diet provides approximately 50 to 60 percent of calories from carbohydrates, approximately 10 to 20 percent of calories from protein, and less than 30 percent of calories from fat. The number of calories required depends on age, weight, and activity level. An RD can also teach the family how to use either the dietary exchange lists or carbohydrate counting system to monitor food intake.

Each food exchange contains a known amount of calories in the form of protein, fat, or carbohydrate. A patient's diet plan will consist of a certain number of exchanges from each food category (meat or protein, fruits, breads and starches, vegetables, and fats) to be eaten at meal times and as snacks. Patients have flexibility in choosing which foods they eat as long as they stick with the number of exchanges prescribed by their RD based on their caloric requirements.

Carbohydrate counting involves totaling the grams of carbohydrates in the foods your child eats to ensure the child does not exceed her goal for the day. In the simple-carb counting method, one carbohydrate choice or unit equals 15 grams of carbohydrates (which is equivalent to one starch or fruit exchange in the exchange method). The number of carb choices allowed daily is based on caloric requirements.

Children with type 1 diabetes who use fast-acting insulin before meals may find that carb counting gives them tighter control of their blood glucose levels, since they can compute the number of insulin units based on both their carbohydrate intake (called the carbohydrate to insulin ratio) and before-meal blood glucose readings.

Dietary changes and moderate exercise are usually the first treatments implemented in type 2 diabetes. Weight loss may be an important goal in helping overweight children and adolescents control their blood sugar levels. Exercise helps keep blood glucose levels down and has other health benefits, as well.

Oral medications

Children with type 2 diabetes may be prescribed oral medications if they are unable to keep their blood glucose levels under control with dietary and exercise measures. As of 2004, metformin was the only oral medication approved by the U.S. FDA for use in children over age ten. Metformin (trade name Glucophage) is in the biguanide class of drugs and works by reducing the amount of glucose the liver produces and the amount of circulating insulin in the body. Other adult type 2 diabetes medications, such as sulfonylureas and meglitinide drugs, which work by increasing insulin production, may be prescribed off-label for pediatric use.

Transplants

Transplantation of a healthy pancreas into a patient with type 1 diabetes can eliminate the need for insulin injections; however, this transplant is typically done only if a kidney transplant is performed at the same time. Although a pancreas transplant is possible, it is not clear if the potential benefits outweigh the risks of the surgery and life-long drug therapy needed to prevent organ rejection, particularly in the case of children.

A second type of transplant procedure, as of 2004 in experimental clinical trials and not available to children, is an islet cell transplant. In this type of treatment, insulin-producing islet cells are harvested from a donor pancreas and injected into the liver of a recipient, where they attach to new blood vessels and (ideally) begin producing insulin. A lifetime regimen of immunosuppressive drugs is required to prevent rejection of the transplanted cells.

Prognosis

As of 2004 diabetes is a chronic and incurable disease. While stem cell research holds great promise for future therapies and potential cures, as of the early 2000s the best hope for keeping children well with diabetes and avoiding long-term complications is maintaining good blood glucose control. The landmark Diabetes Control and Complications Trial (DCCT) found that patients with type 1 diabetes who kept their blood sugar levels as close to normal as possible reduced their risk for developing diabetic eye disease by 76 percent, for diabetic kidney disease by 50 percent, and for diabetic neuropathy by 60 percent.

Diabetes and its related complications was the sixth leading cause of death in 2000. According to the National Institutes of Health, cardiovascular, or heart and blood vessel disease, is the leading cause of diabetes-related death. Uncontrolled diabetes is a leading cause of blindness, end-stage renal disease, and limb amputations. Eye problems including cataracts, glaucoma, and diabetic retinopathy also are more common in people with diabetes.

Diabetic neuropathy is the result of nerve damage caused by uncontrolled diabetes. Autonomic neuropathy affects the autonomic nervous system and can cause gastroparesis (nerve damage of the stomach), neurogenic bladder (nerve damage of the urinary bladder), and a host of other problems with involuntary functions of the nervous system.

In peripheral neuropathy (PN), nerve damage in the extremities (e.g., the legs and feet) causes numbness, pain, and burning. Diabetic foot ulcers are a particular problem since frequently the patient does not feel the pain of a blister, callous, or other minor injury. Poor blood circulation in the legs and feet contribute to delayed wound healing. The inability to sense pain along with the complications of delayed wound healing can result in minor injuries, blisters, or calluses becoming infected and difficult to treat. The most serious consequence of this condition is the potential for amputation of toes, feet, or legs due to severe infection.

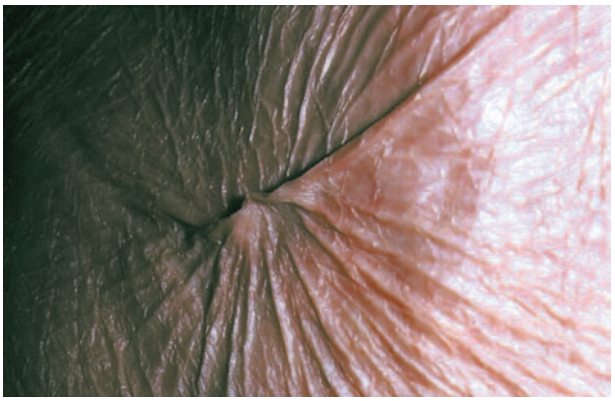
Diabetic kidney disease is another common complications of diabetes. Long- term complications may include the need for kidney dialysis or a kidney transplant due to kidney failure. Diabetes is the number one cause of chronic kidney failure in America.

Children and adults with the autoimmune form of type 1 diabetes are also at greater risk for other autoim-

mune disorders, including thyroid disease, celiac sprue (sometimes called gluten intolerance), autoimmune hepatitis, myasthenia gravis, and pernicious anemia.

Prevention

As of 2004 research continues on diabetes prevention and improved detection of those at risk for developing diabetes. While the onset of type 1 diabetes is unpredictable, the risk of developing type 2 diabetes may be reduced by maintaining ideal weight and exercising regularly. Both physical and emotional stress can cause increases in blood glucose levels, so getting regular immunizations and well-child check-ups, practicing good **sleep** and hygiene habits, encouraging emotional and social growth, and maintaining a stress- controlled lifestyle is important for children with type 1 or type 2 diabetes.



Wrinkled, dehydrated skin of a person in a diabetic coma. Untreated diabetes mellitus results in elevated blood glucose levels, causing a variety of symptoms that can culminate in a diabetic coma. *(© Dr. P. Marazzi/Science Photo Library, National Audubon Society Collection/Photo Researchers, Inc.)*

Parental concerns

Parents of children with diabetes must work with their child's teachers and school administrators to ensure that their child is able to test her blood sugars regularly, take insulin as needed, and have access to food or drink to treat a low. Someone at school should also be trained in how to administer a glucagon injection, an emergency treatment for a hypoglycemic episode when a child loses consciousness.

Section 504 of the Rehabilitation Act of 1973 enables parents to develop both a Section 504 plan (which describes a child's medical needs) and an individualized education plan (IEP) (which describes what special accommodations a child requires to address those needs). An IEP should cover such issues as blood glucose monitoring, dietary plans, and treating highs and lows. If school staff has little to no experience with diabetes, bringing in a certified diabetes educator (CDE) to offer basic training may be useful.

Children with diabetes can lead an active life and enjoy most of the activities and foods their peers do, with a few precautions to avoid blood sugar highs or lows. A certified diabetes educator that has experience working with children can help them understand the importance of regular testing as well as methods for minimizing discomfort. Diabetes summer camps, where children can learn about diabetes care in the company of peers and counselors who also live with the disease, may be useful from both a health and a social standpoint. In addition, peer support groups can sometimes help children come to terms with their diabetes.

Hypoglycemia, or low blood sugar, can be caused by too much insulin, too little food (or eating too late to coincide with the action of the insulin), alcohol consumption, or increased exercise. A child with symptoms of hypoglycemia may be hungry, cranky, confused, and tired. The patient may become sweaty and shaky. Left untreated, a child can lose consciousness or have a seizure. This condition is sometimes called an insulin reaction and should be treated by giving the patient something sweet to eat or drink like candy, juice, glucose gel, or another high sugar snack. A child who loses consciousness due to a low should never be given food or drink due to the risk of **choking**. In these cases, a glucagon injection should be administered and the child should be taken to the nearest emergency care facility.

While exercise can lower blood glucose levels, children with diabetes can and do excel in **sports**. Proper hydration, frequent testing, and a before-game or practice snack can prevent hypoglycemia. Coaches or another onsite adult should be aware of a child's medical condition and be prepared to treat a hypoglycemic attack if necessary.

The other potential danger to a child with diabetes—diabetic ketoacidosis—is uncommon and most likely to occur prior to a diagnosis. It may also happen if insulin is discontinued or if the body is under stress due to illness or injury. Ketones in the urine can be detected using dipstick tests (e.g., Ketostix), or detected using a home ketone blood monitor. Early detection facilitates early treatment and can prevent full-blown DKA.

Because the symptoms of DKA can mimic the flu, and the flu can increase blood sugar levels, a child who comes down with a flu-like illness should be monitored closely and tested regularly. An increase in insulin may also be necessary; parents of children with diabetes

KEY TERMS

Diabetic retinopathy—A condition seen most frequently in individuals with poorly controlled diabetes mellitus where the tiny blood vessels to the retina, the tissues that sense light at the back of the eye, are damaged. This damage causes blurred vision, sudden blindness, or black spots, lines, or flashing light in the field of vision.

Glucagon—A hormone produced in the pancreas that changes glycogen, a carbohydrate stored in muscles and the liver, into glucose. It can be used to relax muscles for a procedure such as duodenography. An injectable form of glucagon is sometimes used to treat insulin shock.

Honeymoon phase—A period of time shortly following diagnosis of type 1 diabetes during which a child's need for insulin may decrease or disappear altogether. The honeymoon phase is transitional, and insulin requirements eventually increases again.

Hyperglycemia—A condition characterized by excessively high levels of glucose in the blood. It occurs when the body does not have enough insulin or cannot use the insulin it does have to turn glucose into energy.

Hypoglycemia—A condition characterized by abnormally low levels of glucose in the blood.

Insulin—A hormone or chemical produced by the pancreas that is needed by cells of the body in order to use glucose (sugar), a major source of energy for the human body.

Ketoacidosis—Usually caused by uncontrolled type I diabetes, when the body isn't able to use glucose for energy. As an alternate source of energy, fat cells are broken down, producing ketones, toxic compounds that make the blood acidic. Symptoms of ketoacidosis include excessive thirst and urination, abdominal pain, vomiting, rapid breathing, extreme tiredness, and drowsiness.

Off-label use—Prescribing a drug for a population (e.g., pediatric) or condition for which it was not originally approved by the U.S. FDA. For example, sulfonylurea drugs are not FDA approved for use in children with type 2 diabetes due to a lack of clinical studies in pediatric populations, but a physician may prescribe them in an off-label use of the drug.

Prediabetes—A precursor condition to type 2 diabetes, sometimes called impaired glucose tolerance or impaired fasting glucose. Prediabetes is clinically defined as individuals who have elevated blood glucose levels that are not diagnostic of type 2 diabetes but are above normal (for the fasting plasma glucose test, this measurement would be 100 to 125 mg/dL (5.6 to 6.9 mmol/L).

should talk with their pediatrician about a sick day plan for their child before they need it.

See also Hypoglycemia.

Resources

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American Dietetic Association. 216 W. Jackson Blvd., Chicago, IL 60606–6995. Web site: <www.eatright.org>.

Children with Diabetes. 5689 Chancery Place, Hamilton, OH 45011. Web site: <www.childrenwithdiabetes.org>.

Juvenile Diabetes Research Foundation. 120 Wall St., 19th Floor, New York, NY 10005. Web site: <www.jdrf.org>.

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Diaper rash

Definition

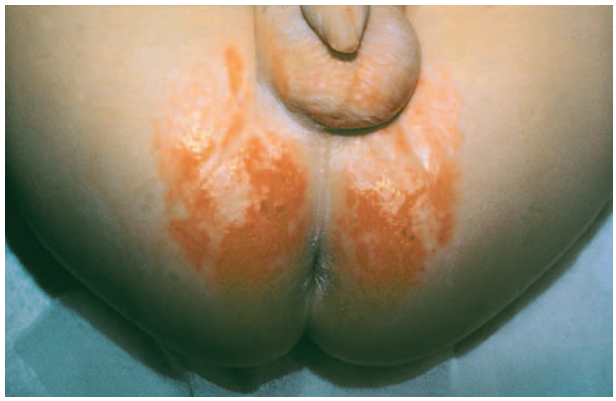
Dermatitis of the buttocks, genitals, lower abdomen, or thigh folds of an infant or toddler is called diaper rash. The outside layer of skin normally forms a protective barrier that prevents infection; when the barrier fails, the child may develop a rash in the area covered by the diaper. Diaper **rashes** occur equally with cloth diapers and disposable diapers.

Description

Diaper dermatitis results from prolonged contact with irritants such as moisture, chemical substances, and friction. Urine ammonia, formed from the breakdown of urea by fecal bacteria, is irritating to sensitive infant skin. Ammonia by itself does not cause skin breakdown. Only skin damaged by infrequent diaper changes and constant urine and feces contact is prone to damage from ammonia in urine. Inadequate fluid intake, heat, and detergents in diapers aggravate the condition. Bouts of **diarrhea** can quickly cause rashes in most children. Diaper rash begins with erythema in the perianal region. Left untreated, the area can quickly excoriate and progress to macules and papules, which form erosions and crust. Under certain circumstances (in infants under the age of six months, toddlers who have been on **antibiotics**, and immune compromised children) diaper dermatitis may become secondarily infected with Candida ablicans. Sometimes severe diaper dermatitis becomes super-infected with bacteria (streptococci or staphylococci).

Demographics

Diaper rashes occur in the diaper-wearing age group (birth to three years of age). Diaper rash occurs in about 10 percent of infants and is most common between the ages of seven and nine months. Some infants seem predisposed to diaper dermatitis. These infants have such sensitive skin that diaper dermatitis is a problem from the first few days of life.



Baby with severe diaper rash. *(© Custom Medical Stock Photo, Inc.)*

Causes and symptoms

When parents and caretakers do not change the children's diapers often, feces is in contact with skin and irritation develops in the perianal area. Urine left in diapers too long breaks down into ammonia, a chemical that is irritating to infant skin. Ammonia dermatitis of this type is a problem in the second half of the first year of life when the infant is producing a larger quantity of urine.

When the diaper area has prolonged skin contact with wetness the natural oils are stripped away, the outer layer of skin is damaged, and there is increased susceptibility to infection by bacteria or yeast.

Frequently a flat, red rash resulting from chafing of the diaper against tender skin causes friction rash. This rash is not in the skin folds. It may be more definite around the edges of the diaper, at the waist and leg bands. The baby does not seem to experience much discomfort.

Sometimes chemicals in detergents contribute to **contact dermatitis**. These rashes should clear up as soon as the chemicals are removed. Ignoring the condition may lead to a secondary infection that is more difficult to resolve.

Another infectious cause of diaper rash is **impetigo**. This bacterial infection is characterized by blisters that ooze and crust.

When to call the doctor

Parents should call the child's healthcare provider for the following reason:

- newborn with rash looks or acts sick or has a fever
- rash looks infected (pimples, blisters, boils, weeping sores, yellow crusts, red streaks)

- rash is not better in three days after treatment for yeast
- rash bright red then peeling off in sheets or raw and bleeding
- rash beyond the diaper area
- rash painful and not responding to home care

Diagnosis

Diagnosis is made by examining the diaper area and taking the history of the onset and duration of the lesions.

The presence of skin lesions means the baby has diaper rash. However, there are several types of rash that may need specific treatment to heal. It is useful to be able to distinguish them by their appearance and causes.

A baby with a rash that does not clear up within two to three days or a rash with blisters or bleeding should receive an evaluation and care from a healthcare professional.

Treatment

Antibiotics are prescribed for rashes caused by bacteria and impetigo. This may be a topical or oral formula, depending on the size of the area involved and the severity of the infection.

Over-the-counter antifungal creams are often used to treat a rash resulting from yeast. If topical treatment is not effective, an oral antifungal is prescribed. Treatment of diaper candida in young infants should include oral drops to treat any candida in the mouth and gut to avoid re-infection.

Mild steroid creams, such as 0.5 to 1 percent hydrocortisone, may be used to treat **seborrheic dermatitis** and intertrigo. Prescription strength creams are needed for short-term treatment of stubborn cases. Intertrigo can be treated with a combination of hydrocortisone and anti-fungal creams.

Complication

The main complication is secondary infection by yeast or bacteria.

Home care

Good diaper hygiene prevents or clears up many simple cases of diaper rash. Many rashes can be treated as follows:

- Change diapers frequently.
- Keep the area dry and clean. Check the diaper often, every hour if the baby has a rash and change the diaper as soon as it is wet or soiled. Check at least once during the night. Good air circulation is also important for healthy skin. Babies should have some time without wearing a diaper. A cotton pad can protect the bed while the baby is diaper free.
- Frequent and vigorous washing with soap can strip the baby's tender skin of natural protective barriers. Wash gently but thoroughly, including the skin folds. Plain water may be the best cleaning agent when there is a rash. Using warm water in a spray bottle (or give a quick bath) and then lightly pat the skin dry to avoid irritation.
- Instead of cleaning the baby's bottom with a moist wipe or washcloth, hold the diaper area over the sink and let warm water wash over the inflamed skin. Then dry the area using a blow-dryer set on cool. Washing with plain water and drying with air is soothing to sore skin; it speeds healing by decreasing friction on the area. Some wipes contain alcohol or chemicals that can be irritating and only make diaper rash worse.
- Parents can sit the baby in a basin or tub of lukewarm water for several minutes with each diaper change. This helps clean and may also be comforting. Or they can pour warm water from a pitcher or use a squirt bottle. They should not use soap unless there is sticky stool, then a mild liquid soap in a basin of warm water is effective; wash gently and rinse well. Baby oil on a cotton ball can also remove stool from small areas.
- Leave diaper off for a while.
- Do not use airtight rubber pants over the diaper area. Some cloth-like disposable diapers promote better air circulation than plastic-type diapers. If disposable diapers are used, it helps to punch holes in them to let in air.
- Petroleum jelly provides a protective coating, even on sore, redden skin, and is easy to clean. Parents should not use talcum powder because of the risk of **pneumonia**. However, cornstarch reduces friction and may prevent future rashes.

Nutrition

What the baby eats can make a difference in stool frequency and acidity. Typically, breast-fed babies have fewer problems with rashes. When adding a new food to the diet, the baby should be watched closely to see whether rashes appear around the baby's mouth or anus. If they do, the new food should be avoided temporarily.

Babies who are taking antibiotics are more likely to get rashes because of yeast. To help bring the good bacterial counts back to normal, *Lactobacillus bifidus* can be added to the diet. It is available in powder form from most health food stores.

Herbal treatment

Some herbal preparations can be useful for diaper rash. Calendula reduces inflammation, tightens tissues, and disinfects. It is recommended for seborrheic dermatitis as well as for general inflammation of the skin. The ointment should be applied at each diaper change. Chickweed ointment can also sooth irritated skin when it is applied once or twice daily.

Prognosis

With proper treatment these rashes are usually better in three days if there is no underlying health problem or skin disease. If the rash does not improve with treatment then the child probably has a yeast infection. In that case, the rash becomes bright red and raw, covers a large area, and is surrounded by red dots. For yeast infection, the child needs a special cream.

Prevention

Changing the diaper immediately and good cleaning are the best action a parent can take to prevent diaper rash. Diaper rashes occur equally with cloth diapers and disposable diapers. Some children will get a rash from certain brands of disposable diapers or from sensitivity to some soaps used in cloth diapers. If cloth diapers are used, always wash them separate from other clothing and add bleach to the soap. After washing, the diapers should be rinsed thoroughly.

Parental concerns

Diapering. There are two choices, cloth or disposable. Parents need to decide what works best for their baby and lifestyle.

In the event of suspected yeast, a tablespoon of cider vinegar in a cup of warm water can serve as a diaper area wash. This is diluted enough that it should not burn, but acidifies the skin pH enough to hamper the yeast growth.

Barrier ointments can be valuable to treat rashes. Those that contain zinc oxide are especially effective. These creams and ointments protect already irritated skin. Cornstarch powder is soothing to rashes that are moist, such as impetigo.

KEY TERMS

Dermatitis—Inflammation of the skin.

Diaper dermatitis (diaper rash)—An inflammatory reaction to irritants in the diaper area.

Impetigo—A bacterial infection of the skin characterized by skin blistering.

Lactobacillus bifidus—A property of breast milk that interferes with the growth of pathogenic bacteria in the gastrointestinal tracts of babies, reducing the incidence of diarrhea. Lactobacillus bifidus can be added to infant formulas to help control diarrhea.

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Aliene S. Linwood, RN, DPA, FACHE

Diarrhea

Definition

To most individuals, diarrhea means an increased frequency or decreased consistency of bowel movements; however, the medical definition is more exact than this. Diarrhea best correlates with an increase in stool weight; this increase is mainly due to excess water, which normally makes up 60 to 85 percent of fecal matter. In this way, true diarrhea is distinguished from diseases that cause only an increase in the number of bowel movements (hyperdefecation) or incontinence (involuntary loss of bowel contents).

Diarrhea is also classified by physicians as acute, which lasts one to two weeks, and as chronic, which continues for longer than two or three weeks. Viral and bacterial infections are the most common causes of acute diarrhea.

Description

In many cases, acute infectious diarrhea is a mild, limited annoyance common to adults and children. Chronic diarrhea, though, can have considerable effect on health and on social and economic well-being. People with **celiac disease**, inflammatory bowel disease, and other prolonged diarrheal illnesses develop nutritional deficiencies that diminish growth and immunity. They affect social interaction and result in the loss of many working hours. Rapid diagnosis and proper treatment can prevent much of the suffering associated with these illnesses.

Demographics

Worldwide, acute infectious diarrhea has a huge impact, causing over 5 million deaths per year. While most deaths are among children under five years of age in developing nations, the impact, even in developed countries, is considerable. For example, over 250,000 individuals are admitted to hospitals in the United States each year because of one of these episodes.

Causes and symptoms

Diarrhea occurs because more fluid passes through the large intestine (colon) than that organ can absorb. As a rule, the colon can absorb several times more fluid than is required on a daily basis. However, when this reserve capacity is overwhelmed, diarrhea occurs.

Diarrhea is caused by infections or illnesses that either lead to excess production of fluids or prevent absorption of fluids. Also, certain substances in the colon, such as fats and bile acids, can interfere with water absorption and cause diarrhea. Rapid passage of material through the colon can also do the same.

Symptoms related to any diarrheal illness are often those associated with any injury to the gastrointestinal tract, such as **fever**, **nausea**, **vomiting**, and abdominal **pain**. All or none of these may be present depending on the disease causing the diarrhea. The number of bowel movements can vary—up to 20 or more per day. In some persons, blood or pus is present in the stool. Bowel movements may be difficult to flush (float) or contain undigested food material.

The most common causes of acute diarrhea are infections (the cause of traveler's diarrhea), **food poisoning**, and medications. Medications are a frequent and often over-looked cause, especially **antibiotics** and antacids. Less often, various sugar-free foods, which sometimes contain poorly absorbable materials, cause diarrhea.

Chronic diarrhea is frequently due to many of the same things that cause the shorter episodes (infections, medications, etc.); however, symptoms last longer. Some infections can become chronic. This occurs mainly with parasitic infections (such as *Giardia*) or when people have altered immunity (such as **AIDS**). In children, more common causes of chronic diarrhea are food allergy and **lactose intolerance**. Toddlers who drink too much juice can have frequent, loose stools.

When to call the doctor

A physician or other healthcare provider should be contacted when the number of bowel movements exceeds three per day for 2 days or more or when fecal material contains blood. A doctor should be called if a person becomes dehydrated. Signs of **dehydration** include decreased urination, lethargy, poor skin tone, and generalized weakness. In very young children, the parents should call a doctor if they observe these symptoms of dehydration:

- dry mouth or tongue
- few or no tears when crying
- no wet diapers for three hours or more
- sunken eyes, cheeks, and fontanel (soft spot on the head of infants)
- irritability and listlessness
- skin that flattens slowly when pinched

Parents should also call the doctor if a child is vomiting so often that he or she cannot keep fluids down, has a high fever, complains of severe abdominal pain, or shows no improvement in symptoms after 24 hours.

Diagnosis

Most cases of acute diarrhea never need diagnosis or treatment, as many are mild and produce few problems. But persons of any age with fever over 102°F (38.9°C), signs of dehydration, bloody bowel movements, severe abdominal pain, known immune disease, or prior use of antibiotics need prompt medical evaluation.

When diagnostic studies are needed, the most useful are stool culture and examination for parasites; however, these are often negative, and a cause cannot be found in a large number of people. The earlier cultures are performed, the greater the chance of obtaining a positive result. For those with a history of antibiotic use in the preceding two months, stool samples need to be examined for the toxins that cause antibiotic-associated colitis. Tests are also available to check stool samples for microscopic amounts of blood and for cells that indicate severe

inflammation of the colon. Examination with an endoscope is sometimes helpful in determining severity and extent of inflammation. Tests to check changes in blood chemistry (potassium, magnesium, etc.) and a complete blood count (CBC) are also often performed.

Chronic diarrhea is quite different, and most persons with this condition receive some degree of testing. Many exams are the same as for an acute episode, as some infections and parasites cause both types of diarrhea. A careful history to evaluate medication use, dietary changes, **family** history of illnesses, and other symptoms is necessary. Key points in determining the seriousness of symptoms are weight loss of over 10 lbs (4.5 kg), blood in the stool, and nocturnal diarrhea (symptoms that awaken an individual from **sleep**).

Both prescription and over-the-counter medications can contain additives, such as lactose and sorbitol, that will produce diarrhea in sensitive individuals. Review of **allergies** or skin changes may also point to a cause. Social history may indicate that stress is playing a role or may identify activities which can be associated with diarrhea (for example, diarrhea that occurs in runners).

A combination of stool, blood, and urine tests may be needed in the evaluation of chronic diarrhea; in addition, a number of endoscopic and x-ray studies are frequently required.

Treatment

Treatment is ideally directed toward correcting the cause; however, the first aim should be to prevent or treat dehydration and nutritional deficiencies. The type of fluid and nutrient replacement depends on whether oral feedings can be taken and on the severity of fluid losses. Oral rehydration solution (ORS) or intravenous fluids are the choices; ORS is preferred if possible.

A physician should be notified if a person is dehydrated. If oral replacement is suggested then commercial (Pedialyte and others) or homemade preparations can be used. The World Health Organization (WHO) has provided this easy recipe for home preparation, which can be taken in small frequent sips:

- table salt, 3/4 tsp
- baking powder, 1 tsp
- orange juice, 1 c
- water, 1 qt

When feasible, food intake should be continued even in those people with acute diarrhea. A physician should be consulted regarding what type and how much food is permitted.

Anti-motility agents (loperamide, diphenoxylate) are useful for those with chronic symptoms; their use is limited or even contraindicated in most individuals with acute diarrhea, especially in those with high fever or bloody bowel movements. They should not be taken without the advice of a physician, and should not be used in children.

Other treatments are available, depending on the cause of symptoms. For example, the bulk agent psyllium helps some people by absorbing excess fluid and solidifying stools; cholestyramine, which binds bile acids, is effective in treating bile-salt-induced diarrhea. Low fat diets or more easily digestible fat is useful in some people. Antidiarrheal drugs that decrease excessive secretion of fluid by the intestinal tract is another approach for some diseases. Avoidance of medications or other products that are known to cause diarrhea (such as lactose) is curative in some people but should be discussed with a physician.

Alternative treatment

It is especially important to find the cause of diarrhea, since stopping diarrhea when it is the body's way of eliminating something foreign is not helpful and can be harmful in the long run.

One effective alternative approach to preventing and treating diarrhea involves oral supplementation of aspects of the normal flora in the colon with the yeasts *Lactobacillus acidophilus*, *L. bifidus*, or *Saccharomyces boulardii*. In clinical settings, these "biotherapeutic" agents have repeatedly been helpful in the resolution of diarrhea, especially antibiotic-associated diarrhea.

Nutrient replacement also plays a role in preventing and treating episodes of diarrhea. Zinc especially appears to have an effect on the immune system, and deficiency of this mineral can lead to chronic diarrhea. Also, zinc replacement improves growth in young persons. To prevent dehydration, individuals suffering from diarrhea should take plenty of fluids, especially water. The BRAT diet also can be useful in helping to resolve diarrhea. This diet limits food intake to bananas, rice, applesauce, and toast. These foods provide soluble and insoluble fiber without irritation. If the toast is slightly burnt, the charcoal can help sequester toxins and pull them from the body.

Acute homeopathic remedies can be very effective for treating diarrhea especially in infants and young children.

KEY TERMS

Antimotility drug—A medication, such as loperamide (Imodium), dephenoxylate (Lomotil), or medications containing codeine or narcotics that decrease the ability of the intestine to contract.

Colitis—Inflammation of the colon (large intestine).

Endoscope—A medical instrument that can be passed into an area of the body (the bladder or intestine, for example) to allow visual examination of that area. The endoscope usually has a fiber-optic camera that allows a greatly magnified image to be shown on a television screen viewed by the operator. Many endoscopes also allow the operator to retrieve a small sample (biopsy) of the area being examined, to more closely view the tissue under a microscope.

Endoscopy—Visual examination of an organ or body cavity using an endoscope, a thin, tubular instrument containing a camera and light source. Many endoscopes also allow the retrieval of a small sample (biopsy) of the area being examined, in order to more closely view the tissue under a microscope.

Lactose intolerance—An inability to properly digest the lactose found in milk and dairy products.

Oral rehydration solution (ORS)—A liquid preparation of electrolytes and glucose developed by the World Health Organization that can decrease fluid loss in persons with diarrhea. Originally developed to be prepared with materials available in the home, commercial preparations have recently come into use.

Steatorrhea—An excessive amount of fat in the feces due to poor fat absorption in the gastrointestinal tract.

Prognosis

Prognosis is related to the cause of the diarrhea; for most individuals in developed countries, a bout of acute, infectious diarrhea is at best uncomfortable. However, in both industrialized and developing areas, serious complications and death can occur.

For those with chronic symptoms, an extensive number of tests are usually necessary to make a proper diagnosis and begin treatment; a specific diagnosis is found in 90 percent of people. In some, however, no specific cause is found and only treatment with bulk agents or anti-motility agents is indicated.

Prevention

Proper hygiene and food handling techniques can prevent many cases. Traveler's diarrhea can be avoided by people using products containing bismuth, such as Pepto-Bismol and/or antibiotics. The most important action is to prevent the complications of dehydration.

Nutritional concerns

Replacement of fluids and electrolytes is important for people experiencing diarrhea. These individuals should take in foods that contain salt, potassium, phosphates, and sugar. Most sodas, sport drinks and non-cream soups are good sources of electrolytes.

Parental concerns

Parents should be sure that their children who experience diarrhea drink plenty of fluids and replace electrolytes with an oral rehydration solution. A doctor should be called if the parent suspects a child is becoming dehydrated. Severe dehydration requires intravenous fluid administration in a medical setting. Antidiarrheal medications should be be given only on the advise of a physician.

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DiGeorge syndrome

Definition

DiGeorge syndrome is a rare congenital disease that affects an infant's immune system and that is due to a large deletion from chromosome 22. The syndrome is marked by absence or underdevelopment of the thymus and parathyroid glands. It is named for the pediatrician who first described it in 1965.

Normally the thymus gland is located below the thyroid gland in the neck and front of the chest and is the primary gland of the lymphatic system, which is necessary for normal functioning of the immune system. The parathyroid glands, located on the sides of the thyroid gland, are responsible for maintenance of normal levels of calcium in the blood. In children with DiGeorge syndrome, the thymus and parathyroid glands are missing or undeveloped. The symptoms of this disorder vary, depending on the extent of missing thymus and parathyroid tissue. The primary problem for children who survive with DiGeorge syndrome is repeated infections due to a defective immune system.

DiGeorge syndrome is sometimes described as a "CATCH 22" disorder, so named because of their characteristics—cardiac defects (C), abnormal facial features (A), thymus underdevelopment (T), **cleft palate** (C), and hypocalcemia due to hypoparathyroidism(H)—all resulting from deletion of portions of chromosome 22. Specific facial features associated with DiGeorge syndrome include low-set ears, wide-set eyes, a small jaw, and a short groove in the upper lip.

DiGeorge syndrome is also called congenital thymic hypoplasia, or third and fourth pharyngeal pouch syndrome, because the congenital abnormalities occur in areas known as the third and fourth pharyngeal pouches, which later develop into the thymus and parathyroid glands.

Demographics

The prevalence of DiGeorge syndrome, is debated; estimates have ranged from one in 4,000 to one in 6,395. Because the symptoms caused by the chromosomal abnormality vary somewhat from child to child, the syndrome probably occurs much more often than was previously thought. In the United States, autopsy studies for DiGeorge syndrome accounted for 0.7 percent of 3469 postmortem examinations in the Seattle, Washington, area over a period of 25 years. Internationally, the incidence of DGS was estimated to be one case per 20,000 persons in Germany and one case per 66,000 persons in Australia. However, with the advent of fluorescence in situ hybridization (FISH) techniques to detect monosomy 22 and the inclusion of related syndromes, more recent estimates place the incidence of DiGeorge syndrome in the range of one case per 3,000 persons.

No major difference is noted in the incidence of DiGeorge syndrome between males and females. The syndrome also appears to be equally common in all racial and ethnic groups.

Causes and symptoms

DiGeorge syndrome is caused either by inheritance of a defective chromosome 22 or by a new defect in chromosome 22 in the fetus. The type of defect that is involved is called deletion. A deletion occurs when the genetic material in the chromosomes does not recombine

properly during the formation of sperm or egg cells. The deletion means that several genes from chromosome 22 are missing in children with DiGeorge syndrome. According to a 1999 study, 6 percent of children with DiGeorge syndrome inherited the deletion from a parent, while 94 percent had a new deletion.

The loss of the genes in the deleted material means that the baby's third and fourth pharyngeal pouches fail to develop normally during the twelfth week of pregnancy. This developmental failure results in a completely or partially absent thymus gland and parathyroid glands. In addition, 74 percent of fetuses with DiGeorge syndrome have severe heart defects. The child is born with a defective immune system and an abnormally low level of calcium in the blood.

These defects usually become apparent within 48 hours of birth. The infant's heart defects may lead to heart failure, or there may be seizures and other evidence of a low level of calcium in the blood (hypocalcemia).

When to call the doctor

Because the immune system of a child with DiGeorge syndrome is defective, a doctor should be consulted at any signs of illness or disease.

Diagnosis

Diagnosis of DiGeorge syndrome can be made by ultrasound examination around the eighteenth week of pregnancy, when abnormalities in the development of the heart or the palate can be detected. Another technique that is used to diagnose the syndrome before birth is called fluorescence in situ hybridization, or FISH. This technique uses DNA probes from the DiGeorge region on chromosome 22. FISH can be performed on cell samples obtained by **amniocentesis** as early as the fourteenth week of pregnancy. It confirms about 95 percent of cases of DiGeorge syndrome.

If the mother has not had prenatal testing, the diagnosis of DiGeorge syndrome is sometimes suggested by the child's facial features at birth. The child is also born with a defective immune system and an abnormally low level of calcium in the blood. These defects usually become apparent within 48 hours after birth. The infant's heart defects may lead to heart failure, or there may be seizures and other evidence of a low level of calcium in the blood. The doctor may make the diagnosis of DiGeorge syndrome during heart surgery when he or she notices the absence or abnormal location of the thymus gland. The diagnosis can be confirmed by blood tests for calcium, phosphorus, and parathyroid hormone levels and by the sheep cell test for immune function.

Treatment

Hypocalcemia

Hypocalcemia in a child with DiGeorge syndrome is unusually difficult to treat. Infants are usually given calcium and vitamin D by mouth. Severe cases have been treated by transplantation of fetal thymus tissue or bone marrow.

Heart defects

Infants with life-threatening heart defects are treated surgically.

Defective immune function

Children with DiGeorge syndrome should be kept away from crowds or other sources of infection. They should not be immunized with vaccines made from live viruses or given corticosteroids.

Nutritional concerns

Children with DiGeorge syndrome should be kept on low-phosphorus diets.

Prognosis

The prognosis is variable; many infants with DiGeorge syndrome die from overwhelming infection, seizures, or heart failure within the first year. A one-month mortality rate of 55 percent and a six-month mortality rate of 86 percent has been reported due to **congenital heart disease**. Advances in heart surgery indicate that the prognosis is most closely linked to the severity of the heart defects and the partial presence of the thymus gland. In most children who survive, the number of T cells, a type of white blood cell, in the blood rises spontaneously as they mature. Survivors are likely to be mentally retarded, however, with mild to moderate learning disabilities, and to have other developmental difficulties, including short stature as well as psychiatric problems in later life

Prevention

Genetic counseling and testing is recommended for a person with DiGeorge syndrome who becomes pregnant, because the disorder can be detected prior to birth. Although most children with DiGeorge syndrome do not inherit the chromosome deletion from their parents, they have a 50 percent chance of passing the deletion on to their own children. Parents should be screened, however, to see if they are carriers, even though inheritance of DiGeorge syndrome is rare.

KEY TERMS

Amniocentesis—A procedure performed at 16-18 weeks of pregnancy in which a needle is inserted through a woman's abdomen into her uterus to draw out a small sample of the amniotic fluid from around the baby for analysis. Either the fluid itself or cells from the fluid can be used for a variety of tests to obtain information about genetic disorders and other medical conditions in the fetus.

Chromosome—A microscopic thread-like structure found within each cell of the human body and consisting of a complex of proteins and DNA. Humans have 46 chromosomes arranged into 23 pairs. Chromosomes contain the genetic information necessary to direct the development and functioning of all cells and systems in the body. They pass on hereditary traits from parents to child (like eye color) and determine whether the child will be male or female.

Congenital—Present at birth.

Deletion—The absence of genetic material that is normally found in a chromosome. Often, the genetic material is missing due to an error in replication of an egg or sperm cell.

Hypocalcemia—A condition characterized by an abnormally low level of calcium in the blood.

Hypoplasia—An underdeveloped or incomplete tissue or organ usually due to a decrease in the number of cells.

T cell—A type of white blood cell that is produced in the bone marrow and matured in the thymus gland. It helps to regulate the immune system's response to infections or malignancy.

Because of an association between DiGeorge syndrome and **fetal alcohol syndrome**, pregnant women should avoid drinking alcoholic beverages.

Parental concerns

Recurrent infections are a major problem in children with DiGeorge syndrome and an important cause of later mortality. Therefore, prevention of infections must be a high priority.

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Diphenhydramine *see* **Antihistamines**

Diphtheria

Definition

Diphtheria is a potentially fatal, contagious disease that usually involves the nose, throat, and air passages but may also infect the skin. Its most striking feature is the formation of a grayish membrane covering the tonsils and upper part of the throat.

Description

Like many other upper respiratory diseases, diphtheria is most likely to break out during the winter months. At one time it was a major childhood killer, but in the early 2000s it is rare in developed countries because of widespread immunization.

Persons who have not been immunized may get diphtheria at any age. The disease is spread most often by droplets from the coughing or sneezing of an infected person or carrier. The incubation period is two to seven days, with an average of three days. It is vital to seek medical help at once when diphtheria is suspected, because treatment requires emergency measures for adults as well as children.

Demographics

Diphtheria is a reportable disease in many countries in the world. Since 1988, all confirmed cases in the United States involved visitors or immigrants. In countries that do not have routine immunization against this infection, the mortality rate varies from 1.5 to 25 percent.

Causes and symptoms

The symptoms of diphtheria are caused by toxins produced by the diphtheria bacillus, *Corynebacterium diphtheriae* (from the Greek for "rubber membrane"). In fact, toxin production is related to infections of the bacillus itself with a particular bacteria virus called a phage (from bacteriophage, a virus that infects bacteria). The intoxication destroys healthy tissue in the upper area of the throat around the tonsils or in open **wounds** in the skin. Fluid from the dying cells then coagulates to form the telltale gray or grayish green membrane. Inside the membrane, the bacteria produce an exotoxin, which is a poisonous secretion that causes the life-threatening symptoms of diphtheria. The exotoxin is carried throughout the body in the bloodstream, destroying healthy tissue in other parts of the body.

The most serious complications caused by the exotoxin are inflammations of the heart muscle (myocarditis) and damage to the nervous system. The risk of serious complications is increased as the time between onset of symptoms and the administration of antitoxin increases, and as the size of the membrane formed increases. The myocarditis may cause disturbances in the heart rhythm and may culminate in heart failure. The symptoms of nervous system involvement can include seeing double (diplopia), painful or difficult swallowing, and slurred speech or loss of voice, which are all indications of the exotoxin's effect on nerve functions. The exotoxin may also cause severe swelling in the neck ("bull neck").

The signs and symptoms of diphtheria vary according to the location of the infection.

Nasal

Nasal diphtheria produces few symptoms other than a watery or bloody discharge. On examination, there may be a small visible membrane in the nasal passages. Nasal infection rarely causes complications by itself, but it is a public health problem because it spreads the disease more rapidly than other forms of diphtheria.

Pharyngeal

Pharyngeal diphtheria gets its name from the pharynx, which is the part of the upper throat that connects the mouth and nasal passages with the voice box. This is the most common form of diphtheria, causing the characteristic throat membrane. The membrane often bleeds if it is scraped or cut. It is important not to try to remove the membrane because the trauma may increase the body's absorption of the exotoxin. Other signs and symptoms of pharyngeal diphtheria are mild **sore throat**, **fever** of 101–102°F (38.3–38.9°C), a rapid pulse, and general body weakness.

Laryngeal

Laryngeal diphtheria, which involves the voice box or larynx, is the form most likely to produce serious complications. The fever is usually higher in this form of diphtheria (103–104°F or 39.4–40°C) and the person is very weak. People may have a severe **cough**, have difficulty breathing, or lose their voice completely. The development of a bull neck indicates a high level of exotoxin in the bloodstream. Obstruction of the airway may result in respiratory compromise and death.

Skin

This form of diphtheria, which is sometimes called cutaneous diphtheria, accounts for about 33 percent of all diphtheria cases. It is found chiefly among people with poor hygiene. Any break in the skin can become infected with diphtheria. The infected tissue develops an ulcerated area, and a diphtheria membrane may form over the wound but is not always present. The wound or ulcer is slow to heal and may be numb or insensitive when touched.

When to call the doctor

A doctor should be called whenever a case of diphtheria is suspected.

Diagnosis

Because diphtheria must be treated as quickly as possible, doctors usually make the diagnosis on the basis of the visible symptoms without waiting for test results.

In making the diagnosis, the doctor examines the affected person's eyes, ears, nose, and throat in order to rule out other diseases that may cause fever and sore throat, such as **infectious mononucleosis**, a sinus infection, or **strep throat**. The most important single symptom that suggests diphtheria is the membrane. When a person develops skin infections during an outbreak of

diphtheria, the doctor will consider the possibility of cutaneous diphtheria and take a smear to confirm the diagnosis.

Laboratory tests

The diagnosis of diphtheria can be confirmed by the results of a culture obtained from the infected area. Material from the swab is put on a microscope slide and stained using a procedure called Gram's stain. The diphtheria bacillus is Gram-positive which means it holds the dye after the slide is rinsed with alcohol. Under the microscope, diphtheria bacilli look like beaded rod-shaped cells, grouped in patterns that resemble Chinese characters. Another laboratory test involves growing the diphtheria bacillus on a special material called Loeffler's medium.

Treatment

Diphtheria is a serious disease requiring hospital treatment in an intensive care unit if the person has developed respiratory symptoms. Treatment includes a combination of medications and supportive care.

Antitoxin

The most important step is prompt administration of diphtheria antitoxin, without waiting for laboratory results. The antitoxin is made from horse serum and works by neutralizing any circulating exotoxin. The doctor must first test people for sensitivity to animal serum. People who are sensitive (about 10%) must be desensitized with diluted antitoxin, since as of 2004 the antitoxin is the only specific substance that counteracts diphtheria exotoxin. No human antitoxin is available for the treatment of diphtheria.

The dose ranges from 20,000 to 100,000 units, depending on the severity and length of time of symptoms occurring before treatment. Diphtheria antitoxin is usually given intravenously.

Antibiotics

Antibiotics are given to wipe out the bacteria, to prevent the spread of the disease, and to protect people from developing **pneumonia**. They are not a substitute for treatment with antitoxin. Both adults and children may be given penicillin, ampicillin, or erythromycin. Erythromycin appears to be more effective than penicillin in treating people who are carriers because of better penetration into the infected area.

Cutaneous diphtheria is usually treated by cleansing the wound thoroughly with soap and water and giving an individual antibiotics for ten days.

Supportive care

Persons with diphtheria require bed rest with intensive nursing care, including extra fluids, oxygenation, and monitoring for possible heart problems, airway blockage, or involvement of the nervous system. People with laryngeal diphtheria are kept in a **croup** tent or high-humidity environment; they may also need throat suctioning or emergency surgery if their airway is blocked.

People recovering from diphtheria should rest at home for a minimum of two to three weeks, especially if they have heart complications. In addition, persons should be immunized against diphtheria after recovery, because having the disease does not always induce antitoxin formation and protect them from reinfection.

Prevention of complications

People with diphtheria who develop myocarditis may be treated with oxygen and with medications to prevent irregular heart rhythms. An artificial pacemaker may be needed. Persons with difficulty swallowing can be fed through a tube inserted into the stomach through the nose. Persons who cannot breathe are usually put on mechanical respirators.

Prognosis

The prognosis depends on the size and location of the membrane and on early treatment with antitoxin; the longer the delay, the higher the death rate. The most vulnerable persons are children under the age of 15 years and those who develop pneumonia or myocarditis. Nasal and cutaneous diphtheria are rarely fatal.

Prevention

Prevention of diphtheria has four aspects: immunization, isolation of infected persons, identification and treatment of contacts, and reporting cases to health authorities.

Immunization

Universal immunization is the most effective means of preventing diphtheria. The standard course of immunization for healthy children is three doses of DPT (diphtheria-tetanus-pertussis) preparation given between two months and six months of age, with booster doses given at 18 months and at entry into school. Adults

KEY TERMS

Antitoxin—An antibody against an exotoxin, usually derived from horse serum.

Bacillus—A rod-shaped bacterium, such as the diphtheria bacterium.

Carrier—A person who possesses a gene for an abnormal trait without showing signs of the disorder. The person may pass the abnormal gene on to offspring. Also refers to a person who has a particular disease agent present within his/her body, and can pass this agent on to others, but who displays no symptoms of infection.

Cutaneous—Pertaining to the skin

Diphtheria-tetanus-pertussis (DTP) vaccine—The standard vaccine used to immunize children against diphtheria, tetanus, and whooping cough. A so-called "acellular pertussis" vaccine (aP) is usually used since its release in the mid-1990s.

Exotoxin—A poisonous secretion produced by bacilli that is carried in the bloodstream to other parts of the body.

Gram stain—A staining procedure used to visualize and classify bacteria. The Gram stain procedure allows the identification of purple (gram positive) organisms and red (gram negative) organisms. This identification aids in determining treatment.

Loeffler's medium—A special substance used to grow diphtheria bacilli to confirm the diagnosis.

Myocarditis—Inflammation of the heart muscle (myocardium).

Toxoid—A preparation made from inactivated exotoxin, used in immunization.

should be immunized at ten-year intervals with Td (tetanus-diphtheria) toxoid. (A toxoid is a bacterial toxin that is treated to make it harmless but still can induce immunity to the disease.)

Isolation of affected persons

Individuals with diphtheria must be isolated for one to seven days or until two successive cultures show that the individuals are no longer contagious. Children placed in isolation are usually assigned a primary nurse for emotional support.

Identification and treatment of contacts

Because diphtheria is highly contagious and has a short incubation period, **family** members and other contacts of persons with diphtheria must be watched for symptoms and tested to see if they are carriers. They are usually given antibiotics for seven days and a booster shot of diphtheria/tetanus toxoid.

Reporting cases to public health authorities

Reporting is necessary for tracking potential epidemics, to help doctors identify the specific strain of diphtheria, and to see if resistance to penicillin or erythromycin has developed.

Parental concerns

Parents in the United States should ensure that their children have full immunizations against diphtheria. Completion of the three-shot series initiates lifelong immunity from diphtheria.

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Diphtheria, tetanus, pertussis vaccine *see* DTP vaccine

Discipline

Definition

The term "discipline" comes from the Latin word "disciplinare," which means "to teach." Many people, however, associate the word with punishment, which falls short of the full meaning of the word. Discipline, properly practiced, uses a multifaceted approach, including models, rewards, and punishments that teach and reinforce desired behavior. Through discipline, children are able to learn self-control, self-direction, competence, and a sense of caring.

Description

The American Academy of Pediatrics suggests that an effective discipline system must contain three elements. If these three aspects are all present in a program of discipline, the result generally is improved child behavior. The elements are:

- a learning environment characterized by positive, supportive parent-child relationships
- a proactive strategy for systematic teaching and strengthening of desired behaviors
- a reactive strategy for decreasing or eliminating undesired behaviors

There are several reasons why children may not behave properly, including a lack of effective disciplinary measures. Children also commonly misbehave when they are deprived of adult attention or when they are tired, bored, or hungry. Children from families affected by **divorce** and separation, poverty, substance abuse, and parental depression seem to be at greater risk for behavior problems. There may also be biologic factors such as **attention-deficit/hyperactivity disorder** (ADHD) and certain temperaments that predispose particular children towards misbehavior. There is also research suggesting that harsh disciplinary measures may actually increase poor behavior.

Ideally, discipline is based on appropriate expectations for each child, based on age and stage of development. It should be used to set reasonable limits in a consistent manner while still allowing some choice among acceptable alternatives. Discipline teaches both social and moral standards and should protect children from harm by teaching what is safe. It should also guide children to respect the rights and property of others.

Though there are a variety of ways in which children may be disciplined, there are some guidelines that all parents should follow:

- Discipline must be age appropriate. While reasoning and verbal explanations may be appropriate for the older child, children younger than 18 months are typically unable to comprehend the reasons for punishment.
- Parents should demonstrate a unified front when it comes to discipline. If parents exhibit opposing approaches, children learn to exploit these differences.
- Rules should be few but simple. Punishment should be a logical or natural consequence of the misbehavior.
- Though consistency is important, parents should remember that it is sometimes appropriate to be flexible and allow for some negotiation, especially with

older children. Doing so can teach decision-making, enhance children's moral judgment, and reinforce independence.

Disciplinary techniques that are most effective take place in the context of a loving and secure relationship between parent and child. Parents' responses to a child's behavior, whether approving or disapproving, are likely to have a greater effect in a secure, loving environment, because children long for their parents' approval. As children respond to this positive relationship and consistent discipline, the need for negative interaction decreases.

Positive reinforcement

Positive reinforcement focuses on good behavior rather than on undesirable behavior. Parents should identify appropriate behaviors and give frequent feedback, rewarding good behavior quickly so that the child associates the "prize" with the wanted behavior. A reward can be a word of praise, a special activity, additional privileges, or material items. Many desirable behavioral patterns start to emerge as a part of the child's normal development. The role of parents is to notice these behaviors and provide positive attention to them. Some other desirable behaviors are not part of a child's normal development and need to be modeled and taught by their parents. These behaviors include sharing, good manners, effective study habits, among others. Parents need to identify those skills and behaviors they want their children to demonstrate and then make a concerted effort to teach and strengthen those behaviors. Children who learn through positive reinforcement tend to internalize the newly learned behaviors.

Extinction

Extinction is a type of discipline that seeks to prevent inadvertent positive reinforcement for negative behavior. "Time-out" is one of the most common methods in this category. For younger children, time out usually involves removing parental attention and praise or placing the child a chair or some other place for a specified time with no parental interaction. The environment should be neutral, boring, and safe. Time-out works well for children from 18 months up to five or six years of age and is particularly useful for temper **tantrums**, yelling, whining, and fighting. The session should end only when the child has been calm and quiet for at least 15 seconds. Time out should last for a specified time, usually one minute per year of life (to a maximum of five minutes). Withholding privileges is another form of extinction that is more appropriate for older children and adolescents. This strategy requires the removal of a valued privilege and works best if it is used infrequently.

Verbal punishment

Parents may express disapproval of a behavior by scolding or yelling. This may be effective if used very sparingly. However, if used too often it can cause **anxiety** in the child and encourage the child to ignore the parent.

Corporal punishment

Corporal punishment involves the application of some sort of physical **pain** in response to a child's undesired behavior. This response can range from a light slapping of a hand to severe beatings that qualify as **child abuse**. Because of this range in form and severity, the use of corporal punishment as a disciplinary method is controversial. In spite of the significant concerns raised by child-care experts, one form of physical punishment—spanking—remains a widely used measure to reduce undesired behavior in children. Over 90 percent of all families report having used spanking at some time as a means of discipline. Despite its common acceptance, research shows that spanking is a less effective form of discipline than others, such as time-out or removal of privileges. Although it may immediately stop a behavior, the effectiveness of spanking tends to decrease with repeated use. The only way to maintain the initial effect of spanking is to increase its intensity, which runs the risk of escalating to abuse. Spanking, at best, is only effective when used in selective, very infrequent situations.

Children who receive corporal punishment tend to grow into angry adults. The use of spanking in older children is associated with higher rates of substance abuse and crime and has been linked to poor **self-esteem**, depression, and poor educational performance.

Infancy

Discipline strategies with infants should be passive. The main goal is for parents to generally structure daily routines but to also demonstrate flexibility in meeting infants' emerging needs. As infants become more mobile, parents need to impose some limitations and structure in order to create a safe environment in which the child can **play** and explore. Parents must protect infants from all potential hazards in the home by instituting **childproofing** practices. If a child does attempt to play with or approach something dangerous or unacceptable, a firm "No" should suffice, along with either removing the child from the area or by distracting the child with an alternative activity. Parents should not

expect that reasoning or reprimands will control the behavior of an infant.

Toddlerhood

Toddlers, like infants, still benefit most from passive types of discipline and a toddler-safe environment. Again, saying "No", along with redirecting behavior, is usually effective if the toddler is doing something unacceptable. At this stage, however, children are starting to test the limits of their power over and over again. It is important for parents to consistently set limits and stick to them. Doing so reduces the child's confusion and his or her need to test. This is also the time when time-outs might be introduced, especially when redirecting the child's attention no longer seems to work.

Preschool

Preschoolers are starting to understand the need for rules, and their behavior should be guided by these rules and the associated consequences. It is very important that children understand what is expected of them and why they are punished for a particular behavior. Preschoolers also learn from having their good behaviors rewarded.

School age

If rules for behavior have been consistently modeled and expected by the parents, children should exhibit an increased sense of responsibility and self-control when they become school age. Timeouts and consequences continue to be effective disciplinary measures in this age group. As children continue to mature and desire more responsibility and independence, teaching them to deal with the consequences of their behavior is an effective method of discipline. By the time they have become teenagers, children should know what is expected of them and what the potential consequences of misbehavior are. However, discipline remains just as important for teens as it does for younger children. Teens require boundaries. This structure continues to provide order and a sense of security for children until they reach adulthood. When teens do break rules, taking away some of their privileges seems to be the most effective type of disciplinary measure.

Common problems

One of the most common problems in child discipline is an inconsistent approach between two parents. It may prove helpful for parents to regularly communicate regarding their child's behavior and decide ahead of time what disciplinary methods are to be used.

KEY TERMS

Punishment—The application of a negative stimulus to reduce or eliminate a behavior. The two types typically used with children are verbal reprimands and punishment involving physical pain, as in corporal punishment.

Time-out—A discipline strategy that entails briefly isolating a disruptive child in order to interrupt and avoid reinforcement of negative behavior.

Parental concerns

Parents may be worried that the disciplinary methods they have decided are appropriate for their child may not be respected or followed by teachers and other adult caregivers. If this is a concern, parents should outline exactly what consequences or punishments they feel are appropriate and communicate openly with the other adults who care for their child.

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Divorce

Definition

Divorce is the legal termination of a marriage.

Description

More than 1 million children each year experience their parents' divorce. Less than 60 percent of American children live with both of their biological parents; about 25 percent live with their biological mother only; and about 4 percent live with their biological father only. The remaining 11 percent live with step-families, adoptive parents, foster homes, or with other relatives.

In 2002 it was estimated that up to 30 percent (19.8 million) of children in the United States, representing 11.9 million families, lived in single-parent households. While the number of single mothers has remained constant through the 1990s and into the early 2000s at 9.9 million, the number of single fathers has grown from 1.7 million in 1995 to 2 million in 2002, according to data from the U.S. Census Bureau. In 2002, 19.8 million children lived with one parent. Of these, 16.5 million lived with their mother and 3.3 million with their father.

In 2002, fewer than half of single-parent children under the age of 18 received any financial support from the non-custodial parent. The income of more than one third of these households fell below the poverty level. The term "deadbeat dads" is often used in discussions about **abandonment** because most of the divorced parents who do not contribute financially to support their offspring are fathers.

Even though divorce rates peaked in 1979–81 and decreased slightly in the years following, half of all first marriages and 60 percent of second marriages end in divorce. The divorce process is often more emotionally traumatic for the children than for the parents, because children are less able to cope with the separation. About half of all children do not see their fathers following a divorce and only a small percentage have spent the night in their fathers' homes in any given month.

Divorce is the termination of the **family** as a unit. The effects of divorce on children can usually be seen long before the divorce itself, when conflict between the parents can cause behavior changes in the children, even in preschoolers. After the divorce, the children's sense of loss often increases, leading to great sadness, depression, and anxieties, especially on special occasions, such as birthdays, holidays, and school events. The children's emotions depend on their age, but common feelings include sadness, anger, and **fear**. Often these feelings are manifested in behavior changes that are also age-related. Children may grieve the loss of the "traditional" family, and they mourn the loss of the noncustodial parent, typically but not always, the father.

Common childhood and adolescent reactions to parental divorce include a continuing desire for the parents to reunite; fears of desertion; feelings of guilt over having been responsible for the divorce; developmental regression; **sleep disorders**; and physical complaints. While researchers have found that some children recover from the trauma of divorce within one to three years, subsequent long-term studies have documented persistent negative effects that can follow a child into **adolescence** and beyond, especially with regard to the formation of intimate relationships later in life. The effects of parental divorce have been linked to phenomena as diverse as emotional and behavioral problems, school dropout rates, crime rates, physical and sexual abuse, and physical health. However, mental health professionals continue to debate whether divorce is more damaging for children than the continuation of a troubled marriage.

Infancy

Infants' reactions to divorce come from interference with the satisfaction of their basic needs. The removal of the noncustodial parent or increased work hours for the custodial parent can cause **separation anxiety**, while the parents' emotional distress tends to be felt by babies, upsetting their own emotional balance. The inability of infants to understand the concept of divorce makes the changes in their situation seem frighteningly unpredictable and confusing. Reactions include irritability, increased crying, fearfulness, separation **anxiety**, and **sleep** problems.

Toddlerhood

Toddlers may revert to an earlier development stage in such areas as eating, sleeping, **toilet training**, motor activity, language, and emotional independence. Other

signs of distress include anger, fearfulness, **nightmares**, fantasies, and withdrawal.

Preschool

In preschool-age children, continued self focus, coupled with a more advanced level of **cognitive development**, leads to feelings of guilt as these children may become convinced that they are the reason for their parents' divorce. Children at this age are also prone to powerful fantasies, which can include imagined scenarios involving abandonment or punishment. The disruption that follows divorce, particularly in the relationship with the father, also becomes an important factor for children at this age. Developmental regression may take the form of insisting on sleeping in the same room or bed as the parent; refusing to eat all but a few types of food; **stuttering** or reverting to baby talk; disruptions in toilet training; and developing an excessive emotional dependence on one parent.

School age

By the early elementary grades, children are better able to handle separation from the noncustodial parent. Their greater awareness of the divorce situation, however, may lead to elaborate and frightening fantasies of abandonment or of being replaced in the affections of the noncustodial parent. Typical reactions at this stage include sadness, depression, anger, and general anxiety. Disruption of basic development in such areas as eating, sleeping, and elimination is possible but less frequent than in younger children. Many children this age suffer a sharp decline in academic performance, which often lasts throughout the entire school year in which the divorce takes place.

Children in the upper elementary grades are capable of better understanding of the divorce. At this age, the simple fears and fantasies of the younger child are replaced by more complex internal conflicts, such as the struggle to preserve one's allegiance to both parents. Older children become adept at erecting defense mechanisms to protect themselves against the **pain** they feel over a divorce. Such defenses include denial, displacement of feelings, and physical complaints such as fatigue, headaches, and stomachaches. Children in the upper elementary grades are most likely to become intensely angry at their parents for divorcing. Other common emotions at this stage of development include loneliness, grief, anxiety, and a sense of powerlessness.

For teenagers, divorce is difficult because it is yet another source of upheaval in their lives. Teenage behavior is affected not only by recent divorces but also by those that occurred when the child was much younger. One especially painful effect of divorce on adolescents is the negative attitude it can produce toward one or both parents, whom they need as role models but are often blamed for disappointing them.

Teens are also prone to internal conflicts over their parents' divorce. They are torn between love for and anger toward their parents and between conflicting loyalties to both parents. Positive feelings toward their parents' new partners come into conflict with anxiety over the intimacy of these relationships, and the teenager's close affiliation with the custodial parent clashes with his or her need for increased social and emotional independence. Although children at all ages are distressed by parental divorce, during the teen years it can result in potentially dangerous behavior, including drug and alcohol abuse, promiscuous sexual activity, violence, and delinquency.

Children ages 12–15 need consistent support from both parents but may not accept equal time-sharing of their living arrangements. They may blame one or both parents and may become controlling by demanding to stay in one place or to switch residences constantly.

Youths ages 15–18 group may become focused on establishing their independence and on social and school activities, and they may become intolerant of their parents' problems. Although teens still needs parental support, they may also tire of worrying about one or both parents. Being able to listen to teens when they are able to talk about their feelings may be helpful. Although teens may want to see their parents happy, they may have mixed feelings about seeing their parents dating other people. They may feel that condoning parental dating would be disloyal to the other parent. Older teens who need help may have behavior problems, exhibit depression, show poor school performance, run away from home, or get into trouble with the law.

Common problems

Not all children react the same way when told their parents are divorcing. Some ask questions, some cry or get angry, and some initially do not react at all. Problems to watch for include trouble sleeping, crying, aggression, deep anger and resentment, feelings of betrayal, difficulty concentrating, chronic fatigue, and problems with friends or at school.

Experts agree that it is important for parents who are divorcing to avoid involving their children in their disputes or forcing them to choose sides, and parents are often advised to avoid criticizing their former mates in front of their children. In order for children to heal from the emotional pain of parental divorce, they need an

outlet for open expression of their feelings, whether it is a sibling, friend, adult mentor or counselor, or a divorce support group. Extended families can be a significant source of support for children, providing them with stability and with the reassurance that others care about them. Although parental divorce is undeniably difficult for children of all ages, loving, patient, and enlightened parental support can make a crucial difference in helping children cope with the experience both immediately and over the long term.

Parental concerns

The custodial parent should be aware of the effects of the divorce on the child and above all, should reassure the child that the remaining parent will not abandon them. It is also important to maintain as much normalcy as possible after a divorce by sticking to regular routines, such as meal times, bedtime, rules of behavior, and methods of **discipline**. Relaxing limits during a time of change can make children feel insecure.

When to call the doctor

Medical help may be needed if a child inflicts self-injury. Psychological counseling may also be needed to help the child understand and cope with the divorce. This is especially true if any of the common reactions last for an unusual amount of time, intensify over time, or if the child talks about or threatens **suicide**.

Resources

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KEY TERMS

Custodial parent—A parent who has legal custody of their child or children.

Deadbeat dad—A father who has abandoned his child or children and does not pay child custody as required by a court.

Noncustodial parent—The parent who does not have legal custody of the child and does not live in the same home with the child. The noncustodial parent has financial responsibility for the child and visitation rights.

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Ken R. Wells

Dizziness

Definition

As a disorder, dizziness is classified into three categories: vertigo, syncope, and nonsyncope nonvertigo. Each category has its own set of symptoms, all related to the sense of balance. In general, syncope is defined by a brief loss of consciousness (fainting) or by dimmed vision and feeling uncoordinated, confused, and lightheaded. Many people experience a sensation like syncope when they stand up too fast. Vertigo is the feeling that either the individual or the surroundings are spinning. This sensation is like being on a spinning amusement park ride. Individuals with nonsyncope nonvertigo dizziness feel as though they cannot keep their balance. This feeling may become worse with movement.

Description

The brain coordinates information from the eyes, the inner ear, and the body's senses to maintain balance. If any of these information sources is disrupted, the brain may not be able to compensate. For example, people sometimes experience **motion sickness** because the information from their body tells the brain that they are sitting still, but information from the eyes indicates that they are moving. The messages do not correspond and dizziness results.

Vision and the body's senses are the most important systems for maintaining balance, but problems in the inner ear are the most frequent cause of dizziness. The inner ear, also called the vestibular system, contains fluid that helps fine tune the information the brain receives from the eyes and the body. When fluid volume or pressure in one inner ear changes, information about balance is altered. The discrepancy gives conflicting messages to the brain about balance and induces dizziness.

Certain medical conditions can cause dizziness because they affect the systems that maintain balance. For example, the inner ear is very sensitive to changes in blood flow. Because medical conditions such as high blood pressure or low blood sugar can affect blood flow, these conditions are frequently accompanied by dizziness. Circulation disorders are the most common causes of dizziness. Other causes are **head injury**, ear infection, **allergies**, and nervous system disorders.

Dizziness often disappears without treatment or with treatment of the underlying problem, but it can be long term or chronic.

Demographics

According to the National Institutes of Health, 42 percent of Americans seek medical help for dizziness at some point in their lives. The costs may exceed a billion dollars and account for 5 million doctor visits annually. Episodes of dizziness increase with age, and are common among the elderly.

Causes and symptoms

Careful attention to symptoms can help determine the underlying cause of the dizziness. Underlying problems may be benign and easily treated, or they may be dangerous and in need of intensive therapy. Not all cases of dizziness can be linked to a specific cause. More than one type of dizziness can be experienced at the same time, and symptoms may be mixed. Episodes of dizziness may last for a few seconds or for days. The length of an episode is related to the underlying cause.

The symptoms of syncope include dimmed vision, loss of coordination, confusion, lightheadedness, and sweating. These symptoms can lead to a brief loss of consciousness or fainting. They are related to a reduced flow of blood to the brain; they often occur when a person is standing up and can be relieved by sitting or lying down. Vertigo is characterized by a sensation of spinning or turning, accompanied by **nausea**, **vomiting**, ringing in the ears, **headache**, or fatigue. An individual may have trouble walking, remaining coordinated, or keeping balance. Nonsyncope nonvertigo dizziness is characterized by a feeling of being off balance that becomes worse if the individual tries moving or performing detail-intense tasks.

A person may experience dizziness for many reasons. Syncope is associated with low blood pressure, heart problems, and disorders in the autonomic nervous system, the system of involuntary functions such as breathing. Syncope may also arise from emotional distress, **pain**, and other reactions to outside stressors. Nonsyncope nonvertigo dizziness may be caused by rapid breathing, low blood sugar, or migraine headache, or by more serious medical conditions.

Vertigo is often associated with inner ear problems called vestibular disorders. A particularly intense vestibular disorder, Meniere's disease, interferes with the volume of fluid in the inner ear. This disease, which affects approximately one in every 1,000 people, causes intermittent vertigo over the course of weeks, months, or years. Meniere's disease is often accompanied by ringing or buzzing in the ear, hearing loss, and a feeling that the ear is blocked. Damage to the nerve that leads from the ear to the brain can also cause vertigo. Such damage can

result from head injury or a tumor. An acoustic neuroma, for example, is a benign tumor that wraps around the nerve. Vertigo can also be caused by disorders of the central nervous system and the circulatory system, such as hardening of the arteries (arteriosclerosis), **stroke**, or multiple sclerosis.

Some medications cause changes in blood pressure or blood flow. These medications can cause dizziness in some people. Prescription medications carry warnings of such side effects, but common drugs, such as **caffeine** or nicotine, can also cause dizziness. Certain **antibiotics** can damage the inner ear and cause hearing loss and dizziness.

Diet may cause dizziness. The role of diet may be direct, as through alcohol intake. It may be also be indirect, as through arteriosclerosis caused by a high-fat diet. Some people experience a slight dip in blood sugar and mild dizziness if they miss a meal, but this condition is rarely dangerous unless the person is diabetic. **Food sensitivities** or allergies can also be a cause of dizziness. Chronic conditions, such as heart disease, and serious acute problems, such as seizures and strokes, can cause dizziness. However, such conditions usually exhibit other characteristic symptoms.

When to call the doctor

A doctor should be called whenever a person experiences dizziness or other unusual state of mental confusion that does not spontaneously resolve within a few minutes.

Diagnosis

During the initial medical examination, an individual with dizziness should provide a detailed description of the type of dizziness experienced, when it occurs, and how often each episode lasts. A diary of symptoms may help track this information. The person should report any symptoms that accompany the dizziness, such as a ringing in the ear or nausea, any recent injury or infection, and any medication taken.

Blood pressure, pulse, respiration, and body temperature are checked, and the ear, nose, and throat are scrutinized. The sense of balance is assessed by moving the individual's head to various positions or by tilt-table testing. (In tilt-table testing, the person lies on a table that can be shifted into different positions and reports any dizziness that occurs.)

Further tests may be indicated by the initial examination. Hearing tests help assess ear damage. **X rays**, **computed tomography** scan (CT scan), and **magnetic resonance imaging** (MRI) can pinpoint evidence of nerve damage, tumor, or other structural problems. If a vestibular disorder is suspected, a technique called electronystagmography (ENG) may be used. ENG measures the electrical impulses generated by eye movements. Blood tests can determine diabetes, **high cholesterol**, and other diseases. In some cases, a heart evaluation may be useful. Despite thorough testing, however, an underlying cause cannot always be determined.

Treatment

Treatment is determined by the underlying cause. If an individual has a cold or **influenza**, a few days of bed rest is usually adequate to resolve dizziness. Other causes of dizziness, such as mild vestibular system damage, may resolve without medical treatment.

If dizziness continues, drug therapy may prove helpful. Because circulatory problems often cause dizziness, medication may be prescribed to control blood pressure or to treat arteriosclerosis. Sedatives may be useful to relieve the tension that can trigger or aggravate dizziness. Low blood sugar associated with diabetes sometimes causes dizziness and is treated by controlling blood sugar levels. An individual may be asked to avoid caffeine, nicotine, alcohol, and those substances that cause allergic reactions. A low-salt diet may also help some people.

When other measures have failed, surgery may be suggested to relieve pressure on the inner ear. If the dizziness is not treatable by drugs, surgery, or other means, physical therapy may be used and the person may be taught coping mechanisms for the problem.

Because dizziness may arise from serious conditions, it is advisable to seek medical treatment. Alternative treatments can often be used alongside conventional medicine without conflict. Relaxation techniques, such as **yoga** and **massage therapy**, that focus on relieving tension are popularly recommended methods for reducing stress. Aroma therapists recommend a warm bath scented with essential oils of lavender, geranium, and sandalwood.

Homeopathic therapies can work very effectively for dizziness and are especially applicable when no organic cause can be identified. An osteopath or chiropractor may suggest adjustments of the head, jaw, neck, and lower back to relieve pressure on the inner ear. Acupuncturists also offer some treatment options for acute and chronic cases of dizziness. Nutritionists may be able to offer advice and guidance in choosing dietary supplements, identifying foods to avoid and balancing nutritional needs.

KEY TERMS

Acoustic neuroma—A benign tumor that grows on the nerve leading from the inner ear to the brain. As the tumor grows, it exerts pressure on the inner ear and causes severe vertigo.

Arteriosclerosis—A chronic condition characterized by thickening, loss of leasticity, and hardening of the arteries and the build-up of plaque on the arterial walls. Arteriosclerosis can slow or impair blood circulation. It includes atherosclerosis, but the two terms are often used synonymously.

Autonomic nervous system—The part of the nervous system that controls so-called involuntary functions, such as heart rate, salivary gland secretion, respiratory function, and pupil dilation.

Computed tomography (CT)—An imaging technique in which cross-sectional x rays of the body are compiled to create a three-dimensional image of the body's internal structures; also called computed axial tomography.

Electronystagmography—A method for measuring the electricity generated by eye movements. Electrodes are placed on the skin around the eye and the individual is subjected to a variety of stimuli so that the quality of eye movements can be assessed.

Magnetic resonance imaging (MRI)—An imaging technique that uses a large circular magnet and radio waves to generate signals from atoms in the body. These signals are used to construct detailed images of internal body structures and organs, including the brain.

Vestibular system—The brain and parts of the inner ear that work together to detect movement and position.

Prognosis

Outcome depends on the cause of dizziness. Controlling or curing the underlying factors usually relieves dizziness. In some cases, dizziness disappears without treatment. In a few cases, dizziness can become a permanent disabling condition and a person's options are limited.

Prevention

Most people learn through experience that certain activities make them dizzy and they learn to avoid them. For example, if reading in a car produces motion sickness, an individual leaves reading materials for after arrival. Changes to the diet can also cut down on episodes of dizziness in susceptible people. Relaxation techniques can help ward off tension and **anxiety** that can cause dizziness.

These techniques can help minimize or even prevent dizziness for people with chronic diseases. For example, persons with Meniere's disease may avoid episodes of vertigo by omitting salt, alcohol, and caffeine from their diets. Reducing blood cholesterol can help diminish arteriosclerosis and indirectly treat dizziness.

Some cases of dizziness cannot be prevented. Acoustic neuromas, for example, were not as of 2004 predictable or preventable. When the underlying cause of dizziness cannot be discovered, it may be difficult to recommend preventive measures. Alternative approaches designed to rebalance the body's energy flow, such as acupuncture and constitutional homeopathy, may be helpful in cases where the cause of dizziness cannot be pinpointed.

Nutritional concerns

Persons who experience dizziness should limit alcohol intake and avoid diets that are high in fat. Persons with diabetes should eat their meals on a regular schedule. People for whom some foods cause allergic reactions or sensitivities or dizziness, should avoid consuming the offending substances.

Parental concerns

Parents should be alert for complaints from their children of dizziness or other states of mental confusion that do not spontaneously resolve within a minute or so.

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Down syndrome

Definition

Down syndrome is the most common cause of **mental retardation** and malformation in a newborn. A genetic disorder, it occurs because of the presence of an extra chromosome.

Description

Chromosomes are units of genetic information that exist within every cell of the body. Twenty-three distinctive pairs, or 46 total chromosomes, are located within the nucleus (central structure) of each cell. When a baby is conceived by combining one sperm cell with one egg cell, the baby receives 23 chromosomes from each parent, for a total of 46 chromosomes. Sometimes, an accident in the production of a sperm or egg cell causes that cell to contain 24 chromosomes. This event is referred to as nondisjunction. When this defective cell is involved in the conception of a baby, that baby will have a total of 47 chromosomes. The extra chromosome in Down syndrome is labeled number 21. For this reason, the existence of three such chromosomes is sometimes referred to as trisomy 21.

In a very rare number of Down syndrome cases (about 1–2%), the original egg and sperm cells are completely normal. The problem occurs sometime shortly after fertilization; during the phase when cells are dividing rapidly. One cell divides abnormally, creating a line of cells with an extra chromosome 21. This form of genetic disorder is called a mosaic. The individual with this type of Down syndrome has two types of cells: those with 46 chromosomes (the normal number) and those with 47 chromosomes (as occurs in Down syndrome). Some researchers have suggested that individuals with this type of mosaic form of Down syndrome have less severe signs and symptoms of the disorder.

Another relatively rare genetic accident which can cause Down syndrome is called translocation. During cell division, the number 21 chromosome somehow breaks. A piece of the number 21 chromosome then becomes attached to another chromosome. Each cell still has 46 chromosomes, but the extra piece of chromosome 21 results in the signs and symptoms of Down syndrome. Translocations occur in about 3–4 percent of cases of Down syndrome.

Demographics

Down syndrome occurs in about one in every 800 to 1,000 births. It affects an equal number of boys and girls. Less than 25 percent of Down syndrome cases occur due to an extra chromosome in the sperm cell. The majority of cases of Down syndrome occur due to an extra chromosome 21 within the egg cell supplied by the mother (nondisjunction). As a woman's age (maternal age) increases, the risk of having a Down syndrome baby increases significantly. For example, at younger ages, the risk is about one in 4,000. By the time the woman is age 35, the risk increases to one in 400; by age 40 the

risk increases to one in 110; and by age 45 the risk becomes one in 35. There is no increased risk of either mosaicism or translocation with increased maternal age.

Causes and symptoms

While Down syndrome is a chromosomal disorder, a baby is usually identified at birth through observation of a set of common physical characteristics. Babies with Down syndrome tend to be overly quiet; less responsive; with weak, floppy muscles. Furthermore, a number of physical signs may be present. These include:

- flat appearing face
- small head
- flat bridge of the nose
- smaller than normal, low-set nose
- small mouth, which causes the tongue to stick out and to appear overly large
- upward slanting eyes
- extra folds of skin located at the inside corner of each eye, near the nose (called epicanthal folds)
- rounded cheeks
- small, misshapen ears
- small, wide hands
- an unusual, deep crease across the center of the palm (called a simian crease)
- a malformed fifth finger
- a wide space between the big and the second toes
- unusual creases on the soles of the feet
- overly flexible joints (sometimes referred to as being double-jointed)
- shorter than normal height

Other types of defects often accompany Down syndrome. About 30 to 50 percent of all children with Down syndrome are found to have heart defects. A number of different heart defects are common in Down syndrome, including abnormal openings (holes) in the walls that separate the heart's chambers (**atrial septal defect**, ventricular septal defect). These result in abnormal patterns of blood flow within the heart. The abnormal blood flow often means that less oxygen is sent into circulation throughout the body. Another heart defect that occurs in Down syndrome is called **tetralogy of Fallot**. Tetralogy of Fallot consists of a hole in the heart, along with three other major heart defects.

Malformations of the gastrointestinal tract are present in about 5–7 percent of children with Down syndrome. The most common malformation is a narrowed, obstructed duodenum (the part of the intestine into which the stomach empties). This disorder, called duodenal atresia, interferes with the baby's milk or formula leaving the stomach and entering the intestine for digestion. The baby often vomits forcibly after feeding and cannot gain weight appropriately until the defect is repaired.

Other medical conditions that occur in patients with Down syndrome include an increased chance of developing infections, especially ear infections and **pneumonia**; certain kidney disorders; thyroid disease (especially low or hypothyroid); hearing loss; vision impairment that requires corrective lenses; and a 20-times greater chance of developing leukemia (a blood disorder).

Development in a baby and child with Down syndrome occurs at a much slower than normal rate. Because of weak, floppy muscles (**hypotonia**), babies learn to sit up, crawl, and walk much later than their normal peers. Talking is also quite delayed. The level of mental retardation is considered to be mild-to-moderate in Down syndrome. The actual IQ range of Down syndrome children is quite varied, but the majority of such children are in what is sometimes known as the trainable range. This means that most people with Down syndrome can be trained to do regular self-care tasks, function in a socially appropriate manner in a normal home environment, and even hold simple jobs.

As people with Down syndrome age, they face an increased chance of developing the brain disease called Alzheimer's (sometimes referred to dementia or senility). Most people have a six in 100 risk of developing Alzheimer's, but people with Down syndrome have a one-in-four chance of the disease. Alzheimer's disease causes the brain to shrink and to break down. The number of brain cells decreases, and abnormal deposits and structural arrangements occur. This process results in loss of brain function. People with Alzheimer's have strikingly faulty memories. Over time, people with Alzheimer's disease lapse into an increasingly unresponsive state. Some researchers have shown that even Down syndrome patients who do not appear to have Alzheimer's disease have the same changes occurring to the structures and cells of their brains.

As people with Down syndrome age, they also have an increased chance of developing a number of other medical difficulties, including cataracts, thyroid problems, diabetes, and seizure disorders.

Children with Down syndrome. The disease is caused by trisomy 21, meaning their bodies' cells have an extra chromosome 21. *(© Lester V. Bergman/Corbis.)*

Diagnosis

Diagnosis is usually suspected at birth, when the characteristic physical signs of Down syndrome are noted. Once this suspicion has been raised, genetic testing (chromosome analysis) can be undertaken in order to verify the presence of the disorder. This testing is usually done on a blood sample, although chromosome analysis can also be done on other types of tissue, including skin. The cells to be studied are prepared in a laboratory. Chemical stain is added to make the characteristics of the cells and the chromosomes stand out. Chemicals are added to prompt the cells to go through normal development, up to the point where the chromosomes are most visible, prior to cell division. At this point, they are examined under a microscope and photographed. The photograph is used to sort the different sizes and shapes of chromosomes into pairs. In most cases of Down syndrome, one extra chromosome 21 will be revealed. The final result of such testing, with the photographed chromosomes paired and organized by shape and size, is called the individual's karyotype.

Treatment

As of 2004 no treatment is available to cure Down syndrome. Treatment is directed at addressing the individual concerns of a particular patient. For example, heart defects often times require surgical repair, as will duodenal atresia. Many Down syndrome patients need to wear glasses to correct vision. Patients with **hearing impairment** benefit from hearing aids.

In the mid 1900s, all Down syndrome children were quickly placed into institutions for lifelong care. Research shows, however, that the best outlook for children with Down syndrome is **family** life in their own home. This arrangement requires careful support and education of the parents and the siblings. Parents and other siblings face a life-changing event in receiving a new baby who has a permanent condition that will affect essentially all aspects of his or her development. Some community groups are committed to helping families deal with the emotional effects of this new situation. Schools are required to provide services for children with Down syndrome, sometimes in separate **special education** classrooms and sometimes in regular classrooms, a practiced called mainstreaming or inclusion.

Prognosis

The prognosis in Down syndrome is quite variable, depending on the types of complications (heart defects, susceptibility to infections, development of leukemia) of

each individual baby. The severity of the retardation can also vary significantly. Without the presence of heart defects, about 90 percent of children with Down syndrome live into their teens. People with Down syndrome appear to go through the normal physical changes of aging more rapidly, however. The average age at death for an individual with Down syndrome is about 50 to 55 years.

Still, in the early 2000s, the prognosis for a baby born with Down syndrome is better than ever before. Because of modern medical treatments, including **antibiotics** to treat infections and surgery to treat heart defects and duodenal atresia, life expectancy has greatly increased. Community and family support allows people with Down syndrome to have rich, meaningful relationships and in some cases to hold jobs.

Men with Down syndrome appear to be uniformly sterile (meaning that they are unable to have offspring). Women with Down syndrome, however, are fully capable of having babies. About 50 percent of these babies, however, will also be born with Down syndrome.

Prevention

Efforts at prevention of Down syndrome are aimed at genetic counseling of couples who are preparing to have babies. A counselor needs to inform a woman that her risk of having a baby with Down syndrome increases with her increasing age. Two types of testing is available during a pregnancy to determine if the baby being carried has Down syndrome.

Screening tests are used to estimate the chance that an individual woman will have a baby with Down syndrome. At 14–17 weeks of pregnancy, measurements of a substance called AFP (alpha-fetoprotein) can be performed. AFP is normally found circulating in the blood of a pregnant woman but may be unusually high or low with certain disorders. Carrying a baby with Down syndrome often causes AFP to be lower than normal. This information alone, or along with measurements of two other hormones, is considered along with the mother's age to calculate the risk of the baby being born with Down syndrome. These results are only predictions and are only correct about 60 percent of the time. Other screening tests measure and compare the levels of other markers present in the mother's blood. A specialized ultrasound exam measures the thickness of the back of the fetus's neck (called nuchal lucency). Thicker measurements correlate with the possibility of Down syndrome or other chromosomal abnormalities.

All of these screening tests are used to decide which mothers will be offered other, more definitive testing to ascertain whether the baby has Down syndrome. These more definitive tests each carry a risk of miscarriage, which is why screening tests are an important first step. The only way to definitively establish (with about 98–99% accuracy) the presence or absence of Down syndrome in a developing baby is to test tissue from the

KEY TERMS

Chromosome—A microscopic thread-like structure found within each cell of the human body and consisting of a complex of proteins and DNA. Humans have 46 chromosomes arranged into 23 pairs. Chromosomes contain the genetic information necessary to direct the development and functioning of all cells and systems in the body. They pass on hereditary traits from parents to child (like eye color) and determine whether the child will be male or female.

Karyotype—A standard arrangement of photographic or computer-generated images of chromosome pairs from a cell in ascending numerical order, from largest to smallest.

Mental retardation—A condition where an individual has a lower-than-normal IQ, and thus is developmentally delayed.

Mosaic—A term referring to a genetic situation in which an individual's cells do not have the exact same composition of chromosomes. In Down syndrome, this may mean that some of the individual's cells have a normal 46 chromosomes, while other cells have an abnormal 47 chromosomes.

Nondisjunction—An event that takes place during cell division in which a chromosome pair does not separate as it should. The result is an abnormal number of chromosmes in the daughter cells produced by that cell division.

Translocation—The transfer of one part of a chromosome to another chromosome during cell division. A balanced translocation occurs when pieces from two different chromosomes exchange places without loss or gain of any chromosome material. An unbalanced translocation involves the unequal loss or gain of genetic information between two chromosomes.

Trisomy—An abnormal condition where three copies of one chromosome are present in the cells of an individual's body instead of two, the normal number.

pregnancy itself. This is usually done either by **amniocentesis** or chorionic villus sampling (CVS). In amniocentesis, a small amount of the fluid in which the baby is floating is withdrawn with a long, thin needle. In chorionic villus sampling, a tiny tube is inserted into the opening of the uterus to retrieve a small sample of the placenta (the organ that attaches the growing baby to the mother via the umbilical cord, and provides oxygen and **nutrition**). Both amniocentesis and CVS allow the baby's own karyotype to be determined. A couple must then decide whether to use this information in order to begin to prepare for the arrival of a baby with Down syndrome or to terminate the pregnancy.

Once a couple has had one baby with Down syndrome, they are often concerned about the likelihood of future offspring also being born with the disorder. Most research indicates that this chance remains the same as for any other woman at a similar age. However, when the baby with Down syndrome has the type that results from a translocation, it is possible that one of the two parents is a carrier of that defect. (A carrier carries the genetic defect but does not actually have the disorder.) When one parent is a carrier of a translocation, the chance of future offspring having Down syndrome is greatly increased. The specific risk will have to be calculated by a genetic counselor.

Parental concerns

Parenting a child with Down syndrome can be both challenging and rewarding. Children with Down syndrome have a wide range of potential outcomes. Early intervention programs have been proven to be of great help in assisting these children in achieving the highest level of functioning possible. There are many support groups available for parents and siblings of Down syndrome children.

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ORGANIZATION

National Down Syndrome Congress. 1370 Center Drive, Suite 102 Atlanta, GA 30338 (800) 232-6372. Web site: <www.ndsccenter.org>

National Down Syndrome Society. 666 Broadway, 8th Floor, New York, NY 10012-2317. Web site: <www.ndss.org>.

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Doxycycline *see* **Tetracyclines**

Drawings

Definition

Children's drawings are visual representations made with crayons, markers, or pencils that are generated for pleasure but can also be used for therapeutic purposes or developmental **assessment**.

Description

Children's art, especially a drawing, represents one of the delights of childhood. The child's artistic endeavors are mainly produced for pleasure and the exploration of art media. They can also be used for developmental and therapeutic assessment.

Children's drawings obviously show artistic development and expression. In educational and clinical settings, they can be vehicles for assessing a child's personality, intellectual development, **communication skills**, and emotional adjustment. Children's drawings can also aid in helping to diagnose learning disabilities. Law enforcement officers, social workers, and counselors often have children draw traumatic events, especially when they lack the communication skills to explain what they have witnessed or experienced. Children may also feel distanced from the traumatic event by

drawing it and talking about what is happening in the picture, as if discussing a character in a book or on television.

Color analysis has often been a means of determining a child's emotional state. A lot of black or red recurring in a child's drawing may be a troublesome sign. Black often is an indication of depression or feeling hopeless or restricted. Red may indicate intense anger. Blues and greens are usually calm colors, and yellows and oranges often indicate cheerfulness. Therapists are not ordinarily concerned if a child does one drawing in one of the troublesome colors, but may want to investigate a series of dark drawings, especially if the content is also frightening or disturbing. Therapists may use the therapeutic session as a means of emotional release and may encourage a child to create drawings that express their deep fears and angers. Drawings in this case are not assessment instruments, but become therapeutic tools.

Stages of creative development

In 1975, Viktor Lowenfeld launched a theory of artistic development based on systematic creative and cognitive stages. Each stage demonstrated specific characteristics and had an age range. He encouraged the use of his artistic development stages in classrooms and as guides for parents.

These stages are as dependent on a child's exposure to art and art media as they are on a child's innate artistic ability or **fine motor skills**. It should be noted that because a child does not seem to go beyond a specific developmental stage, it does not mean that the child has a cognitive or developmental problem. This apparent arrest of development may be due to limited exposure to art, lack of interest, or fine-motor differences. Cultural values can also affect artistic expression and development, influencing content, art media, style, and symbolic meaning as represented in the child's view of the world.

The following stages are generalized from Lowenfeld's work and that of Betty Edwards. Both theories show children moving from scribbling through several stages to realistic art. Children may overlap stages, making drawings with elements of one stage while progressing or regressing to another. Generally, boys and girls will develop similarly in the initial stages. Whether any child progresses to the latter stages usually requires instruction of some kind.

SCRIBBLING STAGE The scribbling stage usually begins around two years old and lasts until the child is about four years of age. In some cases, it can begin as soon as a child can hold a fat crayon and make marks on paper, which is sometimes around 18 months old. At first, the child is interested only in watching the color flow on the paper. Some children are more interested in the marking itself and may even look away while scribbling. What results on the paper is accidental and often delights the child, even though it is indistinguishable to adults.

With about six months of practice, the child will be more deliberate and may start drawing circles. Later, the child will name the drawing, saying, "This is a dog." The child may even look at the drawing of the dog the next day and say, "This is Daddy." The child will also start drawing people that resemble a tadpole or amoeba (a circle with arms and legs, and sometimes eyes).

PRE-SCHEMATIC STAGE The pre-schematic, or pre-symbolic, stage begins around age four; however, it may start earlier or later, depending on the child's cultural and artistic experience. In this stage, the amoeba or tadpole people may have faces, hands, and even toes, but no bodies. These figures face front and often have big smiles. Omission of body details is not a sign that something is developmentally wrong. It just means that other things in the drawing of the person are more important. For example, heads are the first objects drawn and may continue to be bigger than other parts of the body. This is usually done because the child sees the head as being very important. The child eats, speaks, sees, and hears with parts of the head.

Colors are selected on whim and usually have no relationship with what is being drawn. Figures may be scattered all over the page, or the page turned in every direction as the figures fill the paper. Objects and figures may appear to float all over the page because children do not yet know how to express three-dimensional objects on a two-dimensional surface.

The child's self-portrait appears as an amoeba person, but it will usually be the biggest figure, appearing in the center of the page. The child may test different ways to draw a self-portrait before settling on one for a period of time. In this instance, art helps define a child's self image.

SCHEMATIC STAGE The schematic stage usually begins around seven years old and extends through age nine. At this time, the child has developed specific schema, or symbols for people and objects in his or her environment, and will draw them consistently over and over. Human figures have all necessary body parts. Arms and legs also fill out, instead of being stick-like. This is usually due to more body awareness and recognition of what body parts do; e.g. parts of the body help the child run, catch a ball, jump, etc. Adults usually have very long legs because that is how children see them.

Houses and people no longer float on the page. They are grounded by a baseline that acts as a horizon line. As the child continues to draw, there may be two or more baselines to show distance or topography. Children may also draw a series of pictures, like cartoon squares, to show action sequences over time. This seems to reflect a child's desire to tell stories with the drawings. By eight or nine years of age, children will often draw their favorite cartoon characters or superheroes.

REALISTIC OR GANG STAGE The realistic or gang stage begins around nine years old. Here, the child begins to develop more detail in drawing people and in determining perspective (depth or distance) in drawings. Shapes now have form with shadows and shading. The people they draw show varying expressions. Colors are used to accurately depict the environment, and more complex art materials may be introduced.

Children at this stage are eager to conform and are very sensitive to teasing or criticism from classmates. They also are very critical of their work, individually or when it is compared to the work of others. Children at this stage can be easily discouraged about creating art if they are overly criticized, teased by their peers, or become frustrated with art media or problems expressing what they see in their minds. This is the time to begin quality art instruction, where children receive the technical training in mastery of art media, perspective, figure drawing, and rendering (shading).

Somewhere between ages 12 and 16 years, children face a crisis in artistic development. They will either already have enough skill and encouragement to continue a desire to create art, or they will not. If it is only a matter of training, finding appropriate art classes will help the child through this crisis. If the child has been discouraged by criticism or lack of enough art experience or exposure, the child may not continue to draw or partcipate in visual art activities. Some discouraged children may change to a different art medium. For example, a child may not draw or paint again, but may enjoy making clay pots or welding metal sculptures. Other children will find alternate ways to express their **creativity**. For example, a child may become involved with auto detailing, fly-tying, sewing, or needlework. Still others will never participate in any other kind of artistic activity and may ridicule or disdain those who do.

Common problems

When to call the doctor

Generally, children's drawings are no cause of alarm, despite color choice or content. They are merely artistic expressions and may present a variety of emotions, representations, and themes that are explored and then discarded.

Nevertheless, if a young child is repeatedly drawing violent pictures, there may be reason to seek out a therapist for the child to see if deeper emotional issues exist. For teenagers, especially those who are artistic, entertaining a dark period or even a quasi-violent Goth or vampire series of art work may simply be artistic exploration of darker themes. If this period of art work is coupled with risky behaviors or depression, it may represent a cry for help and therapy may be appropriate.

Other indicators of possible emotional problems may be drawings of a particular object or person much bigger than a drawing the child makes of himself or herself, or a drawing of a human figure in disjointed parts. In these cases, a child should be evaluated by a therapist because drawings of this sort usually indicate being overwhelmed by something or feeling fragmented. Drawings with incomplete or hesitant lines may indicate that a child feels unsure or insecure. Children who make these drawings may just need encouragement. Further evaluation may be necessary if these kinds of drawings continue for a long period of time.

Parental concerns

Since artistic expression and appreciation is an element of a balanced life, encouragement by parents and other adults is essential. Adults can encourage art expression by offering art materials to children at an early age. Even toddlers can make drawings with fat crayons, as crayons are non-toxic. Art materials should be good quality. The materials do not need to be expensive, but they should be good enough so that they perform as they are intended. For example, a child may be given a set of colored markers; but if they do not flow well or are dried up, the child can become discouraged because the tools do not function properly.

Children also enjoy experimenting with a variety of art materials. Using chalks, pastels, charcoal, and pencils of different softness expands the artistic possibilities that crayons and markers begin. This variety allows a child to explore different media and how they behave. No child is expected to become the master of any or all of these media, but the experience with each helps them expand their artistic voice and opens up greater appreciation for artwork by others found in museums or created by their fellow classmates.

Adults can encourage artistic expression by allowing children to use the media they have experimented with in ways that are truly unique. Adults can make sure that children know that drawings are not always

Drawing by a young child depicting a family. *(© Royalty-Free/ Corbis.)*

KEY TERMS

Drawings—Visual representations made with crayons, markers, or pencils.

Perspective—The way an artist shows depth or distance in a drawing or painting, usually by drawing figures and buildings larger in the front of the picture and smaller in the back.

Rendering—An artist's term for shading or creating texture or shape with markings, usually made with pencil, charcoal, ink, or paint.

supposed to look like photographs, but are each person's view of the world. Children's drawings become expressions of how and what each child sees. Adults can help children understand that art is self expression and that there is nothing wrong with what the child chooses to express. Artistic risk taking, experimentation, and the development of meaning are intrinsic to making art, and children can begin to understand these concepts through their own artistic efforts.

Exposure to a variety of visual art at an early age can encourage a child's lifelong appreciation of art. This can be in the form of quality children's picture books that have beautiful illustrations. Trips to art galleries and museums can broaden a child's exposure to a variety of artists, styles, and content. Visiting artists at art shows or art fairs can also be a way to show children how artists work or handle different media. Adults can extend this exposure through discussions about the art works and talking about media or content.

Children's responses to their own drawings and their perception of the level of their competence is often affected by the attitudes of their peers and adults who react to their art work. Direct and indirect criticism of a child's drawings should be avoided. When children are very young, it is sometimes difficult for adults to figure out just what a child's drawing is about. In order to avoid quashing young talent or a child's **self-esteem** by commenting on the beautiful bee the child drew when it really was a dog, adults can praise the child for having made something wonderful and then ask the child to tell about the drawing. From the answer, the adult can then praise the child's work in context. For example, if the child brings a drawing of yellow and blue scribbles, the adult can say, "What beautiful colors! Tell me about your picture." If the child says the drawing is about a flying horse, the adult can respond, "What a graceful flying horse! Does he like to fly?" The adult can continue to engage the child in discussion about the horse, choices of color, reasons for drawing a flying horse that day, or how the child felt doing the drawing.

Criticism can occur constructively when children enroll in technical art classes. There is a context in the art education setting for mastery of art media and technique. The normal **preschool** or elementary classroom is not the place for this kind of critique. Many children have been so severely criticized by teachers that they never pick up art materials again and some are even turned away from appreciating anyone else's art.

See also Cognitive development.

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Janie Franz

Drowning *see* **Near-drowning**

Drug abuse *see* **Substance abuse and dependence**

Drug allergies/sensitivities

Definition

A drug allergy is an adverse reaction to a medication, often an antibiotic, that is mediated by the body's immune system. A drug sensitivity is an unusual reaction to a drug that does not involve the immune system.

Description

Adverse reactions to medication may be allergic reactions involving a child's immune system, individual sensitivities to a drug, or side effects of the drug itself. Some children are allergic or sensitive to drugs that are not harmful for most people. Some drugs, such as aspirin and penicillin or related **antibiotics**, may induce allergic reactions in some children and sensitivities in other children.

Drug allergies

Drug **allergies** account for 5–10 percent of all adverse reactions to medications. They occur when the immune system—designed to protect the body from foreign substances such as bacteria and viruses—recognizes a medication as a harmful substance that must be destroyed. Drugs often induce an immune response; however, the symptoms of an allergic reaction occur in only a small number of children. Although most allergic drug reactions have mild symptoms, on rare occasions they can be life-threatening.

Drug allergies are unpredictable. Most drug allergies develop within days or occasionally weeks of beginning a drug treatment. Although it is very unusual to develop an allergy after months of taking a drug, sometimes children develop a drug allergy after having received multiple doses of the drug.

Unlike other types of adverse drug reactions, the frequency and severity of allergic reactions to drugs usually are independent of the amount of drug that is administered. Even a very small amount of a drug can trigger an allergic reaction.

Many classes of drugs can induce allergic reactions, resulting in a wide variety of symptoms affecting various tissues and organs. The likelihood that a drug will cause an allergic reaction depends in part on the chemical properties of the drug. Larger drug molecules are more likely to cause allergic reactions than smaller drug molecules. Larger drug molecules include the following:

- insulin
- antiserum which contains large immune-system proteins called antibodies
- recombinant proteins produced by genetic engineering

Unlike most other allergens, such as pollen or mold spores, drug molecules often are too small to be detected by the immune system. Smaller drugs such as antibiotics cannot induce an immune response unless they combine with a body cell or a carrier protein in the blood. Furthermore, drug allergies often are caused by the breakdown products or metabolites of the drug rather than by the drug itself. Sometimes the same drug, such as penicillin, can induce different types of allergic reactions.

IGE-MEDIATED ALLERGIES Most allergies, including most drug allergies, occur because of a reaction with an immune system antibody called immunoglobulin E (IgE). The first exposure to the drug sensitizes the child's immune system by inducing specialized white blood cells to produce IgE that recognizes the specific drug. On subsequent exposure to the drug, the drug-specific IgE antibodies bind to the drug on the surfaces of certain cells of the immune system. This binding activates the cells to release histamine and other chemicals that can cause a variety of symptoms. Thus, a child who has no reaction on first exposure to a drug may have a severe reaction with subsequent exposure.

Drug-specific IgE antibodies may cross-react with other drugs that have similar chemical properties, thereby triggering an allergic reaction, as is the case in the penicillin **family**. For example, the antibodies of a child allergic to penicillin may cross-react with the antibiotic amoxicillin or nafcillin.

Insect **stings** and the intravenous injection of certain drugs are the most common causes of

anaphylaxis, the most severe and frightening allergic response. Anaphylaxis involves the entire body. Although it is rare, several hundred Americans die of anaphylaxis every year. Anaphylaxis is most common in children who are allergic to **penicillins** and similar drugs. These drugs cause 97 percent of all deaths from drug allergies.

OTHER TYPES OF DRUG ALLERGIES Some drug allergies occur via immune system components other than IgE. Cytotoxic/cytolytic drug allergies occur when a drug allergen that is associated with a cell membrane, usually a blood cell, interacts with other types of antibodies—called immunoglobulin G (IgG) or immunoglobulin M (IgM)—along with other immune system factors. These interactions damage or destroy body cells.

Immune complex drug allergies occur when a drug combines with antibodies and other immune system components to form complexes in the blood. These complexes can be deposited in blood vessels and on membranes, causing inflammatory reactions that may be either localized or throughout the body. For instance, serum sickness typically causes a rash and joint swelling after the offending drug is administered.

T-cell-mediated allergic drug reactions require immune system cells called T-memory cells that are specific for the drug allergen. When exposed to the allergen, the T-cells are activated and cause an inflammatory response. The most common example of this type of reaction is allergic **contact dermatitis** that causes inflammations of the skin.

Drug sensitivities

Drug sensitivities (also called idiosyncratic reactions or unusual adverse reactions) do not involve the child's immune system or the release of histamine. However, the symptoms of drug sensitivities can be very similar to the symptoms of a drug allergy. Unlike drug allergies, sensitivities often occur upon first exposure to a drug and do not lead to anaphylaxis.

Demographics

Anyone can develop an allergy to any drug at any time. It is not clear why some children develop allergies to drugs that are well tolerated by most people. It is estimated that up to 10 percent of all people develop allergies to penicillin or other antibiotics at some point in their lives. Those taking multiple medications or frequent courses of antibiotics appear to be more at risk for developing drug allergies.

The most common drug sensitivity is to aspirin. Nearly 1 million Americans, primarily adults, are sensitive to aspirin. However, many medications, including aspirin and other non-steroidal anti-inflammatory drugs (NSAIDs) such as ibuprofen (Advil, Motrin, and others), can trigger an **asthma** attack in children. Asthma is a common chronic respiratory condition in children. Attacks occur when the air passages from the lungs to the nose and mouth are narrowed causing difficulty breathing. Aspirin and aspirin-like medications are common triggers for asthma attacks in as many as 30 percent of asthmatic children.

Causes and symptoms

Drug allergies

Any drug (either prescription or over-the-counter) can evoke an allergic reaction; however, antibiotics, especially penicillin and related drugs, are the most common cause of drug allergies. Children also frequently (i.e. to these agents more frequently than to other agents) develop allergies to the following:

- aspirin
- sulfa-based drugs
- barbiturates
- anticonvulsants
- insulin preparations, particularly those from animal sources
- dyes that are injected into blood vessels before taking x rays

The symptoms of a drug allergy vary from quite mild to life-threatening anaphylaxis. Unlike other common allergies, drug allergies often affect the entire body. The most common symptoms of a drug allergy are skin conditions including rash, generalized **itching**, and urticaria (**hives**; a very itchy rash with red swellings affecting only a small area of skin or the entire body; possibly the early symptom of anaphylaxis). The type of rash depends on the type of allergic response.

Less common symptoms of drug allergies include runny nose, sneezing, and congestion.

Uncommon but more serious symptoms of a drug allergy include the following:

- nausea, **vomiting**, diarrhea
- abdominal **pain** or cramps
- **fever**
- low blood cell count
- wheezing and difficulty breathing

- inflammation of the lungs, kidneys, and joints
- angioedema (a sudden swelling of the mucous membranes and tissues under the skin, anywhere on the body but especially on the face, eyes, lips, neck, throat, and genitals)

Angioedema occurs within a few minutes of exposure to the drug, often in conjunction with urticaria. Angioedema often is asymmetrical: for example, only one side of the lip may be affected. Swelling of the tongue, mouth, and airways can cause difficulty speaking, swallowing, or breathing. Angioedema can become life-threatening if the swelling affects the larynx (voice box) and the air passages become blocked. Emergency symptoms of a drug allergy include obstruction of the throat from swelling, severe asthma attack, and anaphylaxis.

Allergic reactions to drugs are the most common cause of an inflammation of the kidneys called tubulointerstitial nephritis. The allergic reaction and development of this acute condition may occur between five days and five weeks after exposure to penicillin, **sulfonamides**, diuretics (drugs to increase urination), and aspirin and other NSAIDs.

IGE-MEDIATED ALLERGIES IgE-mediated allergies can be caused by the following:

- penicillin when the allergic reaction is immediate
- blood factors, including antisera
- hormones
- vaccines (usually an allergic reaction to some component of the vaccine such as egg protein, gelatin, or neomycin, an antibiotic)
- very rarely, local anesthetics such as Novocain

The most common symptom of an IgE-mediated drug allergy is a rash that develops after the child has taken the drug for several days and produced antibodies against it.

ANAPHYLAXIS Anaphylaxis is a violent immune system reaction that can occur when a child who has large amounts of drug-specific IgE antibodies is re-exposed to the drug. The antibodies bind to the drug very rapidly causing an immediate, severe response. Anaphylaxis most often is caused by the following:

- penicillin and related antibiotics
- streptomycin
- tetracycline
- insulin

Analphylaxis usually begins within one to 15 minutes following exposure to the drug. Only rarely does the reaction begin an hour or more after exposure. Anaphylaxis can progress very rapidly leading to collapse, seizures, and loss of consciousness within one to two minutes. Without treatment, cessation of breathing, anaphylactic shock, and death can occur within 15 minutes. Any drug that has caused anaphylaxis in a child will probably cause it again on subsequent exposure, unless measures are taken to prevent it.

Symptoms of anaphylaxis include:

- urticaria on various parts of the body
- angioedema
- intense itching
- flushing of the skin
- coughing and sneezing
- nausea, vomiting, diarrhea
- abdominal pain or cramping
- tingling sensations
- ear throbbing
- heart palpitations
- uneasiness or sudden extreme **anxiety**
- swollen throat and/or constricted air passages causing a hoarse voice, wheezing, and difficulty breathing, the most characteristic symptom of anaphylaxis

Constriction of the air passages in the bronchial tract and/or throat, accompanied by shock, can cause a drastic drop in blood pressure that may lead to the following:

- rapid pulse
- paleness
- weakness
- dizziness, lightheadedness
- slurred speech
- mental confusion
- unconsciousness

OTHER DRUG ALLERGIES Cytotoxic/cytolytic-type drug allergies can be caused by the following:

- penicillin
- sulfonamides
- quinidine
- methyldopa

Cytotoxic/cytolytic-type of drug allergy can result in the following:

- immune hemolytic anemia due to the destruction of red blood cells
- thrombocytopenia from the reduction in blood platelets
- granulocytopenia from a deficiency of a type of white blood cell called a granular leukocyte

Drugs that can cause immune complex reactions, such as serum sickness or drug-induced lupus syndromes, include:

- hydralazine
- procainamide
- isoniazid
- phenytoin

Serum sickness (a delayed type of drug allergy that may take one to three weeks to develop) can be caused by an allergic reaction to penicillin or related antibiotics. Serum sickness also can be an allergic response to animal proteins present in an injected drug. Serum sickness is characterized by the following:

- fever
- aching joints
- swelling of the lymph nodes
- rash
- general body swelling
- skin lesions
- nephritis (an inflammation of the kidneys)
- hepatitis (an inflammation of the liver)

Some drugs, including penicillins and sulfonamides, can cause delayed dermatologic allergic reactions. These are various types of skin reactions, including eczema, that do not occur immediately upon exposure to the drug. These types of allergies are thought to be caused by metabolites formed from the breakdown or further reaction of the drug.

Drug allergies can result in hypersensitivity reactions, which in turn can result in liver disorders. Such damage can be caused by the following:

- sulfonamides
- phenothiazines
- halothane
- phenytoin
- isoniazid

Pulmonary hypersensitivity allergic reactions that affect the lungs and result in **rashes** and fever may be caused by nitrofurantoin and sulfasalazine.

Drug sensitivities

Children may have drug sensitivities to aspirin; other NSAIDs; opiates such as morphine and codeine; and some antibiotics, including erythromycin and ampicillin.

Symptoms of drug sensitivities often are very similar to those of drug allergies and include rashes, urticaria, and angioedema.

Anaphylactoid drug reactions are similar to anaphylactic reactions. However, they are caused by a drug sensitivity rather than a drug allergy and can occur upon the first exposure to a drug. Anaphylactoid reactions can occur in response to the following:

- opiates
- radiopaque dyes (radiocontrast media) used in x-ray procedures; 2–3% of patients have immediate generalized reactions to these dyes
- aspirin and other NSAIDs in some people, usually adults
- polymyxin
- pentamidine

When to call the doctor

A physician should be consulted whenever a child has an allergic reaction or sensitivity to a drug. The parent or caregiver should seek emergency assistance if a child has a severe or rapidly worsening allergic reaction to a drug that includes wheezing, difficulty breathing, or other symptoms of anaphylaxis.

Diagnosis

It is important to distinguish between an uncomfortable but mild side effect of a drug and an allergic reaction or sensitivity which could be life-threatening. A drug allergy or sensitivity most often is diagnosed by discontinuing the drug and observing whether the symptoms disappear.

Following a drug reaction the parent should describe the exact course of the reaction; the type of symptoms, when they occurred, and how long they lasted; and whether the child had previously been exposed to the drug. A previous allergic-type reaction to the medication usually is considered diagnostic of a drug allergy. A reaction upon a child's first exposure to the drug is probably a drug sensitivity.

Further diagnosis of a drug allergy may depend on the following:

- a complete medical history, including all drugs taken in the past month, when and how the child received certain drugs, and previous drug reactions
- whether the drug is known to cause allergic reactions
- a family history of drug allergies
- the timing of symptom-onset following drug exposure
- the timing of symptom-disappearance after discontinuing the drug
- the type of rash
- involvement of joints, lymph nodes, or liver
- associated viral infections
- other concurrent medications
- the presence of a chronic disease

Allergy tests

Skin prick tests or intra dermal tests to demonstrate IgE allergies are standardized for very few medications. Penicillin testing is standardized and can be used in extreme situations. Incremental drug challenge tests are also available for several drugs. These tests differ from tests for IgE antibodies but are still useful for demonstrating drug sensitivities. They must be done cautiously as patients are likely to have reactions during the challenge.

The allergist injects a tiny amount of the drug under the skin. If the child is allergic to the drug, swelling and itching occur at the site of injection within 15 to 20 minutes. Skin tests can be used to test for only a few drug allergies, for example, for penicillin and closely related antibiotics. Incremental challenge tests are performed for insulin, streptokinase, chymopapain, and antiserum.

Patch tests may be used to test for allergies to drugs that are applied to the skin such as **topical antibiotics**. A patch containing a small amount of the drug is applied to the skin to test for a localized reaction.

Desensitization is a test in which the allergist gives the child a tiny dose of the drug—as little as 0.001 or 0.00001 of the usual dose—in its usual form—orally, topically, or by injection. Gradually the dose is increased, and the child's reaction of observed. This procedure is done only in life-threatening situations, however, and only under close observation.

Treatment

Mild allergies/sensitivities

Drug allergies and sensitivities most often are treated by discontinuing the medication and replacing it with an alternative one. Mild symptoms usually disappear within a few days after discontinuation of the drug. Hives usually disappear within a few hours. Itchy rashes and hives may be treated with over-the-counter products such as oral **antihistamines**. Occasionally topical corticosteroid drugs are applied to the skin. Angioedema can take hours or days to subside; however, the swelling can be reduced with a corticosteroid or antihistamine.

Severe reactions

Severe immediate reactions occurring within one hour of drug administration, accelerated reactions occurring one to 72 hours after drug exposure, and late reactions (including rash, serum sickness, or fever) that develop more than 72 hours after drug exposure are all treated as follows:

- discontinuation of all nonessential suspect drugs
- antihistamines for hives and rashes
- oral corticosteroids for inflammation

Severe angioedema requires an immediate injection of epinephrine (a form of adrenaline) and further observation in a hospital.

Anaphylaxis requires an immediate injection of epinephrine into a thigh muscle. Epinephrine opens the air passageways and improves blood circulation. Intravenous fluids and injections of antihistamines or corticosteroids such as hydrocortisone also are administered. **Cardiopulmonary resuscitation** (CPR) and intubation may be necessary.

An asthma attack that is triggered by aspirin or other medications can be relieved by quick-relief or rescue medications. These include:

- epinephrine
- short-acting bronchodilators such as albuterol, proventil, ventolin, or xopenex
- prednisone for all moderate to severe reactions

Desensitization

Desensitization or immunotherapy sometimes is used by an allergy/immunology specialist to treat drug allergies to insulin, penicillin, or other antibiotics. Small amounts of the drug are injected or swallowed over a period of hours or a few days or in slowly increasing doses, to reduce sensitivity. Once antibiotic desensitization has been achieved, the full course of antibiotic treatment is followed. The procedure must be repeated if the drug has been discontinued for more than 72 hours.

KEY TERMS

Allergen—A foreign substance that provokes an immune reaction or allergic response in some sensitive people but not in most others.

Anaphylactoid—A non-allergic sensitivity response resembling anaphylaxis.

Anaphylaxis—Also called anaphylactic shock; a severe allergic reaction characterized by airway constriction, tissue swelling, and lowered blood pressure.

Angioedema—Patches of circumscribed swelling involving the skin and its subcutaneous layers, the mucous membranes, and sometimes the organs frequently caused by an allergic reaction to drugs or food. Also called angioneurotic edema, giant urticaria, Quincke's disease, or Quincke's edema.

Antibody—A special protein made by the body's immune system as a defense against foreign material (bacteria, viruses, etc.) that enters the body. It is uniquely designed to attack and neutralize the specific antigen that triggered the immune response.

Antihistamine—A drug used to treat allergic conditions that blocks the effects of histamine, a substance in the body that causes itching, vascular changes, and mucus secretion when released by cells.

Antiserum—Human or animal blood serum containing specific antibodies.

Corticosteroids—A group of hormones produced naturally by the adrenal gland or manufactured synthetically. They are often used to treat inflammation. Examples include cortisone and prednisone.

Cytotoxic—The characteristic of being destructive to cells.

Epinephrine—A hormone produced by the adrenal medulla. It is important in the response to stress and partially regulates heart rate and metabolism. It is also called adrenaline.

Histamine—A substance released by immune system cells in response to the presence of an allergen. It stimulates widening of blood vessels and increased porousness of blood vessel walls so that fluid and protein leak out from the blood into the surrounding tissue, causing localised inflammation of the tissue.

Immunoglobulin E (IgE)—A type of protein in blood plasma that acts as an antibody to activate allergic reactions. About 50% of patients with allergic disorders have increased IgE levels in their blood serum.

Radiopaque dyes, radiocontrast media—Injected substances that are used to outline tissues and organs in some x-ray and other radiation procedures.

Urticaria—An itchy rash usually associated with an allergic reaction. Also known as hives.

Sometimes desensitization is used for non-IgE-mediated drug reactions. Desensitization may take up to a month for the following:

- aspirin
- alloprinol
- gold
- sulfamethoxazole
- sulfasalazine

Prognosis

Mild symptoms of a drug allergy usually disappear without treatment within a few days of discontinuing the drug. Although children may lose their sensitivity to penicillin, if the reaction was urticarial or anaphylaxis, they are not re-challenged with the drug for safety reasons (i.e. it is not possible to predict who has lost sensitivity). In rare cases drug allergies may cause severe asthma attacks, anaphylaxis, or death.

Prevention

Drug allergies are unpredictable because they occur after a child has been exposed to the drug one or more times. The major prevention for known drug allergies and sensitivities is to avoid those drugs and to inform all physicians, hospital personnel, and dentists of the allergies or sensitivities before treatment. In the case of a serious drug allergy, the child should wear a medical alert necklace or bracelet or carry a card (Medic-Alert and others) at all times to alert emergency medical personnel.

Children with allergies or sensitivities to aspirin should avoid all aspirin-containing drugs. Such children usually can tolerate **acetaminophen** and non-acetylated salicylates such as sodium salicylate and salsalate.

If a child is allergic to a drug for which there is no substitute, sometimes the dosage can be reduced to prevent an allergic reaction. If the allergy is mild and the drug cannot be discontinued, the physician may decide to pretreat the allergy, with an antihistamine such as diphenhydramine or a corticosteroid such as prednisone, before the drug is administered to reduce or eliminate the allergic reaction. The physician also may "treat through" the allergy by prescribing antihistamines and corticosteroids during drug administration.

Some disorders cannot be diagnosed without the use of radiopaque dyes. Special dyes that reduce the risk of an anaphylactoid reaction can be used. Children at risk for reaction to such dyes may be premedicated with antihistamines and corticosteroids alone or in combination with beta-adrenergic agents before the dye is injected. Premedications include the following:

- prednisone
- diphenhydramine
- ephedrine

Parental concerns

When a child is given a new medication or starts a new course of treatment with a previous medication, parents should watch closely for symptoms of a drug allergy or sensitivity.

If a child suffers a mild to moderate allergic reaction or sensitivity to a drug, the parent should take the following steps:

- stay calm and reassure the child; anxiety can worsen the symptoms
- apply calamine lotion and cold cloths for an itchy rash; do not use medicated lotions
- observe the child for signs of increasing distress

If a child shows signs of a severe allergic reaction or sensitivity, the parent or caregiver should:

- inject allergy medication if it is available
- check the child's air passage, breathing, and circulation
- call 911 or other emergency assistance if the child is having difficulty breathing, becomes very weak, or loses consciousness
- begin rescue breathing or CPR if necessary
- calm and reassure the child
- inject emergency allergy medicine if available; do not give oral medication if the child is having difficulty breathing
- prevent shock by laying the child flat, elevating the feet, and covering the child with a coat or blanket

In the case of a severe allergic reaction, a parent should not:

- assume that any pretreatment with allergy medication will protect the child
- place a pillow under the child's head if the child is having trouble breathing since this could block the air passage
- give the child anything by mouth

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American Academy of Allergy, Asthma, & Immunology. 555 East Wells Street, Suite 1100, Milwaukee, WI 53202–3823. Web site: <www.aaaai.org>.

American College of Allergy, Asthma & Immunology. 85 West Algonquin Road, Suite 550, Arlington Heights, IL 60005. Web site: <www.acaai.org>.

Asthma and Allergy Foundation of America. 1233 20th Street NW, Suite 402, Washington, D.C. 20036. Web site: <www.aafa.org>.

National Institute of Allergy and Infectious Diseases. 6610 Rockledge Drive, MSC 6612, Bethesda, MD 20892–6612. Web site: <www.niaid.nih.gov>.

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DTP vaccine

Definition

DTP vaccine confers immunity to **diphtheria**, **tetanus**, and pertussis. The vaccine used in the United States is actually multiple diphtheria and tetanus toxoids combined with acellular pertussis (DTaP). The original vaccine, which as of 2004 was still used in other parts of the world, contains whole cells of *Bordatella pertussis*, the organism that causes pertussis, better known as **whooping cough**. The whole cell vaccine is more likely to cause adverse effects and does not provide any greater immunity.

Description

DTP vaccine conveys immunity to three different infectious diseases:

- Diphtheria is a potentially fatal disease that usually involves the nose, throat, and air passages, but may also infect the skin. Its most striking feature is the formation of a grayish membrane covering the tonsils and upper part of the throat. It is caused by *Corynebacterium diphtheriae*. Routine **vaccination** has almost eradicated diphtheria from the United States, but it is still seen in many parts of the world.
- Tetanus, sometimes called lockjaw, is a disease caused by the toxin of *Clostridium tetani*. The disease affects the central nervous system and causes painful muscle contractions. Food is not given by mouth to those with muscle spasm but may be given via nasogastric tube or intravenously. Tetanus is often fatal.
- Pertussis, also called whooping cough, is a respiratory disease caused by *Bordatella pertussis*. The name comes from a typical cough which starts with a deep inhalation, followed by a series of quick, short coughs that continues until the air is expelled from the lungs, and ends with a long shrill, whooping inhalation. Pertussis is very contagious and usually affects young children.

General use

Diphtheria and tetanus toxoids and acellular pertussis, taken together, provides immunity against diphtheria, tetanus, and whooping cough. The vaccine is normally given to children somewhere between the ages of two months and seven years of age (prior to their seventh birthday). Because these diseases can pose a severe problem in early childhood, the shots should be given as early in life as possible.

Precautions

DTP vaccine should not be given to children seven years of age or older. Moreover, children who are allergic to any component of the vaccine should not receive the drug. Because there are several different brands on the market, some children may be allergic to one brand and not to another. Because some of the bacterial cultures are grown in beef broth, the injections may be inadvisable for children who are allergic to beef. Children who have an allergic reaction after the first shot should be referred to an allergist before continuing with the DTP injections. Children who within a week after vaccination develop encephalopathy that cannot be traced to any other cause should not receive further injections. These children may be treated with DT (diphtheria-tetanus) vaccine. Also, DTP vaccine should be used with caution in patients who are receiving anticoagulant therapy. If a patient with a history of fevers and febrile convulsions is to be given DTP, the patient should receive **acetaminophen** at the time of the injection and for the following 24 hours.

Side effects

DPT vaccine has been associated with allergic reactions and with encephalopathy, both of which are rare but severe conditions. Other risks are common but minor:

- redness, irritation, and **itching** at injection site
- fever
- loss of appetite
- drowsiness
- irritability

Interactions

Because DTP vaccine is injected deep into the muscle, it should be given with care to patients receiving anticoagulant therapy. Also, immunosuppressant drugs, including steroids and **cancer** drugs, may reduce the ability of the body to produce antibodies in response to DTP vaccine.

Parental concerns

DTP is given in a series of four doses. Usually, the doses are given at two, four, and six months of age and at 17 to 20 months of age. While the customary age for the first dose is two months, it may be given as early as six weeks of age and up to the seventh birthday. The interval

KEY TERMS

Encephalopathy—Any abnormality in the structure or function of brain tissues.

Larynx—Also known as the voice box, the larynx is the part of the airway that lies between the pharynx and the trachea. It is composed of cartilage that contains the apparatus for voice production–the vocal cords and the muscles and ligaments that move the cords.

Toxin—A poisonous substance usually produced by a microorganism or plant.

between the third and fourth dose should be at least six months.

Although there have been warnings about severe, even fatal reactions to DTP vaccine, these reactions were seen in about one in 140,000 cases with whole cell DTP. The risk with DTaP is considerably lower. Children who had seizures due to the vaccine normally made a full recovery with no neurologic problems afterward. Five well-designed studies failed to show a link between DTP vaccine and any chronic nerve conditions.

The most serious risk of DTaP vaccine is a severe allergic reaction. These reactions, which also occur in response to other vaccines, are potentially fatal. Pre-term infants should be vaccinated according to their chronological age from birth. Interruption of the recommended schedule with a delay between doses should not interfere with the final immunity achieved. There is no need to start the series over again, regardless of the time between doses.

See also Vaccination.

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Duodenal obstruction

Definition

Duodenal obstruction is a partial or complete obstruction of the duodenum, the first part of the small intestine. Obstruction prevents food from passing through the digestive tract, interfering with digestion and **nutrition**.

Description

The duodenum is the first part of the small intestine, extending from the valve at the bottom of the stomach that regulates stomach emptying (pylorus valve) to the second part of the small intestine (jejunum). It is a short but often troublesome section of the digestive tract. The stomach, gallbladder, and pancreas each empty their contents into the duodenum in anticipation of digestion. Obstruction prevents the normal passage of stomach contents into the duodenum and keeps the gallbladder and pancreas from draining their secretions. This problem can lead to a number of conditions and complications involving digestion, nutrition, and fluid balance. In infants and children, congenital defects (anomalies)

usually cause duodenal obstruction, and symptoms are present at birth or shortly after when the infant attempts to feed.

When obstruction occurs, regardless of cause, food, gas, and secretions from within the intestine will accumulate above the point of obstruction, bloating (distending) the affected portion of intestine. Infection of peritoneal tissue lining the intestines and the abdomen (peritonitis) may result from bacteria growing in the accumulation of undigested material. As the distention increases, fluids continue to increase, and the intestine absorbs less. The fluid accumulation and reduced absorption lead to bilious **vomiting**, which is the vomitus will appear greenish, the classic sign of upper intestinal obstruction. Persistent vomiting or **diarrhea** (which can occur in a partial blockage) can result in **dehydration**. Fluid imbalances upset the balance of specific essential chemicals (electrolytes) in the blood, which can cause complications such as irregular heartbeat and, without correction of the electrolyte imbalance, shock.

In newborns, congenital duodenal obstruction can occur when the duodenal channel (duodenal lumen) is not correctly formed (recanalized) during fetal development. The duodenum may have a membrane reducing the channel size (lumen), or two blind pouches instead of one duodenal channel, or a gap or flap of tissue may be present. In each case, the channel is not be sufficiently developed at birth or sufficiently open to allow the passage of food and liquid, resulting in poor digestion and poor nutrition. This condition is known as duodenal atresia, and it results in duodenal obstruction. About 30 to 50 percent of infants born with duodenal atresia also have **Down syndrome**, and some have cardiac abnormalities as well. Duodenal atresia can occur with other conditions such as a narrowing of the duodenal lumen (duodenal stenosis) or twisting of the duodenum around itself (duodenal volvulus). It may also occur in combination with volvulus in another part of the bowel below the duodenum. Inflammation of the pancreas (pancreatitis) may also accompany duodenal atresia.

Malrotation of the duodenum is a more common cause of duodenal obstruction, typically appearing in the first few weeks of life. In malrotation, the duodenum is usually coiled to the right, causing obstruction of the duodenum and failure of the stomach contents to pass through to the next portion of small intestine. Malrotation may also involve the presence of Ladd's bands, abnormal folds or bands of tissue under tension across the lumen of the duodenum. Malrotation can also occur with duodenal volvulus or volvulus lower in the bowel. With volvulus, it can result in serious consequences by cutting off the supply of blood to a portion of bowel (strangulation), reducing the flow of oxygen to bowel tissue (ischemia), and leading to tissue death (gangrene) and shock or to rupture (perforation) of the intestine. Surgery is required immediately to correct this type of duodenal obstruction.

Demographics

Duodenal atresia, one of the causes of duodenal obstruction, affects one in 10,000 live births in the United States and is found equally among boys and girls and more often among premature births. Intestinal malrotation is a more common cause, occurring in one in 500 live births, although only a small percentage of these have duodenal malrotation. The male to female ratio is two to one in the first year of life and then becomes equal.

Causes and symptoms

Obstruction of the duodenum occurs in infants as a result of congenital causes. The duodenal channel may be underdeveloped (duodenal hypoplasia), narrowed (duodenal stenosis), or the duodenum channel may not be properly formed (duodenal atresia). Malrotation or coiling of the duodenum can also obstruct the duodenum, sometimes accompanied by volvulus, a twisting of the duodenum around itself. As of 2004 the specific cause of these congenital defects was not known.

Vomiting is the prevailing symptom of duodenal obstruction and may occur in the first day of life. The vomitus will be greenish (bilious) because it contains bile from the gallbladder. An infant will vomit feedings, lose weight, and be restless and irritable. Other symptoms may include difficulty breathing, excessive salivation and drooling, the presence of a palpable mass in the abdomen, yellow-tinted skin (**jaundice**), and failure to respond (lethargy). If the duodenum is twisted as in volvulus, the newborn may have a distended abdomen and bloody diarrhea.

When to call the doctor

Frequent or constant vomiting, unsuccessful feeding, and poor weight gain should be reported to the pediatrician as soon as noted. If an infant in the first few weeks of life pulls the knees up and intermittently cries in **pain** along with frequent vomiting, the pediatrician should be consulted immediately, and examination in the emergency department of the hospital may be necessary.

KEY TERMS

Anastomosis—Surgical reconnection of two ducts, blood vessels, or bowel segments to allow flow between the two.

Anomaly—Something that is different from what is normal or expected. Also an unusual or irregular structure.

Atresia—The congenital absence of a normal body opening or duct.

Bowel—The intestine; a tube-like structure that extends from the stomach to the anus. Some digestive processes are carried out in the bowel before food passes out of the body as waste.

Congenital—Present at birth.

Contrast agent—Also called a contrast medium, this is usually a barium or iodine dye that is injected into the area under investigation. The dye makes the interior body parts more visible on an x-ray film.

Electrolytes—Salts and minerals that produce electrically charged particles (ions) in body fluids. Common human electrolytes are sodium chloride, potassium, calcium, and sodium bicarbonate. Electrolytes control the fluid balance of the body and are important in muscle contraction, energy generation, and almost all major biochemical reactions in the body.

Gangrene—Decay or death of body tissue because the blood supply is cut off. Tissues that have died in this way must be surgically removed.

Hypoplasia—An underdeveloped or incomplete tissue or organ usually due to a decrease in the number of cells.

Ischemia—A decrease in the blood supply to an area of the body caused by obstruction or constriction of blood vessels.

Lumen—The inner cavity or canal of a tube-shaped organ, such as the bowel.

Peritonitis—Inflammation of the peritoneum. It is most often due to bacterial infection, but can also be caused by a chemical irritant (such as spillage of acid from the stomach or bile from the gall bladder).

Shock—A medical emergency in which the organs and tissues of the body are not receiving an adequate flow of blood. This deprives the organs and tissues of oxygen and allows the build-up of waste products. Shock can be caused by certain diseases, serious injury, or blood loss.

Diagnosis

Abdominal **x rays** will be performed and will typically show what is called the characteristic ''double bubble,'' a combination of air bubbles in the stomach and a dilated duodenum. An echocardiogram and chest x rays may be done to evaluate the infant for any other possible abnormalities, including cardiac defects and abnormal development of the pancreas, which is often associated with duodenal obstruction. If malrotation is suspected, contrast-enhanced x rays of the upper intestinal region are usually able to visualize the twisted duodenum. Ultrasound imaging may also be used to evaluate these conditions.

Diagnostic tests performed in the clinical laboratory will include a complete blood count (CBC), electrolytes (sodium, potassium, chloride), blood urea nitrogen (BUN), and other blood chemistries, especially to evaluate kidney and pancreas function. A urinalysis will be performed. Coagulation tests may be performed if the child is going to have surgery.

Treatment

Duodenal obstruction requires surgery, but it is not always urgent. Treatment may be delayed to evaluate or treat other life-threatening congenital anomalies. A nasogastric tube will first be placed through the infant's nose down into the stomach to decompress both the stomach and duodenum. Intravenous fluids may be given to maintain fluid levels and urine output or to correct dehydration that already has occurred. Electrolyte solutions may be given intravenously to restore electrolyte balance. Surgery to correct duodenal atresia is usually duodeno-duodenostomy. It involves opening the duodenum channel along its length from the stomach to the next portion of intestine, correcting the duodenal lumen end to end (gastrojejunal anastomosis) so that it is a fully open channel.

Ladd's procedure is used to surgically correct malrotation. The abdomen is opened and the large intestine is placed to the left side in order for the doctor to perform the surgery. The appendix is usually removed to avoid a later diagnosis of **appendicitis**. The malrotation, stenosis, or membranous bands are corrected surgically so that

the duodenum has a normal opening and connects properly to the stomach and jejunum. Broad-spectrum **antibiotics** may be given to help avoid infection.

Prognosis

Prognosis will depend on the type and extent of the obstruction, the infant's age at diagnosis, the infant's overall condition, and the presence and severity of any other congenital anomalies. Survival rates for surgical repair of the duodenum is greater than 90 percent, regardless of the cause. Most children do not have continuing digestive problems. Complications occur in 12 to 15 percent of those undergoing surgery. Complications can include other digestive disorders such as intestinal motility, duodenogastric reflux, gastritis, peptic ulcers, and megaduodenum. If malrotation or duodenal volvulus has caused the blood supply to be cut off in a portion of the intestine before surgery, death of intestinal tissue can result and life-threatening gangrene can develop. Widespread infection (peritonitis) may also develop from bacteria growing in the accumulation of undigested material above the obstruction. Mortality in infants who have gangrene or peritonitis is particularly high in those with other defects.

Prevention

No specific measures are recommended to prevent congenital anomalies that result in duodenal obstruction.

Parental concerns

In most cases, parents do not know before the birth of the child that an intestinal obstruction is present, although sometimes examination of the amniotic fluid during pregnancy (**amniocentesis**) alerts the obstetrician of possible abnormalities and prepares parents for the diagnosis. If obstruction is suspected or diagnosed in the first few days of the child's life, parents may be concerned about the risks associated with surgery and possible complications in infancy or early childhood. Parents can be reassured that newer surgical techniques have constantly improved the outcome of surgeries for intestinal obstruction, including duodenal obstruction. Diagnosed early, intestinal obstruction can be corrected with few complications, and a child who does not have other congenital problems usually is able to resume normal development.

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L. Lee Culvert

Dwarfism

Definition

Dwarfism is a term applied broadly to a number of conditions resulting in unusually short stature.

Description

While dwarfism is sometimes used to describe achondroplasia, a condition characterized by short stature and disproportionately short arms and legs, it is also used more broadly to refer to a variety of conditions resulting in unusually short stature in both children and adults. In some cases physical development may be disproportionate, as in achondroplasia, but in others the parts of the body develop proportionately. Short stature may be unaccompanied by other symptoms, or it may occur together with other problems, both physical and mental.

There are many conditions and diseases that can cause short stature. Some of these conditions involve a primary bone disorder, namely that the bones do not grow and develop normally. These conditions are called skeletal dysplasia. Over 500 specific skeletal dysplasias have been identified. Of these, Achondroplasia is the most common, affecting about 80 percent of all little people. An individual with achondroplasia has disproportionate short stature.

The four most common causes of dwarfism in children are achondroplasia, **Turner syndrome**, inadequate pituitary function (**pituitary dwarfism**), and lack of emotional or physical nurturance. Achondroplasia (short-limbed dwarfism) is a genetic disorder that

impairs embryonic development, resulting in abnormalities in bone growth and cartilage development. It is one of a class of illnesses called chondrodystrophies, all of which involve cartilage abnormalities and result in short stature. In achondroplasia, the long bones fail to develop normally, making the arms and legs disproportionately short and stubby (and sometimes curved). Overly long fibulae (one of two bones in the lower leg) cause the bowlegs that are characteristic of the condition. In addition, the head is disproportionately large and the bridge of the nose is depressed. Persons with achondroplasia are 3–5 feet (91–152 cm) tall and of normal **intelligence**. Their reproductive development is normal, and they have greater than normal muscular strength. The condition occurs in one out of every 10,000 births, and its prevalence increases with the age of the parents, especially the father. Many infants with the condition are stillborn.

Turner syndrome is a chromosomal abnormality occurring only in females in whom one of the X chromosomes is missing or defective. Girls with Turner syndrome are usually between 4.5 and 5 feet (137–152 cm) tall. Their ovaries are undeveloped, and they do not undergo **puberty**. Besides short stature, other physical characteristics include a stocky build and a webbed neck.

Pituitary dwarfism is a result of growth hormone deficiency. The deficiency may be genetic or the result of a severe brain injury. When untreated, skeletal growth is extremely slow, and puberty may or may not occur. Development can be normalized with the regular administration of synthetic hormones.

Parental neglect and malnourishment can cause a child to fail to grow properly. Infants in particular need physical comfort as well as caloric nourishment in order to thrive.

Demographics

Adult males under 5 feet (1.5 m) tall and females under 4 feet 8 inches (1.4 m) are classified as being short-statured. Children are considered unusually short if they fall below the third percentile of height for their age group. In 2004 there were approximately 5 million people of short stature (for their age) living in the United States, of whom 40 percent were under the age of 21.

Achondroplasia occurs in all races and with equal frequency in males and females and affects about one in every 40,000 children. The prevalence of Turner syndrome is widely reported as being approximately one per 2,500 live female births.

In 2004, more than 20,000 children in United States were receiving supplemental growth hormone (GH) therapy. It is estimated that about one-fourth of them had organic causes of GH deficiencies. There appears to be no racial or ethnic component to pituitary dwarfism, but males seem to be afflicted more often than females.

Causes and symptoms

Some prenatal factors known to contribute to growth retardation include a variety of maternal health problems, including toxemia, kidney and heart disease, infections such as **rubella** and maternal **malnutrition**. Maternal age is also a factor (adolescent mothers are prone to have undersize babies), as is uterine constraint (which occurs when the uterus is too small for the baby). Possible causes that center on the fetus rather than the mother include chromosomal abnormalities, genetic and other syndromes that impair skeletal growth, and defects of the placenta or umbilical cord. Environmental factors that influence intrauterine growth include maternal use of drugs (including alcohol and tobacco). Some infants who are small at birth (especially **twins**) may attain normal stature within the first year of life, while others remain small throughout their lives.

Endocrine and metabolic disorders are another important cause of growth problems. Growth can be impaired by conditions affecting the pituitary, thyroid, parathyroid, and adrenal glands (all part of the endocrine system). Probably the best known of these conditions is growth hormone deficiency, which is associated with the pituitary and hypothalamus glands. If the deficiency begins prenatally, the baby will still be of normal size and weight at birth but will then experience slowed growth. Weight gain still tends to be normal, leading to overweight and a higher than average proportion of body fat. The facial structures of children with this condition are immature, making them look younger than their actual age. Adults in whom growth hormone deficiency has not been treated attain a height of only about 2.5 feet (76 cm). They also have high-pitched voices, high foreheads, and wrinkled skin. Another endocrine disorder that can interfere with growth is **hypothyroidism**, a condition resulting from insufficient activity of the thyroid gland. Affecting one in 4,000 infants born in the United States, it can have a variety of causes, including underdevelopment, absence, or removal of the thyroid gland, lack of an enzyme needed for adequate thyroid function, iodine deficiency, or an under-active pituitary gland. In addition to retarding growth, it can cause **mental retardation** if thyroid hormones are not administered in the first months of an infant's life. If the condition goes untreated, it causes impaired mental development in 50 percent of affected children by the age of six months.

About 15 percent of cases of short stature in children is caused by chronic diseases, of which endocrine disorders are only one type. Many of these conditions do not appear until after the fifth year of life. Children with renal disease often experience growth retardation, especially if the condition is congenital. **Congenital heart disease** can cause slow growth, either directly or through secondary problems. Short stature can also result from a variety of conditions related to inadequate **nutrition**, including malabsorption syndromes (in which the body is lacking a substance—often an enzyme—necessary for proper absorption of an important nutrient), chronic inflammatory bowel disorders, caloric deficiencies, and zinc deficiency. A form of severe malnutrition called marasmus retards growth in all parts of the body, including the head (causing mental retardation as well). Marasmus can be caused by being weaned very early and not adequately fed afterwards; if the intake of calories and protein is limited severely enough, the body wastes away. Although the mental and emotional effects of the condition can be reversed with changes in environment, the growth retardation it causes is permanent. On occasion, growth retardation may also be caused solely by emotional deprivation.

When to call the doctor

Growth problems should be tracked and addressed by a doctor at a child's regular check-ups. If the child is consistently below the fifth percentile on standard growth charts or if a child stops growing at all, the parent(s) should discuss the implications with the child's pediatrician.

Diagnosis

Dwarfism is determined by direct measurement of a person's height. Achondroplasia can be detected through prenatal screening. **X rays** of the long bones may be performed in a newborn. Pituitary dwarfism can be diagnosed with blood tests for growth hormones or MRI of the head.

Treatment

Since growth problems are so varied, there is a wide variety of treatments for them, including nutritional changes, medications to treat underlying conditions, and, where appropriate, hormone replacement therapy. There is no specific treatment for achondroplasia, besides treating any orthopedic problems that may arise.

More than 150,000 children in the United States receive growth hormone therapy to remedy growth retardation caused by endocrine deficiencies. Growth hormone for therapeutic purposes was originally derived from the pituitary glands of deceased persons. However, natural growth hormone, aside from being prohibitively expensive, posed health hazards due to contamination. In the 1980s, men who had received growth hormone therapy in childhood were found to have developed Kreuzfeldt-Jakob disease, a fatal neurological disorder. Since then, natural growth hormone has been replaced by a biosynthetic hormone that received FDA approval in 1985.

Young female dwarf standing next to a boy of normal stature. *(Photograph by Dr. Richard Pauli. U. of Wisconsin, Madison, Clinical Genetics Center.)*

Prognosis

People who are short statured have approximately normal life expectancy. Administration of human growth

KEY TERMS

Achondroplasia—A congenital disturbance of growth plate development in long bones that results in a person having shortened limbs and a normal trunk.

Midget—An individual who is short statured but has normal body proportions. The term is considered to be offensive.

hormone may increase their adult height although they are unlikely to attain normal height. Those with achondroplasia seldom reach 5 feet (1.5 m) in height.

Prevention

There is no known way to prevent dwarfism because it results from genetic causes. Short stature as a result of parental neglect can be prevented. Education of the parents on the needs of the child is necessary, or the child may be removed from parental custody.

Nutritional concerns

Persons who have short stature should eat nutritionally sound, balanced meals. Their caloric requirements are slightly less than those of people who have normal height.

Parental concerns

Parents of children who are short statured should provide the same love and support as they would to any other child. In addition, they should offer counseling to help their children cope with their smaller stature. Adequate medical treatment should be provided to assure the best possible outcome.

See also Pituitary dwarfism; Turner syndrome.

Resources

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Little People of America Inc. PO Box 745, Lubbock, TX 79408. Web site: <www.lpaonline.org/>.

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Dyslexia

Definition

Dyslexia is a learning disability characterized by problems in reading, spelling, writing, speaking, or listening. It results from the inability to process graphic symbols. In many cases, dyslexia appears to be inherited.

Description

The word dyslexia is derived from the Greek word, *dys* (meaning poor or inadequate) and the word *lexis* (meaning words or language). Dyslexic children seem to have trouble learning early reading skills, problems hearing individual sounds in words, analyzing whole words in parts, and blending sounds into words. Letters such as "d" and "b" may be confused. Often a child with dyslexia has a problem translating language into thought (such as in listening or reading), or translating thought into language (such as in writing or speaking). Dyslexia is also referred to as developmental reading disorder (DRD).

Dyslexia is a problem involving higher (cortical) processing of symbols in the brain. Most children with dyslexia are of normal **intelligence**; many have above-average intelligence. However, when a child is dyslexic, there is often an unexpected difference between achievement and aptitude. Each child with dyslexia has different strengths and weaknesses, although many have unusual talents in art, athletics, architecture, graphics, drama, music, or engineering. These special talents are often in areas that require the ability to integrate sight, spatial skills, and coordination.

Common characteristics of a child with dyslexia include problems with:

- identifying single words
- understanding sounds in words, sound order, or rhymes
- spelling
- transposing letters in words
- handwriting
- reading comprehension
- the spoken language
- understanding directions
- understanding opposites, such as up/down or early/late

Social and emotional difficulties often accompany this disorder, as children are unable to meet expectations of parents and teachers and feel frustrated at their inability to achieve their goals. They may have a negative self-image and become angry, anxious, and depressed.

Demographics

About 15–20 percent of the population of the United States has a language-based learning disability. Of students with specific learning disabilities receiving **special education** services, 70–80 percent have deficits in reading. With such a high incidence, there is a question as to whether this is really a difference in learning style rather than a true "disability." The condition affects males more than females, and appears in all ages, races, and income levels.

Causes and symptoms

The underlying cause of dyslexia is not known, although research suggests the condition is often inherited. In 1999, The Centre for Reading Research in Norway presented the first research to study the largest **family** with reading problems ever known. By studying the reading and writing abilities of close to 80 family members across four generations, the researchers reported, for the first time, that chromosome 2 can be involved in the inheritability of dyslexia. When a fault occurs on this gene, it leads to difficulties in processing written language. Previous studies have pointed out linkages of other potential dyslexia genes to chromosome 1, chromosome 15 (DYX1 gene), and to chromosome 6 (DYX2 gene). The researchers who pinpointed the localized gene on chromosome 2 (DYX3) hope that this finding will lead to earlier and more precise diagnoses of dyslexia.

Research suggests a possible link with a subtle visual problem that affects the speed with which affected people can read. Anatomical and brain imagery studies show differences in the way the brain of a dyslexic child develops and functions.

Indicators of dyslexia include:

- possible family history of **learning disorders**
- difficulty learning to recognize written words
- difficulty rhyming
- difficulty determining the meaning (idea content) of a single sentence
- writing or arithmetic learning problems

When to call the doctor

The doctor should be called if a child appears to have difficulty learning to read or exhibits any symptoms of dyslexia.

Diagnosis

Anyone who is suspected to have dyslexia should have a comprehensive evaluation, including medical, psychological, behavioral, hearing, vision, and intelligence testing. The test should include all areas of learning and learning processes, not only reading. Other causes of learning disabilities, such as attention deficit hyperactivity disorder (ADHD), affective disorders (e.g. depression or **anxiety**), central auditory processing dysfunction, **pervasive developmental disorders**, and physical or sensory impairments, must be ruled out before the diagnosis of dyslexia can be confirmed. A child of any age may be evaluated for dyslexia using an age-appropriate battery of tests.

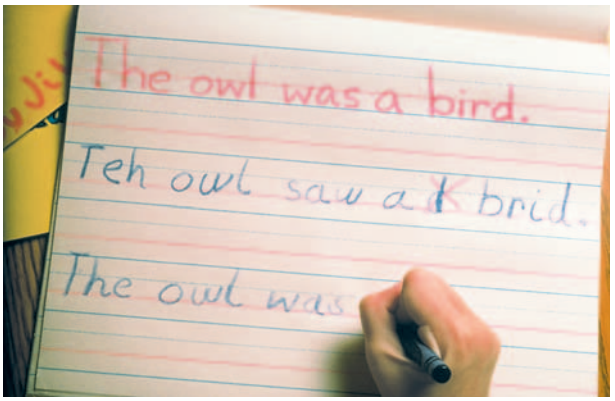
Test results are used to determine eligibility for special education services in many states as well as eligibility for programs in colleges and universities. They provide a basis for making educational recommendations, and determine the baseline for evaluation of improvement in the child's performance. In the United States, the Individuals with Disabilities Education Act (IDEA), Section 504 of the Rehabilitation Act of 1973 and the Americans with Disabilities Act (ADA) define the rights of children with dyslexia and other learning disabilities. These children are legally entitled to special services to help them overcome and accommodate their learning problems, including education programs to meet their needs. The Acts also protect people with dyslexia against unfair and illegal discrimination.

As further research pinpoints the genes responsible for some cases of dyslexia, there is a possibility that earlier testing will be established to allow for timely interventions to prevent the onset of the condition and treat it when it does occur. Unfortunately, in many schools, a child is not identified as having dyslexia until after repeated failures.

Treatment

Dyslexia is a life-long condition, but with proper intervention, a child can learn to read and/or write well. When a child is diagnosed with dyslexia, the parents should find out from the school or the diagnostician exactly what the problem is, what method of teaching is recommended, and why a particular method is suggested.

The primary focus of treatment is aimed at solving the specific learning problem of each affected child. Most often, this may include modifying teaching methods and the educational environment, since traditional educational methods will not always be effective with a dyslexic child. An Individual Education Plan (IEP) should be created for each child, reflecting his or her specific requirements. Special education services may include specialist help by an instructor specifically trained to teach dyslexic students through individualized tutoring or special day classes. It is important to teach these students using all the senses—hearing, touching, writing, and speaking—through a multi-sensory program.



A student with dyslexia has difficulty copying words. *(© Will & Deni McIntyre/Science Source, National Audubon Society Collection/Photo Researchers, Inc.)*

People with dyslexia need a structured language program, with direct instruction in the letter-sound system. Teachers must provide the rules governing written language. Most experts agree that the teacher should emphasize the association between simple phonetic units with letters or letter groups, rather than an approach that stresses memorizing whole words.

To assist with associated social and emotional difficulties, teachers must use strategies that will help the child find success in academics and personal relationships. Such strategies include rewarding efforts and not just the results, helping the child set realistic goals, and encouraging the child to do volunteer work that requires empathy and a social conscience (for example, a child with dyslexia who does well in science or math could serve as a peer tutor in those subjects or could tutor a younger child with dyslexia). Psychological counseling may also be helpful.

Prognosis

There is a great deal of variation among different people with dyslexia, producing different symptoms and degrees of severity. The prognosis depends on the severity of the disability, but is usually good if the condition is diagnosed early, the intervention used is effective and appropriate for the specific child, and if the child has a

KEY TERMS

Individualized educational plan (IEP)—A detailed description of the educational goals, assessment methods, behavioral management plan, and educational performance of a student requiring special education services.

Learning disorders—Academic difficulties experienced by children and adults of average to above-average intelligence that involve reading, writing, and/or mathematics, and which significantly interfere with academic achievement or daily living.

Spatial skills—The ability to locate objects in a three-dimensional world using sight or touch.

strong self-image and supportive family, friends, and teachers. However, difficulties with reading may persist throughout adulthood, which may result in occupational problems in certain careers. However, many successful people, such as Erin Brockovich and Whoopi Goldberg, have dyslexia.

Prevention

Since learning disorders often run in families, affected families should try to recognize learning disability problems early. For families without a previous history of learning disabilities, an intervention can begin as early as **preschool** or kindergarten if teachers detect early signs.

Parental concerns

There are many resources available to aid parents in helping their children. For example, the International Dyslexia Association (<http://interdys.org>) provides extensive information for parents, teachers, and children. Parents are encouraged to utilize these resources to ensure their child's success in school and in interactions with their peers and later as working adults. They must also guard against feeling that the child is lazy or not trying hard. Instead, they should provide a supportive and loving environment.

Dyslexia may have an impact upon the child's family. Non-dyslexic siblings may be jealous of the attention, time, and money the dyslexic child receives from the parents. Since dyslexia runs in families, one or both parents may have had similar school problems. The child's problems may bring back feelings of frustration and failure for parents, which may interfere with their parenting skills.

See also Language delay; Language disorders.

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ORGANIZATIONS

International Dyslexia Association. Suite 382, Chester Bldg., 8600 LaSalle Rd., Ste. 382, Baltimore, MD 21286-2044. (800) ABC-D123 or (410) 296-0232. <http://interdys.org/index.jsp/bibcit.composed>

Learning Disabilities Association. 4156 Library Rd., Pittsburgh, PA 15234-1349. (412) 341-1515; Fax: (412) 344-0224. <www.ldanatl.org>

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Dysmenorrhea

Definition

Dysmenorrhea refers to the **pain** or discomfort associated with **menstruation**. Although not a serious medical problem, the term describes a woman adolescent girl with menstrual symptoms severe enough to keep her from functioning for a day or two each month.

Description

Menstrual cramps are a common problem for adolescent girls and women. They may be mild, moderate, or severe and are the single most common cause of days missed from school and work. About 10 percent of girls are incapacitated for up to three days each month. Although many teens do not suffer from dysmenorrhea because their uterus is still growing, they may get it several years after their first period begins. The symptoms

may begin one to two days before menses, peak on the first day of flow, and subside during that day or over several days.

Causes and symptoms

Primary dysmenorrhea is the more common type of dysmenorrhea and is due to the production of prostaglandins. Prostaglandins are natural substances made by cells in the inner lining of the uterus and other parts of the body. Those made in the uterus make the uterine muscles contract and help the uterus to shed the lining that has built up during the menstrual cycle. It appears, however, that the level of prostaglandins has nothing to do with how strong a woman's cramps are. Some women have high levels of prostaglandins and no cramps, whereas other women with low levels have severe cramps. Thus cramps must also be related to something other than prostaglandins, such as genetics, stress, and different body types. The first year or two of a girl's periods are not usually very painful; however, once ovulation begins, the blood levels of the prostaglandins rise, leading to stronger contractions during menstruation. Prostaglandins can also cause headaches, **nausea**, **vomiting**, and **diarrhea**. The likelihood that a woman will have cramps increases if the following apply to her:

- She has a **family** history of painful periods.
- She leads a stressful life.
- She does not get enough **exercise**.
- She uses **caffeine**.
- She has pelvic inflammatory disease.

Primary dysmenorrhea usually presents during **adolescence**, within three years of menarche. It is unusual for symptoms to start within the first six months after menarche. Affected young women experience sharp, intermittent spasms of pain, usually centered in the suprapubic area. Pain may radiate to the back of the legs or the lower back. Systemic symptoms of nausea, vomiting, diarrhea, fatigue, **fever**, **headache**, or lightheadedness are fairly common. Pain usually develops within hours of the start of menstruation and peaks as the flow becomes heaviest during the first day or two of the cycle. Some women notice that painful periods disappear after having their first child. This could be due to the stretching of the opening of the uterus or the fact that birth improves the uterine blood supply and muscle activity.

Secondary dysmenorrhea is defined as menstrual pain due to pelvic pathology. This condition usually occurs after a woman has had normal menstrual periods for some time. It differs from primary dysmenorrheal in that the pain is caused by an abnormality or disease of the uterus, tubes, or ovaries. The most common causes are:

- pelvic inflammatory disease
- fibroids (intracavitary or intramural)
- intrauterine contraceptive devices
- endometriosis
- inflammation and scarring(adhesions)
- functional ovarian cysts
- benign or malignant tumors of ovary, bowel or bladder, or other site
- inflammatory bowel disease

Diagnosis

A focused history and physical examination are usually sufficient to make the diagnosis of primary dysmenorrhea. The history reveals the typical cramping pain with menstruation, and the physical examination is completely normal. A doctor should perform a thorough pelvic exam and take a patient history to rule out an underlying condition that could cause cramps. It is usually possible to differentiate dysmenorrhea from **premenstrual syndrome** (PMS) based on the patient's history. The pain associated with PMS is generally related to breast tenderness and abdominal bloating, rather than a lower abdominal cramping pain. PMS symptoms begin before the menstrual cycle and resolve shortly after menstrual flow begins.

Circumstances that may indicate secondary dysmenorrheal include the following:

- dysmenorrhea occurring during the first one or two cycles after menarche (congenital outflow obstruction)
- dysmenorrhea beginning after 25 years of age
- late onset of dysmenorrhea after a history without previous pain with menstruation (possibly caused by complications of pregnancy: ectopic or threatened spontaneous abortion)
- pelvic abnormality on physical examination; infertility (possible endometriosis, pelvic inflammatory disease or other causes of scarring); heavy menstrual flow or irregular cycles (consider adenomyosis, fibroids, polyps); dyspareunia
- little or no response to therapy with **nonsteroidal anti-inflammatory drugs**, **oral contraceptives**, or both

Treatment

Secondary dysmenorrhea is controlled by treating the underlying disorder.

The appropriate choice of therapy for most women with primary dysmenorrheal is a nonsteroidal anti-inflammatory drug (NSAIDs), which prevents the forma-

KEY TERMS

Adenomyosis—Uterine thickening caused when endometrial tissue, which normally lines the uterus, extends outward into the fibrous and muscular tissue of the uterus.

Endometriosis—A condition in which the tissue that normally lines the uterus (endometrium) grows in other areas of the body, causing pain, irregular bleeding, and frequently, infertility.

Hormone—A chemical messenger secreted by a gland or organ and released into the bloodstream. It travels via the bloodstream to distant cells where it exerts an effect.

Ovary—One of the two almond-shaped glands in the female reproductive system responsible for producing eggs and the sex hormones estrogen and progesterone.

Ovulation—The monthly process by which an ovarian follicle ruptures releasing a mature egg cell.

Progesterone—The hormone produced by the ovary after ovulation that prepares the uterine lining for a fertilized egg.

Uterus—The female reproductive organ that contains and nourishes a fetus from implantation until birth. Also called the womb.

tion and release of prostaglandins. Aspirin is not used for the treatment of dysmenorrheal because it is not potent enough in the usual dosage. Response to NSAIDs usually occurs within 30 to 60 minutes, but since individual response may vary, it is sometimes necessary to try different NSAIDs if the pain is not relieved with the first drug after one or two menstrual cycles. The NSAIDs include ibuprofen, naproxen (Aleve), and Motrin.

If an NSAID is not available, **acetaminophen** (Tylenol) may help ease the pain. Heat applied to the painful area may bring relief, and a warm bath twice a day also may help. Birth control pills are 90 percent effective in easing the pain of dysmenorrheal. They work by a twofold action: they reduce the menstrual fluid volume and suppress ovulation. They are generally not prescribed initially because it is a daily medication unless the woman also wants a birth control method. They may be chosen as a first line of therapy.

Alternative treatment

Simply changing the position of the body can help ease cramps. The simplest technique is assuming the fetal position, with knees pulled up to the chest while hugging a heating pad or pillow to the abdomen. Likewise, several **yoga** positions are popular ways to ease menstrual pain. In the "cat stretch," position, the woman rests on her hands and knees, slowly arching the back. The pelvic tilt is another popular yoga position, in which the woman lies on her back with knees bent and then lifts the pelvis and buttocks.

Dietary recommendations to ease cramps include increasing fiber, calcium, and complex carbohydrates, cutting fat, red meat, dairy products, caffeine, salt, and sugar. **Smoking** also has been found to worsen cramps. Some research suggests that vitamin B supplements, primarily vitamin B6 in a complex, magnesium, and fish oil supplements (omega-3 fatty acids) also may help relieve cramps.

Other women find relief through visualization, concentrating on the pain as a particular color, and gaining control of the sensations. Aromatherapy and massage may ease pain for some women. Others find that imagining a white light hovering over the painful area can actually lessen the pain for brief periods.

Exercise may be a way to reduce the pain of menstrual cramps through the brain's production of endorphins, the body's own painkillers. And orgasm can make a woman feel more comfortable by releasing tension in the pelvic muscles.

Acupuncture and Chinese herbs are additional alternative treatments for cramps.

Prognosis

Medication should lessen or eliminate pain by the end of three menstrual cycles. If it does not work, then a re-evaluation is necessary.

Prevention

NSAIDs taken one to two days before a period begins should eliminate cramps for some women.

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Linda K. Bennington

Dystonia *see* **Spasticity**

E

E. coli infection *see* **Enterobacterial infections**

Ear exam with otoscope

Definition

An otoscope is a hand-held instrument with a tiny light and a cone-shaped attachment called an ear speculum. It is used to examine the ear canal.

Purpose

An ear examination is a normal part of most routine physical examinations by a doctor or nurse. It is also done when an ear infection or other type of ear problem is suspected. An otoscope allows the doctor to look into the ear canal to see the ear drum. Redness or fluid in the eardrum can indicate an ear infection. Some otoscopes (called pneumatic otoscopes) can deliver a small puff of air to the eardrum to see if the eardrum will vibrate (which is normal). An ear examination with an otoscope can also detect a build-up of wax in the ear canal or a rupture or puncture of the eardrum.

Description

An ear examination with an otoscope is usually done by a doctor or a nurse as part of a complete physical examination. The ears may also be examined if an ear infection is suspected due to **fever**, ear **pain**, or hearing loss. The child will often be asked to tip the head slightly toward the shoulder opposite of the ear being examined, so the ear to be examined is pointing up. The doctor or nurse may hold the ear lobe as the speculum is inserted into the ear and may adjust the position of the otoscope to get a better view of the ear canal and eardrum. Both ears are usually examined, even if there seems to be a problem with just one ear.

The ear canal is normally skin-colored and is covered with tiny hairs. It is normal for the ear canal to have some yellowish-brown earwax. The eardrum is typically thin, shiny, and pearly-white to light gray in color. The tiny bones in the middle ear can be seen pushing on the eardrum membrane like tent poles. The light from the otoscope will reflect off of the surface of the ear drum.

An ear infection will cause the eardrum to look red and swollen. In cases where the eardrum has ruptured, there may be fluid draining from the middle ear. A doctor may also see scarring, retraction of the eardrum, or bulging of the eardrum.

Precautions

No precautions are required. However, if an ear infection is present, an ear examination may cause some discomfort or pain. If there is an object lodged in the ear canal, pushing on the otoscope may push the object further into the ear and damage the eardrum.

Preparation

No preparation is required prior to an ear examination with an otoscope. The ear speculum, which is inserted into the ear, is cleaned and sanitized before it is used. Speculums come in various sizes, and the doctor or nurse selects the size that is most comfortable for the child's ear. Sometimes if there is an excessive build up of wax in the ear, the doctor or nurse will remove some of it so that the eardrum can be seen more clearly.

Aftercare

If an ear infection is diagnosed, the patient may require treatment with **antibiotics**. If there is a buildup

KEY TERMS

Ear speculum—A cone- or funnel-shaped attachment for an otoscope that is inserted into the ear canal to examine the eardrum.

Otoscope—A hand-held instrument with a tiny light and a funnel-shaped attachment called an ear speculum, which is used to examine the ear canal and eardrum.

Pneumatic otoscope—An otoscope that can also produce a small puff of air that vibrates the eardrum.

Doctor examining a boy's ear canal with an otoscope, an instrument with a tiny light and cone shaped attachment called an ear speculum. *(Photograph by SPL. Custom Medical Stock Photo, Inc.)*

of wax in the ear canal, it might be rinsed or scraped out.

Risks

This type of ear examination is simple and generally harmless. Caution should always be used any time an object is inserted into the ear. This process can irritate an infected external ear canal and can rupture an eardrum if performed improperly or if the patient moves suddenly. If an object lodged in the ear is what is causing discomfort, pushing in the otoscope without checking first may result in the object being pushed further into the ear, possibly causing damage to the eardrum or further irritating the ear canal.

Parental concerns

An ear exam with an otoscope can occasionally cause some discomfort if there is an ear infection or other ear problem. If a child has frequent ear infections a doctor may recommend getting an otoscope designed for in home use. In this case, the doctor will show the parent how to use it, and the parent is encouraged to practice on healthy adults before attempting to use it on the child.

See also Otitis media.

Resources

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Ear infection, middle *see* **Otitis media**
Ear infection, outer *see* **Otitis externa**
Ear tubes *see* **Myringotomy and ear tubes**
Ear wax impaction *see* **Cerumen impaction**
Eardrum perforation *see* **Perforated eardrum**

Early childhood education

Definition

Early childhood education consists of activities and/or experiences that are intended to effect developmental

changes in children prior to their entry into elementary school.

Description

Early childhood education (ECE) programs include any type of educational program that serves children in the **preschool** years and is designed to improve later school performance. In the second half of the twentieth century, the early education system in the United States grew substantially. This trend allowed the majority of American children to have access to some form of early childhood education.

There are several types of programs that represent early childhood education. They are also known by a variety of names, including preschool and pre-kindergarten (pre-K). One of the first early childhood education initiatives in the United States was the Head Start program, started in 1965. Head Start is a federal government education initiative that has provided children from low-income families free access to early education. It targets children of low socioeconomic status or those who qualify in some at-risk category. **Head Start programs** are funded by the federal Department of Health and Human Services.

Many early childhood education programs operate under the auspices of Title I of the Elementary and Secondary Education Act. Under Title I, local educational agencies apply to state agencies for approval of their program, and when approved, the programs are then funded with federal money. The No Child Left Behind Act (NCLB) of 2001 encourages the use of Title I, Part A funds for preschool programs, recognizing the importance of preparing children for entering school with the language, cognitive, and early reading skills that help them meet later academic challenges. In the school year of 2001–2002 approximately 300,000 children benefiting from Title I services were enrolled in preschool.

Other early childhood education programs may be run by private for-profit companies, churches, or as part of a private school curriculum. These programs are normally tuition-based.

Since the early 1990s, many states have developed options for children from middle- and upper-income families for receiving free preschool education. Georgia introduced the first statewide universal pre-K program, offering free early childhood education to all four-year-old children. New York and Oklahoma have also developed universal pre-K programs, and Florida voters have approved a constitutional amendment for a free preschool program to be available for all four-year-olds by 2005.

Nearly three-fourths of young children in the United States are involved in some sort of early childhood education. Some groups of children have higher rates of participation in early childhood education programs than others. Children living in low-income households are less likely to be enrolled in ECE than those children in families living above the poverty line. Black and white children enroll in these programs in higher numbers than Hispanic American children. Children with better-educated mothers are more likely than other children to participate.

Benefits of early childhood education

Early childhood education can produce significant gains in children's learning and development. High quality early childhood education assists many at-risk children in avoiding poor outcomes, such as dropping out of school. Although the benefits seem to cross all economic and social lines, the most significant gains are almost always noted among children from families with the lowest income levels and the least amount of formal education. However, whether these benefits are long lasting is disputed. Some studies focused on the IQ score gains of disadvantaged children in Head Start programs, but these gains seemed to be short-term. However, studies also indicate that ECE produces persistent gains on achievement test scores, along with fewer occurrences of being held back a grade and being placed in **special education** programs. Other long-term benefits include decreased crime and delinquency rates and increased high school graduation. One extensive study found that people who participated in ECE were less likely to be on welfare as adults compared to those who had not received any early childhood education.

All programs in early childhood education are not equally effective in promoting the learning and development of young children. Long-term benefits are usually seen only in high-quality early childhood education programs. A significant problem with early childhood education is that most programs available cannot be considered high quality. In addition, the most effective ones are unaffordable for most American families. The overall effectiveness of an early childhood program is dependent upon several factors: quality staff, an appropriate environment, proper grouping practices, consistent scheduling, and parental involvement. According to the U.S. Department of Education, some additional characteristics of a high-quality early education program are as follows:

- Children have a safe, nurturing and stimulating environment, with the supervision and guidance of competent, caring adults.

- Teachers plan a balanced schedule in which the children do not feel rushed or fatigued.
- The school provides nutritious meals and snacks.
- The program includes a strong foundation in **language development**, early literacy, and early math.
- The program contains a clear statement of goals and philosophy that is comprehensive and addresses all areas of child development.
- The program engages children in purposeful learning activities and **play**, instructed by teachers who work from lesson and activity plans.
- Balance exists between individual, small-group, and large-group activities.
- Teachers frequently check children's progress.
- The staff regularly communicate with parents and caregivers so that caregivers are active participants in their children's education.
- Preschools that operate for a full day on a year-round basis, thus providing children with two years of preschool, achieve better results than those that offer less intense services.

In high-quality preschool programs, observers should see children working on the following:

- learning the letters of the alphabet
- learning to hear the individual sounds in words
- learning new words and how to use them
- learning early writing skills
- learning about written language by looking at books and by listening to stories
- becoming familiar with math and science

Because of the potential benefits to children, some people support the idea of government-sponsored universal early childhood education programs. Those who support this movement do so for the following reasons:

- The private and social costs of failing children early in their lives can be high. The lifetime social costs associated with one high school dropout may be as high as $350,000. Even modest improvements may justify the costs of ECE.
- Some studies show that for every dollar invested in quality ECE citizens save about $7 or more on investment later on.
- There is a potential for less reliance on welfare and other social services. Government receives more tax revenue because there are more taxpaying adults.
- People should rethink the value of early childhood education because of increasing needs for a more highly educated workforce in the twenty-first century.
- Early intervention may prevent intergenerational poverty.

Opponents of universal government early childhood education give the following reasons for objecting to it:

- Evidence indicates that the positive effects from the fairly expensive and intensive pre-K programs tend to be short-term.
- The public schools are already fraught with problems, and providing a downward extension to three- and four-year-olds is ill conceived.
- Some studies show that premature schooling may potentially slow or reduce a child's overall development by reducing valuable play time.
- Additional studies show that quality early education could as of 2004 cost more than $5,800 per year. The government would be taxing many people who may not wish to pay for preschool for another family's children.

In spite of the controversies, demographic trends in the early 2000s indicate that early childhood education has become, and will continue to be, an important aspect of the U.S. educational system.

Parental concerns

Parents are often understandably concerned about the quality of the early childhood education programs available to them. By taking the time to investigate several schools, most parents find a program with which they and their child are comfortable.

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Deanna M. Swartout-Corbeil, RN

Early puberty *see* **Precocious puberty**

Ecstasy *see* **Stimulant drugs**

Eczema *see* **Dermatitis**

Edwards' syndrome

Definition

Edwards' syndrome is caused by an extra (third) copy of chromosome 18. The extra chromosome is lethal for most babies born with this condition. It causes major physical abnormalities and severe **mental retardation**. Very few children afflicted with this syndrome survive beyond the first year.

Description

Edwards' syndrome is associated with the presence of a third copy of chromosome number 18. Humans normally have 23 pairs of chromosomes. Chromosomes are numbered 1–22, and the 23rd pair is composed of the sex chromosomes, X and Y. A person inherits one set of 23 chromosomes from each parent. Occasionally, a genetic error occurs during egg or sperm cell formation. A child conceived with such an egg or sperm cell may inherit an incorrect number of chromosomes. In the case of Edwards' syndrome, the child inherits three (trisomy), rather than two, copies of chromosome 18. Ninety-five percent of the children are full trisomies, 2 percent are due to translocations, where only part of an extra chromosome is present (this may be hereditary), while 3 percent are mosaic trisomies, where the extra chromosome is present in some but not all of the cells.

Edwards' syndrome is usually fatal, with most babies dying before birth. Of those who do make it to birth, 20–30 percent die within one month. However, a small number of babies (less than 10 percent) live at least one year.

Edwards' syndrome is also referred to as trisomy 18, trisomy E, and trisomy 16–18. It is the second most common trisomy, after trisomy 21 (**Down syndrome**).

Demographics

Edwards' syndrome occurs in approximately one in every 5,000 live births and one in every 5,000 stillborn births; it affects girls more often than boys. Women older than their early thirties have a greater risk of conceiving a child with Edwards' syndrome, but it can also occur with younger mothers.

Causes and symptoms

Most children born with Edwards' syndrome appear weak and fragile, and they are often underweight. The head is unusually small and the back of the head is prominent. The ears are malformed and low-set, and the mouth and jaw are small (micrognathia). The baby may also have a **cleft lip** or **cleft palate**. Frequently, the hands are clenched into fists, and the index finger overlaps the other fingers. The child may have clubfeet, and toes may be webbed or fused.

Numerous problems involving the internal organs may be present. Abnormalities often occur in the lungs and diaphragm (the muscle that controls breathing), and blood vessel malformations are common. Various types of **congenital heart disease**, including ventricular septal defect (VSD), atrial septic defect (ASD), or PDA (**patent ductus arteriosus**), may be present. The child may have an umbilical or inguinal **hernia**, malformed kidneys, and abnormalities of the urogenital system, including undescended testicles in a male child (cryptochordism).

When to call the doctor

A child with Edwards' syndrome is likely to have many medical and development needs. Parents should develop good working relationships with their doctor, other specialists, and therapists, and should consult them as needed.

If a woman gives birth to a child with Edwards' syndrome and plans to have another child, a doctor as well as a genetic counselor should be consulted so that prenatal screening and genetic counseling can be conducted.

Diagnosis

Edwards' syndrome at birth may be diagnosed by the physical abnormalities characteristic to the syndrome. Physical examination of the infant may show arched type finger print patterns, while **x rays** may reveal a short breast bone (sternum). Definitive diagnosis is achieved through karyotyping, which involves drawing the baby's blood for a microscopic examination of the chromosomes. Using special stains and microscopy, individual chromosomes are identified, and the presence of an extra chromosome 18 is revealed.

Edwards' syndrome can be detected before birth. If a pregnant woman is older than 35, has a **family** history of genetic abnormalities, has previously conceived a child with a genetic abnormality, or has suffered earlier miscarriages, she may undergo tests to determine whether her child carries genetic abnormalities. Potential tests include maternal serum alpha-fetal protein analysis or screening, ultrasonography, **amniocentesis**, and chorionic villus sampling.

In addition, a pregnant woman carrying a child with Edwards' syndrome may have an unusually large uterus during pregnancy, due to the presence of extra amniotic fluid (polyhydramnios). An unusually small placenta may be noted during the birth of the child.

Treatment

There is no cure for Edwards' syndrome. Since babies with Edwards' syndrome frequently have major physical abnormalities, doctors and parents face difficult choices regarding treatment. Abnormalities can be treated to a certain degree with surgery, but extreme invasive procedures may not be in the best interests of an infant whose lifespan is measured in days or weeks. Medical therapy often consists of supportive care with the goal of making the infant comfortable, rather than prolonging life.

However, 5–10 percent of children with Edwards' syndrome do survive past the first year of life, and require appropriate treatment for the many chronic effects associated with the syndrome. Problems with muscle tone and nervous system abnormalities will affect the development of motor skills, possibly resulting in **scoliosis** (curvature of the spine) and esotropia (crossed eyes). Surgical interventions may be limited by child's cardiac health.

Constipation due to poor abdominal muscle tone is often a life-long problem for babies and children with Edwards' syndrome, resulting in fretfulness, discomfort, and feeding problems. Anti-gas medication, special milk formulas, stool softener medicines, **laxatives**, and suppositories are all possible treatments that the doctor may recommend to ease the discomfort of gas in the bowels or constipation. An enema should not given to the baby or child because it can deplete electrolytes and alter body fluid composition.

Children with Edwards' syndrome will exhibit severe developmental delays, but with early intervention through **special education** and therapy programs, they can attain some developmental milestones.

Children with Edwards' syndrome appear to have increased risk of developing a **Wilms' tumor**, a **cancer** of the kidney that primarily affects children. Therefore, it is recommended that older infants and children with Edwards' syndrome have a routine ultrasound of the abdominal cavity.

Other illnesses that may affect a child with Edwards' syndrome and that may require treatment include congenital heart disease, pulmonary **hypertension**, elevated blood pressure, apnea episodes, pneumonias, sinus infections, seizures, urinary tract infections, ear infections, and eye infections. Other abnormalities that may require consideration of medical or surgical intervention include club foot, facial clefts, **spina bifida**, and **hydrocephalus**.

Nutritional concerns

Babies with Edwards' syndrome generally have feeding problems related to difficulties in coordination of breathing, sucking, and swallowing. Many have a weak suck and uncoordinated swallow resulting in **choking** and sometimes **vomiting**. **Gastroesophageal reflux disease**, or GERD (the upward movement of small amounts of stomach contents to the esophagus or throat), aspiration (inhalation or trickle of fluids into the lungs), and oral facial clefts may also contribute to feeding difficulties. The baby should be referred to a feeding specialist to help with feeding problems. The specialist can show the parents how to position the baby's head up, in good body alignment, because a baby with Edwards' syndrome may have hyperextension of the head. This is a common condition that occurs before the baby has developed head control. It results in the elongation of throat muscles, making swallowing more difficult. Because of feeding difficulties, many babies with Edwards' syndrome are fed through a tube inserted through the nose or mouth, down through the esophagus, and into the stomach. Some babies eventually progress to bottle or breastfeeding, while others have a gastrostomy (G-tube) placed abdominally to prevent the trauma of tube insertion. Some children are fed both orally and through the tube.

The baby should be fed with pre-softened preemie nipples and given small amounts frequently. To help prevent reflux, the baby's head should be elevated about 30 degrees or more during feeding and for one to two hours after a feeding. If tolerated, high calories formulas or supplements may be fed to help the baby gain weight.

Prognosis

Most children born with Edwards' syndrome die within their first year of life. The average lifespan is less than two months for 50 percent of the children, and 90–

KEY TERMS

Amniocentesis—A procedure performed at 16-18 weeks of pregnancy in which a needle is inserted through a woman's abdomen into her uterus to draw out a small sample of the amniotic fluid from around the baby for analysis. Either the fluid itself or cells from the fluid can be used for a variety of tests to obtain information about genetic disorders and other medical conditions in the fetus.

Atrial septal defect—An opening between the right and left atria (upper chambers) of the heart.

Chorionic villus sampling—A procedure performed at 10 to 12 weeks of pregnancy in which a needle is inserted either through the mother's vagina or abdominal wall into the placenta to withdraw a small amount of chorionic membrane from around the early embryo. The amniotic fluid can be examined for signs of chromosome abnormalities or other genetic diseases.

Chromosome—A microscopic thread-like structure found within each cell of the human body and consisting of a complex of proteins and DNA. Humans have 46 chromosomes arranged into 23 pairs. Chromosomes contain the genetic information necessary to direct the development and functioning of all cells and systems in the body. They pass on hereditary traits from parents to child (like eye color) and determine whether the child will be male or female.

Karyotyping—A laboratory test used to study an individual's chromosome make-up. Chromosomes are separated from cells, stained, and arranged in order from largest to smallest so that their number and structure can be studied under a microscope.

Maternal serum analyte screening—A medical procedure in which a pregnant woman's blood is drawn and analyzed for the levels of certain hormones and proteins. These levels can indicate whether there may be an abnormality in the unborn child. This test is not a definitive indicator of a problem and is followed by more specific testing such as amniocentesis or chorionic villus sampling.

Patent ductus arteriosus—A congenital defect in which the temporary blood vessel connecting the left pulmonary artery to the aorta in the fetus doesn't close after birth.

Trisomy—An abnormal condition where three copies of one chromosome are present in the cells of an individual's body instead of two, the normal number.

Ultrasonography—A medical test in which sound waves are directed against internal structures in the body. As sound waves bounce off the internal structure, they create an image on a video screen. Ultrasonography is often used to diagnose fetal abnormalities, gallstones, heart defects, and tumors. Also called ultrasound imaging.

Ventricular septal defect—An opening between the right and left ventricles of the heart.

95 percent die before their first birthday. The 5–10 percent of children who survive their first year have severe developmental disabilities. They need support to walk, and learning is limited. Verbal communication is also limited, but they can respond to comforting and can learn to recognize, smile, and interact with caregivers and others, and acquire such skills as rolling over and self-feeding. They will have many physical abnormalities that require constant care as doctors and parents work together to prevent and treat various problems.

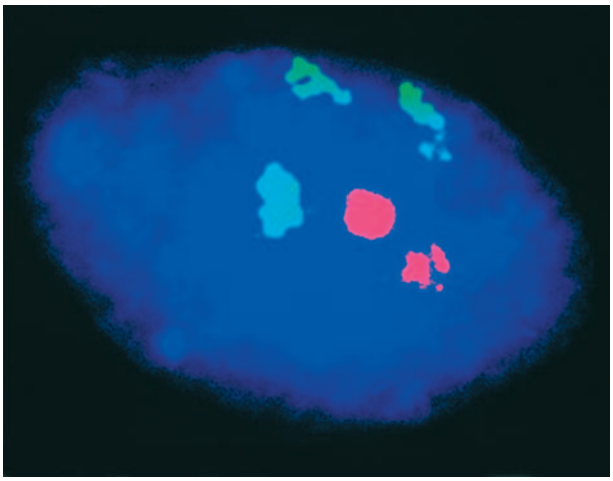
Prevention

Most cases of Edwards' syndrome are not hereditary and cannot be prevented. However, parents who have had a child with Edwards' syndrome are at increased risk of having another child with the syndrome.

Parental concerns

Following the birth of a child with Edwards' syndrome, families may wish to seek counseling regarding the effects on relationships within the family. Many people respond with guilt, **fear**, or blame when a genetic disorder is manifested within a family. Support groups are good sources of information about Edwards' syndrome and can offer emotional and psychological support.

For those families whose child does survive the first weeks of life, the doctor should review with the parents the expected clinical course of the condition, and prepare a management plan for use when the child goes home. Each case must be considered on an individual basis, and the doctor should acknowledge the personal feelings of the parents, as well as the individual circumstances of each child. The theme of "best interest of the child" helps



Micrograph showing trisomy 18, three copies of chromosome 18 (green) in cell's nucleus (blue) versus the normal two. The two fuschia spots are the sex chromosomes, XX, a female. *(© Department of Clinical Cytogenetics/Addenbrookes Hospital/Science Photo Library/Photo Researchers, Inc.)*

as a guiding principle in decision-making throughout the life of the child. There will be many challenges associated with the care of a child with Edwards' syndrome. As medical crises occur, parents will face decisions and emotions related to the possibility of the child dying.

See also Atrial septal defect; Congenital heart disease.

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EEG *see* **Electroencephalogram**

Ehlers-Danlos syndrome

Definition

The Ehlers-Danlos syndrome (EDS) refers to a group of inherited disorders that affect collagen structure and function. Genetic abnormalities in the manufacturing of collagen within the body affect connective tissues, causing them to be abnormally weak. Ehlers-Danlos syndrome is also referred to as inherited connective tissue disorder.

Description

Collagen is a strong, fibrous protein that lends strength and elasticity to connective tissues such as the skin, tendons, organ walls, cartilage, and blood vessels. Each of these connective tissues requires collagen tailored to meet its specific purposes. The many roles of collagen are reflected in the number of genes dedicated to its production. There are at least 28 genes in humans that encode at least 19 different types of collagen. Mutations in these genes can affect basic construction as well as the fine-tuned processing of the collagen.

EDS is a group of inherited connective tissue disorders that usually affects the skin, ligaments, joints, and blood vessels. Classification of EDS types was revised in 1997. The new classification involves categorizing the different forms of EDS into six major sub-types, including classical, hypermobility, vascular, kyphoscoliosis, arthrochalasia, and dermatosparaxis, and a collection of rare or poorly defined varieties. This new classification is simpler than the previous classification system and is primarily based on descriptions of the actual symptoms.

Classical type

Under the old classification system, EDS classical type was divided into two separate types: type I and type II. The major symptoms associated with EDS classical type involve the skin and joints. The skin has a smooth, velvety texture, and bruises easily. Affected individuals typically have extensive scarring, particularly at the knees, elbows, forehead, and chin. The joints are hyperextensible, giving a tendency toward dislocation of the hip, shoulder, elbow, knee, or clavicle. Due to decreased

muscle tone, affected infants may experience a delay in reaching motor milestones. Children may have a tendency to develop hernias or other organ shifts within the abdomen. **Sprains** and partial or complete joint dislocations are also common. Symptoms can range from mild to severe. EDS classical type is inherited in an autosomal dominant manner.

There are three major clinical diagnostic criteria for EDS classical type: skin hyperextensibility, unusually wide scars, and joint hypermobility. There is no definitive test for the diagnosis of classical EDS. Both DNA and biochemical studies have been used to help identify affected individuals. In some cases, skin biopsy has been found useful in confirming a diagnosis. Unfortunately, these tests are not sensitive enough to identify all individuals with classical EDS. If there are multiple affected individuals in a **family**, it may be possible to perform prenatal diagnosis using a DNA information technique known as a linkage study.

Hypermobility type

Excessively loose joints are the hallmark of this EDS type, formerly known as EDS type III. Both large joints, such as the elbows and knees, and small joints, such as toes and fingers, are affected. Partial and total joint dislocations are common, particularly involving the jaw, knee, and shoulder. Many individuals experience chronic limb and joint **pain**, although **x rays** of these joints appear normal. The skin may also bruise easily. Osteoarthritis is a common occurrence in adults. EDS hypermobility type is inherited in an autosomal dominant manner.

There are two major clinical diagnostic criteria for EDS hypermobility type. These include skin involvement (either hyperextensible skin or smooth and velvety skin) and generalized joint hypermobility. There is no test for this form of EDS.

Vascular type

Formerly called EDS type IV, EDS vascular type is the most severe form. The connective tissue in the intestines, arteries, uterus, and other hollow organs may be unusually weak, leading to organ or blood vessel rupture. Such ruptures most likely occur between ages 20 and 40, although they can occur any time, and may be life-threatening.

There is a classic facial appearance associated with EDS vascular type. Affected individuals tend to have large eyes, a thin pinched nose, thin lips, and a slim body. The skin is thin and translucent, with veins dramatically visible, particularly across the chest.

The large joints have normal stability, but small joints in the hands and feet are loose, showing hyperextensibility. The skin bruises easily. Other complications may include collapsed lungs, premature aging of the skin on the hands and feet, and ruptured arteries and veins. After surgery, there may be poor wound healing, a complication that tends to be frequent and severe. Pregnancy also carries the risk of complications. During and after pregnancy, there is an increased risk of the uterus rupturing and of arterial bleeding. Due to the severe complications associated with EDS type IV, death usually occurs before the fifth decade. A study of 419 individuals with EDS vascular type, completed in 2000, found that the median survival rate was 48 years, with a range of six to 73 years. EDS vascular type is inherited in an autosomal dominant manner.

There are four major clinical diagnostic criteria for EDS vascular type. These include thin translucent skin, arterial/intestinal/uterine fragility or rupture, extensive bruising, and characteristic facial appearance. EDS vascular type is caused by a change in the gene COL3A1, which codes for one of the collagen chains used to build Collagen type III. Laboratory testing is available for this form of EDS. A skin biopsy may be used to demonstrate the structurally abnormal collagen. This type of biochemical test identifies more than 95 percent of individuals with EDS vascular type. Laboratory testing is recommended for individuals with two or more of the major criteria.

DNA analysis may also be used to identify the change within the COL3A1 gene. This information may be helpful for genetic counseling purposes. Prenatal testing is available for pregnancies in which an affected parent has been identified and the DNA mutation is known or the biochemical defect has been demonstrated.

Kyphoscoliosis type

The major symptoms of kyphoscoliosis type, formerly called EDS type VI, are general joint looseness. At birth, the muscle tone is poor, and motor skill development is subsequently delayed. Also, infants with this type of EDS have an abnormal curvature of the spine (**scoliosis**). The scoliosis becomes progressively worse with age; affected individuals are usually unable to walk by age 20. The eyes and skin are fragile and easily damaged, and blood vessel involvement is a possibility. The bones may also be affected as demonstrated by a decrease in bone mass. Kyphoscoliosis type is inherited in an autosomal recessive manner.

There are four major clinical diagnostic criteria for EDS kyphoscoliosis type. These include generally loose joints, low muscle tone at birth, scoliosis at birth (which worsens with age), and a fragility of the eyes, which may give the white area of the eye a blue tint or cause the eye to rupture. This form of EDS is caused by a change in the PLOD gene on chromosome 1, which encodes the enzyme lysyl hydroxylase. A laboratory test is available in which urinary hydroxylysyl pyridinoline is measured. This test, performed on urine, is extremely sensitive and specific for EDS kyphoscolios type. Laboratory testing is recommended for infants with three or more of the major diagnostic criteria.

Prenatal testing is available if a pregnancy is known to be at risk and an identified affected family member has had positive laboratory testing. An **amniocentesis** may be performed in which fetal cells are removed from the amniotic fluid and enzyme activity is measured.

Arthrochalasia type

Dislocation of the hip joint typically accompanies arthrochalasia type EDS, formerly called EDS type VIIB. Other joints are also unusually loose, leading to recurrent partial and total dislocations. The skin has a high degree of stretchability and bruises easily. Individuals with this type of EDS may experience mildly diminished bone mass, scoliosis, and poor muscle tone. Arthrochalasia type is inherited in an autosomal dominant manner.

There are two major clinical diagnostic criteria for EDS arthrochalasia type. These include severe generalized joint hypermobility and bilateral hip dislocation present at birth. This form of EDS is caused by a change in either of two components of Collagen type I, called proa1(I) type A and proa2(I) type B. A skin biopsy may be performed to demonstrate an abnormality in either component. Direct DNA testing is also available.

Dermatosparaxis type

Individuals with this type of EDS, once called type VIIC, have extremely fragile skin that bruises easily but does not scar excessively. The skin is soft and may sag, leading to an aged appearance even in young adults. Individuals may also have hernias. Dermatosparaxis type is inherited in an autosomal recessive manner.

There are two major clinical diagnostic criteria for EDS dematosparaxis type. These include severe skin fragility and sagging or aged-appearing skin. This form of EDS is caused by a change in the enzyme called procollagen I N-terminal peptidase. A skin biopsy may be performed for a definitive diagnosis of dermatosparaxis type.

Other types

There are several other forms of EDS that have not been as clearly defined as the aforementioned types. Symptoms of EDS within this category may include soft, mildly stretchable skin, shortened bones, chronic **diarrhea**, joint hypermobility and dislocation, bladder rupture, or poor wound healing. Inheritance patterns within this group include X-linked recessive, autosomal dominant, and autosomal recessive.

Demographics

EDS was originally described by Dr. Van Meekeren in 1682. Dr. Ehlers and Dr. Danlos further characterized the disease in 1901 and 1908, respectively. According to the Ehlers-Danlos National Foundation, one in 5,000 to one in 10,000 people are affected by some form of EDS.

Causes and symptoms

There are numerous types of EDS, all caused by changes in one of several genes. The manner in which EDS is inherited depends on the specific gene involved. There are three patterns of inheritance for EDS: autosomal dominant, autosomal recessive, and X-linked (extremely rare).

Chromosomes are made up of hundreds of small units known as genes, which contain the genetic material necessary for an individual to develop and function. Humans have 46 chromosomes, which are matched into 23 pairs. Because chromosomes are inherited in pairs, each individual receives two copies of each chromosome and likewise two copies of each gene.

Changes or mutations in genes can cause genetic diseases in several different ways, many of which are represented within the spectrum of EDS. In autosomal dominant EDS, only one copy of a specific gene must be changed for a person to have EDS. In autosomal recessive EDS, both copies of a specific gene must be changed for a person to have EDS. If only one copy of an autosomal recessive EDS gene is changed, the person is referred to as a carrier, meaning he or she does not have any signs or symptoms of the disease itself, but carries the possibility of passing the disorder to a future child. In X-linked EDS, a specific gene on the X chromosome must be changed. However, this affects males and females differently because males and females have a different number of X chromosomes.

The few X-linked forms of EDS fall under the category of X-linked recessive. As with autosomal recessive, this implies that both copies of a specific gene must be changed for a person to be affected. However, because males only have one X-chromosome, they are affected if an X-linked recessive EDS gene is changed on their single X-chromosome. That is, they are affected even though they have only one changed copy. On the other hand, that same gene must be changed on both of the X-chromosomes in a female for her to be affected.

Although there is much information regarding the changes in genes that cause EDS and their various inheritance patterns, the exact gene mutation for all types of EDS is not known.

When to call the doctor

The doctor should be called if a child has symptoms of Ehlers-Danlos syndrome. Medical advice should also be sought if a person has a family history of Ehlers-Danlos syndrome and is planning to conceive a child.

Diagnosis

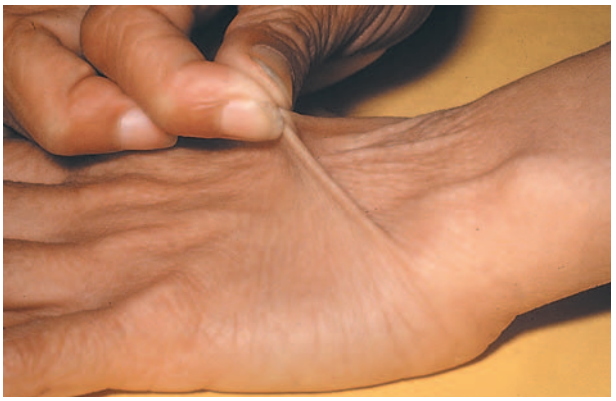
Clinical symptoms such as extreme joint looseness and unusual skin qualities, along with family history, can lead to a diagnosis of EDS. Specific tests, such as skin biopsies, are available for diagnosis of certain types of EDS, including vascular, arthrochalasia, and dermatosparaxis types. A skin biopsy involves removing a small sample of skin and examining its microscopic structure. A urine test is available for the kyphoscoliosis type.

Treatment

Medical therapy relies on managing symptoms and trying to prevent further complications. There is no cure for EDS.

Braces may be prescribed to stabilize joints, although surgery is sometimes necessary to repair joint damage caused by repeated dislocations. Physical therapy teaches individuals how to strengthen muscles around joints and may help to prevent or limit damage. Elective surgery is discouraged due to the high possibility of complications.

There are anecdotal reports that large daily doses (0.04–0.14 oz, or 1–4 g) of vitamin C may help decrease bruising and aid in wound healing. Constitutional homeopathic treatment may be helpful in maintaining optimal health in persons with a diagnosis of EDS. Before beginning these types of therapies, an individual with EDS should discuss them with his or her doctor. Therapy that does not require medical consultation involves protecting the skin with sunscreen and avoiding activities that place stress on the joints. **Wounds** and infections must be treated with care because tissue healing may be poor. Suturing can be difficult, for the skin can be extremely fragile.



Elasticity of the skin is one characteristic of Ehlers-Danlos syndrome. *(Photo Researchers, Inc.)*

Prognosis

The outlook for individuals with EDS depends on the type of EDS with which they have been diagnosed. Symptoms vary in severity, even within one sub-type. Some individuals have negligible symptoms, while others are severely restricted in their daily life. Extreme joint instability and scoliosis may limit a person's mobility. Most individuals will have a normal lifespan. However, those with blood vessel involvement, particularly persons with EDS vascular type, have an increased risk of fatal complications.

EDS is a lifelong condition. Affected individuals may face social obstacles related to their disease on a daily basis. Some individuals with EDS have reported living with fears of significant and painful skin ruptures, becoming pregnant (especially those with EDS vascular type), experiencing worsening of their condition, becoming unemployed due to physical and emotional burdens, and undergoing social stigmatization in general.

Some people with EDS are not diagnosed until well into adulthood and, in the case of EDS vascular type, occasionally not until after death due to complications of the disorder. The diagnosis may be devastating to the individual and, in many cases, to other family members when they learn they are at risk for being affected.

Although children with EDS face significant challenges, it is important to remember that each child is unique with his or her own distinguished qualities and

KEY TERMS

Arthrochalasia—Excessive loosness of the joints.

Blood vessels—General term for arteries, veins, and capillaries that transport blood throughout the body.

Cartilage—A tough, elastic connective tissue found in the joints, outer ear, nose, larynx, and other parts of the body.

Collagen—The main supportive protein of cartilage, connective tissue, tendon, skin, and bone.

Connective tissue—A group of tissues responsible for support throughout the body; includes cartilage, bone, fat, tissue underlying skin, and tissues that support organs, blood vessels, and nerves throughout the body.

Dermatosparaxis—Skin fragility caused by abnormal collagen.

Hernia—A rupture in the wall of a body cavity, through which an organ may protrude.

Homeopathy—A holistic system of treatment developed in the eighteenth century. It is based on the idea that substances that produce symptoms of sickness in healthy people will have a curative effect when given in very dilute quantities to sick people who exhibit those same symptoms. Homeopathic remedies are believed to stimulate the body's own healing processes.

Hyperextensibility—The ability to extend a joint beyond the normal range.

Hypermobility—Unusual flexibility of the joints, allowing them to be bent or moved beyond their normal range of motion.

Joint dislocation—The displacement of a bone from its socket or normal position.

Kyphoscoliosis—Abnormal front-to-back and side-to-side curvature of the spine.

Ligament—A type of tough, fibrous tissue that connects bones or cartilage and provides support and strength to joints.

Osteoarthritis—A noninflammatory type of arthritis, usually occurring in older people, characterized by degeneration of cartilage, enlargement of the margins of the bones, and changes in the membranes in the joints. Also called degenerative arthritis.

Scoliosis—An abnormal, side-to-side curvature of the spine.

Tendon—A tough cord of dense white fibrous connective tissue that connects a muscle with some other part, especially a bone, and transmits the force which the muscle exerts.

Uterus—The female reproductive organ that contains and nourishes a fetus from implantation until birth. Also called the womb.

Vascular—Pertaining to blood vessels.

potential. Persons with EDS go on to have families and careers, and to be respected citizens, surmounting the challenges of their disease.

Prevention

If a couple has had a child diagnosed with EDS, the chance that they will have another child with the same disorder depends on the form of EDS the child has and if either parent is affected by the same disease.

In classical autosomal dominant EDS, the risk for recurrence in the parents' other children is one in four.

X-linked recessive EDS is accompanied by a slightly more complicated pattern of inheritance. If a father with an X-linked recessive form of EDS passes a copy of his X chromosome to his children, the sons will be unaffected and the daughters will be carriers. If a mother is a carrier for an X-linked recessive form of EDS, she may have affected or unaffected sons, or carrier or unaffected daughters, depending on the second sex chromosome inherited from the father.

Prenatal diagnosis is available for specific forms of EDS, including kyphosocliosis type and vascular type. However, prenatal testing is only a possibility in these types if the underlying defect has been found in another family member.

Parental concerns

Constant bruises, skin wounds, and trips to the hospital take their toll on affected children and their parents. Prior to diagnosis, parents of children with EDS have found themselves under suspicion of **child abuse**.

Management of all types of EDS may include genetic counseling to help the affected individual and his

or her family understand the disorder and its impact on other family members and future children.

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Ehlers-Danlos Support Group - UK. P.O. Box 335, Farnham, Surrey, GU10 1XJ. UK. +44 1252 690 940. Web site: <www.ehlers- danlos.org>.

Ehlers-Danlos National Foundation. 6399 Wilshire Blvd., Ste. 203, Los Angeles, CA 90048 (323) 651-3038. Fax: (323) 651-1366. Web site: <www.ednf.org>.

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GeneClinics. Available online at: <www.geneclinics.org>.

Judith Sims
Java O. Solis, M.S.

Elbow injury *see* **Nursemaid's elbow**

Electric shock injuries

Definition

Electric shock injuries are caused by lightning or electric current passing through the body. In infants, electric shock injuries occur most often when they put metal objects in their mouths.

Description

Electric current can be described as the flow of microscopic particles called electrons through wires and electrical appliances. Materials like metal and water through which electric current (electricity) travels easily are called conducting materials or conductors. The body is an excellent conductor, and electric current from any source passing through the body produces electric shock injuries.

The severity of electric shock injuries depends on the current's voltage, the amount of current (amperage), the type of current (direct or alternating), the body's resistance to the current, the current's path through the body, and the length of time the body remains in contact with the current. The interplay of these factors can produce effects ranging from a mild **tingling** to instant death. How electric shocks affect the skin depends on the skin's resistance to current, which in turn depends on the wetness, thickness, and cleanliness of the skin. Thin or wet skin is much less resistant than thick or dry skin. When the skin's resistance to current is low, the current may cause little or no skin damage but severely burn internal organs and tissues. By contrast, high skin resistance can produce severe skin **burns** but prevent the current from entering the body.

The nervous system (brain, spinal cord, and nerves) is very sensitive to electric shock injury, and neurological problems are the most common consequences suffered by electric shock victims. Neurological damage can be minor and clear up on its own or with medical treatment or can be severe and permanent. Damage to the respiratory and cardiovascular systems is highest at time of injury. Electric shocks can paralyze the respiratory system or disrupt heart action, causing instant death. Also at risk are the smaller veins and arteries, which can develop blood clots. Damage to the smaller vessels is often followed by amputation after high-voltage injuries. Many other injuries are possible after an electric shock, including cataracts, kidney failure, and destruction of muscle tissue. The victim may also suffer a fall or an electric arc may set clothing or nearby flammable substances on fire. Strong shocks are often accompanied by violent **muscle spasms** that can break and dislocate bones.

Demographics

Electric shocks are responsible for about 1,000 deaths in the United States each year, or about 1 percent

of all accidental deaths. Children are not often seriously injured by electricity, but they are prone to electric shock by the low voltage (110–220 volts) of typical household current. In children aged 12 years and younger, household appliance electrical cords and extension cords are reported to cause more than 63 percent of injuries. Wall outlets are responsible for 15 percent of electric injuries.

Causes and symptoms

Electric shocks are caused by the passage of electric current through the body. They are caused in infants and young children by their playing with electrical appliances or cords and in older children by mischievous exploration of electrical systems or use of faulty electrical appliances or tools.

A child who has suffered an electric shock may have very little external injury or may have obvious severe burns. Burns are usually most severe at the points of contact with the electrical source. The hands, heels, and head are common points of contact. Other injuries are also possible if the child has been thrown clear of the electrical source by forceful muscular contractions. The child may have internal injuries especially if he or she is experiencing any shortness of breath, chest **pain**, or abdominal pain. In children, the typical electrical mouth burn from biting an electric cord appears as a burn on the lip. The area has a red or dark, charred appearance.

When to call the doctor

A person shocked by high voltage (500 volts or more) should be evaluated in the emergency department of a hospital or clinic. Any person present at the scene of the accident should immediately call 911.

Brief low-voltage shocks (110–220 volts or less) that do not result in any symptoms or burns of the skin do not require care. However, following a low-voltage shock, parents should consult their healthcare provider if the child has any noticeable burn to the skin, any **numbness**, tingling, or vision, hearing, or speech problems, no matter how mild. A doctor should also always evaluate electric cord burns to the mouth of a child.

Diagnosis

Diagnosis relies on the information gathered about the circumstances of the electric shock, a thorough physical examination, and monitoring of cardiovascular and kidney activity. The physician's primary concern is to determine if significant unseen injury exists. Injury may occur to muscles, the heart, or the brain from the electricity or to any bones or other organs from being thrown from the electric source. Tests may include any of the following:

- electrocardiogram (ECG) to check the heart
- complete blood count (CBC)
- urine test for muscle enzymes, to screen for muscle injury
- x rays or **magnetic resonance imaging** (MRI) to look for **fractures** or dislocations, both of which may be caused by electrocution
- CT scan for internal injuries

Treatment

When a severe electric shock injury happens at home, the main power should immediately be shut off. If that cannot be done, and current is still flowing through the child, the alternative is to stand on a dry, nonconducting surface such as a folded newspaper, flattened cardboard carton, or plastic or rubber mat and use a nonconducting object such as a wooden broomstick (never a damp or metallic object) to push the child away from the source of the current. The victim and the source of the current must not be touched while the current is still flowing, for doing so can electrocute the rescuer. Emergency medical help should be summoned as quickly as possible. People who are trained to perform **cardiopulmonary resuscitation** (CPR) should, if appropriate, begin first aid while waiting for emergency medical help to arrive.

At the clinic or hospital, treatment depends on the severity of the burns and/or the nature of other injuries found. Minor burns are usually treated with topical antibiotic ointment and dressings. More severe burns may require surgery to clean the **wounds** or even to perform skin grafting. Severe burns on the arms, legs, or hands may require surgery to remove damaged muscle or even amputation.

Prognosis

Recovery from electric shock depends on the nature and severity of the injuries. The percentage of the body surface area burned is the most important factor affecting prognosis. Electric shocks cause death in 3–15 percent of cases, with infection being the most common cause of death in people hospitalized following electrical injury. Electrical damage to the brain may result in a permanent **seizure disorder**, depression, **anxiety**, or other personality changes. Injuries from household appliances and other low-voltage sources are less likely to produce extreme damage.

KEY TERMS

Computed tomography (CT)—An imaging technique in which cross-sectional x rays of the body are compiled to create a three-dimensional image of the body's internal structures; also called computed axial tomography.

Conducting materials—Materials that conduct electricity, materials through which electric current travels easily. Examples are metals and water.

Electric current—The rate of flow of electric charge, measured in amperes. Electric current can also be described as the flow of microscopic particles called electrons flowing through wires and electronic components and appliances.

Electrical resistance—Resistance to the flow of electrical current.

Magnetic resonance imaging (MRI)—An imaging technique that uses a large circular magnet and radio waves to generate signals from atoms in the body. These signals are used to construct detailed images of internal body structures and organs, including the brain.

Non-conducting materials—Also called insulators, materials through which electric current does not propagate. Examples are ceramics, rubber, wood.

Skin grafting—A surgical procedure by which skin or a skin substitute is placed over a burn or non-healing wound to permanently replace damaged or missing skin or to provide a temporary wound covering.

Prevention

Parents and other adults need to be aware of possible electric dangers in the home. Damaged electric appliances, wiring, cords, and plugs should be repaired, replaced or discarded. Hair dryers, radios, and other electric appliances should never be used in the bathroom or where they may accidentally come in contact with water. Young children need to be kept away from electrical appliances and should be taught about the dangers of electricity as early as possible. They should not be allowed to **play** with any electrical cord. Electric outlets also require **safety** covers in homes with young children. In children older than 12 years, most electrical injuries result from exploring and playing around high-power systems. Teenagers should accordingly be warned not to climb on power towers, play near transformer systems, or explore electrified train rails or other electrical systems.

See also Computed tomography.

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Monique Laberge, Ph.D.

Electroencephalogram

Definition

An electroencephalogram (EEG), also called a brain wave test, is a diagnostic test which measures the electrical activity of the brain (brain waves) using highly sensitive recording equipment attached to the scalp by fine electrodes.

Purpose

EEG is performed to detect abnormalities in the electrical activity of the brain which may help diagnose the presence and type of various brain disorders, to look for causes of confusion, and to evaluate head injuries, tumors, infections, degenerative diseases, and other disturbances that affect the brain. The test is also used to investigate periods of unconsciousness. EEG may also confirm brain death in someone who is in a coma. EEG cannot be used to measure **intelligence** or diagnose mental illness. Specifically, EEG is used to diagnose the following:

- seizure disorders (such as epilepsy or convulsions)
- structural brain abnormality (such as a brain tumor or brain abscess)
- head injury, **encephalitis** (inflammation of the brain)
- hemorrhage (abnormal bleeding caused by a ruptured blood vessel)
- cerebral infarct (tissue that is dead because of a blockage of the blood supply)
- **sleep** disorders (such as narcolepsy)

Description

Brain cells communicate by producing tiny electrical impulses, also called brain waves. These electrical signals have certain rhythms and shapes, and EEG is a technique that measures, records, and analyzes these signals to help make a diagnosis. Electrodes are used to detect the electrical signals. They come in the shape of small discs that are applied to the head and connected to a recording device. The recording machine then converts the electrical signals into a series of wavy lines that are drawn onto a moving piece of graph paper. An EEG test causes no discomfort. Although having electrodes pasted on the skin may feel strange, they only record activity and do not produce any sensation. The patient needs to lie still with eyes closed because any movement can affect results. The patient may also be asked to do certain things during the EEG recording, such as breathing deeply and rapidly for several minutes or looking at a bright flickering light.

An EEG is performed by an EEG technician in a specially designed room that may be in the doctor's office or at a hospital. The patient is asked to lie on a bed or in a comfortable chair so that a relaxed EEG recording can be done. The technician either measures the scalp and marks the spots where small discs (electrodes) will be placed or fits the head with a special cap containing between 16 and 25 of these discs. The scalp is then rubbed with a mild, scratchy cleanser that may cause mild discomfort for a short while. The discs are attached to the body with a cream or gel. Alternatively, the technician may secure the discs to the skin with an adhesive. The heart may also be monitored during the procedure.

Precautions

Before an EEG, care should be taken to avoid washing hair with an oily scalp product 24 hours before the test. Doctors usually recommend that patients eat a meal or light snack some four hours before the test. Caffeinated drinks should be avoided for eight hours before the test. Sometimes, the EEG gives better results when the patient has had less than the usual amount of sleep. The doctor may ask that the child be kept awake for all or part of the night before the EEG. The healthcare provider may also discontinue some medications before the test.

Preparation

The physical and psychological preparation required for this test depends on the child's age, interests, previous experiences, and level of trust. For older children, research has shown that preparing ahead can reduce crying or resisting the test. In addition, children report less **pain** and show less distress when prepared. Proper preparation for the test can reduce a child's **anxiety**, encourage cooperation, and help develop coping skills.

Some general guidelines for preparing a toddler or preschooler for an EEG include the following:

- Explain the EEG procedure in words that the child understands, avoiding abstract terminology.
- Ensure that the child understands the exact body part involved and that the procedure will be limited to that area.
- Describe how the test is likely to feel.
- Give the child permission to yell, cry, or otherwise express any pain or discomfort verbally.
- Stress the benefits of the EEG procedure and list things that the child may find pleasurable after the test, such as feeling better or going home.

The above guidelines also apply to school age children. Additionally, for older children, parents can try the following:

- Suggest ways to keep calm and reduce anxiety such as counting, deep breathing, or thinking pleasant thoughts.
- Include the child in the decision-making process, such as the time of day where the EEG is performed.

KEY TERMS

Electrode—A medium for conducting an electrical current.

Encephalitis—Inflammation of the brain, usually caused by a virus. The inflammation may interfere with normal brain function and may cause seizures, sleepiness, confusion, personality changes, weakness in one or more parts of the body, and even coma.

Epilepsy—A neurological disorder characterized by recurrent seizures with or without a loss of consciousness.

Hemorrhage—Severe, massive bleeding that is difficult to control. The bleeding may be internal or external.

Hyperventilation—Rapid, deep breathing, possibly exceeding 40 breaths/minute. The most common cause is anxiety, although fever, aspirin overdose, serious infections, stroke, or other diseases of the brain or nervous system. Also refers to a respiratory therapy involving deeper and/or faster breathing to keep the carbon dioxide pressure in the blood below normal.

Narcolepsy—A life-long sleep disorder marked by four symptoms: sudden brief sleep attacks, cataplexy (a sudden loss of muscle tone usually lasting up to 30 minutes), temporary paralysis, and hallucinations. The hallucinations are associated with falling asleep or the transition from sleeping to waking.

Seizure—A sudden attack, spasm, or convulsion.

Sleep disorder—Any condition that interferes with sleep. Sleep disorders are characterized by disturbance in the amount of sleep, in the quality or timing of sleep, or in the behaviors or physiological conditions associated with sleep.

- Suggest that the child hold the hand of the technician or someone else helping with the procedure.

As for adolescents, detailed information about the EEG should be provided and the reasons for the procedure should be explained in correct medical terminology. When the EEG is required for a **seizure disorder**, there is the potential risk that the test will trigger a seizure. This possibility should be openly discussed. Adolescents commonly have high concerns about risks and the best way to prepare them is to fully inform them. The healthcare provider could also be asked to limit the number of strangers entering and leaving the room during the EEG procedure, since they can raise the patient's anxiety level.

Patient receiving an electroencephalogram, a neurological test to measure and record electrical activity in the brain. *(© Richard T. Nowitz/Corbis.)*

Aftercare

There are no side effects or special procedures required after an EEG. The technician simply removes the gel with water and the adhesive, if used, with a special cleanser. Shampooing will rid the hair of any other material. A few patients are mildly sensitive to the gel or may get irritation from the rubbing of their scalps.

Risks

The EEG test is very safe. However, if a patient has a seizure disorder, a seizure may be triggered by the flashing lights or hyperventilation. The healthcare

provider performing the EEG is trained to take care of the patient if this happens.

Normal results

An EEG returns normal results when brain waves have normal frequency and amplitude and other characteristics are typical.

Parental concerns

Before the test, parents should know that the child probably will cry, and restraints may be used. The most important way to help a child through an EEG procedure is by being there and caring. Crying is a normal response to the strange environment, unfamiliar people, restraints, and separation from the parent. Infants and young children will cry more for these reasons than because the test or procedure is uncomfortable. Knowing this from the onset may help parents feel less anxiety about what to expect. Having specific information about the test may further reduce anxiety.

See also Encephalitis; Narcolepsy; Sleep disorders.

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Electronic fetal monitoring

Definition

Electronic fetal monitoring (EFM) involves the use of an electronic fetal heart rate (FHR) monitor to record the baby's heart rate. Elastic belts are used to hold sensors against the pregnant woman's abdomen. The sensors are connected to the monitor and detect the baby's heart rate as well as the uterine contractions. The monitor then records the FHR and the contractions as a pattern on a strip of paper. Electronic fetal monitoring is performed late in pregnancy or continuously during labor to ensure normal delivery of a healthy baby. EFM can be utilized either externally or internally in the womb.

Purpose

All electronic fetal monitors detect the FHR and maternal uterine activity (UA), and both are displayed for interpretation since the pattern of the baby's heartbeat during labor often reflects the baby's condition. During contractions, the normal pattern is for the FHR to slow somewhat, picking up again as the contraction ends. The EFM continuously prints out a record of both the FHR and the duration and frequency of the uterine contractions, so that deviations from normal patterns can be identified. Certain variations in this pattern, such as precipitous drops in the FHR at the end of a contraction can constitute a true life or death situation requiring emergency delivery of the baby. Prior to the use of EFMs, nurses and doctors periodically monitored the baby's heartbeat manually by placing a stethoscope on

the mother's abdomen. It is important to note that the EFM is a screening tool and not diagnostic of any particular disorder.

Fetal asphyxia (an impaired exchange of oxygen and carbon dioxide) is recognized as an important cause of stillbirth and neonatal death. Asphyxia has also been implicated as a cause of **cerebral palsy**, although many cases of cerebral palsy have occurred without evidence of birth asphyxia. Most fetuses, however, tolerate intrauterine hypoxia during labor and are delivered without complications. If the interruption to the supply of oxygen is short, the baby may recover without any damage. If the time is longer, there may be some injury that is reversible. If the time period without oxygen is especially long, there may be permanent injury to one or more organs of the body.

Fetal monitoring can be helpful in a variety of different situations. During pregnancy, fetal monitoring can be used as a part of **antepartum testing**. If the practitioner feels that a baby may be at risk for problems during pregnancy, non-stress tests, biophysical profiles, or even contraction stress tests are performed twice a week to monitor fetal well-being. In this test, changes in the baby's heart rate are noted with the fetus's own movements. The heart rate of a healthy baby should go up whenever she or he moves.

Description

Using the external fetal monitor is simple and painless. Two belts are placed around the pregnant woman's abdomen. One is to hold the transducer that picks up the FHR and the other is to hold a tocodynanometer, which picks up uterine activity or contractions. External monitoring of the fetus is accomplished by means of a transducer that emits continuous sound waves (ultrasound). A water-soluble gel is placed on the underside of the transducer to permit the conduction of fetal heart sounds. When the transducer is placed correctly on the maternal abdomen, the sound waves bounce off the fetal heart and are picked up by the electronic monitor. The actual moment-by-moment FHR can be simultaneously viewed on a screen while being printed on graph paper. Incorrect placement of the transducer may detect a pulsating maternal vessel with a resultant swooshing sound (uterine soufflé), and the rate will be the same as the maternal pulse. Maternal uterine activity is noted and recorded when the pressure of a contraction pushes on a sensor, which is on the underside of a tocodynanometer. Once again, incorrect placement may not completely detect contractions. The sensor on the tocodynanometer must be placed on that part of the uterus that can be palpated easily. If it is too high or too low, the contractions may not be detected.

If it becomes difficult to detect the FHR with the external monitor or if there are subtle signs of a developing problem, the practitioner may recommend the use of an internal monitor. The measurement of fetal heart activity is performed most accurately by attaching an electrode directly to the fetal scalp. This is an invasive procedure that requires the rupture of membranes (amniotomy) and is associated with occasional complications.

An internal monitor may also be used to determine the actual strength of the contraction as well as the resting tone of the uterus. A woman may appear to be having strong contractions but not be progressing in labor. Progress in labor is determined by cervical dilation. The insertion of an intrauterine pressure catheter (IUPC) permits the determination of the strength of the contractions in millimeters of Hg, a measurement used for pressure. A good labor pattern that facilitates cervical dilation can be calculated by taking the difference in pressure between the peak of the contraction and the resting tone and adding them up over a ten-minute period. The unit of measurement for this calculation is called a Montevideo unit, and ideally the sum total of the pressures should be between 150 and 250 Montevideo units to achieve cervical dilation. If the calculation is in this range and the woman's cervix is not changing, then and only then can a diagnosis of failure to progress be made. The IUPC also provides an accurate measurement of the resting tone of the uterus. It is important that the uterus relax between contractions in order for the baby to receive oxygen. If the uterus is not relaxing or if the resting tone is rising, this can be an indication of a placental abruption (the tearing away of the placenta from the wall of the uterus).

Another use of an IUPC is for amnioinfusion. This is a procedure in which a physiologic solution (such as normal saline) is infused into the uterine cavity to replace the amniotic fluid. It is used to relieve cord compression, reduce fetal distress caused by meconium staining, and as a correction of decreased amniotic fluid.

Preparation

There are no special preparations needed for fetal monitoring. An explanation of the procedure and its risks should be provided by the healthcare provider and a consent form may be signed for the procedure.

KEY TERMS

Amnioinfusion—A procedure whereby a physiologic solution such as normal saline or lactated ringer's solution is infused through a lumen in an intrauterine pressure catheter into the uterus to alleviate cord compression and to help dilute meconium staining.

Amniotic fluid—The liquid in the amniotic sac that cushions the fetus and regulates temperature in the placental environment. Amniotic fluid also contains fetal cells.

Amniotomy—Rupturing or breaking the amniotic sac (bag of waters) to permit the release of fluid.

Asphyxia—Lack of oxygen.

Deceleration—A decrease in the fetal heart rate that can indicate inadequate blood flow through the placenta.

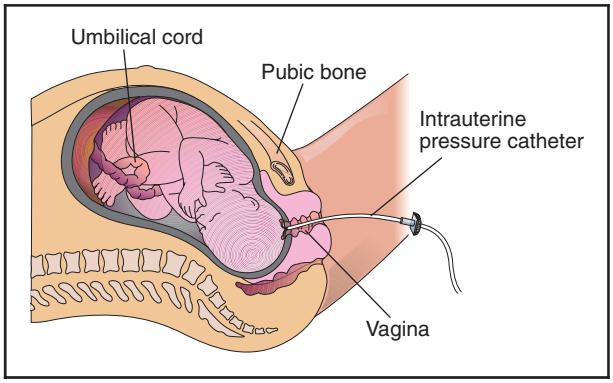
Hypoxia—A condition characterized by insufficient oxygen in the cells of the body

Meconium—A greenish fecal material that forms the first bowel movement of an infant.

Perinatal—Referring to the period of time surrounding an infant's birth, from the last two months of pregnancy through the first 28 days of life.

Risks

Besides the risk of an unnecessary **cesarean section**, other risks posed to the mother by EFM include her **immobilization** in bed. Immobilization simultaneously limits changing positions for comfort and causes changes in blood circulation, which decreases the oxygen supply to the fetus and can lead to abnormal changes in the FHR on the EFM that was applied to detect these changes. Another problem with the use of the EFM is that practitioners have a tendency to focus on it instead of the laboring woman. For these and other reasons, the United States Preventive Services Task Force states that there is some evidence that using EFM on low-risk women in labor might not be indicated. EFM, however, has become an accepted standard of care in many settings in the United States for management of labor. Interestingly, there has not been a reported reduction in perinatal morbidity in the United States with the use of EFM. There is a benefit to using EFM in women with complicated labors, such as those induced or augmented with oxytocin, prolonged labors, vaginal birth after having a cesarean section, abnormal presentation, and twin pregnancy.



Electronic fetal monitoring (EFM) is performed late in pregnancy or continuously during labor to ensure normal delivery of a healthy baby. EFM can be utilized either externally or internally in the womb. This illustration shows the internal procedure, in which an electrode is attached directly to the baby's scalp to monitor the heart rate. Uterine contractions are recorded using an intrauterine pressure catherter which is inserted through the cervix into the uterus. *(Illustration by Electronic Illustrators Group.)*

Generally the insertion of a fetal scalp electrode is a safe procedure, but it may occasionally cause umbilical cord prolapse or infection due to early amniotomy. Problems could also occur if the electrode or IUPC causes trauma to the eye, fetal vessels, umbilical cord, or placenta. Scalp infections with the herpes virus or group B streptococcus are possible, and concern has been raised regarding the potential for enhancing transmission of the human **immunodeficiency** virus (HIV). As with any procedure, the potential benefit of EFM must be weighed against the potential risks.

Normal results

The average fetal heart rate is in the range of 110 to 160 beats per minute (bpm). A baby who is receiving sufficient oxygen through the placenta moves around and the monitor strip will show the baby's heart rate rising briefly as he or she moves. The monitor strip is considered to be reactive when the baby's heart rate elevates at least 15 bpm above the baseline heart rate for at least 15 seconds twice in a 20-minute period. Other indicators of fetal well-being include short term variability (STV), which constitutes changes in the FHR from one beat to another, and long term variability (LTV), which is changes in the FHR over a long period of time.

If the baby's heart rate drops very low or rises very high, this can signal a serious problem if it occurs for longer than a ten-minute period. During a contraction, the flow of oxygen (from the mother) through the placenta (to the baby) is temporarily blocked. The baby should be capable of withstanding this condition since it

Fetal monitor belt around a pregnant woman's torso to record the heart rate of her baby. *(© Custom Medical Stock Photo, Inc.)*

is receiving sufficient oxygen between contractions. The first sign that a baby is not getting enough oxygen between contractions is often a drop in the baby's heart rate after a contraction, called a late deceleration. The baby's heart rate recovers to a normal level between contractions, only to drop again after the next contraction. This is a more subtle sign of distress. These babies will do fine if they are delivered in a short period of time following the onset of the late decelerations. Sometimes, these signs develop long before delivery is expected. In that case, a c-section may be necessary.

One of the worst indications of fetal distress, however, is a tracing that shows no variability at all. It is a flat tracing and indicates that the baby has sustained a severe assault on its central nervous system. It is not capable of responding to stimuli in its environment. The mother may report that she has experienced decreased fetal movement as the baby has only enough oxygen to keep the heart beating. It is for this reason that all pregnant women should be taught to keep track of fetal movement every day and to report any significant changes.

See also Apgar testing; Cesarean section.

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Elimination diet

Definition

An elimination diet is the systematic elimination of foods or group of foods from the diet suspected in causing a food allergy. It is used as a means to diagnose an allergic reaction to foods.

Purpose

While people of all ages can develop an allergic sensitivity to certain foods, such **allergies** are especially common among children. In the United States, one child in six develops an allergic reaction to certain substances, and foods are among the prime offenders. (Many **food allergies** are outgrown during adolescence.) Food elimination is considered only when no other cause can be found for the symptoms the child is experiencing. Common symptoms of food allergies include **hives**, angioedema (swelling), **rashes**, respiratory congestion, and gastrointestinal problems such as **constipation**, **diarrhea**, and/or gas. Food allergies are also known to play a secondary role in many chronic conditions, such as **asthma**, **acne**, ear infections, eczema, headaches, and hay fever. The most effective means of treating food allergies is to avoid the foods that produce allergic reactions.

Description

There are two main ways of diagnosing food allergies by the elimination method. A casual approach involves eliminating, one at a time, foods from the diet suspected of causing allergic reactions and observing the person to see if there is a reduction in symptoms in the absence of particular foods. This method is often recommended for chil-

dren, as it is easier to follow than the standard elimination diet. The more rigorous method (which is a true elimination diet) reverses this strategy by eliminating many foods at the outset and then reintroducing suspected allergens (allergy-producing substances) one at a time. Elimination diets often include a rotation component, by which even the limited foods allowed at the beginning are allocated in such a way that no single food is eaten more than once within a three-day period. This feature has two purposes. First, it alleviates the monotony of a limited diet. Second, it allows for the possibility that some persons may even be allergic to the relatively safe foods allowed initially. If there is an allergic reaction at this stage, rotating foods makes it possible to identify the cause of the problem.

An elimination diet is divided into two parts: the elimination phase and the reintroduction (or food challenge) phase. During the elimination phase, which generally lasts between one and two weeks, as many known allergy-producing foods as possible are eliminated from the diet. Foods commonly known to cause allergies include: citrus fruits, strawberries, corn, peas, tomatoes, peanuts, nuts, legumes, soy products, wheat, oats, chicken, shellfish, eggs, dairy products, cow's milk, vinegar and other products of fermentation, coffee and tea, cane sugar, chocolate, and food additives. The elimination diet can be very strict or more liberalized depending on the severity of the symptoms. It is important to personalize the diet whenever possible.

During the elimination phase of the diet, which clears the body of allergens, ingredient labels for all processed foods should be carefully scrutinized to make sure that none of the proscribed foods makes its way into the diet. Different diets handle the reintroduction phase differently. In some cases, the "test" foods are introduced at three-day intervals while in others, a new food is reintroduced every day for 15 days. Foods should be reintroduced in as pure a form as possible (for example, cream of wheat rather than bread) for maximum certainty that the resulting effects are produced by the substance in question rather than by some other ingredient added during the manufacturing process.

A strict elimination diet should not be undertaken without the supervision of a physician and/or dietitian.

Another way to identify food allergies is to keep a food diary, recording everything eaten for a period of three or four weeks and noting any allergic reactions during that period.

Risks

The elimination phase of the diet should not last more than two weeks, since this restricted regimen will lack some essential nutrients. Also, a child's growth may be affected if placed on an elimination diet for an extended period. For example, a child with a cow's milk allergy must obtain his calcium, vitamin D, and other essential nutrients found in milk from other sources. Children with food allergies need to be followed by a physician and or dietitian to ensure they are not at risk for growth problems or inadequate intake of nutrients.

Normal results

Persons with chronic food allergies should see their symptoms subside during the elimination period. Sometimes they may experience withdrawal, an episode in which symptoms may actually worsen before they subside. These include: bloating, food cravings, **headache**, fatigue, and general aches and pains. This condition may last for a few days. Upon reintroduction of the offending food, these symptoms should return and are often worse than previously reported.

Parental concerns

In order to effectively eliminate the offending foods, a parent should be educated on label reading, cross-contact, and selecting alternate foods for an allergic child.

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American Dietetic Association. 120 South Riverside Plaza, Suite 2000, Chicago, IL 60606–6995. (Web site: <www.eatright.org>.

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Embryonic development *see* **Prenatal development**

Encephalitis

Definition

Encephalitis is an inflammation of the brain, usually caused by a direct viral infection or a hypersensitivity reaction to a virus or foreign protein. Brain inflammation caused by a bacterial infection is sometimes called cerebritis. When both the brain and spinal cord are involved, the disorder is called encephalomyelitis. An inflammation of the brain's covering, or meninges, is called **meningitis**.

Description

Encephalitis is an inflammation of the brain. The inflammation is a reaction of the body's immune system to infection or invasion. During the inflammation, the brain's tissues become swollen. The combination of the infection and the immune reaction to it can cause **headache** and a **fever**, as well as more severe symptoms in some cases.

The viruses causing primary encephalitis can be epidemic or sporadic. The **polio** virus is an epidemic cause. Arthropod-borne viral encephalitis is responsible for most epidemic viral encephalitis. The viruses live in animal hosts and mosquitoes that transmit the disease. The most common form of non-epidemic or sporadic encephalitis is caused by the **herpes simplex** virus, type 1 (HSV-1) and has a high rate of death. **Mumps** is another example of a sporadic cause.

Demographics

Approximately 2,000 cases of encephalitis are reported to the Centers for Disease Control and Prevention in Atlanta, Georgia, each year. Encephalitis can strike anyone, at any age, although some kinds of encephalitis are more common in children. Other kinds of encephalitis can affect anyone, but may affect children more severely.

Causes and symptoms

There are more than a dozen viruses that can cause encephalitis, spread by either human-to human contact or by animal **bites**. Encephalitis may occur with several common viral infections of childhood. Viruses and viral diseases that may cause encephalitis include:

- **chickenpox**
- **measles**
- mumps
- Epstein-Barr virus (EBV)
- cytomegalovirus infection
- HIV
- herpes simplex
- herpes zoster (shingles)
- herpes B
- polio
- **rabies**
- mosquito-borne viruses (arboviruses)

Primary encephalitis is caused by direct infection by the virus, while secondary encephalitis is due to a post-infectious immune reaction to viral infection elsewhere in the body. Secondary encephalitis may occur with measles, chickenpox, mumps, **rubella**, and EBV. In secondary encephalitis, symptoms usually begin five to ten days after the onset of the disease itself and are related to the breakdown of the myelin sheath that covers nerve fibers.

In rare cases, encephalitis may follow **vaccination** against some of the viral diseases listed above. Creutzfeldt-Jakob disease, a very rare brain disorder caused by an infectious particle called a prion, may also cause encephalitis.

Mosquitoes spread viruses responsible for equine encephalitis (eastern and western types), St. Louis encephalitis, California encephalitis, and Japanese encephalitis. **Lyme disease**, spread by ticks, can cause encephalitis, as can Colorado tick fever. Rabies is most often

spread by animal bites from dogs, cats, mice, raccoons, squirrels, and bats and may cause encephalitis.

Equine encephalitis is carried by mosquitoes that do not normally bite humans but do bite horses and birds. It is occasionally picked up from these animals by mosquitoes that do bite humans. Japanese encephalitis and St. Louis encephalitis are also carried by mosquitoes. The risk of contracting a mosquito-borne virus is greatest in mid- to late summer, when mosquitoes are most active, in those rural areas where these viruses are known to exist. Eastern equine encephalitis occurs in eastern and southeastern United States; western equine and California encephalitis occur throughout the West; and St. Louis encephalitis occurs throughout the country. Japanese encephalitis does not occur in the United States but is found throughout much of Asia. The viruses responsible for these diseases are classified as arbovirus, and these diseases are collectively called arbovirus encephalitis.

Herpes simplex encephalitis, the most common form of sporadic encephalitis in western countries, is a disease with significantly high mortality. It occurs in children and adults and both sides of the brain are affected. It is theorized that brain infection is caused by the virus moving from a peripheral location to the brain via two nerves, the olfactory and the trigeminal (largest nerves in the skull).

Herpes simplex encephalitis is responsible for 10 percent of all encephalitis cases and is the main cause of sporadic, fatal encephalitis. In untreated people, the rate of death is 70 percent while the mortality is 15 to 20 percent in persons who have been treated with acyclovir. The symptoms of herpes simplex encephalitis are fever, rapidly disintegrating mental state, headache, and behavioral changes.

The symptoms of encephalitis range from very mild to very severe and may include:

- headache
- fever
- lethargy (sleepiness, decreased alertness, and fatigue)
- malaise
- **nausea and vomiting**
- visual disturbances
- tremor
- decreased consciousness (drowsiness, confusion, delirium, and unconsciousness)
- stiff neck
- seizures

Symptoms may progress rapidly, changing from mild to severe within several days or even several hours.

When to call the doctor

A physician should be called whenever a headache does not respond to medication or when a person experiences a fever over 104°F (40.0°C), **nausea and vomiting**, visual disturbances, a stiff neck, or seizures.

A doctor should be called when an infant's temperature rises above 100°F (37.8°C) and cannot be brought down within a few minutes. Infants whose temperatures exceed 102°F (38.9°C) should be sponge-bathed in cool water while waiting for emergency help to arrive.

Diagnosis

Diagnosis of encephalitis includes careful questioning to determine possible exposure to viral sources. Tests that can help confirm the diagnosis and rule out other disorders include:

- blood tests (to detect antibodies to viral antigens and foreign proteins)
- cerebrospinal fluid analysis, or spinal tap (to detect viral antigens and provide culture specimens for the virus or bacteria that may be present in the cerebrospinal fluid)
- electroencephalogram (EEG)
- CT and MRI scans

A brain biopsy (surgical gathering of a small tissue sample) may be recommended in some cases in which treatment has thus far been ineffective and the cause of the encephalitis is unclear. Definite diagnosis by biopsy may allow specific treatment that would otherwise be too risky.

Treatment

Choice of treatment for encephalitis depends on the cause. Bacterial encephalitis is treated with **antibiotics**. Viral encephalitis is usually treated with **antiviral drugs**, including acyclovir, ganciclovir, foscarnet, ribavirin, and AZT. Viruses that respond to acyclovir include herpes simplex, the most common cause of sporadic (non-epidemic) encephalitis in the United States.

The symptoms of encephalitis may be treated with a number of different drugs. Corticosteroids, including prednisone and dexamethasone, are sometimes prescribed to reduce inflammation and brain swelling. Anticonvulsant drugs, including phenytoin, are used to control seizures. Fever may be reduced with **acetaminophen** or other fever-reducing drugs.

A person with encephalitis must be monitored carefully, since symptoms may change rapidly. Blood tests may be required regularly to track levels of fluids and salts in the blood.

KEY TERMS

Cerebrospinal fluid analysis—A laboratory test, important in diagnosing diseases of the central nervous system, that examines a sample of the fluid surrounding the brain and spinal cord. The fluid is withdrawn through a needle in a procedure called a lumbar puncture.

Computed tomography (CT)—An imaging technique in which cross-sectional x rays of the body are compiled to create a three-dimensional image of the body's internal structures; also called computed axial tomography.

Electroencephalogram (EEG)—A record of the tiny electrical impulses produced by the brain's activity picked up by electrodes placed on the scalp. By measuring characteristic wave patterns, the EEG can help diagnose certain conditions of the brain.

Inflammation—Pain, redness, swelling, and heat that develop in response to tissue irritation or injury. It usually is caused by the immune system's response to the body's contact with a foreign substance, such as an allergen or pathogen.

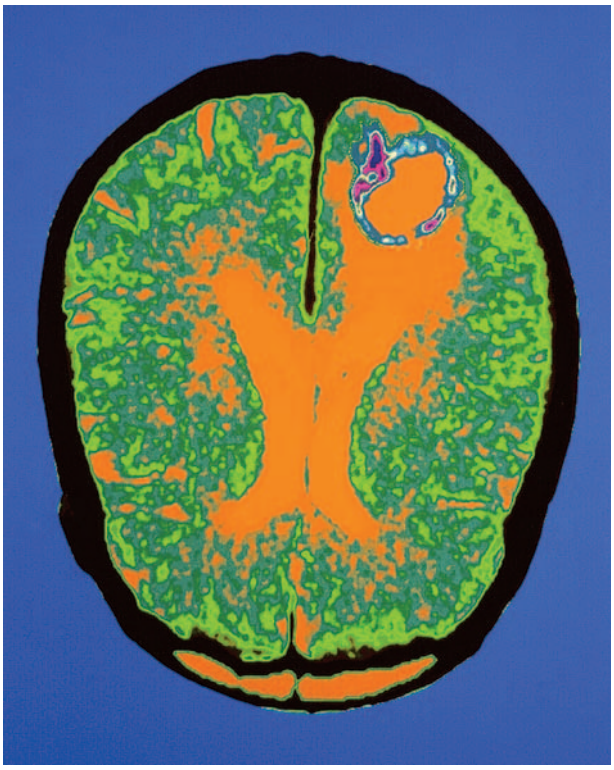
Magnetic resonance imaging (MRI)—An imaging technique that uses a large circular magnet and radio waves to generate signals from atoms in the body. These signals are used to construct detailed images of internal body structures and organs, including the brain.

Vaccine—A substance prepared from a weakened or killed microorganism which, when injected, helps the body to form antibodies that will prevent infection by the natural microorganism.

Virus—A small infectious agent consisting of a core of genetic material (DNA or RNA) surrounded by a shell of protein. A virus needs a living cell to reproduce.

Prognosis

Encephalitis symptoms may last several weeks. Most cases of encephalitis are mild, and recovery is usually quick. Mild encephalitis usually leaves no residual neurological problems. Overall, approximately 10 percent of those with encephalitis die from their infections or complications such as secondary infection. Some forms of encephalitis have more severe courses, including herpes encephalitis, in which mortality is 15 to 20 percent with treatment, and 70 to 80 percent without. Antiviral treatment is ineffective for eastern equine encephalitis, and mortality is approximately 30 percent.



Computed tomography scan (CT scan) of a child with encephalitis. The right side of the brain shows abnormal dilation of the ventricles (orange). *(© Airelle-Joubert/Photo Researchers, Inc.)*

Permanent neurological consequences may follow recovery in some cases. Consequences may include personality changes, memory loss, language difficulties, seizures, and partial paralysis.

Prevention

Because encephalitis is caused by infection, it may be prevented by avoiding the infection. Minimizing contact with others who have any of the viral illnesses listed above may reduce one's chances of becoming infected. Most infections are spread by hand-to-hand or hand-to-mouth contact; frequent hand washing may reduce the likelihood of infection if contact cannot be avoided.

Mosquito-borne viruses may be avoided by preventing mosquito bites. Mosquitoes are most active at dawn and dusk and are most common in moist areas with standing water. Covering skin and using mosquito repellents on exposed skin can reduce the chances of being bitten.

Vaccines are available against some viruses, including polio, herpes B, Japanese encephalitis, and equine encephalitis. **Rabies vaccine** is available for animals; it is also given to people after exposure. Japanese encephalitis

vaccine is recommended for those traveling to Asia and staying in affected rural areas during transmission season.

Nutritional concerns

Adequate **nutrition** and fluids improve the chances for a full recovery from encephalitis.

Parental concerns

Parents should carefully monitor their infants and young children for symptoms of fever. Any fever that exceeds 103°F (39.4°C) for more than a few minutes should be promptly treated. Any complaints of a stiff neck, loss of consciousness, unexplained vomiting, or seizure activity should be promptly brought to competent medical attention.

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Encopresis

Definition

Encopresis is defined as repeated involuntary defecation somewhere other than a toilet by a child age four or older that continues for at least one month.

Description

Soiling, fecal soiling, and fecal incontinence are alternate terms used for this behavior. Whatever the cause, parents should talk openly about the problem with the child. When parents treat a bowel problem as a cause for embarrassment or shame, they may unintentionally aggravate or prolong it.

Demographics

About 1 to 3 percent of children are affected by encopresis. More boys than girls are affected.

Causes and symptoms

Encopresis can be one of two types, nonretentive encopresis and retentive encopresis. About 80 to 95 percent of all cases are retentive encopresis. Children with this disorder have an underlying medical reason for soiling. The remaining cases have no physical condition that bars normal toileting behaviors. This type, nonretentive encopresis, is a behavioral condition in which the child refuses to defecate in a toilet.

Retentive encopresis is most often the result of chronic **constipation** and fecal impaction. In these children, feces have become impacted in the child's colon, causing it to distend. This causes the child to not feel the urge to defecate. The anal sphincter muscle becomes weak and unable to contain the soft stools that pass around the impaction. Despite the constipation, these children actually do have regular, though soft, bowel movements that they are unable to control. The child may not even be aware that he or she has defecated until the fecal matter has already passed. Many children have a history of constipation that extends back as far as five years before the problem is brought to medical attention.

A child may exhibit nonretentive encopresis, or functional encopresis, for several reasons. First, he or she may not be ready for **toilet training**. When a child is learning appropriate toilet habits during toddlerhood and **preschool** years, involuntary or inappropriate bowel movements are common. Second, the child may be afraid of the toilet or of defecating in public places like school. Others may use fecal incontinence to manipulate their parent or other adults. These children often have other serious behavioral problems.

When to call the doctor

A doctor should be called whenever children experience unresolved constipation or difficulty controlling their stools.

Diagnosis

Before beginning treatment for encopresis, the pediatrician first looks for any physical cause for the inappropriate bowel movements. The doctor asks parents about the child's earlier toilet training and typical toileting behaviors and inquires about a history of constipation. The doctor will digitally examine the child's anal area to check the strength of the anal sphincter muscle and look for a fecal impaction. An abdominal x ray may be needed to confirm the size and position of the impaction.

Treatment

If the pediatrician makes a diagnosis of retentive encopresis, the physician may recommend **laxatives**, stool softeners, or an enema to free the impaction. Subsequently, the doctor may make several suggestions for to avoid chronic constipation. Children should eat a high-fiber diet, with lots of fruits, vegetables, and whole grains. They should be encouraged to drink larger amounts of water and get regular **exercise**. Children should be taught to not feel ashamed of toileting behaviors, and psychotherapy may help decrease the sense of shame and guilt that many children feel.

If no fecal impaction is found, the pediatrician works with a counselor or psychiatrist to analyze the variables that characterize the encopresis. If the child is not physically or cognitively ready for toilet training, it should be postponed.

In the remainder of nonretentive encopresis cases, treatment should then center on making sure the child has comfortable bowel movements, since some cases of nonretentive encopresis involve some level of discomfort associated with constipation.

Prognosis

The prognosis for most children with encopresis is good, assuming that all underlying problems are identified and appropriately treated.

Prevention

There is no known way to prevent encopresis. Experienced counselors suggest that early identification of problems and accurate diagnosis are useful in limiting the severity and duration of encopresis.

Nutritional concerns

A high-fiber diet may be recommended for persons with encopresis. Affected persons should consume lots of fruits, vegetables, and whole grains. Adequate to copious intake of fluids are also recommended.

Parental concerns

Parents of a child with a serious behavior disorder like **oppositional defiant disorder** should work with their child's therapist to deal with encopresis in the

KEY TERMS

Constipation—Difficult bowel movements caused by the infrequent production of hard stools.

Impaction—A condition in which earwax has become tightly packed in the outer ear to the point that the external ear canal is blocked.

context of other behavioral problems. Parents should work with their children to establish appropriate stooling behaviors and institute a system of rewards for successful toileting.

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Enterobacterial infections

Definition

Enterobacterial infections are disorders of the digestive tract and other organ systems produced by a group of rod-shaped bacteria called Enterobacteriaceae.

Description

Enterobacterial infections can be produced by bacteria that normally live in the human digestive tract without causing serious disease or by bacteria that enter from the outside. The most troublesome organism in this group is *Escherichia coli*. Other examples of enterobacteria are species of *Salmonella*, *Shigella*, *Klebsiella*, *Enterobacter*, *Serratia*, *Proteus*, and *Yersinia*.

Transmission

Enterobacterial infections in the digestive tract typically start when the organisms invade the mucous tissues that line the digestive tract. They may be bacteria that are already present in the stomach and intestines, or they may be transmitted by contaminated food and water. It is also possible for enterobacterial infections to spread by person-to-person contact. In many cases these infections are nosocomial, which means that they can be acquired in the hospital. The usual incubation period is 12 to 72 hours.

Demographics

Diarrhea caused by enterobacteria is a common problem in the United States. It is estimated that each person has an average of 1.5 episodes of diarrhea each

year, with higher rates in children, institutionalized people, and Native Americans. This type of enterobacterial infection can range from a minor nuisance to a life-threatening disorder, especially in infants; elderly persons; patients with **immunodeficiency**, including acquired immunodeficiency syndrome (**AIDS**); and malnourished people. Enterobacterial infections are one of the two leading killers of children in developing countries.

Causes and symptoms

E. coli infections cause most of the enterobacterial infections in the United States. The organisms are categorized according to whether they are invasive or noninvasive. Noninvasive types of *E. coli* include what are called enteropathogenic *E. coli* (EPEC), and enterotoxigenic *E. coli* (ETEC). EPEC and ETEC types produce a bacterial poison (toxin) in the stomach that interacts with the digestive juices and causes the patient to lose large amounts of water through the intestines.

The invasive types of *E. coli* are called enterohemorrhagic *E. coli* (EHEC), and enteroinvasive *E. coli,* (EIEC). These subtypes invade the stomach tissues directly, causing tissue destruction and bloody stools. EHEC can produce complications leading to hemolytic-uremic syndrome (HUS), a potentially fatal disorder marked by the destruction of red blood cells and kidney failure. EHEC has become a growing problem in the United States because of outbreaks caused by contaminated food. A particular type of EHEC known as O157:H7 has been identified since 1982 in undercooked hamburgers and unpasteurized milk and apple juice. Between 2 and 7 percent of infections caused by O157:H7 develop into HUS.

Klebsiella and *Proteus* sometimes cause urinary tract infections; **pneumonia** occurs generally in immunocompromised hosts or alcoholics, and ear and sinus infections in immunocompromised hosts. *Enterobacter* and *Serratia* can cause bacterial infection of the blood (bacteremia), particularly in patients with weakened immune systems.

Symptoms

The symptoms of enterobacterial infections are sometimes classified according to the type of diarrhea they produce.

WATERY DIARRHEA Patients infected with ETEC and some types of EPEC develop watery diarrhea. Rarely *Shigella* and *Salmonella* cause watery diarrhea. These infections are located in the small intestine, result from bacterial toxins interacting with digestive juices, do not produce inflammation, and do not usually need treatment with **antibiotics**.

BLOODY DIARRHEA (DYSENTERY) Bloody diarrhea, sometimes called dysentery, is produced by EHEC, EIEC, some types of *Salmonella*, some types of *Shigella*, and *Yersinia*. In dysentery, the infection is located in the colon, cells and tissues are destroyed, inflammation is present, and antibiotic therapy is usually required.

NECROTIZING ENTEROCOLITIS (NEC) **Necrotizing enterocolitis** (NEC) is a disorder that begins in newborn infants shortly after birth. Although NEC was not as of 2004 fully understood, it is thought that it results from a bacterial or viral invasion of damaged intestinal tissues. The disease organisms then cause the death (necrosis) of bowel tissue or gangrene of the bowel. NEC is primarily a disease of **prematurity**; 60–80 percent of cases occur in high-risk preterm infants. NEC is responsible for 2–5 percent of cases in newborn intensive care units (NICU). Enterobacteriaceae that have been identified in infants with NEC include *Salmonella*, *E. coli*, *Klebsiella*, and *Enterobacter*.

When to call the doctor

A healthcare professional should be called if an infected child exhibits any of the following symptoms:

- symptoms of **dehydration** such as decreased urination, dry mouth, irritability, and few or no tears when crying
- **vomiting** for more than three days
- **fever** greater than 102.2°F (39°C) in a toddler or older child, or any fever in an infant less than six months old
- fever that cannot be controlled with **acetaminophen** (Tylenol) or ibuprofen (Motrin), or that lasts more than three days
- presence of blood in diarrhea
- hard, swollen belly

Diagnosis

In order to confirm a diagnosis of enterobacterial infection, physicians usually rely on patient history, physical examination, and laboratory tests.

Patient history

The diagnosis of enterobacterial infections is complicated by the fact that viruses, protozoa, and other types of bacteria can also cause diarrhea. In most cases of mild diarrhea, it is not critical to identify the organism because the disorder is self-limiting. Some groups of patients, however, should have stool tests. They include:

- patients with bloody diarrhea
- patients with watery diarrhea who have become dehydrated
- patients with watery diarrhea that has lasted longer than three days without decreasing in amount
- patients with disorders of the immune system

The patient history is useful for public health reasons as well as a help to the doctor in determining what type of enterobacterium may be causing the infection. The doctor will ask about the frequency and appearance of the diarrhea as well as about other digestive symptoms. If the patient is nauseated and vomiting, the infection is more likely to be located in the small intestine. If the patient is running a fever, a diagnosis of dysentery is more likely. The doctor will also ask if anyone else in the patient's **family** or workplace is sick. Some types of enterobacteriaceae are more likely to cause group outbreaks than others. Other questions pertain to the patient's food intake over the previous few days and recent travels to countries with typhoid fever or cholera outbreaks.

Physical examination

The most important parts of the physical examination are checking for signs of severe fluid loss and examining the abdomen. The doctor will look at the inside of the patient's mouth and evaluate the skin for signs of dehydration. The presence of a skin rash and an enlarged spleen suggests typhoid fever rather than a bacterial infection. If the patient's abdomen hurts when the doctor examines it, a diagnosis of dysentery is more likely.

Laboratory tests

The stool test is most commonly used for identifying the cause of diarrhea. Examining a stool sample under a microscope can help to rule out parasitic and protozoal infections. Routine stool cultures, however, cannot be used to identify any of the four types of *E. coli* that cause intestinal infections. ETEC, EPEC, and EIEC are unusual in the United States and can usually be identified only by specialists in research laboratories. Because of concern about EHEC outbreaks, however, most laboratories in the United States as of 204 screen for O157:H7 with a test that identifies its characteristic toxin. All patients with bloody diarrhea should have a stool sample tested for *E. coli* O157:H7.

Treatment

The initial treatment of enterobacterial diarrhea is usually empiric. Empiric means that the doctor treats the patient on the basis of the visible symptoms and professional experience in treating infections, without waiting for laboratory test results. In uncomplicated cases, symptoms usually go away within five to ten days without treatment of antibiotics. In other cases, antibiotics may be necessary to overcome the infection. Newborn infants and patients with immune system disorders are given antibiotics once the organism has been identified. Gentamicin, tobramycin, and amikacin are in the early 2000s used more frequently to treat enterobacterial infections because many of the organisms are becoming resistant to ampicillin and cephalosporin antibiotics.

Alternative treatment

Alternative treatments for diarrhea are intended to relieve the discomfort of abdominal cramping. Most alternative practitioners advise consulting a medical doctor if the patient has sunken eyes, dry eyes or mouth, or other signs of dehydration.

HERBAL MEDICINE Herbalists may recommend cloves taken as an infusion or ginger given in drop doses to control intestinal cramps, eliminate gas, and prevent vomiting. Peppermint (*Mentha piperita*) or chamomile (*Matricaria recutita*) tea may also ease cramps and intestinal spasms.

HOMEOPATHY Homeopathic practitioners frequently recommend *Arsenicum album* for diarrhea caused by contaminated food and *Belladonna* for diarrhea that comes on suddenly with mucus in the stools. *Veratrum album* would be given for watery diarrhea, and *Podophyllum* for diarrhea with few other symptoms.

Nutritional concerns

Because of the extensive loss of water through diarrhea, it is important to prevent dehydration. The affected child should be encouraged to drink fluids such as water, breast milk or formula (if applicable), electrolyte replacement drinks, or clear broths. Diluted juice should be avoided because juice can worsen diarrhea. Drinks with **caffeine** should be avoided because of caffeine's diuretic effects (i.e., causes water to be lost through urine).

Prognosis

The prognosis for most enterobacterial infections is good; most patients recover in about a week or ten days without needing antibiotics. HUS, on the other hand, has a mortality rate of 35 percent even with intensive care. About one-third of the survivors have long-term problems with kidney function, and another 8 percent develop high blood pressure, seizure disorders, and blindness.

KEY TERMS

Dysentery—A disease marked by frequent watery bowel movements, often with blood and mucus, and characterized by pain, urgency to have a bowel movement, fever, and dehydration.

Empirical treatment—Medical treatment that is given on the basis of the doctor's observations and experience.

Escherichia coli—A type of enterobacterium that is responsible for most cases of severe bacterial diarrhea in the United States.

Hemolytic-uremic syndrome (HUS)—A potentially fatal complication of *E. coli* infection characterized by kidney failure and destruction of red blood cells.

Necrotizing enterocolitis—A serious bacterial infection of the intestine that occurs primarily in sick or premature newborn infants. It can cause death of intestinal tissue (necrosis) and may progress to blood poisoning (septicemia).

Nosocomial infection—An infection acquired in a hospital setting.

Toxin—A poisonous substance usually produced by a microorganism or plant.

Prevention

The World Health Organization (WHO) offers the following suggestions for preventing enterobacterial infections, including *E. coli* O157:H7 dysentery:

- Cook ground beef or hamburgers until the meat is thoroughly done. Juices from the meat should be completely clear, not pink or red. All parts of the meat should reach a temperature of 158°F (70°C) or higher.
- Do not drink unpasteurized milk or fruit juices or use products made from raw milk.
- Wash hands thoroughly and frequently, especially after using the toilet.
- Wash fruits and vegetables carefully or peel them. Keep all kitchen surfaces and serving utensils clean.
- If drinking water is not known to be safe, boil it or drink bottled water.
- Keep cooked foods separate from raw foods and avoid touching cooked foods with knives or other utensils that have been used with raw meat.

Parental concerns

Because of the extensive media coverage that often follows outbreaks of enterobacterial infections such as *E. coli*, many parents associate such infections with eating undercooked meat such as hamburger. It is important, however, that other modes of transmission be considered, such as poorly or infrequently washed hands. Frequent hand washing is encouraged for caregivers and children alike, particularly in settings such as daycares and schools.

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Enuresis *see* **Bed-wetting**

Eosinophilic gastroenteropathies

Definition

Eosinophilic gastroenteropathies are gastrointestinal (GI) diseases (enteropathies) in which one or more layers of the GI tract (most commonly the stomach and small

intestine) are selectively infiltrated with a type of white blood cell called eosinophils, as part of an allergic response.

Description

Eosinophilic gastroenteropathies are characterized by the accumulation of an abnormally large number of eosinophils (eosinophilic infiltration) in one or more specific places anywhere in the digestive system and associated lymph nodes resulting in **nausea**, difficulty swallowing, abdominal **pain**, **vomiting** and **diarrhea**, excessive loss of proteins in the GI tract, and **failure to thrive**. All gastroenteropathies are characterized by the presence of abnormal GI symptoms, eosinophilic infiltration in one or more areas of the GI tract, and the absence of an identified cause for the formation of an abnormally large number of eosinophils in the blood (eosinophilia). Some patients also suffer loss of protein from the body that often results in low blood levels of albumin and total protein (protein-losing enteropathy) due to increased GI tract permeability. As the GI tract wall becomes infiltrated with large numbers of eosinophils, its normal architecture is disrupted, and so is its function. Eosinophils are immune system white blood cells that destroy parasitic organisms and play a major role in allergic reactions. For this reason, the gastroenteropathies are often considered as food-related gastrointestinal allergy syndromes.

Eosinophilic gastroenteropathies have a specific name corresponding to the area of the digestive system where the highest numbers of eosinophils are found. They include the following:

- eosinophilic **gastroenteritis** (EG), in which eosinophilic infiltration occurs in one or more layers of the stomach and/or small intestine
- eosinophilic esophagitis (EE), in which eosinophilic infiltration is confined to the muscular tube that carries food from the throat to the stomach (esophagus)
- eosinophilic colitis (EC), in which the infiltration is confined to the large intestine (colon)
- eosinophilic duodenitis (ED), in which the infiltration is confined to the small intestine

Eosinophilic gastroenteritis (EG) is the best characterized gastroenteropathy. It is classified according to the layer of the GI tract involved, and mixed forms also occur. The walls of the GI tract have four layers of tissue, called mucosa, submucosa, muscularis externa, and serosa. The innermost layer is the mucosa, a membrane that forms a continuous lining of the GI tract from the mouth to the anus. In the large bowel, this tissue contains cells that produce mucus to lubricate and protect the smooth inner surface of the bowel wall. Connective tissue and muscle separate the muscosa from the second layer, the submucosa, which contains blood vessels, lymph vessels, nerves, and glands. Next to the submucosa is the muscularis externa, consisting of two layers of muscle fibers, one that runs lengthwise and one that encircles the bowel. The fourth layer, the serosa, is a thin membrane that produces fluid to lubricate the outer surface of the bowel so that it can slide against adjacent organs. The different types of EG are:

- Pattern I eosinophilic gastroenteritis: Children affected with Pattern I EG have extensive infiltration of eosinophils in the area below the submucosa and muscularis layers. It is more commonly seen in the stomach (gastric antrum) but may also affect the small intestine or colon. Patients typically have intestinal obstruction. Cramping and abdominal pain associated with **nausea and vomiting** occur frequently. Food allergy and past history of allergy are less common in these patients than in patients with Pattern II EG.
- Pattern II eosinophilic gastroenteritis: In this the most prevalent form of EG, extensive infiltration of eosinophils occurs in the mucosal and submucosal layers. These patients have colicky abdominal pain, nausea, vomiting, diarrhea, and weight loss. Infants with Pattern II EG also commonly have a history of allergy. The condition may also be associated with protein-losing enteropathy, low levels of iron in the blood serum or in the bone marrow (iron-deficiency anemia), or impaired absorption of nutrients by the intestines (malabsorption). Growth retardation, delayed **puberty**, or abnormal menstruations has also been reported in children and adolescents with Pattern II EG.
- Pattern III eosinophilic gastroenteritis: This least common form of eosinophilic gastroenteropathy involves the serosal layer and the entire GI wall is usually affected. Its inflammation leads to an accumulation of fluid in the abdomen (ascites). This fluid contains many eosinophils and can infiltrate the membrane of the lungs (pleural effusion). A history of allergy also appears to be common in this group. Symptoms may include chest pain, **fever**, shortness of breath, and limited motion of the chest wall.

Eosinophilic esophagitis (EE) is characterized by the abnormal accumulation of eosinophils localized in the esophagus. In EE, high levels of eosinophils are detected in the esophagus but not in any other parts of the digestive tract. The presence of the eosinophils in the esophagus causes inflammation of its walls, which makes digestion extremely painful. Unlike that of normal children, the esophagus of an individual with EE does

not have a smooth, uniform pink surface but displays lines (furrowing) and white patches. Children with EE have classic signs of gastroesophageal reflux (abdominal pain, difficulty swallowing, and vomiting) but fail to respond to antireflux medications. The danger of failing to diagnose this disorder is that children may be referred for unnecessary surgery because of their reflux symptoms.

Eosinophilic colitis (EC) is characterized by eosinophilic infiltration localized only in the large bowel, resulting in fever, diarrhea, bloody stools, **constipation**, obstruction/strictures, acute abdominal pain, and tenderness often localized in the right lower abdomen. EC often follows the onset of EG.

Eosinophilic duodenitis (ED) is characterized by eosinophilic inflammation of the small bowel that results in the production of leukotrienes, substances that participate in defense reactions and contribute to hypersensitivity and inflammation. Malabsorption of nutrients always results along with severe cramping, bowel obstruction, and intestinal bleeding with passage of bloody stools.

Related diseases

Other diseases feature enteropathies with symptoms similar to that of eosinophilic gastroenteropathies:

- Whipple's disease: This rare digestive disease of unknown origin affects the lining of the small intestine and results in malabsorption of nutrients. It may also affect other organs of the body.
- **Celiac disease** (celiac sprue): This chronic, hereditary, intestinal malabsorption disorder is caused by an intolerance to gluten, the insoluble component of wheat and other grains. Clinical improvement of symptoms follows withdrawal of gluten-containing grains in the diet.
- Mastocytosis: This genetic disorder is characterized by abnormal accumulations of a type of cell (mast cells) normally found in connective tissue. The liver, spleen, lungs, bone, skin, and sometimes the membrane surrounding the brain and spine (meninges) may be affected.
- Tropical sprue: This disorder unknown cause is characterized by malabsorption, multiple nutritional deficiencies, and abnormalities in the small bowel mucosa. It appears to be acquired and related to environmental and nutritional conditions and is most prevalent in the Caribbean, South India, and Southeast Asia.
- Crohn's disease: Also known as ileitis, regional enteritis, or granulomatous colitis, this disease is a form of inflammatory bowel disease characterized by severe, chronic inflammation of the wall of the GI tract.
- Dietary protein enteropathy: This disease is characterized by persistent diarrhea and vomiting with resulting malabsorption and failure to thrive with onset most commonly in infancy. Protein-losing enteropathy may lead to abnormally large amounts of fluid in the intercellular tissue spaces of the body (edema), abdominal distension, and lack of red blood cells (anemia).
- Dietary protein-induced proctocolitis: In generally healthy infants this disease of unknown origin causes visible specks or streaks of blood mixed with mucus in the stool. Blood loss is usually minimal, and anemia is rare. The disorder appears in the first months of life, with a mean age at diagnosis of two months.

Transmission

Although many factors have been identified as causing eosinophilic gastroenteropathies, some researchers suspect that undiscovered infections may also play a role. Thus, as of 2004, researches also believe an unknown mode of transmission may possibly be involved.

Demographics

In the United States, eosinophilic gastroenteritis is very rare, and the incidence is difficult to estimate. However, since Kaijser's description in 1937, more than 280 cases have been reported in the medical literature. Although cases have also been reported worldwide, the exact incidence is unclear because of a lack of diagnostic precision. Cases of EG are reported mostly in Caucasians, with some cases occurring among Asians. A slight male preponderance has also been documented. People with a history of **allergies**, eczema, and seasonal **asthma** are more likely to have this disease. Eosinophilic esophagitis was long thought to be a variant of stomach reflux disease. It is as of 2004 known to be a distinct disorder predominately occurring in children. A study published in the *New England Journal of Medicine* in 2004 shows that EE rates have risen so dramatically in recent years that they may be at higher levels than that of other inflammatory gastrointestinal disorders. At Cincinnati Children's hospital, cases of EE were examined in patients from Hamilton County. Between 1991 and 2003, 315 cases met diagnostic criteria for EE. Only 2.8 percent of these cases were identified prior to 2000. Of the 315 cases, 103 resided within the county. Incidence rates of EE have not been reported in any other region of the United States, so national incidence rates are impossible to determine. However, if rates were the same as they are in Hamilton County, the annual occurrence of EE would be one in every 10,000 children, or approximately 22,000 children in the United States. The exact

incidence and prevalence of EC and ED are not known, but these diseases are occurring or being diagnosed with increasing frequency and are especially prominent in children.

Causes and symptoms

The eosinophil is a component of the immune system and is particularly involved with defense against parasites, but as of 2004 no parasite had been found responsible for any of the eosinophilic gastroenteropathies. The cause or mechanism of eosinophilic infiltration is also unknown, although some scientists suspect that the condition, first identified in Europe in the mid 1940s, is genetic, as it seems that in about 16 percent of known cases, an immediate **family** member is also diagnosed with an eosinophilic GI disorder. Various factors have been shown to trigger eosinophilic infiltration of the GI tract, and it has been shown that this, in turn, causes tissue damage by loss of cell granules (degranulation) and the untimely release of small proteins specialized in cell-to-cell communication (cytokines) that directly damage the GI tract wall. Examples of factors that are believed to have an incriminating role in triggering a flare-up include foods that trigger an allergic reaction (allergens) and **immunodeficiency** disorders caused by very low levels of immunoglobulins that result in an increased susceptibility to infection. Honey intolerance and bee pollen administration have also been suggested as a causative agent for EG. Researchers have confirmed a familial pattern to EE, which suggests either a genetic predisposition or a relationship to an unknown environmental exposure.

Gastroenteropathy symptoms vary depending on where the eosinophils are found and in what layer of the digestive system their numbers are highest. Symptoms therefore tend to be highly specific to each individual case. They may only appear when certain foods are ingested, or only during certain seasons of the year, or every few weeks, or in severe cases, every time any food is eaten. Infants with eosinophilic gastroenteropathies usually have acute reactions after food intake (within minutes to in one to two hours) that generally include nausea, vomiting and severe abdominal pain, later followed by diarrhea. These symptoms may occur alone or as part of a shock reaction. Symptoms vary depending on the type of gastroenteropathy (EG, EE, EC, or ED) and on the precise location of eosinophilic infiltration within the digestive system, as well as which layer or layers of the digestive system wall is infiltrated with eosinophils. Symptoms include, but are not limited to, the following:

- abdominal pain (EG, EE, ED)
- anorexia (EG, EE)
- asthma (EE)
- bloating (EG, ED, EC)
- cramps (EG, EC, ED)
- feeling full before finishing a meal (early satiety) (EG)
- milk/formula regurgitation (EG, EE)
- nausea, vomiting (EG, EE, EC, ED)
- weight loss (EG, EE, EC, ED)
- diarrhea (EG, EC, ED)
- presence of fluid (edema) in ankles (EG, EE)
- choking (EE)
- difficulty swallowing (dysphagia) (EG, EE)
- strictures (EE, EC)
- passage of dark stools (melena) (EG, EC, ED)
- constipation (EC, ED)
- bowel obstruction (EC, ED)
- intestinal bleeding (EC, ED)

When to call the doctor

Parents should call their healthcare provider to test for eosinophilic involvement if their child has recurrent symptoms of gastrointestinal disorder and feeding problems.

Diagnosis

Eosinophilic gastroenteropathies are diseases that can be easily misdiagnosed. EE has long been misdiagnosed as gastroesophageal reflux, another digestive disease in which partially digested food from the stomach regurgitates and backs up (reflux). However, EE differs from esophageal reflux in the large numbers of eosinophils that are present in the GI tract. Diagnosis for eosinophilic gastroenteropathies is therefore only established on microscopic analysis of a tissue specimen (biopsy) revealing eosinophilic infiltration. Additionally, diagnosis is based on the following:

- Complete blood count (CBC): CBC reveals the presence of blood eosinophilia, found in 20 to 80 percent of cases. CBC also appears to differentiate between different types of eosinophilic gastroenteropathies, since they have different total eosinophil counts.
- Mean corpuscular volume test: This test can determine the presence of iron-deficiency anemia and serum albumin levels that vary according to disorder type.
- Fecal protein loss test: This test is used to identify the inability to digest and absorb proteins in the GI tract.

- Imaging tests: Ultrasound and CT scan may show thickened intestinal walls and ascitic fluid in patients with Pattern III EG, as well as the degree of involvement of the different layers.
- Barium studies: In this test, the patient ingests a barium sulfate solution that makes for contrast on **x rays**. Barium studies can reveal mucosal edema and thickening of the small intestinal wall in EC and ED.
- Exploratory abdominal surgery (laparotomy): In some cases, laparotomy may be indicated, especially in patients with Pattern III EG.

Treatment

Treatment of eosinophilic gastroenteropathies is mainly symptomatic and supportive. Surgery may be necessary in severe EC cases in which there is obstruction of the intestines.

As of 2004 there is no known cure for eosinophilic gastroenteropathies, so medications are used to relieve symptoms and prevent full-blown attacks (or flare-ups). The only known medication to successfully stop eosinophilic inflammation is the corticosteroid drug, prednisone. Oral glucocorticosteroids are usually prescribed for those with EC or ED obstructive symptoms. Children with Pattern II EG may benefit from anti-inflammatory medications (for example, oral glucocorticoids or oral cromolyn) and specialized diet therapy, particularly in the case of food intolerance or allergy. Fluticasone propionate (Flonase, Flovent) is reported to be helpful in most cases of EG, if the medicine is swallowed so that it comes directly in contact with the esophageal tissues that are infiltrated by eosinophils. There is also reported success with use of a drug (Montelukast) that stops the production of the inflammatory leukotrienes associated with EC. Elemental formulas are also very effective. Cromolyn sodium (Gastrocrom) has been used with some success but does not work in all cases.

Clinical trials

Parents may consider enrolling their EG diagnosed child or adolescent in a clinical trial sponsored by the National Institute of Allergy and Infectious Diseases (NIAID). For example, in as of 2004 on-going omalizumab clinical study, participants undergo the following procedures:

- Leukapheresis: This procedure is performed to collect quantities of white blood cells to study the effects of the drug omalizumab on eosinophils and other immune system substances. Blood flows from a needle inserted in an arm vein through a catheter (plastic tube) into a machine that separates the blood into its components by centrifugation (spinning). Some of the white cells are removed, and the rest of the blood (red cells, plasma, and platelets) is returned to the body through a needle placed in the other arm.
- Skin testing: Participants are tested for allergies to specific allergens. A small amount of various allergens are placed on the arm. The skin is pricked at the sites of the allergens and the skin reaction after several minutes is observed.
- Upper and lower endoscopy: These procedures are performed so doctors can examine the specific part of the GI tract involved in the disorder. If both endoscopies are required, they are performed at the same time. For the upper endoscopy procedure, the throat is sprayed with an anesthetic (numbin) medicine and a long, flexible tube is passed through the esophagus, stomach, and small intestine. For the lower endoscopy procedure, the tube is passed through the rectum into the large intestine. Subjects are given a medication to cause relaxation through a vein before the procedure. Biopsies are performed during both endoscopies.
- Omalizumab therapy: This procedure is given as an injection under the skin. Following the first injection, subjects stay in the NIH clinical center hospital for 24 hours for **safety** monitoring. The remaining doses are scheduled as outpatient visits. Two weeks after the last dose, patients are admitted to the clinical center for two to three days for repeat endoscopy, leukapheresis, skin testing, and physical examination. Additional follow-up visits are scheduled.

Alternative treatment

As of 2004, a study is being conducted on the effectiveness of the membrane stabilizing drug sodium chromoglycate. More research is required to determine long-term safety and effectiveness of this drug. An experimental steroid called Budesonide may be helpful, but no clinical trials had been performed as of 2004. The severity of EG flare-ups has been reduced in some patients with **antihistamines** (such as Claritin, Allegra, or Zyrtec). A new class of asthma medications called leukotriene inhibitors has shown some mixed results in clinical trials for asthma patients and has been used experimentally in cases of EG but without conclusive results.

Nutritional concerns

It is believed that whole food proteins are the most common triggers of an EG attack. Most infants with this condition are, therefore, put on a restricted diet and

KEY TERMS

Allergic reaction—An immune system reaction to a substance in the environment; symptoms include rash, inflammation, sneezing, itchy watery eyes, and runny nose.

Anemia—A condition in which there is an abnormally low number of red blood cells in the bloodstream. It may be due to loss of blood, an increase in red blood cell destruction, or a decrease in red blood cell production. Major symptoms are paleness, shortness of breath, unusually fast or strong heart beats, and tiredness.

Antibody—A special protein made by the body's immune system as a defense against foreign material (bacteria, viruses, etc.) that enters the body. It is uniquely designed to attack and neutralize the specific antigen that triggered the immune response.

Antigen—A substance (usually a protein) identified as foreign by the body's immune system, triggering the release of antibodies as part of the body's immune response.

Ascites—An abnormal accumulation of fluid within the abdominal cavity.

Biopsy—The surgical removal and microscopic examination of living tissue for diagnostic purposes or to follow the course of a disease. Most commonly the term refers to the collection and analysis of tissue from a suspected tumor to establish malignancy.

Complete blood count (CBC)—A routine analysis performed on a sample of blood taken from the patient's vein with a needle and vacuum tube. The measurements taken in a CBC include a white blood cell count, a red blood cell count, the red cell distribution width, the hematocrit (ratio of the volume of the red blood cells to the blood volume), and the amount of hemoglobin (the blood protein that carries oxygen).

Corticosteroids—A group of hormones produced naturally by the adrenal gland or manufactured synthetically. They are often used to treat inflammation. Examples include cortisone and prednisone.

Cytokines—Chemicals made by the cells that act on other cells to stimulate or inhibit their function. They are important controllers of immune functions.

Dysphagia—Difficulty in swallowing.

Enteropathy—A disease of the intestinal tract.

Eosinophil—A type of white blood cell containing granules that can be stained by eosin (a chemical that produces a red stain). Eosinophils increase in response to parasitic infections and allergic reactions.

Eosinophilia—An abnormal increase in the number of eosinophils, a type of white blood cell.

Gastroesophageal reflux disease (GERD)—A disorder of the lower end of the esophagus in which the lower esophageal sphincter does not open and close normally. As a result the acidic contents of the stomach can flow backward into the esophagus and irritate the tissues.

Gastrointestinal (GI) system—The body system involved in digestion, the breaking down and use of food. It includes the stomach, small intestine, and large intestine. Also known as the gastrointestinal tract.

Gastrostomy tube—A tube that is inserted through a small incision in the abdominal wall and that extends through the stomach wall into the stomach for the purpose of introducing parenteral feedings. Also called a gastric tube, gastrointestinal tube, or stomach tube.

Glucocorticoids—A general class of adrenal cortical hormones that are mainly active in protecting against stress and in protein and carbohydrate metabolism. They are widely used in medicine anti-inflammatories and immunosuppresives.

Immunodeficiency disease—A disease characterized chiefly by an increased susceptibility to infection. It is caused by very low levels of immunoglobulins that result in an impaired immune system. Affected people develop repeated infections.

Immunoglobulin G (IgG)—Immunoglobulin type gamma, the most common type found in the blood and tissue fluids.

Leukotrienes—Substances that are produced by white blood cells in response to antigens and contribute to inflammatory and asthmatic reactions.

Malabsorption—The inability of the digestive tract to absorb all the nutrients from food due to some malfunction or disability.

Melena—The passage of dark stools stained with blood pigments or with altered blood.

Protein-losing enteropathy—Excessive loss of plasma and proteins in the gastrointestinal tract.

Shock—A medical emergency in which the organs and tissues of the body are not receiving an adequate flow of blood. This deprives the organs and tissues of oxygen and allows the build-up of waste products. Shock can be caused by certain diseases, serious injury, or blood loss.

Stricture—An abnormal narrowing or tightening of a body tube or passage.

provided with elemental formulas containing no whole food proteins, such as Neocate or Elecare. For older children, physicians usually start by recommending a trial **elimination diet** that excludes milk, eggs, wheat, gluten, soy, and beef, because a link has been established with food intolerance and food allergy. Most patients improve significantly on diets avoiding foods to which they are allergic. Radioallergosorbent assay test (RAST) or skin testing can identify food hypersensitivity. If an exceptionally high number of food reactions are found, an amino-acid-based diet or elemental diet is often considered. Some patients with EE/EG/EC are even fed elemental formulas via a gastrostomy tube or are limited to TPN (blood-vessel feeding) if the disease is severe with many complications.

Prognosis

As of 2004 there is no cure for eosinophilic gastroenteropathies, and outcomes depend on the specific enteropathy. A small subset of patients are partly or totally disabled by the effects of the disorder, but most can have active and fulfilling lives. Eosinophilic gastroenteropathies are not known to be fatal, but some types cause such severe bleeding or nutritional deficiency that the condition may be life-threatening if not treated with appropriate medications and support measures. Some younger children who have been diagnosed at an early age are known to outgrow the most severe symptoms. Children with EG have a good prognosis. Mild and sporadic symptoms can be managed with observation, and disabling GI flare-ups can be controlled with prednisone. Most patients also respond well to oral glucocorticosteroids.

Prevention

No specific prevention measures can be recommended for eosinophilic gastroenteropathies since their specific cause was unknown as of 2004.

Parental concerns

Because of the high risk of misdiagnosis for eosinophilic gastroenteropathies, parents of infants who have persistent feeding problems and do not respond well to classical digestive disorder medications should request biopsies to test for possible eosinophilic involvement.

When diagnosis is confirmed, certain lifestyle changes are usually required, such as avoidance of certain foods or making sure that medication is taken every day. Parents should also be aware that commonly prescribed corticosteroid medications have side effects that are potentially serious. People who use them tend to become overweight, with swollen faces. Long-term use has also been shown to damage kidneys.

See also Food allergies/sensitivities; Gastroenteritis; Gastroesophageal reflux disease.

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American College of Gastroenterology. 4900–B South 31st St., Arlington, VA 22206. Web site: <www.acg.gi.org>.

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Monique Laberge, Ph.D.

Epigastric hernia *see* **Abdominal wall defects**

Epiglottitis

Definition

Epiglottitis is an infection of the epiglottis, which can lead to severe airway obstruction.

Description

When air is inhaled (inspired), it passes through the nose and the nasopharynx or through the mouth and the oropharynx. These are both connected to the larynx, a tube made of cartilage. The air continues down the larynx to the trachea. The trachea then splits into two branches, the left and right bronchi (bronchial tubes). These bronchi branch into smaller air tubes that run within the lungs, leading to the small air sacs of the lungs (alveoli).

Either food, liquid, or air may be taken in through the mouth. While air goes into the larynx and the respiratory system, food and liquid are directed into the tube leading to the stomach, the esophagus. Because food or liquid in the bronchial tubes or lungs could cause a blockage or lead to an infection, the airway is protected. The epiglottis is a leaf-like piece of cartilage extending upwards from the larynx. The epiglottis can close down over the larynx when someone is eating or drinking, preventing these food and liquids from entering the airway.

Epiglottitis is an infection and inflammation of the epiglottis. Because the epiglottis may swell considerably, there is a danger that the airway will be blocked off by the very structure designed to protect it. Air is then unable to reach the lungs. Without intervention, epiglottitis has the potential of being fatal. Because epiglottitis involves swelling and infection of tissues, which are all located at or above the level of the epiglottis, it is sometimes referred to as supraglottitis (supra meaning above). About 25 percent of all children with this infection also have **pneumonia**.

Demographics

In the twentieth century, epiglottitis was primarily a disease of two- to seven-year-old children, with boys twice as likely to become ill as girls. In the early 2000s vaccines have greatly reduced the incidence of *Haemophilus influenzae* type b (Hib) epiglottitis, and the disease is more frequently seen in adults. In children, epiglottitis is an incredibly rare disease, thanks to timely Hib **vaccination** in childhood.

KEY TERMS

Epiglottis—A leaf-like piece of cartilage extending upwards from the larynx, which can close like a lid over the trachea to prevent the airway from receiving any food or liquid being swallowed.

Extubation—The removal of a breathing tube.

Intubation—A procedure in which a tube is inserted through the mouth and into the trachea to keep the airway open and to help a patient breathe.

Laryngospasm—Spasmodic closure of the larynx.

Larynx—Also known as the voice box, the larynx is the part of the airway that lies between the pharynx and the trachea. It is composed of cartilage that contains the apparatus for voice production–the vocal cords and the muscles and ligaments that move the cords.

Nasopharynx—One of the three regions of the pharynx, the nasopharynx is the region behind the nasal cavity.

Oropharynx—One of the three regions of the pharynx, the oropharynx is the region behind the mouth.

Supraglottitis—Another term for epiglottitis.

Trachea—The windpipe. A tube composed of cartilage and membrane that extends from below the voice box into the chest where it splits into two branches, the bronchi, that lead to each lung.

Tracheostomy—A procedure in which a small opening is made in the neck and into the trachea or windpipe. A breathing tube is then placed through this opening.

Causes and symptoms

The most common cause of epiglottitis is infection with the bacteria called *Haemophilus influenzae* type b. Other types of bacteria are also occasionally responsible for this infection, including some types of *Streptococcus* bacteria and the bacteria responsible for causing **diphtheria**.

A patient with epiglottitis typically experiences a sudden **fever** and begins having severe throat and neck **pain**. Because the swollen epiglottis interferes significantly with air movement, every breath creates a loud, harsh, high-pitched sound referred to as **stridor**. Because the vocal cords are located in the larynx just below the area of the epiglottis, the swollen epiglottis makes the patient's voice sound muffled and strained. Swallowing becomes difficult, and the patient may drool. The patient often leans forward and juts out his or her jaw, while struggling for breath.

Epiglottitis strikes suddenly and progresses quickly. A child may begin complaining of a **sore throat** and within a few hours be suffering from extremely severe airway obstruction.

Diagnosis

Diagnosis begins with a high level of suspicion that a quickly progressing illness with fever, sore throat, and airway obstruction is very likely to be epiglottitis. If epiglottitis is suspected, no efforts should be made to look at the throat or to swab the throat in order to obtain a culture for identification of the causative organism. These maneuvers may cause the larynx to go into spasm (laryngospasm), completely closing the airway. These procedures should only be performed in a fully equipped operating room, so that if laryngospasm occurs, a breathing tube can be immediately placed in order to keep the airway open.

An instrument called a laryngoscope is often used in the operating room to view the epiglottis, which will appear cherry-red and quite swollen. An x ray picture taken from the side of the neck should also be obtained. The swollen epiglottis has a characteristic appearance, called the "thumb sign."

Treatment

Treatment almost always involves the immediate establishment of an artificial airway: inserting a breathing tube into the throat (intubation) or making a tiny opening toward the base of the neck and putting a breathing tube into the trachea (tracheostomy). Because the patient's apparent level of distress may not match the actual severity of the situation, and because the disease's progression can be quite surprisingly rapid, it is preferable to go ahead and place the artificial airway, rather than adopting a wait-and-see approach.

Because epiglottitis is caused by a bacteria, **antibiotics** such as cefotaxime, ceftriaxone, or ampicillin with sulbactam should be given through a needle placed in a vein (intravenously). This prevents the bacteria that are circulating throughout the bloodstream from causing infection elsewhere in the body.

Prognosis

With treatment (including the establishment of an artificial airway), only about 1 percent of children with

epiglottitis die. Without the artificial airway, this figure jumps to 6 percent. Most patients recover from the infection and can have the breathing tube removed (extubation) within a few days.

Prevention

Prevention involves the use of a vaccine against *H. influenzae* type b (called the **Hib vaccine**). It is given to babies at two, four, six, and 15 months. Use of this vaccine has made epiglottitis a very rare occurrence.

Parental concerns

Parents should be aware of the advantages of the Hib vaccine. They should also call the doctor immediately if a child has a sudden, high fever and neck or throat pain.

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American Academy of Otolaryngology-Head and Neck Surgery Inc. One Prince St., Alexandria VA 22314–3357. Web site: <www.entnet.org>.

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Epilepsy *see* **Seizure disorder**

Erb's palsy *see* **Brachial plexopathy, obstetric**

Erythema *see* **Fifth disease**

Erythroblastosis fetalis

Definition

Erythroblastosis fetalis, also known as hemolytic disease of the newborn or immune hydrops fetalis, is a disease in the fetus or newborn caused by transplacental transmission of maternal antibody, usually resulting from maternal and fetal blood group incompatibility. Rh incompatibility may develop when a woman with Rh-negative blood becomes pregnant by a man with Rh-positive blood and conceives a fetus with Rh-positive blood. Red blood cells (RBCs) from the fetus leak across the placenta and enter the woman's circulation throughout pregnancy with the greatest transfer occurring at delivery. This transfer stimulates maternal antibody production against the Rh factor, which is called isoimmunization. In succeeding pregnancies, the antibodies reach the fetus via the placenta and destroy (lyse) the fetal RBCs. The resulting anemia may be so profound that the fetus may die in utero. Reacting to the anemia, the fetal bone marrow may release immature RBCs, or erythroblasts, into the fetal peripheral circulation, causing erythroblastosis fetalis. Maternal-fetal incompatibilities of ABO blood types leading to neonatal erythroblastosis are less severe and less common than those of the Rh factor.

Description

Red blood cells (RBCs) carry several types of proteins, called antigens, on their surfaces. The A, B, and O antigens represent the classification of an individual's blood as type A, B, AB, or O. Depending on the genetic predisposition of the parents, an A, B, or O antigen gene can be passed to a child. How the genes are paired determines the person's blood type.

A person who inherits an A antigen gene from each parent has type A blood; receiving two B antigen genes corresponds with type B blood; and inheriting A and B antigen genes means a person has type AB blood. If the O antigen gene is inherited from both parents, the child has type O blood; however, the pairing of A and O antigen genes corresponds with type A blood; and if the B antigen gene is matched with the O antigen gene, the person has type B blood.

Another red blood cell antigen, called the Rh factor, also plays a role in describing a person's blood type. A person with at least one copy of the gene for the Rh factor has Rh-positive blood; if no copies are inherited, the person's blood type is Rh-negative. In blood typing, the presence of A, B, and O antigens plus the presence or absence of the Rh-factor determine a person's specific blood type, such as A-positive, B-negative, and so on.

A person's blood type has no effect on health. However, an individual's immune system considers only that person's specific blood type, or a close match, acceptable. If a radically different blood type is introduced into the bloodstream, the immune system produces antibodies, proteins that specifically attack and destroy any cell carrying the foreign antigen.

Determining a woman's blood type is very important when she becomes pregnant. Blood cells from the unborn baby (fetal red blood cells) can cross over into the mother's bloodstream, and this risk is higher at delivery. If the mother and her baby have compatible blood types, the crossover does not present any danger. However, if the blood types are incompatible, the mother's immune system produces antibodies against the baby's blood.

Usually, this incompatibility is not a factor in a first pregnancy, because few fetal blood cells reach the mother's bloodstream until delivery. The antibodies that form after delivery cannot affect the first child. In subsequent pregnancies, however, the fetus may be at greater risk. The threat arises from the possibility that the mother's antibodies will attack the fetal red blood cells. If this happens, the fetus can suffer severe health effects and may die.

There are two types of incompatibility diseases: Rh incompatibility disease and ABO incompatibility disease. Both diseases have similar symptoms, but Rh disease is much more severe, because anti-Rh antibodies cross over the placenta more readily than anti-A or anti-B antibodies. (The immune system does not form antibodies against the O antigen.) As a result, a greater percentage of the baby's blood cells may be destroyed by Rh disease.

Both incompatibility diseases are uncommon in the United States due to medical advances since the 1950s. Prior to 1946 (when newborn blood transfusions were introduced) 20,000 babies were affected by Rh disease yearly. Further advances, such as suppressing the mother's antibody response, have reduced the incidence of Rh disease to approximately 4,000 cases per year.

Rh disease only occurs if a mother is Rh-negative and her baby is Rh-positive. For this situation to occur, the baby must inherit the Rh factor gene from the father. Most people are Rh-positive. Only 15 to 16 percent of the Caucasian population is Rh-negative, compared to approximately 8 percent of the African-American population and significantly lower in Asian populations. Interestingly, the Basque population of Spain has an incidence of 30 to 32 percent Rh-negativity.

ABO incompatibility disease is almost always limited to babies with A or B antigens whose mothers have type O blood. Approximately one third of these babies show evidence of the mother's antibodies in their bloodstream, but only a small percentage develop symptoms of ABO incompatibility disease.

Cause and symptoms

Rh disease and ABO incompatibility disease are caused when a mother's immune system produces antibodies against the red blood cells of her unborn child. The antibodies cause the baby's red blood cells to be destroyed and the baby develops anemia. The baby's body tries to compensate for the anemia by releasing immature red blood cells, called erythroblasts, from the bone marrow.

The overproduction of erythroblasts can cause the liver and spleen to become enlarged, potentially causing liver damage or a ruptured spleen. The emphasis on erythroblast production is at the cost of producing other types of blood cells, such as platelets and other factors important for blood clotting. Since the blood lacks clotting factors, excessive bleeding can be a complication. If this condition develops in the fetus in utero, the pregnant woman will generally notice a decrease in fetal movement, which should be immediately reported to her clinician.

The destroyed red blood cells release the blood's red pigment (hemoglobin) which degrades into a yellow substance called bilirubin. Bilirubin is normally produced as red blood cells die, but the body is only equipped to handle a certain low level of bilirubin in the bloodstream at one time. Erythroblastosis fetalis overwhelms the removal system, and high levels of bilirubin accumulate, causing hyperbilirubinemia, a condition in which the baby becomes jaundiced. The **jaundice** is apparent from the yellowish tone of the baby's eyes and skin. If hyperbilirubinemia cannot be controlled, the baby develops kernicterus. The term kernicterus means that bilirubin is being deposited in the brain, possibly causing permanent damage.

Other symptoms that may be present include high levels of insulin and low blood sugar, as well as a condition called hydrops fetalis. Hydrops fetalis is characterized by an accumulation of fluids within the baby's body, giving it a swollen appearance. This fluid accumulation inhibits normal breathing, because the lungs cannot expand fully and may contain fluid. If this condition continues for an extended period, it can interfere with lung growth. Hydrops fetalis and anemia can also contribute to heart problems.

Diagnosis

Erythroblastosis fetalis can be predicted before birth by determining the mother's blood type. If she is Rh-negative, the father's blood is tested to determine whether he is Rh-positive. If the father is Rh-positive, an antibody screen is done to determine whether the

Rh-negative woman is sensitized to the Rh antigen (developed isoimmunity). The indirect Coombs test measures the number of antibodies in the maternal blood. If the Rh-negative woman is not isoimmunized, a repeat antibody determination is done around 28 weeks' gestation, and the expectant woman should receive an injection of an anti-Rh (D) gamma globulin called Rhogham.

In cases in which incompatibility is not identified before birth, the baby suffers recognizable characteristic symptoms such as anemia, hyperbilirubinemia, and hydrops fetalis. The blood incompatibility is uncovered through blood tests such as the direct Coombs test, which measures the level of maternal antibodies attached to the baby's red blood cells. Other blood tests reveal anemia, abnormal blood counts, and high levels of bilirubin.

Treatment

Negative antibody titers can consistently identify the fetus that is not at risk; however, the titers cannot reliably point out the fetus which is in danger because the level of titer does not always correlate with the severity of the disease. For example, a severely sensitized woman may have antibody titers that are moderately high and remain at the same level while the fetus is being more and more severely affected. Conversely, a woman sensitized by previous Rh-positive fetuses may have a high antibody titer during her pregnancy while the fetus is Rh-negative.

When a mother has antibodies against her unborn infant's blood, the pregnancy is watched very carefully. Fetal **assessment** includes percutaneous umbilical cord blood sampling (PUBS) (cordocentesis), **amniocentesis**, amniotic fluid analysis, and ultrasound. Ultrasound should be done as early as possible in the first trimester to determine gestational age. Following that, serial ultrasounds and amniotic fluid analysis should be done to follow fetal progress. Complications are indicated by high levels of bilirubin in the amniotic fluid or baby's blood or if the ultrasound reveals hydrops fetalis. If bilirubin levels in amniotic fluid remain normal, the pregnancy can be allowed to continue to term and spontaneous labor. If bilirubin levels are elevated, indicating impending intrauterine death, the fetus can be given intrauterine transfusions at ten-day to two-week intervals, generally until 32 to 34 weeks gestation, when delivery should be performed.

There are two techniques that are used to deliver a blood transfusion to a baby before birth. The original intrauterine fetal transfusion, an intraperitoneal transfusion technique was first performed around 1963. With this method, a needle is inserted through the mother's abdomen and uterus and into the baby's abdomen. Red blood cells injected into the baby's abdominal cavity are absorbed into its bloodstream. In early pregnancy if the baby's bilirubin levels are gravely high, PUBS (cordocentesis) is performed. This procedure involves sliding a very fine needle through the mother's abdomen and, guided by ultrasound, into a vein in the umbilical cord to inject red blood cells directly into the baby's bloodstream.

After birth, the baby's symptoms are assessed. One or more transfusions may be necessary to treat anemia, hyperbilirubinemia, and bleeding. Hyperbilirubinemia is also treated with phototherapy, a treatment in which the baby is placed under a special light. This light causes changes in how the bilirubin molecule is shaped, which makes it easier to excrete. The baby may also receive oxygen and intravenous fluids containing electrolytes or drugs to treat other symptoms.

Prognosis

In many cases of blood type incompatibility, the symptoms of erythroblastosis fetalis are prevented with careful monitoring and blood type screening. Treatment of minor symptoms is typically successful, and the baby does not suffer long-term problems.

Nevertheless, erythroblastosis is a very serious condition for approximately 4,000 babies annually. In about 15 percent of cases, the baby is severely affected and dies before birth. Babies who survive pregnancy may develop kernicterus, which can lead to deafness, speech problems, **cerebral palsy**, or **mental retardation**. Extended hydrops fetalis can inhibit lung growth and contribute to heart failure. These serious complications are life threatening, but with good medical treatment, the fatality rate is very low.

Prevention

With any pregnancy, whether it results in a live birth, miscarriage, stillbirth, or abortion, blood typing is a universal precaution against blood compatibility disease. Blood types cannot be changed, but adequate forewarning allows precautions and treatments that limit the danger to unborn babies.

Parental concerns

If an Rh-negative woman gives birth to an Rh-positive baby, she is given an injection of Rhogam within 72 hours of the birth. This immunoglobulin destroys any

KEY TERMS

Amniocentesis—A procedure performed at 16-18 weeks of pregnancy in which a needle is inserted through a woman's abdomen into her uterus to draw out a small sample of the amniotic fluid from around the baby for analysis. Either the fluid itself or cells from the fluid can be used for a variety of tests to obtain information about genetic disorders and other medical conditions in the fetus.

Amniotic fluid—The liquid in the amniotic sac that cushions the fetus and regulates temperature in the placental environment. Amniotic fluid also contains fetal cells.

Anemia—A condition in which there is an abnormally low number of red blood cells in the bloodstream. It may be due to loss of blood, an increase in red blood cell destruction, or a decrease in red blood cell production. Major symptoms are paleness, shortness of breath, unusually fast or strong heart beats, and tiredness.

Antibody—A special protein made by the body's immune system as a defense against foreign material (bacteria, viruses, etc.) that enters the body. It is uniquely designed to attack and neutralize the specific antigen that triggered the immune response.

Antigen—A substance (usually a protein) identified as foreign by the body's immune system, triggering the release of antibodies as part of the body's immune response.

Bilirubin—A reddish yellow pigment formed from the breakdown of red blood cells, and metabolized by the liver. When levels are abnormally high, it causes the yellowish tint to eyes and skin known as jaundice. Levels of bilirubin in the blood increase in patients with liver disease, blockage of the bile ducts, and other conditions.

Hemoglobin—An iron-containing pigment of red blood cells composed of four amino acid chains (alpha, beta, gamma, delta) that delivers oxygen from the lungs to the cells of the body and carries carbon dioxide from the cells to the lungs.

Hemolysis—The process of breaking down of red blood cells. As the cells are destroyed, hemoglobin, the component of red blood cells which carries the oxygen, is liberated.

Hydrops fetalis—A condition in which a fetus or newborn baby accumulates fluids, causing swollen arms and legs and impaired breathing.

Hyperbilirubinemia—A condition characterized by a high level of bilirubin in the blood. Bilirubin is a natural byproduct of the breakdown of red blood cells, however, a high level of bilirubin may indicate a problem with the liver.

Isoimmunization—The development of antibodies in a species in response to antigens from the same species.

Percutaneous umbilical blood sampling (PUBS)—A technique used to obtain pure fetal blood from the umbilical cord while the fetus is in utero and also called cordocentesis.

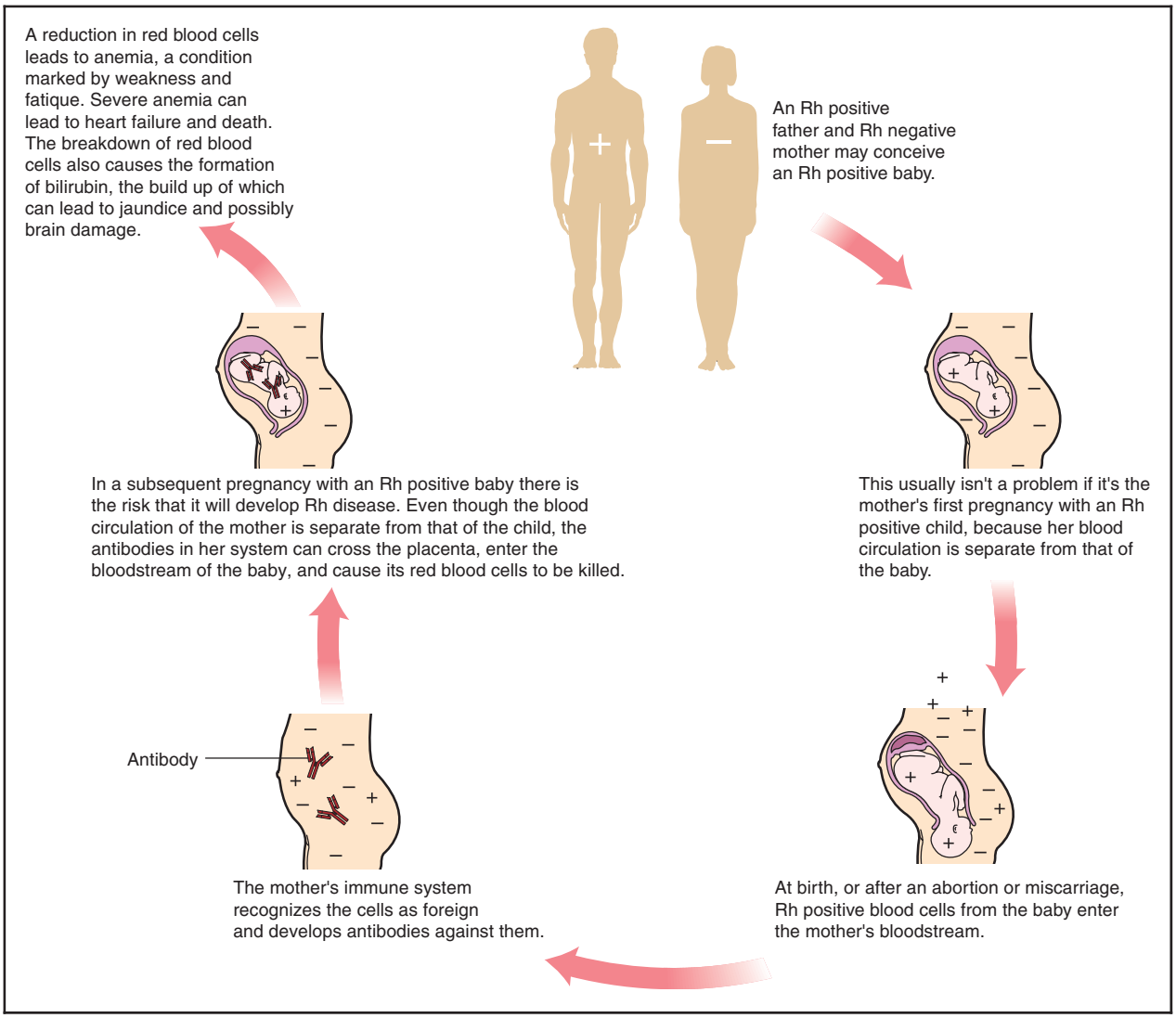
Placenta—The organ that provides oxygen and nutrition from the mother to the unborn baby during pregnancy. The placenta is attached to the wall of the uterus and leads to the unborn baby via the umbilical cord.

Platelet—A cell-like particle in the blood that plays an important role in blood clotting. Platelets are activated when an injury causes a blood vessel to break. They change shape from round to spiny, "sticking" to the broken vessel wall and to each other to begin the clotting process. In addition to physically plugging breaks in blood vessel walls, platelets also release chemicals that promote clotting.

Rh factor—An antigen present in the red blood cells of 85% of humans. A person with Rh factor is Rh positive (Rh+); a person without it is Rh negative (Rh-). The Rh factor was first identified in the blood of a rhesus monkey and is also known as the rhesus factor.

Transplacental—Passing through or occurring across the placenta.

fetal blood cells in her bloodstream before her immune system can react to them. In cases where this precaution is not taken, antibodies are created, and future pregnancies may be complicated. Because antibody production does not usually begin in a previously unsensitized mother until after delivery, erythroblastosis in subsequent children can be prevented by giving the mother an injection of Rhogam within 72 hours of delivery. The preparation must be given after each pregnancy—whether it ends in delivery, ectopic pregnancy,



Flow chart demonstrating how Rh disease is carried to fetus through mother. *(Illustration by Hans & Cassidy.)*

miscarriage, or abortion. The anti-Rh antibodies from the preparation destroy fetal RBCs in the mother's blood before they can sensitize the maternal immune system. If a massive fetomaternal hemorrhage has occurred, additional injections of the preparation may be necessary. This treatment has a failure rate of about 1–2 percent, apparently due to the mother's sensitization during pregnancy rather than at delivery. Therefore, all mothers who have Rh-negative blood and no apparent sensitization (as indicated by antibody titer) should be treated with a standard 300g dose of Rh(D) immune globulin (Rhogam) at about 28 weeks of gestation. The exogenous antibodies in the mother's circulation are gradually destroyed over the next three to six months, and the mother remains unsensitized. Rhogam should also be given after any episode of bleeding and after amniocentesis or chorionic villus sampling.

Delivery should be as nontraumatic as possible. The placenta should not be removed manually to avoid squeezing fetal cells into the maternal circulation. A newborn born with erythroblastosis should be attended to immediately by a pediatrician who is prepared to perform an exchange transfusion at once if required.

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Erythromycins

Definition

Erythromycins, also called macrolides, are a group of **antibiotics**, medicines that kill bacteria or prevent their growth.

Description

The antibiotics in this group are:

- azithromycin (Zithromax)
- clarithromycin (Biaxin)
- clindamycin (Cleocin)
- erythromycin (EES, Pediazole)
- lincomycin (Lincocin)

These drugs are chemically related and have similar uses, but because they are distributed differently in the body, they may be used for different purposes. There are other, older drugs in this group, but they are no longer in general use.

General use

Erythromycin is similar in use to penicillin and is widely used for patients who are allergic to penicillin. Penicillin has advantages over erythromycin in that it kills bacteria, while erythromycin only stops bacterial growth and relies on the body's immune system to kill bacteria. Also, erythromycin is more likely to cause stomach upset than is penicillin. Sometimes erythromycin may be used to treat a microorganism that is resistant to penicillin.

Azithromycin and clarithromycin both reach the lungs and respiratory tract better than does erythromycin. These two drugs may be preferred for respiratory tract infections.

Clindamycin and lincomycin are similar to each other and are more effective than erythromycin for treatment of infections caused by anaerobic bacteria. Anaerobic bacteria can grow in the absence of oxygen.

Precautions

Symptoms should begin to improve within a few days of beginning to take this medicine. If they do not, or if they get worse, parents should check with the physician who prescribed the medicine.

Erythromycins may cause mild **diarrhea** that usually goes away during treatment. However, severe diarrhea could be a sign of a very serious side effect. Anyone who develops severe diarrhea while taking erythromycin or related drugs should stop taking the medicine and call a physician immediately.

Side effects

The most common side effects are mild diarrhea, **nausea**, **vomiting**, and stomach or abdominal cramps. These problems usually go away as the body adjusts to the drug and do not require medical treatment. Less common side effects, such as sore mouth or tongue and vaginal **itching** and discharge also may occur and do not need medical attention unless they persist or are bothersome.

More serious side effects are not common but may occur. If any of the following side effects occurs, check with a physician immediately:

- severe stomach **pain**, nausea, vomiting, or diarrhea
- fever
- skin rash, redness, or itching
- unusual tiredness or weakness

Although rare, very serious reactions to azithromycin (Zithromax) are possible, including extreme swelling of the lips, face, and neck, and **anaphylaxis** (a violent allergic reaction which can potentially include shock). If children develop these symptoms after taking azithromycin, they should stop taking the medicine and parents should get them immediate medical help.

Other rare side effects may occur with erythromycins and related drugs.

Interactions

Erythromycins may interact with many other medicines. When interaction happens, the effects of one or both of the drugs may change or the risk of side effects may be greater. Parents of children taking erythromycins should let the physician know all other medicines their children are taking. Among the drugs that may interact with erythromycins are:

- acetaminophen (Tylenol)
- medicine for overactive thyroid
- male hormones (androgens)
- female hormones (estrogens)
- other antibiotics
- blood thinners
- antiseizure medicines such as valproic acid (Depakote, Depakene)
- caffeine
- antihistamine such as astemizole (Hismanal)
- antiviral drugs such as zidovudine (Retrovir)

The list above does not include every drug that may interact with erythromycins. Parents should be sure to check with a physician or pharmacist before combining erythromycins with any other prescription or nonprescription (over-the-counter) medicine.

Some of the stomach upset caused by erythromycin can be minimized by changing the dosage form. Erythromycin is available as enteric-coated tablets, which are released in the intestine rather than the stomach; as a liquid; and as bead-filled capsules. These forms are less likely to cause stomach upset than traditional tablets.

Parental concerns

If a child has had an allergic reaction to erythromycin or any of its related drugs, the prescriber should be notified.

It is very important for patients to take erythromycins for as long as they have been prescribed. Patients must not stop taking the drug just because symptoms begin to improve. This point is important with all types of infections, but it is especially important in strep infections, which can lead to serious heart problems if they are not cleared up completely.

Erythromycins work best when they are at constant levels in the blood. To help keep levels constant, patients should take the medicine in doses spaced evenly through the day and night. No doses should be missed. Some of these medicines are most effective when taken with a full glass of water on an empty stomach, but they may be taken with food if stomach upset is a problem. Others work equally well when taken with or without food. Check package directions or ask the physician or pharmacist for instructions on how to take the medicine.

Liquid forms of erythromycin should be administered with a medicinal teaspoon or other measuring device. Household teaspoons vary in size and may give either too much or too little of the medication.

Bead-filled capsules may be opened and sprinkled on pudding or applesauce for ease of administration. Enteric-coated tablets should never be split or crushed, since doing so will destroy the effectiveness of the coating.

Parents should never ask physicians to prescribe antibiotics for children's illnesses. Antibiotics are important for appropriate infections but are seriously overprescribed. Overuse leads to needless expense for the parents, some discomfort and risk for the child, and the development of antibiotic-resistant bacteria, which have become a public health problem.

Children have complained about the bitter taste of clarithromycin oral liquid. This factor should not be considered a problem. Liquid medications that taste good may be mistaken for candy or sweets, and children may overdose themselves. All medications should be kept away from children.

See also Pneumonia; Penicillins.

KEY TERMS

Anaphylaxis—Also called anaphylactic shock; a severe allergic reaction characterized by airway constriction, tissue swelling, and lowered blood pressure.

Enteric coating—A coating or shell placed on a tablet that breaks up and releases the medicine into the intestine rather than the stomach.

Microorganism—An organism that is too small to be seen with the naked eye, such as a bacterium, virus, or fungus.

Respiratory system—The organs that are involved in breathing: the nose, the throat, the larynx, the trachea, the bronchi and the lungs. Also called the respiratory tract.

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Esophageal atresia

Definition

Esophageal atresia (EA) is a birth defect (congenital anomaly) in which the esophagus, which connects the mouth to the stomach, is shortened and closed off (dead ended) at some point along its length. This defect almost always occurs in conjunction with **tracheoesophageal fistula** (TEF), a condition in which the esophagus is improperly attached to the trachea, the "windpipe" that carries air into the lungs. It is believed that these defects occur around the fourth week of pregnancy when the digestive tract is forming. There is no known cause for the defects.

Description

Failure of an unborn child (fetus) to develop properly can result in birth defects. These defects typically involve organs whose function is either incidental or not necessary at all before birth, meaning that the defects will not be detected until the baby is born. The digestive tract is unnecessary for fetal growth, since all **nutrition** comes from the mother through the placenta and umbilical cord. During fetal development, the esophagus and trachea arise from the same original tissue, forming into two side-by-side passageways, the esophagus leading from the throat to the stomach and digestive tract, and the trachea leading from the larynx to the lungs and respiratory system. Normally, the two tubes form separately (differentiate); however, in the case of EA/TEF, they do not differentiate, which results in various malformed configurations. There are five configurations, as follows:

- Type A (7.7%): Esophageal atresia in which both segments of the esophagus end in blind pouches. Neither segment is attached to the trachea.
- Type B (0.8%): Esophageal atresia with tracheoesophageal fistula in which the upper segment of the esophagus forms a fistula to the trachea. The lower segment of the esophagus ends in a blind pouch. This condition is very rare.
- Type C (86.5%): Esophageal atresia with tracheoesophageal fistula, in which the upper segment of the esophagus ends in a blind pouch (EA) and the lower segment of the esophagus is attached to the trachea (TEF).
- Type D (0.7%): Esophageal atresia with tracheoesophageal fistula, in which both segments of the esophagus are attached to the trachea. This is the rarest form of EA/TEF.
- Type H (4.2%): Tracheoesophageal fistula in which there is no esophageal atresia because the esophagus is continuous to the stomach. Fistula is present between the esophagus and the trachea.

Normally, the esophagus moves food from the mouth to the stomach. When the esophagus ends in a pouch instead of emptying into the stomach, food, liquids, and saliva cannot pass through. The combination of EA with TEF compromises digestion, nutrition, and respiration (breathing), creating a life-threatening

condition that requires immediate medical attention. All babies with EA/TEF require surgical repair to correct the condition and allow proper nutrition and swallowing. Many children have surgeries performed in separate stages over a period of years.

Demographics

Esophageal atresia alone or with tracheoesophageal fistula (EA/TEF) occurs in approximately one in 4,000 live births. Children' s hospitals in the United States report from five to 20 babies undergo surgery each year for EA/TEF.

Causes and symptoms

The cause of esophageal atresia, like that of most birth defects, was as of 2004 unknown.

An infant born with EA/TEF may at first appear to swallowing normally. However, the first signs of EA/TEF may be the presence of tiny, white, frothy bubbles of mucous in the infant's mouth and sometimes in the nose as well. When these bubbles are suctioned away, they reappear. This symptom occurs when the blind pouch begins to fill with mucus and saliva that would normally pass through the esophagus into the stomach. Instead these secretions back up into the mouth and nasal area, causing the baby to drool excessively. Although the infant may swallow normally, a rattling sound may be heard in the chest along with coughing and **choking**, especially when the infant tries to nurse. Some infants, depending on the severity of the defect, may appear blue (cyanosis), a sign of insufficient oxygen in the circulatory system. The infant' s abdomen may be swollen and firm (distended) because the abnormal trachea allows air to build up in the stomach, filling the abdominal space that holds the surrounding organs. Aspiration **pneumonia**, an infection of the respiratory system caused by inhalation of the contents of the digestive tract, may also develop.

When to call the doctor

EA is suspected when an infant drools excessively, accompanied by choking and sneezing and difficulty feeding. This condition may be detected within the first few days of life while the infant is still in the hospital or birthing center. If a newborn being cared for at home shows excessive drooling or begins to **cough** and struggle when nursing or swallowing, it is essential to contact the pediatrician immediately and to go to an emergency department for immediate care. If respiratory distress develops, it is critical to obtain immediate care to reduce the risk of aspiration of material (saliva or milk) into the trachea and the lungs.

Diagnosis

When a physician suspects esophageal atresia after being presented with the typical symptoms, diagnosis usually begins with gently passing a catheter through the nose and into the esophagus. Esophageal atresia is indicated if the catheter stops at the blind pouch, indicating that it has hit an obstruction. If EA is present, the catheter will typically stop at 4 to 5 inches (10–12 cm) from the nostrils. Barium-enhanced x-ray examination may reveal a dilated esophageal pouch, made larger by the collection of amniotic fluid in the pouch. During fetal development, the enlarged esophagus may also have pressed on and narrowed the trachea, a condition in the fetus that can contribute to fistula development. Air in the stomach may confirm the presence of fistula; gas in the large intestine rules out intestinal (duodenal) atresia. The physician will also perform a comprehensive physical examination, looking for other congenital anomalies that are known to accompany EA/TEF. Chest **x rays** may be taken to look for skeletal and cardiac abnormalities. Abdominal x rays may be taken as well to look for intestinal obstruction and abnormalities. An echocardiogram (ECG) may be performed to evaluate heart function and ultrasound of the kidneys performed to evaluate kidney function.

Treatment

Infants with EA, with or without TEF, are unlikely to survive without surgery to reconnect the esophagus. The procedure is done as soon as possible; however, **prematurity**, the presence of other birth defects, or complications of aspiration pneumonia may delay surgery. Once diagnosed, the baby may be fed intravenously until surgery is performed. Mucus and saliva will also be continuously removed via a catheter. Healthy infants who have no complications, such as heart or lung problems or other types of intestinal malformations, can usually have surgery within the first 24 hours of life. Surgery techniques used to treat the five types of EA/TEF defects are similar.

Surgery is conducted while the infant is under general anesthesia; a tube is placed through the mouth to continuously suction the esophageal pouch during the procedure. An intravenous line (IV tubing into the veins) is established to allow fluids to be administered as needed during surgery. Oxygen therapy is administered if needed. In infants with pulmonary problems, tracheal intubation (an airway placed in the trachea) may be

KEY TERMS

Anastomosis—Surgical reconnection of two ducts, blood vessels, or bowel segments to allow flow between the two.

Anomaly—Something that is different from what is normal or expected. Also an unusual or irregular structure.

Atresia—The congenital absence of a normal body opening or duct.

Congenital—Present at birth.

Esophagus—The muscular tube that leads from the back of the throat to the entrance of the stomach. It is coated with mucus and surrounded by muscles, and pushes food to the stomach by sequential waves of contraction. It functions to transport food from the throat to the stomach and to keep the contents of the stomach in the stomach.

Fetal—Refers to the fetus. In humans, the fetal period extend from the end of the eight week of pregnancy to birth.

Fistula—An abnormal channel that connects two organs or connects an organ to the skin.

Trachea—The windpipe. A tube composed of cartilage and membrane that extends from below the voice box into the chest where it splits into two branches, the bronchi, that lead to each lung.

Tracheoesophageal fistula—An abnormal connection between the trachea and esophagus, frequently associated with the esophagus ending in a blind pouch.

performed. If lung infection is suspected, the infant is given broad-spectrum **antibiotics** intravenously, either pre- or post-operatively.

The surgeon makes an incision in the right chest wall between the ribs, allowing access to the esophagus and the trachea for repair of one or both as needed. If the gap between the two portions of the esophagus is short, the surgeon may join both ends of the esophagus (anastomosis). If the upper portion of the esophagus is short and a long gap exists between upper and lower portions, reconstructive surgery cannot be performed, and the infant must receive nutrition in some way to allow several months of growth. In this case, a gastrostomy (stomach tube) may be surgically placed directly into the stomach for feeding. In the most typical EA/TEF repair, the fistula is first closed off, creating a separate airway. Then the blind esophageal pouch is opened and connected with suturing (stitching) to the other portion of the esophagus, creating a normal "food pipe" directly into the stomach. The esophagus is separated from the trachea if necessary. If the two ends of the esophagus are too far apart to be reattached, tissue from the large intestine is used to join them.

Nutritional concerns

If an infant is unable to nurse normally before surgery can be performed, nutrition is provided intravenously (parenteral) or directly through a tube into the stomach (gastronomy). After the surgery, infants should be able to swallow normally and resume nursing or feeding.

Prognosis

Surgery to correct esophageal atresia is usually successful, with survival rates close to 100 percent in otherwise healthy infants after the condition is corrected. Postoperative complications may include difficulty swallowing, since the esophagus may not contract efficiently, and gastrointestinal reflux, in which the acidic contents of stomach back up into the lower part of the esophagus, possibly causing ulcers.

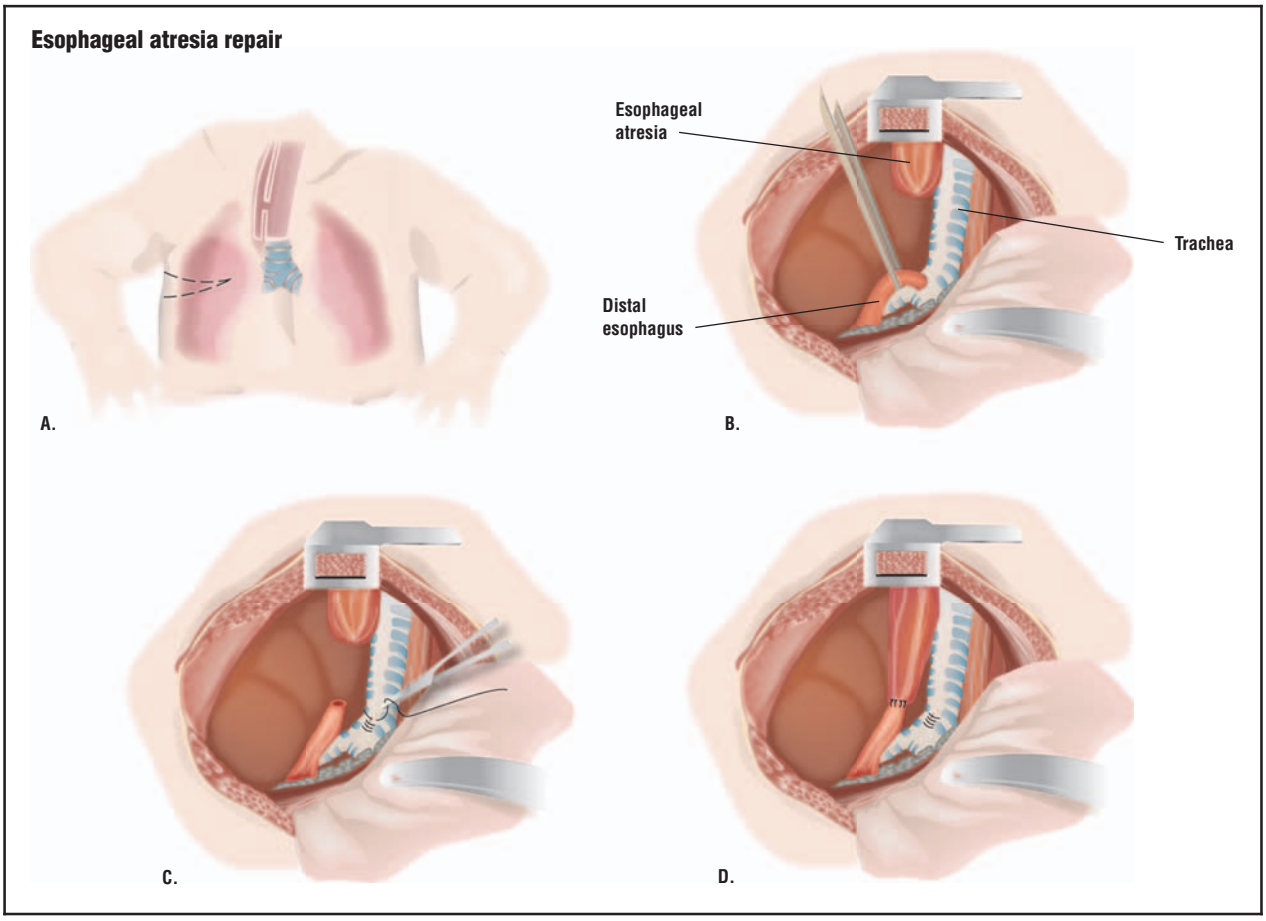
Prevention

No preventive measures are recommended because the cause of these birth defects was as of 2004 unknown and their occurrence not predictable.

Parental concerns

Despite a difficult beginning for infants with esophageal atresia with or without TEF, parents can be reassured that the defect can usually be corrected with surgery, allowing normal digestion, nutrition, and breathing to take place in their child. Concerns about complications are well founded, including increased susceptibility to colds and infections, as well as the presence of chronic conditions. Ongoing medical care helps manage these conditions and maintain good health in children who have had EA/TEF. Parents can seek advice about strengthening the child's immune system through appropriate nutrition and supplements.

See also Tracheoesophageal fistula.



To repair esophageal atresia, an opening is cut into the chest (A). The two parts of the existing esophagus are identified (B). The lower esophagus is detached from the trachea (C) and connected to the upper part of the esophagus (D). *(Illustration by GGS Information Services.)*

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Exercise

Definition

Exercise is physical activity that is planned, structured, and repetitive for the purpose of conditioning the body. Exercise consists of cardiovascular conditioning, strength and resistance training, and flexibility.

Purpose

Exercise is essential for improving overall health, maintaining fitness, and helping to prevent the development of **obesity**, **hypertension**, and cardiovascular disease. Surveys conducted by the Centers for Disease Control and Prevention (CDC) indicate that 61.5 percent of children aged nine to 13 years do not participate in any organized physical activity (for example, **sports**, dance classes) and 22.6 percent are not physically active during their free time. According to the American Obesity Association, approximately 30 percent of children and adolescents aged six to 19 years are overweight and 15 percent are obese.

A sedentary lifestyle and excess caloric consumption are the primary causes of this increase in overweight and obesity; regular exercise is considered an important factor in controlling weight. Overweight and obese children and adolescents are at higher risk of developing several medical conditions, including the following:

- **asthma**
- diabetes
- hypertension
- orthopedic complications, such as hip and knee **pain** and limited range of motion
- cardiovascular disease
- high cholesterol
- sleep apnea
- psychosocial disorders, such as depression, negative body image, and eating disorders

Clinical studies have shown that regular exercise has numerous benefits, including the following:

- preventing weight gain and maintaining healthy weight
- reducing blood pressure and cholesterol
- improving coordination
- improving **self-esteem** and self-confidence
- decreasing the risk of developing diabetes, cardiovascular disease, and certain types of cancer
- increased life expectancy

Description

Exercise consists of cardiovascular conditioning, strength and resistance training, and flexibility to improve and maintain the fitness of the body's heart, lungs, and muscles.

Cardiovascular conditioning

Cardiovascular conditioning involves moderate to vigorous physical activity that results in an elevated heart rate for a sustained period of time. Regular cardiovascular exercise improves the efficiency of the functioning of the heart, lungs, and circulatory system. For adults, aerobic exercise within a target heart rate range calculated based on a maximum heart rate by age is recommended. For healthy children, cardiovascular exercise that elevates the heart rate to no greater than a maximum heart rate of 200 beats per minute is recommended.

In general, the American Heart Association recommends at least 60 minutes of moderate to vigorous physical activity every day for children and adolescents. Cardiovascular conditioning activities should be appropriate for the age, gender, and emotional status of the child. Examples of exercise that elevates the heart rate are bicycle riding, running, swimming, jumping rope, brisk walking, dancing, soccer, and basketball.

Strength and resistance training

Strength and resistance training increases muscle strength and mass, bone strength, and the body's metabolism. Strengthening exercises increase muscle strength by putting more strain on a muscle than it is normally accustomed to receiving. Strength training can be performed with or without special equipment. Strength/resistance training equipment includes handheld dumbbells, resistance machines (Nautilus, Cybex), and elastic bands. Strength training can also be performed without equipment; exercises without equipment include push-ups, abdominal crunches, and squats. Children as young as six years can participate in strength training with weights, provided they are supervised by a fitness professional trained in youth strength training. Child-sized resistance machines may be available at some fitness facilities. According to youth strength training guidelines, children and adolescents should perform strength training for approximately 20 minutes two or three times weekly on nonconsecutive days.

Flexibility

Flexibility is important to improve and maintain joint range of motion and reduce the likelihood of muscle **strains**. Most young children are naturally more flexible than older children and adults and will instinctively perform movements that promote flexibility. As children age, they should be encouraged to continue to stretch. Flexibility is especially important for children and adolescents engaged in vigorous exercise (running, competitive sports). Stretching is best performed following a

warm-up and/or at the completion of an exercise session or sport. One activity that promotes flexibility that is increasing in popularity for children is **yoga**, in the form of children's yoga classes or exercise videos.

Precautions

Before a child begins any exercise program, he or she should be evaluated by a physician in order to rule out any potential health risks. Children and adolescents with physical restrictions or certain medical conditions may require an exercise program supervised by a health-care professional, such as a physical therapist or exercise physiologist. If **dizziness**, **nausea**, excessive shortness of breath, or chest pain occur during any exercise program, the activity should be stopped, and a physician should be consulted before the child resumes the activity. Children and adolescents who use any type of exercise equipment should be supervised by a knowledgeable fitness professional, such as a personal trainer.

Preparation

A physical examination by a physician is important to determine if strenuous exercise is appropriate or detrimental. Prior to beginning exercise, a proper warm-up is necessary to help prevent the possibility of injury resulting from tight muscles, tendons, ligaments, and joints. Appropriate warm-up exercises include walking, light calisthenics, and stretching.

Aftercare

Proper cool-down after exercise is important and should include a gradual decrease in exercise intensity to slowly bring the heart rate back to the normal range, followed by stretches to increase flexibility and reduce the likelihood of muscle soreness. Following vigorous activities that involve sweating, lost fluids should be replaced by drinking water.

Risks

Improper warm-up and inappropriate use of weights can lead to muscle strains. Overexertion without enough time between exercise sessions to recuperate also can lead to muscle strains, resulting in inactivity due to pain. Some children and adolescents may be susceptible to exercise-induced asthma. For children and adolescents who perform high-impact activities, such as running, stress **fractures** may occur. **Dehydration** is a risk during longer activities that involve sweating; children and adolescents should be supplied with water during and after activity.

KEY TERMS

Aerobic—An organism that grows and thrives only in environments containing oxygen.

Calisthenics—Exercise involving free movement without the aid of equipment.

Normal results

Significant health benefits are obtained by including at least a moderate amount of physical exercise for 30 to 60 minutes daily. Regular physical activity plays a positive role in preventing disease and improving overall health status. For children and adolescents just beginning an exercise program, results (including weight loss, increased muscle strength, and aerobic capacity) will be noticeable in four to six weeks.

Parental concerns

Given the increasing prevalence of overweight and obesity in children and adolescents, it is important for parents to encourage regular exercise and also serve as role models by exercising themselves. Television, computers, and **video games** have replaced physical activity for playtime for the majority of children. Parents should make a commitment to replacing sedentary activities with active indoor and outdoor games. For busy families, exercise can be performed in multiple 10- to 15-minute sessions throughout the day.

For children aged two to five years, physical activities should emphasize basic movement skills, imagination, and **play**. Examples of appropriate activities for this age group include rolling and bouncing a ball, jumping, hopping, skipping, mimicking animal movements, and pedaling a tricycle.

For children aged five to eight years, physical activities should emphasize basic motor skills and more complex movements (eye-hand coordination). Non-competitive group sports or classes are appropriate for this age, and parents should focus on helping their children find an enjoyable physical activity.

For children aged eight to ten years, physical activities should emphasize the benefits of regular exercise. Team sports and group classes are appropriate for this age. Experts have found that physical activity decreases in this age group, so parents should focus on being

Mother and daughter practicing yoga. *(© Ariel Skelley/Corbis.)*

supportive and encouraging their children to be physically active.

For children aged 11 to 14 years, physical activities should continue to emphasize the benefits of regular exercise. Participation in team sports, as well as individual activities, such as dance or martial arts, is appropriate for this age. Peer influence and hormonal changes can affect participation in group physical activities, so parents should consider encouraging exercise at home for children reluctant to participate with peers.

Community centers, local YMCAs, health clubs, and other organizations offer age-appropriate exercise programs for children and adolescents led by experienced and knowledgeable instructors. In addition, home exercise videos geared toward children are available in stores and from Web sites.

For children and adolescents with medical conditions that may limit exercise or place them at higher risk for exercise-related complications, supervised exercise programs may be available at hospital-based wellness centers.

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American College of Sports Medicine. 401 W. Michigan Street, Indianapolis, IN 46202–3233. Web site: <www.acsm.org/>.

American Council on Exercise. 4851 Paramount Drive San Diego, California 92123. Web site: <www.acefitness.org>.

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KEY TERMS

Asthma—A disease in which the air passages of the lungs become inflamed and narrowed, causing wheezing, coughing, and shortness of breath.

Bronchitis—Inflammation of the air passages of the lungs.

Chronic—Refers to a disease or condition that progresses slowly but persists or recurs over time.

Cough suppressant—A medication that stops or prevents coughing.

Emphysema—A chronic respiratory disease that involves the destruction of air sac walls to form abnormally large air sacs that have reduced gas exchange ability and that tend to retain air within the lungs. Symptoms include labored breathing, the inability to forcefully blow air out of the lungs, and an increased susceptibility to respiratory tract infections. Emphysema is usually caused by smoking.

Mucus—The thick fluid produced by the mucous membranes that line many body cavities and structures. It contains mucin, white blood cells, water, inorganic salts, and shed cells, and it serve to lubricate body parts and to trap particles of dirt or other contaminants.

Phlegm—Thick mucus produced in the air passages.

Respiratory system—The organs that are involved in breathing: the nose, the throat, the larynx, the trachea, the bronchi and the lungs. Also called the respiratory tract.

Secretion—A substance, such as saliva or mucus, that is produced and given off by a cell or a gland.

Trachea—The windpipe. A tube composed of cartilage and membrane that extends from below the voice box into the chest where it splits into two branches, the bronchi, that lead to each lung.

Expectorants

Definition

Expectorants are drugs that loosen and clear mucus and phlegm from the respiratory tract.

Description

There are two drugs that are routinely used to clear mucus from the respiratory tract: guaifenesin and acetylcysteine. Guaifenesin may be taken by mouth and is an ingredient in many over-the-counter **cough** and cold remedies. Although acetylcysteine is by far the more reliable of the two, it must be administered with special inhalation equipment or instilled directly into the trachea.

Other drugs have been used as expectorants, but lack evidence of either efficacy or **safety** or both:

- ammonium chloride
- bromhexine
- ipecacuanha
- potassium iodide
- wild cherry syrup

These drugs, and others, are not in common use, although wild cherry syrup may be used as a flavoring agent in some liquid cough preparations. Some home remedies, including chicken soup and hot tea, may also be useful in breaking up mucus.

General use

Guaifenesin, the only medicinal product in common use as an expectorant, is an ingredient in many cough medicines, such as Anti-Tuss, Dristan Cold & Cough, Guaifed, GuaiCough, and some Robitussin products. Some products that contain guaifenesin are available only with a physician's prescription; others can be bought without a prescription. They come in several forms: capsules, tablets, and liquids. There is some dispute, even among experts, about whether guaifenesin is an effective expectorant. In some studies, it has been useful, while in others it has not shown any value. Guaifenesin should not be given to children under the age of two years unless directed by a physician.

Precautions

Guaifenesin is not meant to be used for coughs associated with **asthma**, emphysema, chronic **bronchitis**, or **smoking**. It also should not be used for coughs that are producing a large amount of mucus. A lingering cough could be a sign of a serious medical condition. Coughs that last more than seven days or are associated with **fever**, rash, **sore throat**, or lasting **headache** should have medical attention. Parents should call a physician as soon as possible.

Side effects

Side effects are rare but may include **vomiting**, **diarrhea**, stomach upset, headache, skin rash, and **hives**.

Interactions

Guaifenesin is not known to interact with any foods or other drugs. However, cough medicines that contain guaifenesin may contain other ingredients that do interact with foods or drugs. Parents should check with a physician or pharmacist for details about specific products.

Because the value of guaifenesin is uncertain, while the adverse effects have been documented, parents should consider using alternatives to guaifenesin-containing cough remedies for children.

Parental concerns

There is no good evidence either for or against the use of over-the-counter products for treatment of coughs. Parents may wish to avoid using cough remedies for children unless instructed to do so by a physician. Expectorants are for use only in coughs with mucus production, sometimes called productive coughs. They are of no value in coughs without mucus, sometimes called dry coughs or non-productive coughs. The recommended dosage on the product label should not be exceeded. A calibrated medicinal teaspoon, not a household teaspoon, should be used to measure any medication.

See also Cough supressants; Decongestants.

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Extracorporeal membrane oxygenation

Definition

Extracorporeal membrane oxygenation (ECMO) is a procedure that uses an artificial heart-lung machine to take over the work of the lungs (and sometimes the heart). ECMO is used most often in newborns and young children, but it also can be used as a last resort for adults whose heart or lungs are failing.

Purpose

In newborns, ECMO is used to support or replace an infant's undeveloped or failing lungs by providing oxygen and removing carbon dioxide waste products so the lungs can rest. Infants who need ECMO may include those with the following problems:

- Meconium aspiration syndrome: Breathing in of a newborn's first stool by a fetus or newborn, which can block air passages and interfere with lung expansion.
- Persistent pulmonary **hypertension**: A disorder in which the blood pressure in the arteries supplying the lungs is abnormally high.
- Respiratory distress syndrome: A lung disorder usually of premature infants that causes increasing difficulty in breathing, leading to a life-threatening deficiency of oxygen in the blood.
- Congenital diaphragmatic **hernia**: The profusion of part of the stomach and/or intestines through an opening in the diaphragm.
- Pneumonia
- Blood poisoning
- Inborn errors of metabolism: Some genetic diseases.

ECMO is also used to support a child's damaged, infected, or failing lungs for a few hours to allow treatment or healing. It is effective for those children with severe, but reversible, heart or lung problems who have not responded to treatment with a ventilator, drugs, or extra oxygen. Children who need ECMO usually have one of the following problems:

- immature or underdeveloped lungs
- heart failure
- pneumonia
- respiratory failure caused by trauma or severe infection
- status asthmaticus (severe **asthma** attack)

The ECMO procedure can help a patient's lungs and heart rest and recover, but it will not cure the underlying disease. Any patient who requires ECMO is seriously ill and will likely die without the treatment. Because there is some risk involved, this method is used only when other means of support have failed.

Precautions

Typically, ECMO patients have daily chest **x rays** and blood work, and constant vital sign monitoring. They are usually placed on a special rotating bed that is designed to decrease pressure on the skin and help move secretions from the lungs.

After the child is stable on ECMO, the breathing machine settings are lowered to "rest" settings, which allows the lungs to rest without the risk of too much oxygen or pressure from the ventilator.

Description

There are two types of ECMO. Venoarterial (V-A) ECMO supports the heart and lungs and is used for patients with blood pressure or heart functioning problems in addition to respiratory problems. Venovenous (V-V) ECMO supports the lungs only.

V-A ECMO requires the insertion of two tubes, one in the jugular and one in the carotid artery. In the V-V ECMO procedure, the surgeon places a plastic tube into the jugular vein through a small incision in the neck.

Once in place, the tubes are connected to the ECMO circuit, and then the machine is turned on. The child's blood flows out through the tube and may look very dark because it contains very little oxygen. A pump pushes the blood through an artificial membrane lung, where oxygen is added and carbon dioxide is removed. The size of the artificial lung depends on the size of the child. The blood is then warmed and returned to the patient. A steady amount of blood (called the flow rate) is pushed through the ECMO machine every minute. As the patient improves, the flow rate is lowered.

Many patients require heavy sedation while they are on ECMO to lessen the amount of oxygen needed by the muscles.

As the patient improves, the amount of ECMO support is decreased gradually, until the machine is turned off for a brief trial period. If the patient does well without ECMO, the treatment is stopped.

Typically, newborns remain on ECMO for three to seven days, although some babies need more time (especially if they have a diaphragmatic hernia). Once the baby is off ECMO, he or she will still need a ventilator (breathing machine) for a few days or weeks.

Preparation

Before ECMO is begun, the patient receives medication to ease **pain** and restrict movement.

Aftercare

Because infants on ECMO may have been struggling with low oxygen levels before treatment, they may be at higher risk for developmental problems. They will need to be monitored as they grow. Some infants may have difficulty feeding after ECMO treatment.

Risks

Bleeding is the biggest risk for ECMO patients, since blood thinners (most often heparin) are given to guard against blood clots. Bleeding can occur anywhere

KEY TERMS

Carotid artery—One of the major arteries supplying blood to the head and neck.

Congenital diaphragmatic hernia—A profusion of part of the stomach through an opening in the diaphragm that is present at birth.

Meconium aspiration syndrome—Breathing in of meconium (a newborn's first stool) by a fetus or newborn, which can block air passages and interfere with lung expansion.

Membrane oxygenator—The artificial lung that adds oxygen and removes carbon dioxide.

Pulmonary hypertension—A disorder in which the pressure in the blood vessels of the lungs is abnormally high.

Respiratory distress syndrome—A lung disorder usually of premature infants that causes increasing difficulty in breathing, leading to a life-threatening deficiency of oxygen in the blood.

Venoarterial (V-A) bypass—The type of extracorporeal membrane oxygenation that provides both heart and lung support, using two tubes (one in the jugular vein and one in the carotid artery).

Venovenous (V-V) bypass—The type of extracorporeal membrane oxygenation that provides lung support only, using a tube inserted into the jugular vein.

in the body but is most serious when it occurs in the brain. This is why doctors periodically perform ultrasound brain scans of anyone on ECMO. **Stroke**, which may be caused by bleeding or blood clots in the brain, has occurred in some children undergoing ECMO.

If bleeding becomes a problem, the patient may require frequent blood or platelet transfusions or operations to control the bleeding. If the bleeding cannot be stopped, ECMO is withdrawn.

Other risks include infection or vocal cord injury. Some patients develop severe blood infections that cause irreversible damage to vital organs.

There is a small chance that some part of the complex equipment may fail, which could introduce air into the system or affect the patient's blood levels, causing damage or death of vital organs (including the brain). For this reason, the ECMO circuit is constantly monitored by a trained technologist, nurse, or respiratory therapist.

Normal results

Normal results include the lungs and/or heart returning to healthy functioning while on ECMO treatment.

Parental concerns

ECMO is used only for severely ill children. Parents need to talk with the nurse and doctor on a daily basis for updates on the condition of the child. The child may appear slightly swollen.

When to call the doctor

Notify a doctor if the child on ECMO is not behaving as expected (sedated and quiet), appears less pink (or bluer than normal), or is bleeding.

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ECMO Moms and Dads. Rt. 1, Box 176AA, Idalou, TX 79329. Web site: <www.medhelp.org/amshc/amshc341.htm>.

Extracorporeal Life Support Organization. 1327 Jones Dr., Ste. 101, Ann Arbor, MI 48105. Web site: <www.elso.med.umich.edu>.

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Extracurricular activities

Definition

Extracurricular activities are those sponsored by and usually held at school but that are not part of the academic curriculum. They often involve some time commitment outside of the regular school day.

Description

Extracurricular activities range from **sports** to newspaper editing to music and theater. Many activities, like

football and drama, enjoy extreme longevity, serving as a part of their school's program over a number of years. Others, like an ecology club or writers' workshop, may be offered for a shorter time span to reflect a community interest or involvement by a particular sponsoring faculty member or class of students. For many students, extracurricular activities present an opportunity to practice social skills and to experiment in activities that may represent a career interest. For a child who is not gifted academically, the opportunity to excel in the arts or sports may make a big difference in his or her **self-esteem**.

Many extracurricular activities, such as the school newspaper, photography, and drama, can lead to careers. Extracurricular activities also help to form the student's profile for consideration in college admissions. A student's academic record and scores on standardized tests form the core of his or her college application profile. However, admissions officers consider other factors, such as a demonstrated talent in athletics or the arts or leadership in school or extracurricular activities. After-school activities can also include scouting and volunteering, such as working with the Red Cross, a local animal shelter, a homeless shelter, or in a political campaign. Through these diverse activities, students can have fun, build a resume for college, increase **creativity**, improve organizational skills, learn time management, and develop people skills.

A 2001 survey of more than 50,000 high school students in Minnesota published in the March 2003 issue of the *Journal of School Health* found that those who participated in extracurricular activities had higher levels of social, emotional, and healthy behavior than students who did not participate. Students were classified into four groups based on their participation in sports and other activities, such as clubs, volunteer work, band, choir, or music lessons: neither activity, both, other activities only, and sports only. Odds ratios for the group involved in both types of activities were significantly higher than those for all the other groups for all healthy behaviors and measures of connectedness and significantly lower for all but one of the unhealthy behaviors.

Students involved in sports alone or in combination with other activities had significantly higher odds than the other two groups for **exercise**, milk consumption, and healthy self-image, and significantly lower odds for emotional distress, **suicidal behavior**, **family** substance abuse, and physical and sexual abuse victimization. Students involved in other activities alone or in combination with sports had significantly higher odds than the other two groups for doing homework and significantly lower odds for alcohol consumption, marijuana use, and vandalism.

Among male students in the Minnesota study, 19.1 percent engaged in neither sports nor other activities, 23.4 percent in other activities only, 15.1 percent in sports only, and 42.4 percent in both. Among female students, 12.6 percent were involved in neither, 31.6 percent in other activities only, 7.3 percent in sports only, and 48.6 percent in both. Analyses by race/ethnicity showed that white students were more likely than students of color to be involved in both sports and other activities (48.1% versus 33.6%) and sports only (11.4% versus 9.5%), while students of color were more likely to be involved in other activities only (33.8% versus 26.3%) and neither activity (23.1% versus 14.2%). Combining categories to look specifically at involvement in sports shows, that while participation rates for males (57.5%) and females (55.8%) are similar, a substantially higher proportion of white students participated in sports than students of color (59.5% versus 43.1%), according to the *Journal of School Health* article.

Preschool

Preschoolers are often enrolled in classes or activities outside of **preschool**. These activities include dance, swimming, T-ball, soccer, and gymnastics. Children this age can benefit from these activities, but the number of activities should be limited. Parents or other primary caregivers should consider how much time their children spend on these activities and the impact they have. Before enrolling children in activities outside the home and preschool, they should first attend a session to make sure it is appropriate for the child and that the child will benefit from it. A schedule that is too demanding can be stressful on a child and can lead to behavioral problems. Studies have shown that young children who feel stressed due too many extracurricular activities are more prone to illness.

School age

Studies show that children who participate in one or more after-school activities are less prone to negative **peer pressure** and have higher levels of self-esteem than children who do not participate. Studies have also shown that extracurricular activities can boost a child's academic performance and provide students with a way to feel proud of themselves and their capabilities. They can help a child release pent-up frustration and energy, develop social skills, and discover talents, abilities, and interests.

In the early school age years, it is important for parents to let the child chose the activity or activities.

Parents should not to press the child to win or excel. They should make sure the child does not overdo it, either by taking on an activity he or she cannot handle or by taking on too many activities. Parents need to insure the extracurricular activities do not interfere with school work or time spent with the family.

Once children reach middle or high school, there are usually many extracurricular activities available, including team sports such as soccer, baseball, basketball, and volleyball, and academic interests such as foreign language club, debate team, chess club, student government, student publications, 4-H Club, environmental clubs, choir, band, photography, politics, and business. Students may also have the chance to join clubs made up of students with a similar heritage or culture, including African American, Latino, Jewish, and gay and lesbian, such as the Gay-Straight Student Alliance groups found at some high schools.

Most school teachers, counselors, or principals provide a list of activities for student participation. The lists are often posted on student bulletin boards, and announcements are sometimes made in appropriate classes, such as history teachers promoting the history club or teachers promoting the group that they advise. Information can also be found in the school's student newspaper. Some students like to join clubs that one or more friends are joining while others join clubs to make friends. Students may want to keep in mind the following issues when they consider joining an extracurricular activity:

- Age: Students may have to be a certain age or in a certain grade to participate in an activity.
- Money: Some clubs or activities require students to pay a fee. There may also be costs involved with group outings, uniforms, or other items. Some groups require members to participate in fundraising activities.
- Physical exam: Students who want to join a sports team may be required to take a physical, and some schools require drug tests before students qualify to participate in sports or other extracurricular activities. Students with concerns or specific health problems, such as **asthma** or diabetes, should check with their family doctor before joining a team that requires physical activity, such as a sport or cheerleading.
- Grades: Some clubs, teams, or schools may require a minimum grade point average to join.
- Time: In competitive sports, time must be set aside for practice and competition. Team members are sometimes required to set up a game or help in other ways. Clubs can meet weekly, every other week, or monthly while athletic teams sometimes practice every day after school and on weekends.

Common problems

A common problem for many students involved in extracurricular activities is that they take on too much. Students should make out a schedule in advance of a semester that balances school, work, after-school activities, and home life. Also, activities should be fun rather than stressful for students. School grades should not suffer because of time spent at work or in after-school activities.

In sports, injuries are not uncommon but can sometimes be prevented with proper conditioning. Every child who plans to participate in organized athletic activity should have a pre-season sports physical. This special examination is performed by a pediatrician or family physician who carefully evaluates the site of any previous injury, may recommend special stretching and strengthening exercises to help growing athletes create and preserve proper muscle and joint interaction, and pays special attention to the cardiovascular and skeletal systems.

Telling the physician which sport the athlete plays helps that physician determine which parts of the body will be subjected to the most stress. The physician then is able to suggest steps to take for minimizing the chance of injury. Other injury-reducing game plans include:

- being in shape
- knowing and obeying the rules that regulate the activity
- not playing when tired, ill, or in **pain**
- not using steroids, which can improve athletic performance but cause life-threatening problems
- taking good care of athletic equipment and using it properly
- wearing appropriate protective equipment

Parental concerns

Parents must stay focused and make sure the extracurricular activities do not interfere with their child's schoolwork. If a child is tense, irritable, has difficulty concentrating in class, is often fatigued, or is shirking homework, it may be due to the extracurricular activities taking too much of his or her time. Parents may find it helpful to require a certain realistic level of academic achievement in order for the student to continue participation in extracurricular activities. Parents must also be

KEY TERMS

Cardiovascular—Relating to the heart and blood vessels.

Peer pressure—Social pressure exerted by a group or individual in a group on someone to adopt a particular type of behavior, dress, or attitude in order to be an accepted member of a group or clique.

Primary caregiver—A person who is responsible for the primary care and upbringing of a child.

Self-esteem—A sense of competence, achievement, and self-respect. Maslow felt that the most stable source of self-esteem is genuine accomplishment rather than public acclaim or praise.

Steroids—Hormones, including aldosterone, cortisol, and androgens, that are derived from cholesterol and that share a four-ring structural characteristic.

willing to put in a certain amount of time and effort, such as taking the child to and from after-school activities and events if the student does not drive.

It is important, especially with younger children, that parents check the activities to make sure there is adequate adult supervision for any team or club in which their children participate.

When to call the doctor

Children who refuse to join any extracurricular activities yet appear unhappy or have no friends may be suffering from emotional problems such as depression or low self-esteem. Professional help, such as counseling, may be needed. Sometimes a lack of self-esteem or other problems are too much for a student to handle alone. Parents may need to seek professional psychological help for children suffering from low self-esteem when the child is depressed or shows overly **aggressive behavior**.

When a child is hurt while playing sports, the family physician, pediatrician, or an orthopedic surgeon, should evaluate symptoms that persist, intensify, or reduce the athlete's ability to **play** without pain. Prompt diagnosis often can prevent minor injuries from becoming major problems or causing long-term damage.

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Association for Supervision and Curriculum Development. 1703 Beauregard St., Alexandria, VA 22311. Web site: <www.acsd.org>.

National Institute on Out-Of-School Time. 106 Central St., Wellesley, MA 02481. Web site: <www.niost.org>.

Society for Research in Child Development. University of Michigan, 3131 S. State St., Suite 302, Ann Arbor, MI 48108. Web site: <www.srcd.org>.

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Eye abrasion *see* **Corneal abrasion**

Eye and vision development

Definition

The visual system is the most complex sensory system in the human body. However, it is the least mature system at birth. Though they have the anatomical structures needed for sight, infants have not learned to use them yet. Much of their first weeks and months are spent learning to see. As children grow, more complex skills, like visual perception, develop.

Description

At birth, the sense of hearing is much more dominant than the sense of sight. Normal visual development is the change from just responding to simple brightness or high contrast, toward the organization of details into patterns and the ability to apply meaning to an object or picture.

Infancy

At birth, babies are capable of seeing shapes by following lines where light and dark meet. They can see variations of light and dark and shades of gray. Newborns can only focus between 8–12 inches (20–30 cm), so much of their vision is blurred. Full-term babies should be able to see their mother's facial expressions within a week of birth.

Eye muscle coordination in a newborn is also very immature. Babies' eyes often turn in or out or do not work together, a condition called **strabismus**. Babies initially learn to focus their eyes by looking at faces. They then gradually move out to objects brought close to them. Tracking and eye teaming skills begin to develop when infants start following moving objects. This usually happens by three months of age. Brightly colored moving objects, such as a mobile, can help stimulate visual development. Babies then start to learn how to coordinate their eye movements. At four months of age, babies can see the full range of colors. Between four and six months, the child normally begins batting at or reaching for the mobile or **toys** held in front of him or her.

During the first three to six months, the retina is fairly well-developed, and babies can visualize small objects. Depth perception also develops. By six months of age, the eye has reached about two-thirds of what will be its adult size. At this stage, the two eyes are most likely working together. The result is good binocular, or two-eyed, vision. It is during the first year of life that the eyes' greatest physical development occurs.

As babies start controlling their own physical movements, their eye/body coordination develops. By the fourth or fifth month, babies' brains have finished learning how to blend the images coming in from both their left and right eyes into a single image, with strong depth perception. Spatial and dimensional awareness keep improving as the baby learns to aim accurately when reaching for objects. Babies at this age also learn to change focus quickly and accurately between near and far distances.

A child's clarity of vision (visual acuity) has usually developed to 20/20 by the time the child reaches six months of age. At this time, babies achieve fairly precise eye movement control. At ages eight to 12 months, babies are judging distances well. Their eye/hand/body coordination continues to evolve, allowing them to grasp and throw objects with some accuracy. The integration of their fine motor abilities and their vision permits the child to manipulate smaller objects, and many begin feeding themselves. Once children begin to walk, they learn to use their eyes to guide and manage their bodies' large muscle groups to direct their whole movements.

Infancy

The following timeline discussion highlights some of the developmental milestones of vision development in a child's first year. Between birth and one month, a baby shows preference for familiar faces and objects, pays attention to the human face for short periods of time, has acuity of about 20/400 but can detect a black line on a white background that is only 1/16 of an inch (1.6 mm) wide, and possesses color vision, with the exception of blue.

At two months, a baby will visually lock onto a human face, watches people who are some distance away, is able to alternate his or her gaze between two people or objects, and demonstrates simple visual preferences.

Between four and six months, a baby is enthralled with other baby's faces, and he or she enjoys looking in a mirror. At this age the baby recognizes a person on sight and smiles. The baby also shifts from preferring what is familiar to that which is new, with the exception of people. The child will also look for objects when they fall from view.

From six to 12 months, a baby continues to "see" objects even when they are no longer visible. At this age, the baby also responds to words a parent uses to label familiar objects and people, by gazing in their direction.

KEY TERMS

Nystagmus—An involuntary, rhythmic movement of the eyes.

Strabismus—A disorder in which the eyes do not point in the same direction.

Toddlerhood

After the first year, children's eyes and vision continue to develop. Their eye muscles gain strength, and the connections between nerves multiply. This development is aided by providing visual stimulation. Activities such as stacking building blocks, coloring, and cutting all assist in improving eye/hand/body coordination, eye teaming, and depth perception. By age three, most children have developed the necessary language and motor skills that allow them to participate in some traditional vision tests.

Preschool

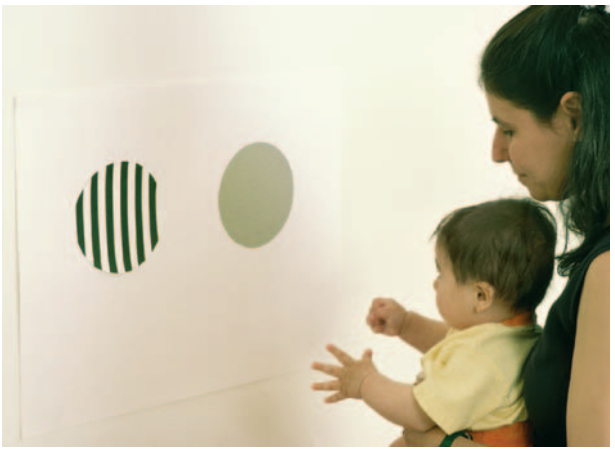
During the **preschool** years, a child's vision keeps developing. The child develops visually guided eye/hand/body coordination, the **fine motor skills** and visual motor skills required for reading. The following can facilitate a preschooler's visual development:

- reading aloud to the child and letting him or her see what is being read
- providing a chalkboard or finger paints and demonstrating how to use them in play
- allowing time for interacting with other children and for playing alone

School age

It is important that children have a complete eye examination before beginning school. The optometrist or ophthalmologist needs to determine if a child's vision is prepared to handle reading, writing, and other close-up activities. While toddlers use their eyes primarily for distance sight, school requires that the child's eyes focus on very close work for hours every day. This activity occasionally causes eye problems to arise. It is important to note that children rarely report vision problems. They believe their vision is normal and believe others see the way they do. The basic vision skills needed for school work are:

- near vision (the ability to see clearly at 10–13 inches [25-32 cm])



Child being tested for visual acuity. *(© Laura Dwight/Corbis.)*

- distance vision (the ability to see clearly beyond arm's reach)
- binocular vision (using both eyes together for depth perception)
- focusing skills (the ability to keep both eyes accurately focused at the proper distance)
- eye movement skills (the ability to aim the eyes accurately)
- peripheral awareness (the awareness of objects located to the side while looking straight ahead)
- eye-hand coordination

Common problems

Infants born prematurely have more difficulty integrating and interpreting visual information even when their acuity is normal. In some cases, children develop their visual reflex later than normal. This is called visual maturation delay. A condition, **nystagmus**, which sometimes develops in infancy, causes the eyes to jump, dance, wiggle, or oscillate. Babies with this problem may or may not have normal vision.

Parental concerns

Parents need to assess their child frequently for any signs that the child's visual development is not progressing as expected. Some vision disorders are untreatable at later ages, so it is important to have the child seen by an optometrist or ophthalmologist no later than the age of three.

When to call the doctor

At the first signs of eye and vision problems, parents should consult their pediatrician, optometrist, or ophthalmologist. Some of these signs are:

- an eye that is crossed far into the nasal area
- eyes turned grossly in or out or which do not move normally before the age of three months
- an eye that moves while the other remains still
- an eye that appears considerably different from the other
- the inability at three months of age of an infant to follow a toy passed in front of him from side to side

During the preschool years, parents should continue looking for signs that a vision development problem exists. These signs may include a short attention span for the child's age; difficulty with eye/hand/body coordination; or the avoidance of coloring, puzzles, and other activities.

A child should have his first eye exam by the age of three (or sooner if vision problems run in the **family**), so the practitioner can assess if vision is developing normally. Vision should be checked again when the child enters school.

Some of the signs of visual problems in the school age child are:

- frequently losing his or her place while reading
- frequently avoiding close work
- holding reading material closer than usual
- frequently rubbing the eyes
- complaining of headaches
- turning or tilting the head to use one eye only

Since vision changes may occur without the parents or the child noticing them, a child should visit an eye doctor at least every two years, more frequently if specific problems or risk factors exist.

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Eye cancer *see* **Retinoblastoma**

Eyeglasses and contact lenses

Definition

Eyeglasses and contact lenses are devices that correct refractive errors in vision. Eyeglass lenses are mounted in frames that are worn on the face, sitting mostly on the ears and nose, so that the lenses are positioned in front of the eyes. Contact lenses appear to be worn in direct contact with the cornea, but they actually float on a layer of tears that separates them from the cornea.

Purpose

The purpose of eyeglasses and contact lenses is to correct or improve the vision of people with nearsightedness (**myopia**), farsightedness (hyperopia), presbyopia, and astigmatism.

Description

Eyes are examined by optometrists (OD) or by ophthalmologists (MD or DO). Prescriptions, if necessary, are then given to patients for glasses. Eyeglasses are generally made by an optician. A separate contact lens-fitting exam is necessary if an individual wants contact lenses, because an eyeglass prescription can differ from a contact lens prescription.

Eyeglasses

More than 140 million people in the United States wear eyeglasses. People whose eyes have refractive errors do not see clearly without glasses, because the light emitted from the objects they are observing does not come into focus on their retinas. For people who are farsighted, images come into focus behind the retina; for people who are nearsighted, images come into focus in

front of the retina. For both, the result is a blurring of vision.

LENSES Lenses work by changing the direction of light so that images come into focus on the retina. The greater the index of refraction of the lens material and the greater the difference in the curvature between the two surfaces of the lens, the greater the change in direction of light that passes through it and the greater the correction.

Lenses can be unifocal, with one correction for all distances, or they can correct for more than one distance (multifocal). One type of multifocal, the bifocal, has an area of the lens (usually at the bottom) that corrects for nearby objects (about 14 in [35 cm] from the eyes); the remainder of the lens corrects for distant objects (about 20 ft [6 m] from the eyes). Another type of multifocal, a trifocal, has an area in-between that corrects for intermediate distances (usually about 28 in [71 cm]) Conventional bifocals and trifocals have visible lines between the areas of different correction; however, lenses in which the correction gradually changes from one area to the other, without visible lines, have been available since the 1970s. Such lenses are sometimes called progressives or no-line bifocals.

To be suitable for eyeglass lenses, a material must be transparent, without bubbles, and have a high index of refraction. The greater the index of refraction, the thinner the lens can be. Lenses are made from either glass or plastic (hard resin). The advantage of plastic is that it is lightweight and more impact-resistant than glass. The advantage of glass is that it is scratch resistant and provides the clearest possible vision.

Glass was the first material to be used for eyeglass lenses and was used for several hundred years before plastic was introduced. The crown glass used for eyeglass lenses has an index of refraction of 1.52.

In the early 2000s eyeglass wearers can choose polycarbonate or polyurethane materials for their lenses. Polycarbonate is the most impact-resistant material available for eyewear, and polyurethane has exceptional optical qualities and an index of refraction of up to 1.66, much higher than the conventional plastics used for lenses and even higher than glass. Parents whose children have high prescriptions should ask about high-index material options for their lenses. Aspheric lenses are also useful for high prescriptions. They are flatter and lighter than conventional lenses.

There are many lenses and lens-coating options for individual needs, including coatings that block the ultraviolet (UV) light and/or blue light which have been found to be harmful to the eyes. Such coatings are not needed on polycarbonate lenses, which already have UV protection. UV coatings are particularly important on sunglasses and ski goggles. Sunglasses, when nonprescription, should be labeled with an indication that they block out 99 to 100 percent of both UV-A and UV-B rays.

There are anti-scratch coatings that increase the surface hardness of lenses (an important feature when using plastic lenses) and anti-reflective (AR) coatings that eliminate almost all glare and allow other people to see the eyes of the wearer. AR coatings may be particularly helpful to people who use computers or who drive at night. Mirror coatings that prevent other people from seeing the wearer's eyes are also available. There is a whole spectrum of tints, from light to dark, used in sunglasses. Tint, however, does not block-out UV rays, so a UV coating is needed. Polaroid lenses that block out much of the reflected light also allow better vision in sunny weather and are helpful for people who enjoy boating. Photosensitive (photochromatic) lenses that darken in the presence of bright light are handy for people who do not want to carry an extra set of glasses. Photochromatic lenses are available in glass and plastic.

FRAMES Frames can be made from metal or plastic, and they can be rimless. There is an almost unlimited variety of shapes, colors, and sizes. The type and degree of refractive correction in the lens determine to some extent the type of frame most suitable. Some lenses are too thick to fit in metal rims, and some large-correction prescriptions are best suited to frames with small-area lenses.

Rimless frames are the least noticeable type, and they are lightweight because the nosepiece and temples are attached directly to the lenses, eliminating the weight of the rims. They tend not to be as sturdy as frames with rims, so they are not a good choice for children, or for people who frequently remove their glasses and put them on again. They are also not very suitable for lenses that correct a high degree of farsightedness, because such lenses are thin at the edges.

Metal frames are less noticeable than plastic, and they are lightweight. They are available in solid gold, gold-filled, anodized aluminum, nickel, silver, stainless steel, and titanium and titanium alloy. Until the late 1980s, when titanium-nickel alloy and titanium frames were introduced, metal frames were, in general, more fragile than plastic frames. Titanium frames, however, are very strong and lightweight. An alloy of titanium and nickel, called Flexon, is strong, lightweight, and returns to its original shape after being twisted or dented. It is not perfect for everyone, though, because some young

people are sensitive to its nickel content. Flexon frames are also relatively expensive.

Plastic frames are durable, can accommodate just about any lens prescription, and are available in a wide range of prices. They are also offered in a variety of plastics, including acrylic, epoxy, cellulose acetate, cellulose propionate, polyamide, and nylon, and in different colors, shapes, and levels of resistance to breakage. Epoxy frames are resilient and return to their original shape after being deformed, so they do not need to be adjusted as frequently as other types. Nylon frames are almost unbreakable. They revert to their original shape after extreme trauma and distortion; because of this property, though, they cannot be readjusted after they are manufactured.

FIT An individual should have the distance between the eyes (PD) measured, so that the optical centers of the lenses will be in front of the person's pupils. Bifocal heights also have to be measured with the chosen frame in place and adjusted on the person. Again, this is so that the lenses will be positioned correctly. If not positioned correctly, the individual may experience eyestrain, **headache**, or other problems.

Children may sometimes need a few days to adjust to a new prescription. However, problems should be reported, because the glasses may need to be rechecked.

Contact lenses

Over 32 million people in the United States wear these small lenses that fit on top of the cornea. They provide a field of view unobstructed by eyeglass frames. They do not fog up or get splattered, so it is possible to see well while walking in the rain. They are less noticeable than any style of eyeglass. On the other hand, they take time to get accustomed to; require more measurements for fitting; require many follow-up visits to an eye doctor; can lead to complications such as infections and corneal damage; and may not correct astigmatism as well as eyeglasses, especially if the astigmatism is severe.

Originally, hard contact lenses were made of a material called PMMA. The more common types of contact lenses are listed below:

- Rigid gas-permeable (RGP) daily-wear lenses are made of plastic that does not absorb water but allows oxygen to get from the atmosphere to the cornea. (This is important because the cornea has no blood supply and needs to get its oxygen from the atmosphere through the film of tears that moves beneath the lens.) These lenses must be removed and cleaned each night.
- Rigid gas-permeable (RGP) extended-wear lenses are made from plastic that also does not absorb water but is more permeable to oxygen than the plastic used for daily-wear lenses. They can be worn up to a week.
- Daily wear soft lenses are made of plastic that is permeable to oxygen and absorbs water; therefore, they are soft and flexible. These lenses must be removed and cleaned each night, and they do not correct all vision problems. Soft lenses are easier to get used to than rigid lenses but are more prone to tears and do not last as long.
- Extended-wear soft lenses are highly permeable to oxygen, are flexible by virtue of their ability to absorb water, and can usually be worn for up to one week. They do not correct all vision problems. There is more risk of infection with extended-wear lenses than with daily-wear lenses.
- Extended-wear disposable lenses are soft lenses worn continually for up to six days and then discarded, with no need for cleaning.
- Planned-replacement soft lenses are daily-wear lenses that are replaced on a regular schedule, which is usually every two weeks, monthly, or quarterly. They must also be cleaned.

Soft contact lenses come in a variety of materials. There are also different kinds of RGP and soft multifocal contact lenses available. Monovision, where one contact lens corrects for distance vision while the other corrects for near vision, may be an option for persons with presbyopia. Monovision, however, may affect depth perception and may not be appropriate for everyone. Contact lenses also come in a variety of tints. Soft contacts are available that can change the color of dark-colored eyes. Even though such lenses have no prescription, they must be fitted and checked to make sure that an eye infection does not occur. People should never wear someone else's contact lenses. Doing so can lead to infection or damage to the eye.

Risks

Young people allergic to certain plastics should not wear contact lenses or eyeglass frames or lenses manufactured from that type of plastic. People allergic to nickel should not wear Flexon frames. Children and teens at risk of being in accidents that might shatter glass lenses should wear plastic lenses, preferably polycarbonate. (Lenses made from polycarbonate, the same type of plastic used for the space shuttle windshield, are about 50 times stronger than other lens materials.) Also, young people whose work places them at risk of receiving electric shock should avoid metal frames.

KEY TERMS

Astigmatism—An eye condition in which the cornea doesn't focus light properly on the retina, resulting in a blurred image.

Cornea—The clear, dome-shaped outer covering of the eye that lies in front of the iris and pupil. The cornea lets light into the eye.

Index of refraction—A constant number for any material and any given color of light that is an indicator of the degree of bending of the light caused by that material.

Lens—The transparent, elastic, curved structure behind the iris (colored part of the eye) that helps focus light on the retina. Also refers to any device that bends light waves.

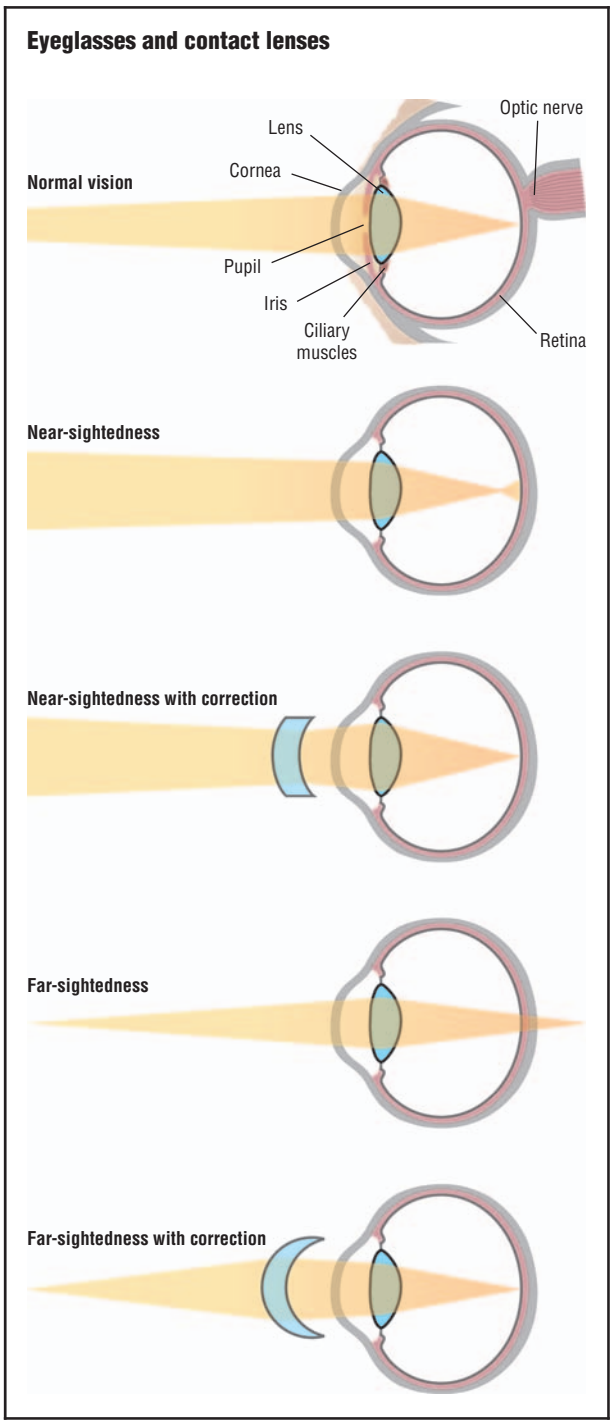
Permeable—A condition in which fluid or certain other substances are allowed to pass through.

Polycarbonate—A very strong type of plastic often used in safety glasses, sport glasses, and children's eyeglasses. Polycarbonate lenses have approximately 50 times the impact resistance of glass lenses.

Presbyopia—A condition affecting people over the age of 40 where the system of accommodation that allows the eyes to focus on near objects fails to work because of age-related hardening of the lens of the eye.

Retina—The inner, light-sensitive layer of the eye containing rods and cones. The retina transforms the image it receives into electrical signals that are sent to the brain via the optic nerve.

Ultraviolet (UV) radiation—A portion of the light spectrum with a wavelength just below that of visible light. UV radiation is damaging to DNA and can destroy microorganisms. It may be responsible for sunburns, skins cancers, and cataracts in humans. Two bands of the UV spectrum, UVA and UVB, are used to treat psoriasis and other skin diseases.



When the eyes function properly, the lens can focus images perfectly on the retina. However, nearsightedness causes the image to focus before it reaches the retina, and far-sightedness causes the image to focus past the retina. In each case, a corrective lens is used to adjust the focus of the image to bring a clear picture to retina, optic nerve, and eventually to the brain. *(Illustration by GGS Information Services.)*

People employed in certain occupations may be prohibited from wearing contact lenses or may be required to wear **safety** eyewear over the contact lenses. Some occupations, such as construction or auto repair, may require safety lenses and safety frames. Physicians and employers should be consulted for recommendations.

Contact lens wearers must be examined periodically by their eye doctors to make sure that the lenses fit prop-

erly and that there is no infection. Both infection and lenses that do not fit properly can damage the cornea. People can be allergic to certain solutions that are used to clean or lubricate lenses. For that reason, individuals should not randomly switch products unless they speak with their doctor. Contact lens wearers should seek immediate attention if they experience eye **pain**, a burning sensation, red eyes, intolerable sensitivity to light, cloudy vision, or an inability to keep the eyes open.

To avoid infection, it is important for contact lens wearers to exactly follow their instructions for lens insertion and removal, as well as cleaning. Soft contact lens wearers should never use tap water to rinse their lenses or to make-up solutions. All contact lens wearers should also always have a pair of glasses and a carrying case for their contacts with them, in case the contacts have to be removed due to eye irritation. Wearing contact lenses increases the risk of corneal damage and eye infections.

Normal results

Improved vision is the primary result of corrective lenses. The normal expectation is that people will achieve 20/20 vision while wearing corrective lenses. Contact lenses may contribute to improved cosmetic appearance for some users.

Parental concerns

Parents of young children requiring corrective lenses should be prepared for broken or lost glasses. Parents of adolescents who wear contact lenses should help their children to maintain a regular cleaning schedule. Glasses normally last one to two years. Growing children may require changes of prescription more frequently than people in other age groups.

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F

Fabry's disease *see* **Lipidoses**

Facial nerve palsy *see* **Bell's palsy**

Factor IX deficiency *see* **Hemophilia**

Factor VIII deficiency *see* **Hemophilia**

Failing a grade *see* **Retention in school**

Failure to thrive

Definition

Failure to thrive (FTT) is a term used to describe children whose physical growth over time is inadequate when compared to a standard growth chart.

Description

There is no universally accepted definition of failure to thrive, though it has been recognized as a medical condition since the early 1900s. It describes a condition rather than a specific disease. Children are considered as failing to thrive when their rate of growth does not meet the expected growth rate for a child their age. The difficulty lies in knowing what rate of growth is expected for any individual child, since many factors, including race and genetics, may influence growth.

Recognizing abnormal growth requires an understanding of normal infant growth. Infants normally lose up to 10 percent of their weight in the first few days of life. However, this weight should be regained within two weeks. The average full-term baby doubles its birth weight by six months and has tripled it by one year. Children with failure to thrive are often not meeting those milestones. If a baby continues to lose weight or does not gain weight as expected, he or she is probably not thriving.

Children who fail to thrive are either not receiving or have an inability to take in or retain adequate **nutrition** in order to gain weight and grow. If the condition progresses, the undernourished child may become irritable and/or apathetic and may not reach typical developmental markers such as sitting up, walking, and talking at the usual ages.

Demographics

The incidence of growth failure of American children is difficult to assess. Failure to thrive is believed to affect up to 5 percent of the population but is most common in the first six months of a child's life. It is commonly seen in babies born prematurely. Most diagnoses of failure to thrive are made in infants and toddlers in the first few years of life. An estimated 10 percent of children seen in primary care settings have symptoms of failure to thrive. The condition can appear in all socioeconomic groups, although it is seen more frequently in those families experiencing poverty. There is an increased incidence among children receiving Medicaid, those living in rural areas, and in children who are homeless.

Causes and symptoms

Failure to thrive may have several underlying causes. The causes of failure to thrive are typically differentiated into organic and non-organic. Organic causes are those caused by an underlying medical disorder. Inorganic causes are those caused by a caregiver's actions. However, these definitions are simplified, as both medical and behavioral causes often appear together.

Organic causes of failure to thrive may include:

- premature birth, especially if the fetus had intrauterine growth retardation
- maternal **smoking**, alcohol use, or illicit drugs during pregnancy
- mechanical problems present, resulting from a poor ability to suck or swallow, for example, presence of **cleft lip** and cleft palate
- unexplained poor appetites that are unrelated to mechanical problems or structural abnormalities, for example, breathing difficulties that can result from congestive heart failure (Any difficulty in breathing makes eating more difficult and can result in FTT. Inadequate intake also can result from metabolic abnormalities, excessive **vomiting** caused by obstruction of the gastrointestinal tract, or kidney dysfunction. In addition, gastroesophageal reflux causing regurgitation of formula or refusal of feeding.)
- poor absorption of food, inability of the body to use absorbed nutrients, or increased loss of nutrients

Some examples of non-organic causes of failure to thrive are:

- poor feeding skills on the part of the parent
- dysfunctional **family** interactions
- difficult parent-child interactions
- lack of social support
- lack of parenting preparation
- family dysfunction, such as abuse or divorce
- child neglect
- emotional deprivation

Studies show that only between 5 percent and 26 percent of FTT cases are due to a purely organic cause. Children in abusive or neglectful families are at higher risk of FTT, but these cases make up only a small proportion of the total. The most common cause of failure to thrive is **malnutrition**, either as part of an organic problem or simply because of an energy imbalance.

The following symptoms are possible indications of failure to thrive:

- delayed social and mental skills
- delayed development of secondary sexual traits in adolescents
- height, weight, and head circumference in an infant or young child not progressing as expected on growth charts
- edema (swelling)
- wasting
- enlarged liver
- rashes or changes in the skin
- changes in hair texture

When to call the doctor

Parents should notify their physician if their child does not seem to be developing at a normal pace. If parents notice a drop in weight or if the baby does not want to eat, the doctor should be notified. A major change in eating patterns also warrants contact.

Diagnosis

If a child fails to gain weight for three months in a row during the first year of life, physicians normally become concerned. The most important part of a physician's evaluation is taking a detailed history. Prenatal history is important, and the doctor will want to know if the pregnant mother smoked, consumed alcohol, used any medications, or had any illness during the pregnancy. The doctor will also want a dietary history, to determine if there have been any feeding problems. A history of how formula is mixed is important, because improperly prepared formula can result in failure to thrive. Parents will also be asked about whether the child had any illnesses, as some can cause a problem with the growth potential of children. A family and social history will also be done.

Doctors diagnose failure to thrive by plotting the child's weight, length, and head circumference on standard growth charts. Children who fall below a particular weight range for their age or who dip below two or more percentile curves on the chart over a short period of time will likely have a more thorough evaluation to find out if there is a problem. A complete blood count, various serum chemical and electrolyte tests, and a urinalysis may be helpful in discovering any underlying medical disorders. The doctor will want to determine if the child is receiving enough nourishment. To do this, the parents will be asked to record what the child eats each day, and a subsequent calorie count will be done. The doctor may also talk to the parents to help identify any home problems like financial difficulties, household stress, or neglect.

It is important to remember that some children will normally fall below the standards on growth charts. If children are full of energy, interacting normally with their parents, and show no signs of illness, then they are

probably not failing to thrive and are just smaller children.

Once the diagnosis of failure to thrive has been made, the physician will attempt to determine if it is from an organic or non-organic cause.

KEY TERMS

Inorganic causes—Cases of failure to thrive brought on by a caregiver's actions.

Organic causes—Underlying medical or physical disorders causing failure to thrive.

Treatment

Because there are numerous factors that may contribute to a failure to thrive diagnosis, children diagnosed with the disorder sometimes have an entire medical team working on the case. If there is an underlying physical cause, correcting that problem may reverse the condition. The doctor will recommend high-calorie foods and place the child on a high-density formula like Pediasure. More severe cases may involve tube feedings, which can take place at home. A child with extreme failure to thrive may need **hospitalization**, during which he or she can be fed and monitored continuously. This will give the treatment team an opportunity to also observe the caregiver's interactions with the child.

The duration of treatment will vary from child to child. Weight gain takes time, so several months may go by before a child returns to his normal weight range. Children requiring hospitalization usually stay for approximately two weeks or more to get them out of danger, but many months can pass before the symptoms of malnutrition disappear.

Nutritional concerns

The long-term goal for every child with FTT is to provide adequate energy intake for growth. Therefore, even if no causative factor is uncovered for a child with FTT, aggressive dietary management is the key to successful treatment. Proper feeding can be achieved through infant formulas that are adjusted to meet the child's specific nutrient needs. Infants may be given concentrated formulas, assuming their kidney function is normal. In cases of kidney disorders, increasing the fat content of the formula may be useful as a way of delivering additional calories. Older children with FTT may benefit from adding cheese, sour cream, butter, margarine, or peanut butter to meals. Also, high-calorie shakes can be used to supplement meals. Multivitamin and mineral supplements, including iron and zinc, usually are recommended to all undernourished children. Tube feeding is usually not indicated except for severe cases of malnutrition.

Prognosis

Whether FTT results from organic or non-organic reasons, children with this condition require aggressive calorie supplementation. Some cases may lead to significant developmental delays in children. The cognitive outcome of children who have had FTT is not clear, and this may lead to emotional and behavioral problems later. However, carefully looking for the causes of failure to thrive and implementing calorie supplementation is important for obtaining a positive outcome in these children.

Prevention

Initial failure to thrive caused by physical defects cannot be prevented but can often be corrected before they become a danger to the child. Maternal education as well as emotional and economic support systems may help to prevent failure to thrive in those cases where is no physical deformity.

Parental concerns

Parents who note any of the symptoms of failure to thrive should report them to their child's physician so that treatment can begin.

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Fallot's tetralogy *see* **Tetralogy of Fallot**

Familial Mediterranean fever

Definition

Familial Mediterranean fever (FMF) is an inherited disorder characterized by an inflammatory response recurring with attacks of fever accompanied by intense **pain** in the abdomen, chest, or joints. Attacks usually last 12–72 hours and can occasionally involve a skin rash. Kidney disease is a serious complication that may develop if the disorder is not treated. FMF is most prevalent in people of Armenian, Sephardic-Jewish, Arabic, and Turkish ancestry. The disorder takes its name from the fact that these ethnic groups live in countries along or near the eastern coast of the Mediterranean Sea.

FMF is sometimes grouped together with other periodic fevers in a category called autoinflammatory disorders. The term was invented to describe illnesses caused by genetic defects in the human immune response. Other disorders in this group are TNF-receptor associated periodic syndrome (TRAPS), hyper-IgD syndrome (HIDS), and familial Hibernian fever.

Description

FMF has been described as a disorder of "inappropriate" inflammation or an autoinflammatory syndrome. That is, an event that causes a mild or unnoticeable inflammation in most people might cause a severe inflammatory response in someone with FMF. Certain areas of the body are at risk for FMF-related symptoms. A serosa is a serous (fluid-producing) membrane that can be found inside the abdominal cavity (peritoneum), around the lungs (pleura), around the heart (pericardium), and inside the joints (synovium). The symptoms of FMF are due to inflammation of one or more of the serosal membranes (serositis). Thus, FMF is sometimes called recurrent polyserositis. Other names for the disorder are periodic peritonitis, periodic fever, periodic disease, familial paroxysmal polyserositis, periodic amyloid syndrome, periodic peritonitis syndrome, Reimann periodic disease, Reimann syndrome, Siegel-Cattan-Mamou syndrome, and Armenian syndrome.

During an attack of FMF, large numbers of neutrophils, a type of white blood cell, move into the affected areas of the body, where they cause painful inflammation and fever. These episodes may be accompanied by a skin rash (erythema) or joint pain. In a few cases, chronic arthritis is a problem. Amyloidosis is a potentially serious condition in which proteins called amyloids are mistakenly produced and deposited in organs and tissues throughout the body. Left untreated, amyloidosis often leads to kidney failure, which is the major long-term health risk in FMF.

In most cases, patients diagnosed with FMF first notice the attacks of fever and pain in childhood or **adolescence**. The intervals between these episodes may extend for days or months and are unpredictable. People with FMF typically lead normal lives during these pain-free intervals. It is not entirely clear what brings on an attack, although people with FMF often report mild physical trauma, physical exertion, or emotional stress just prior to the onset of symptoms. The mainstay of treatment for FMF is an oral medication called colchicine, which is highly effective for the fever and pain that accompany the disorder, as well as for amyloidosis and the kidney disease that can result from it.

Demographics

Estimates of the incidence of FMF in specific eastern Mediterranean populations range from one in 2,000 Arabs to one in 250 Sephardic Jews, one in 500 Armenians, and one in 1,000 Turks. Specific mutations in the MEFV gene are more common in certain ethnic groups and may cause a somewhat different course of the disease. Researchers think that a few mutations in the MEFV gene likely became common in a small population in the eastern Mediterranean several thousand years ago. The mutation was transmitted to later generations because people who carried a single copy of the mutated gene had a modified (but not abnormal) inflammatory response that may have protected them against some infectious agent at that time. Those who carried a single "beneficial" mutation in the MEFV gene were more likely to survive and reproduce, which may explain

the high carrier frequency (up to one in five) in some populations. A better understanding and recognition of the symptoms of FMF in the late 1990s and early 2000s has resulted in more reports of the condition in other ethnic groups such as Ashkenazic Jews, Italians, Armenian-Americans, and Japanese. About 50 percent of patients diagnosed with FMF, however, have no **family** history of the disease.

With regard to sex, FMF is more common in men than in women, with a gender ratio of two men for every one woman. In terms of age groups, FMF is more common in younger people. One researcher states that 50–60 percent of patients are younger than 10 years, 80–95 percent are below the age of 20, with the remainder between 20 and 40 years of age. FMF is rare in people older than 40.

Causes and symptoms

Causes

FMF is a genetic condition inherited in an autosomal recessive fashion. Mutations in the MEFV gene (short for Mediterranean fever) on chromosome number 16 are the underlying cause of FMF. Autosomal recessive inheritance means that a person with FMF has mutations in both copies of the MEFV gene. All genes come in pairs, and one copy of each pair is inherited from each parent. If neither parent of a child with FMF has the condition, it means they carry one mutated copy of the MEFV gene, but also one normal copy, which is enough to protect them from disease. If both parents carry the same autosomal recessive gene, there is a one in four chance with each pregnancy that the child will inherit both recessive genes and develop FMF.

The MEFV gene carries the instructions for production of a protein called pyrin, named for pyrexia, a medical term for fever. The research group in France that codiscovered the protein in 1997 named it marenostrin, after *mare nostra*, the Latin words that the ancient Romans used for the Mediterranean Sea. Research has shown that pyrin has some function in controlling neutrophils, which are the white blood cells that move into an area of the body affected by stress or trauma. In a person with a normal immune system, some inflammation may follow stress or trauma, but the pyrin protein is responsible for shutting down the response of neutrophils once they are no longer needed. An abnormal pyrin protein associated with FMF may be partly functional but unstable. In some instances, the abnormal pyrin itself seems to be ''stressed'', and loses its ability to regulate the inflammatory response to trauma. Without such regulation, a normal inflammatory response spirals out of control. Exactly what causes pyrin in FMF to lose its ability to control neutrophils in some situations is not fully understood as of 2004.

Symptoms

The recurrent acute attacks of FMF typically begin in childhood or adolescence. These acute episodes of fever and painful inflammation usually last 12–72 hours. About 90 percent of people with FMF have their first attack by age 20. The group of symptoms that characterizes FMF includes the following:

- Fever: An FMF attack is nearly always accompanied by a fever, but it may not be noticed in every case. Fevers are typically 100 to 104°F (38–40°C). Some people experience chills prior to the onset of fever.
- Abdominal pain: Nearly all people with FMF experience abdominal pain at one point or another, and for most it is the most common complaint. The pain can range from mild to severe and can be diffuse or localized. It can mimic **appendicitis**, and many people with undiagnosed FMF have had appendectomies or exploratory surgery of the abdomen only to have the fever and abdominal pain return.
- Chest pain (pleuritis): Pleuritis, also called pleurisy, occurs in up to half of the affected individuals in certain ethnic groups. The pain is usually felt on one side of the chest. Pericarditis, an inflammation of the membrane surrounding the heart, would also be felt as chest pain.
- Joint pain: About 50 percent of people with FMF experience joint pain during attacks. The pain is usually confined to one joint at a time, and often involves the hip, knee, or ankle. For some people, however, the recurrent joint pain eventually becomes chronic arthritis.
- Muscle pain (myalgia): Up to 20 percent of individuals with FMF report muscle pain. These episodes typically last less than two days and tend to occur in the evening or after physical exertion. Rare cases of muscle pain and fever lasting as long as one month have been reported.
- Skin rash. A rash described as an erythema (skin reddening) resembling erysipelas accompanies FMF attacks in a minority of people. The rash typically occurs on the front of the lower leg or top of the foot, and appears as a red, warm, swollen area about 4–6 inches (10–15 cm) in diameter.
- Amyloidosis: FMF is associated with high levels in the blood of a protein called serum amyloid A (SAA). Over time, excess SAA tends to be deposited in tissues and organs throughout the body. The presence and

deposition of excess SAA is known as amyloidosis. Amyloidosis may affect the gastrointestinal tract, liver, spleen, heart, and (in males) testes, but its effects on the kidneys are of greatest concern. The frequency of amyloidosis varies among the different ethnic groups, and its overall incidence is difficult to determine because of the use of colchicine to avert the problem. Left untreated, however, those individuals who do develop amyloidosis of the kidneys may require a kidney transplant or may even die of renal failure. The frequency and severity of a person's attacks of fever and serositis seem to have no relation to the risk of developing amyloidosis. In fact, a few people with FMF have been described who have had amyloidosis but apparently no other FMF-related symptoms.

- Other symptoms: A small percentage of boys with FMF develop painful inflammation around the testes, while girls may experience episodes of inflammation in the pelvis. In other patients, headaches are a common occurrence during attacks. Lastly, certain types of vasculitis (inflammation of the blood vessels) seem to be more common in people diagnosed with FMF.

When to call the doctor

Parents should consult a doctor for their child if they have Mediterranean ancestry and their child develops recurrent attacks of fever and pain consistent with the symptoms of FMF as described above.

In general, symptoms involving one or more of the following broad groups should lead to suspicion of FMF. Unexplained recurrent fevers, polyserositis, skin rash, and/or joint pain; abnormal blood studies (see below); and kidney or other disease associated with amyloidosis. A family history of FMF or its symptoms would obviously be an important clue, but the recessive nature of FMF means there usually is no family history. The diagnosis may be confirmed when a person with unexplained fever and pain responds to treatment with colchicine, since colchicine is not known to have a beneficial effect on any other condition similar to FMF. Abnormal results on a blood test typically include leukocytosis (elevated number of neutrophils in the blood); an increased erythrocyte sedimentation rate (the rate at which red blood cells form a sediment in a blood sample); and increased levels of proteins associated with inflammation (called acute phase reactants) such as SAA.

Diagnosis

The diagnosis of FMF is often delayed because the symptoms that define the condition are common to many other disorders. Fevers occur for many reasons, and nonspecific pains in the abdomen, chest, and joints are also frequent ailments. Several infections can result in symptoms similar to FMF (Mallaret **meningitis**, for instance), and many people with FMF undergo exploratory abdominal surgery and ineffective treatments before they are finally diagnosed. Membership in a less commonly affected ethnic group may also delay or hinder the correct diagnosis. In many cases the doctor is able to diagnose the disorder only by a slow process of eliminating other diagnostic possibilities. He or she may order **x rays** or other imaging studies in order to rule out certain types of arthritis.

Direct analysis of the MEFV gene for FMF mutations is the only method to be certain of the diagnosis. While it was as of 2004 not yet possible to detect all MEFV gene mutations that might cause FMF, successful cloning of the MEFV gene has led to a rapid test that can identify the most common mutations of the gene. Thus, if DNA analysis is negative, clinical methods must be relied upon. If both members of a couple were proven to be FMF carriers through genetic testing, highly accurate prenatal diagnosis would be available in any subsequent pregnancy.

Similar syndromes of periodic fever and inflammation include familial Hibernian fever and hyperimmunoglobulinemia D syndrome, but both are much less common than FMF.

Treatment

Colchicine is an anti-inflammatory chemical compound that can be used as a medication and is frequently prescribed for gout. In the late twentieth century, colchicine was discovered to also be effective in reducing the frequency and severity of attacks in FMF. Treatment for FMF in the early 2000s consists of taking colchicine daily. Studies have shown that about 75 percent of FMF patients achieve complete remission of their symptoms, and about 95 percent show marked improvement when taking colchicine. Compliance with taking colchicine every day may be hampered by its side effects, which include **diarrhea**, **nausea**, abdominal bloating, and gas. Colchicine is also effective in preventing, delaying, or reversing kidney disease associated with amyloidosis.

Other medications may be used as needed to treat the pain and fever associated with FMF attacks. The most common drugs used are narcotics for severe pain and **nonsteroidal anti-inflammatory drugs** (NSAIDs) for arthritis and muscle pain. Dialysis and/or a kidney transplant might become necessary in someone with advanced kidney disease.

KEY TERMS

Acute phase reactants—Blood proteins whose concentrations increase or decrease in reaction to the inflammation process.

Amyloid—A waxy, translucent, starch-like protein that is deposited in tissues during the course of certain chronic diseases such as rheumatoid arthritis and Alzheimer's disease.

Amyloidosis—The accumulation of amyloid deposits in various organs and tissues in the body so that normal functioning is compromised. Primary amyloidosis usually occurs as a complication of multiple myeloma. Secondary amyloidosis occurs in patients suffering from chronic infections or inflammatory diseases such as tuberculosis, rheumatoid arthritis, and Crohn's disease.

Colchicine—A drug used to treat painful flare-ups of gout. It is also effective in reducing the frequency and severity of attacks in familial Mediterranean fever.

Erythema—A diffuse red and inflamed area of the skin.

Leukocyte—A white blood cell that defends the body against invading viruses, bacteria, and cancer cells. There are five types of leukocytes–neutrophils, basophils, eosinophils, lymphocytes, and monocytes.

Leukocytosis—An increased level of white cells in the blood. Leukocytosis is a common reaction to infections.

Neutrophil—The primary type of white blood cell involved in inflammation. Neutrophils are a type of granulocyte, also known as a polymorphonuclear leukocyte. They increase in response to bacterial infection and remove and kill bacteria by phagocytosis.

Pericarditis—Inflammation of the pericardium, the sac that surrounds the heart and the roots of the great blood vessels.

Peritonitis—Inflammation of the peritoneum. It is most often due to bacterial infection, but can also be caused by a chemical irritant (such as spillage of acid from the stomach or bile from the gall bladder).

Pleuritis—Inflammation of the pleura, the membrane surrounding the lungs. Also called pleurisy.

Pyrexia—A medical term meaning fever.

Pyrin—A protein that regulates the body's inflammatory response to stress or trauma. The MEFV gene involved in FMF produces an unstable form of pyrin that fails to adequately control the inflammatory response.

Serositis—Inflammation of a serosal membrane (any membrane that lines a body cavity that does not open to the outside of the body). Polyserositis refers to the inflammation of two or more serosal membranes.

Synovitis—Inflammation of the synovial membrane, the membrane that lines the inside of the articular capsule of a joint.

Alternative treatment

Some researchers have reported on herbal compounds that appear to be useful in managing patients with FMF. One Japanese case report concerned a patient who was helped by Kampo formulations, which are the traditional herbal medicines of Japan. A larger study of 24 Armenian patients diagnosed with FMF reported that the 14 patients who were given ImmunoGuard, a herbal preparation containing licorice root and schisandra along with several other herbs, had fewer and milder attacks of FMF than the 10 patients who were given a placebo.

Prognosis

Children who are diagnosed early enough and take colchicine consistently have an excellent prognosis. Most will have very few, if any, attacks of fever and polyserositis and will likely not develop serious complications of amyloidosis. Future research should provide doctors with a better understanding of the inflammation process, focusing on how neutrophils are genetically regulated. That information could then be used to develop treatments for FMF with fewer side effects and might also assist in developing therapies for other autoinflammatory diseases.

With regard to long-term effects of FMF, about 5 percent of patients will develop severe arthritis in adult life. Girls with FMF are likely to have fertility problems as adults; about 30 percent will be unable to have children at all, and those who can conceive have a 20 to 30 percent chance of miscarriage.

Prevention

Given the genetic nature of FMF, there is as of 2004 no cure for the disorder. Any couple that has a child

diagnosed with FMF or anyone with a family history of the condition (especially those in high-risk ethnic groups) should be offered genetic counseling to obtain the most up-to-date information on FMF and testing options.

Parental concerns

Parental concerns about a child with FMF depend to some extent on the frequency and severity of attacks, as the frequency can range from two episodes per week to one per year. Families whose children have relatively infrequent attacks will be much less affected by the disorder than those whose plans are frequently disrupted by a child's acute attack. In most cases, however, children diagnosed with FMF have excellent health between attacks, can keep up with their schoolwork, participate in **sports**, and enjoy a normal social life. They do not require special diets, educational programs, isolation from other children, or other modifications of the family's routine. One concern that parents may wish to discuss with the doctor, however, is the narcotic medications that are often prescribed to ease the pain that accompanies acute attacks of FMF. Some of these drugs are potentially addictive, and their use should be carefully supervised.

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Family

Definition

A family is a group of two people or more related by marriage, blood relation, or **adoption** and who live together. The immediate family traditionally consists of parents and their offspring.

Description

No other factor influences children as deeply as their families. As a social unit with genetic, emotional, and legal dimensions, the family can foster the child's growth, development, health, and well-being. The family can provide the child with affection, a sense of belonging, and validation. Every area of a child's life is affected by the family.

Family functions

The family has basic functions. In order for the family to meet a child's psychological needs, its members must be nurturing, convey mutual respect, provide for intimacy, and engage in **bonding** and attachment. The family also socializes the child, guiding the child to be members of the society beyond the family. The family conveys religious and cultural beliefs and traditions to the next generation. The family is the child's source of economic resources, which meet the child's various physical needs for food, shelter, and clothing. Then, too, the family sees to it that the child receives health and dental care.

Demographics

Family structure is dynamic. In 1970, traditional nuclear families made up 40 percent of all households, but only 26 percent of all households in 1991. In addition, roles have changed within the nuclear family. The role of provider, once assigned mainly to the father, gradually came in the early 2000s to be shared by both parents, and as of 2004 many fathers are more active in parenting their children than their fathers were in parenting them.

Toward the end of the twentieth century, increasing numbers of families did not fit the nuclear profile. Some families have only one parent; others are combinations based on second marriages; still others are comprised of unmarried couples living with or without children.

The number of **single-parent families** increased from 12.9 percent of all families in 1970 to 29 percent in 1991 and 28 percent (20 million) in 1997. The increases are mostly the result of the increase in the **divorce** rate and the increase in births to single mothers. Many women are single parents and heads of households, and many of these live in poverty.

Common problems

Poverty is the single most powerful risk for families and children and affects families in many ways. Poverty exerts its greatest impact during children's **preschool** years, the age group in which children are most likely to live in poverty. Poor families are less likely or able to provide educational and cultural experiences for their children.

Parents' economic status (education, occupation, and income) controls the parent's ability provide adequate housing, a safe environment, and responsible child care while the parents work. Availability of and quality of social support influence family life and well-being as individuals cope with raising children in poverty.

Unemployment by either or both parents causes financial hardships and social and emotional strain, which can disrupt family life. The quality of the parent's work and the satisfaction the parent gets from it affects the parents and, in turn, their effectiveness in parenting.

Child abuse and neglect is a destructive force in families and results in children's **anxiety**, depression, lowered **self-esteem**, and a decline in school performance. Similarly, children who witness domestic violence suffer some of these same consequences.

KEY TERMS

Coping—In psychology, a term that refers to a person's patterns of response to stress.

Culture—The system of communal beliefs, values behaviors, customs, and materials that members of a society use to understand their world and each other, and which are passed down among suceeding generation.

Custodial parent—A parent who has legal custody of their child or children.

Extended family—Traditionally defined as the biological relatives of a nuclear family (the parents, sisters, and brothers of both members of a married couple); sometimes used to refer to the people living in the household as partners and parents with children.

Noncustodial parent—The parent who does not have legal custody of the child and does not live in the same home with the child. The noncustodial parent has financial responsibility for the child and visitation rights.

Stress—A physical and psychological response that results from being exposed to a demand or pressure.

Parental concerns

Parents are troubled by children who are out of control and have problem behaviors such as **running away**, **truancy**, school failings or suspensions, and delinquency. Youths who are habitually truant may need school counseling. Truancy specialists provide topic-focused workshops and referrals to family counseling; court intervention is sometimes necessary.

The frequency of divorce and remarriage produces **stepfamilies** with their own difficulties and challenges. The new stepfamily members may have no shared family history or common lifestyle, and members may have different beliefs. In addition, children may feel torn between the custodial parent, with whom they live, and the noncustodial parent, with whom they visit.

Economic stress affects the whole family. When financial problems occur, the family may be forced to move for employment. Families have lifestyle commitments and ties to their community. When these ties break-up, children, especially adolescents, are likely to experience a loss of hope. Parental career disappointments can cause anxiety and self-

A traditional family of father, mother, daughter, and son. *(© Ariel Skelley/Corbis.)*

doubt in children. Family economic stress may cause parental substance abuse, which can lead to school distraction and declining academic performance in young people.

Finally, children may feel isolated from parents and friends. The most stressful time for a family can be the period preceding a possible foreclosure or business failure. Parents may make desperate attempts to save their source of income. They may also be trying to keep their loss hidden from the rest of the community. Such behaviors can isolate children from parents who are too busy to notice and from neighbors who are not even aware of their trouble.

The long-term effects of continued family stress can cause physical and psychological problems for children. If problems occur often and if several problems appear at the same time, serious attention should be given the child.

When to call the doctor

Parents should call their pediatrician if the child shows unhealthy physical or emotional symptoms that may be in response to family problems or transitions. Physical problems may include weight gain or weight loss, or unexplained stomachaches or headaches. Emotional problems may cause a decline in academic performance, breaking curfews, or getting into trouble in school or with the law.

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Family therapy

Definition

Family therapy is a type of psychotherapy that involves all members of a nuclear family or stepfamily and, in some cases, members of the extended family (e.g., grandparents). A therapist or team of therapists conducts multiple sessions to help families deal with important issues that may interfere with the functioning of the family and the home environment.

Purpose

The goal of family therapy is to help family members improve communication, solve family problems, understand and handle special family situations (for example, death, serious physical or mental illness, or child and adolescent issues), and create a better functioning home environment. For families with one member who has a serious physical or mental illness, family therapy can educate families about the illness and work out problems associated with care of the family member. For children and adolescents, family therapy most often is used when the child or adolescent has a personality, **anxiety**, or mood disorder that impairs their family and social functioning, and when a stepfamily is formed or begins having difficulties adjusting to the new family life. Families with members from a mixture of racial, cultural, and religious backgrounds, as well as families made up of same-sex couples who are raising children, may also benefit from family therapy.

Description

Family therapy is generally conducted by a therapist or team of therapists who are trained and experienced in family and group therapy techniques. Therapists may be psychologists, psychiatrists, social workers, or counselors. Family therapy involves multiple therapy sessions, usually lasting at least one hour each, conducted at regular intervals (for example, once weekly) for several months. Typically, family therapy is initiated to address a specific problem, such as an adolescent with a psychological disorder or adjustment to a death in the family. However, frequently, therapy sessions reveal additional problems in the family, such as communication issues. In a therapy session, therapists seek to analyze the process of family interaction and communication as a whole and do not take sides with specific family members. Therapists who work as a team can model new behaviors for the family through their interactions with each other during a session.

Family therapy is based on family systems theory, in which the family is viewed as a living organism rather than just the sum of its individual members. Family therapy uses systems theory to evaluate family members in terms of their position or role within the system as a whole. Problems are treated by changing the way the system works rather than trying to fix a specific member. Family systems theory is based on several major concepts.

Concepts in family therapy

THE IDENTIFIED PATIENT The identified patient (IP) is the family member with the symptom that has brought the family into treatment. Children and adolescents are frequently the IP in family therapy. The concept of the IP is used by family therapists to keep the family from scapegoating the IP or using him or her as a way of avoiding problems in the rest of the system.

HOMEOSTASIS (BALANCE) Homeostasis means that the family system seeks to maintain its customary organization and functioning over time, and it tends to resist change. The family therapist can use the concept of homeostasis to explain why a certain family symptom has surfaced at a given time, why a specific member has become the IP, and what is likely to happen when the family begins to change.

THE EXTENDED FAMILY FIELD The extended family field includes the immediate family and the network of grandparents and other relatives of the family. This concept is used to explain the intergenerational transmission of attitudes, problems, behaviors, and other issues.

Children and adolescents often benefit from family therapy that includes the extended family.

DIFFERENTIATION Differentiation refers to the ability of each family member to maintain his or her own sense of self, while remaining emotionally connected to the family. One mark of a healthy family is its capacity to allow members to differentiate, while family members still feel that they are members in good standing of the family.

TRIANGULAR RELATIONSHIPS Family systems theory maintains that emotional relationships in families are usually triangular. Whenever two members in the family system have problems with each other, they will "triangle in" a third member as a way of stabilizing their own relationship. The triangles in a family system usually interlock in a way that maintains family homeostasis. Common family triangles include a child and his or her parents; two children and one parent; a parent, a child, and a grandparent; three siblings; or, husband, wife, and an in-law.

In the early 2000s, a new systems theory, multisystemic therapy (MST), has been applied to family therapy and is practiced most often in a home-based setting for families of children and adolescents with serious emotional disturbances. MST is frequently referred to as a "family-ecological systems approach" because it views the family's ecology, consisting of the various systems with which the family and child interact (for example, home, school, and community). Several clinical studies have shown that MST has improved family relations, decreased adolescent psychiatric symptoms and substance use, increased school attendance, and decreased re-arrest rates for adolescents in trouble with the law. In addition, MST can reduce out-of-home placement of disturbed adolescents.

Preparation

In some instances the family may have been referred to a specialist in family therapy by their pediatrician or other primary care provider. It is estimated that as many as 50 percent of office visits to pediatricians have to do with developmental problems in children that are affecting their families. Some family doctors use symptom checklists or psychological screeners to assess a family's need for therapy. For children and adolescents with a diagnosed psychological disorder, family therapy may be added to individual therapy if family issues are identified as contributing factors during individual therapy.

Family therapists may be either psychiatrists, clinical psychologists, or other professionals certified by a specialty board in marriage and family therapy. They usually evaluate a family for treatment by scheduling a series of interviews with the members of the immediate family, including young children, and significant or symptomatic members of the extended family. This process allows the therapist(s) to find out how each member of the family sees the problem, as well as to form first impressions of the family's functioning. Family therapists typically look for the level and types of emotions expressed, patterns of dominance and submission, the roles played by family members, communication styles, and the locations of emotional triangles. They also note whether these patterns are rigid or relatively flexible.

Preparation also usually includes drawing a genogram, which is a diagram that depicts significant persons and events in the family's history. Genograms include annotations about the medical history and major personality traits of each member. Genograms help uncover intergenerational patterns of behavior, marriage choices, family alliances and conflicts, the existence of family secrets, and other information that sheds light on the family's present situation.

Precautions

Individual therapy for one or more family members may be recommended to avoid volatile interaction during a family therapy session. Some families are not considered suitable candidates for family therapy. They include:

- families in which one, or both, of the parents is psychotic or has been diagnosed with antisocial or paranoid personality disorder
- families whose cultural or religious values are opposed to, or suspicious of, psychotherapy
- families with members who cannot participate in treatment sessions because of physical illness or similar limitations
- families with members with very rigid personality structures (Here, members might be at risk for an emotional or psychological crisis.)
- families whose members cannot or will not be able to meet regularly for treatment

Risks

The chief risk in family therapy is the possible unsettling of rigid personality defenses in individuals or relationships that had been fragile before the beginning of therapy. Intensive family therapy may also be difficult for family members with diagnosed psychological disorders. Family therapy may be especially difficult and stressful for children and adolescents who may not fully

KEY TERMS

Blended family—A family formed by the remarriage of a divorced or widowed parent. It includes the new husband and wife, plus some or all of their children from previous marriages.

Differentiation—The ability to retain one's identity within a family system while maintaining emotional connections with the other members.

Extended family field—A person's family of origin plus grandparents, in-laws, and other relatives.

Family systems theory—An approach to treatment that emphasizes the interdependency of family members rather than focusing on individuals in isolation from the family. This theory underlies the most influential forms of contemporary family therapy.

Genogram—A family tree diagram that represents the names, birth order, sex, and relationships of the members of a family. Therapists use genograms to detect recurrent patterns in the family history and to help the family members understand their problem(s).

Homeostasis—The balanced internal environment of the body and the automatic tendency of the body to maintain this internal "steady state." Also refers to the tendency of a family system to maintain internal stability and to resist change.

Identified patient (IP)—The family member in whom the family's symptom has emerged or is most obvious.

Nuclear family—The basic family unit, consisting of a father, a mother, and their biological children.

Stepfamily—A family formed by the marriage or long-term cohabitation of two individuals, where one or both have at least one child from a previous relationship living part-time or full-time in the household. The individual who is not the biological parent of the child or children is referred to as the stepparent.

Triangling—A process in which two family members lower the tension level between them by drawing in a third member.

understand interactions that occur during family therapy. Adding individual therapy to family therapy for children and adolescents with the same therapist (if appropriate) or a therapist who is aware of the family therapy can be helpful.

Normal results

Normal results vary, but in good circumstances, they include greater insight, increased differentiation of individual family members, improved communication within the family, loosening of previously automatic behavior patterns, and resolution of the problem that led the family to seek treatment.

Parental concerns

Stepfamilies, which are increasing in prevalence, are excellent candidates for family therapy. Children and adolescents in stepfamilies often have difficulties adjusting, and participating in family therapy can be beneficial. Stepfamilies, increasingly referred to as "blended families," experience unique pressures within each new family unit. Stepfamily researchers, family therapists, and the Stepfamily Association of America (SAA) view the term as inaccurate because it seems to suggest that members of a stepfamily blend into an entirely new family unit, losing their individuality and attachment to other outside family members. Because other family types (biological, single-parent, foster, adoptive) are defined by the parent-child relationship, the SAA believes that the term "stepfamily" more accurately reflects that relationship and is consistent with other family definitions. Viewing the stepfamily as a blended family can lead to unrealistic expectations, confused and conflicted children, difficult adjustment, and in many cases, failure of the marriage and family. Family therapy can help family members deal with these issues.

Children and adolescents and, in some cases even the parents, may be reluctant to participate in family therapy. Home-based family therapy has in the early 2000s become available as an option for families with severely disturbed adolescents and family members reluctant to see a therapist. In home-based therapy, a therapist or team of therapists comes directly to the family's home and conducts therapy sessions there.

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Jennifer E. Sisk, M.A.

FAS *see* **Fetal alcohol syndrome**

Father-child relationships *see* **Parent-child relationships**

Fear

Definition

Fear is an intense aversion to or apprehension of a person, place, activity, event, or object that causes emotional distress and often avoidance behavior. Fears are common in childhood.

Description

More than 50 percent of children experience normal **phobias**, which is the fear of a specific object, or more general worries, called anxieties, before they are 18 years old. For adults it may be helpful to distinguish between rational fears, such as fear of snakes or guns, which are survival mechanisms and serve to protect a person from danger; and irrational fears, or phobias, which cannot be traced to any reasonable cause.

Most children have some fears. Fears are normal, and can be a good thing. For example, children need to know they should not run into a street. They need to know not to **play** with knives. A little fear is good, but too much fear is a problem. So is too little fear. A child with too much fear may not want to leave the house. A child with not enough fear may get into a stranger's car. Children's personalities also will influence their fears. One child may be scared of more things than another child. Some children are braver, while others are more shy and fearful.

Many childhood fears fall somewhere between the rational and irrational, occurring in phases as the child or adolescent is exposed to new experiences and as both cognitive reasoning and the capacity for imagination develop. Whether a child's fear is considered normal generally depends on his or her age, background, and most importantly on how much it interferes with his or her normal daily activities. Fear of water may be considered normal in a child who has never learned how to swim, but it might be considered abnormal in the adolescent son of a coastal fisherman.

The most significant factors in overcoming fear are identifying the fear, developing a sense of control over the feared environment, and envisioning alternatives to the feared negative outcomes. Forcing children to perform activities they are afraid to do destroys, rather than builds, autonomy and self-confidence. If a child refuses to do something or explicitly voices fear, those feelings should be taken seriously and explored through questions and discussion. Parents can ask the child or adolescent what change can be made to accommodate the fear in order to make him or her feel more in control.

Some research suggests that reading scary picture books functions as a courage-building tool for children and helps them face their fears in a controlled environment; they are free to turn the page or to remind themselves that the monster is not real. Horror stories or movies may serve the same purpose for teens but not for children who cannot exercise the same level of choice by leaving the theater and should not be exposed to disturbing movies.

Infancy

Babies fear falling, being dropped, and loud noises. A fear of strangers is also common in infants starting at the age of seven to nine months and lasting until about 18 months, when it begins to decrease. Fear symptoms in infants are primarily crying, stiffening, and sometimes shaking.

Toddlerhood

Fears among toddlers include strangers, animals, bugs, storms, sirens, large objects, dark colors, darkness, people with masks, monsters, and "bad" people, such as burglars. Children at this age also commonly fear being separated from their parents. Fear symptoms in toddlers include crying and avoidance of the feared person or object.

Preschool

Preschoolers fear being separated from parents, being left alone or sleeping alone, and imaginary figures, such as ghosts, monsters, and supernatural beings. Symptoms may be physical, such as a stomachache or **headache**.

School age

In younger school-age children, fears include **separation anxiety**; death; violence, such as in war or murder; kidnapping and physical injury; natural disasters such as floods, earthquakes, and tornados; and **anxiety** about academic achievement and other forms of school performance. Children at age seven often have a fear of not being liked while children ages eight and nine may worry about personal inabilities.

In older adolescents, common fears include anxiety about school achievement, social rejection and related worries, and sexual anxieties, including dating and **sexually transmitted diseases**, especially human **immunodeficiency** virus (HIV).

Symptoms in adolescents and teens include anger, avoidance, and denial of the fear, and panic reactions, such as sweating, trembling, fast heartbeat, and rapid breathing.

Nearly all fears have a scientific name, such as triskaidekaphobia, the fear of the number 13. In the classic Christmas television special, "A Charlie Brown Christmas," Charlie Brown had pantophobia, the fear of everything. Other common fears include:

- ailurophobia (fear of cats)
- didaskaleinophobia (fear of going to school)
- entomophobia (fear of insects)
- glossophobia (fear of speaking)
- myctophobia (fear of darkness)
- ophidiaphobia (fear of snakes)
- xenophobia (fear of strangers or foreigners)
- zoophobia (fear of animals)

Common problems

Research shows that most children report having several fears at any given age. Some research shows that 90 percent of children ages two to 14 have at least one specific fear. If the fear does not interfere with the child's daily life, such as sleeping, going to school, and engaging in social activities, then professional help is generally not needed.

Phobias belong to a large group of mental problems known as anxiety disorders and can be divided into three specific types: specific phobias (formerly called simple phobias), social phobias, and agoraphobia.

A specific phobia is the fear of a particular situation or object, including anything from airplane travel to dentists. Found in one out of every ten Americans, specific phobias seem to run in families and are roughly twice as likely to appear in women. If the person rarely encounters the feared object, the phobia does not cause much harm. However, if the feared object or situation is common, it can seriously disrupt everyday life. Common examples of specific phobias, which can begin at any age, are fear of snakes, flying, dogs, escalators, elevators, high places, or open spaces.

People with social phobia have deep fears of being watched or judged by others and of being embarrassed in public. Common social phobias in children include reading aloud in front of a class; participating in a musical, drama, or athletic event; starting or joining in a conversation; talking to adults; attending social events, such as dances and parties; taking tests; attending physical education class; using school or public bathrooms; and asking a teacher for help.

Social phobia is not the same as **shyness**. Shy people may feel uncomfortable with others, but they do not experience severe anxiety, they do not worry excessively about social situations beforehand, and they do not avoid events that make them feel self-conscious. On the other hand, people with social phobia may not be shy; they may feel perfectly comfortable with people except in a public place. This feeling usually begins about age 15 and affects three times as many women as men.

An episode of spontaneous panic is usually the initial trigger for the development of agoraphobia. After an initial panic attack, the person becomes afraid of experiencing a second one. Patients literally "fear the fear," and worry incessantly about when and where the next attack may occur. As they begin to avoid the places or situations in which the panic attack occurred, their fear generalizes. Eventually the person completely avoids public places. In severe cases, people with agoraphobia can no longer leave their homes for fear of experiencing a panic attack.

Agoraphobia is the intense fear of feeling trapped and having a panic attack in specific situations. Social phobias may be only mildly irritating, or they may significantly interfere with daily life. It is not unusual for people with social phobia to turn down job offers or avoid relationships because of their fears.

Parental concerns

While normal fears tend to be experienced in phases and tend to be outgrown by adulthood, abnormal fears are those that are persistent and recurrent or fears that interfere with daily activities for at least a month. Abnormal fears, including extreme separation anxiety, being afraid to go to school, or extreme social fears, may indicate an anxiety disorder.

When to call the doctor

When children's fears persist beyond the age when they are appropriate, they can begin to interfere with their daily lives. Typically, children who experience this type of irrational fear, or phobia, should get treatment from a psychologist.

The most popular and effective treatment for phobias is behavior therapy, which approaches the phobia as an undesirable behavior to be unlearned. Most often it takes the form of desensitization, a technique by which the fearful person is exposed to the feared stimulus in an extremely mild form and then with gradually increasing degrees of intensity. For example, a child who fears dogs may first be asked to look at pictures of dogs, then perhaps play with a stuffed dog or view a dog from afar, ultimately getting to the point when she is able to pet and play with dogs.

Phobias also respond to treatment by medication, including anti-anxiety drugs such as Xanax and BuSpar and selective serotonin reuptake inhibitors (SSRIs), such as Prozac and Zoloft. Medication is especially helpful for social phobia, where it can help the child overcome her aversion to social interaction sufficiently to work with a therapist. When agoraphobia accompanies panic attacks, it also responds to cognitive-behavioral treatment for panic disorder, often in conjunction with anti-anxiety and antidepressant medications similar to those prescribed for other phobias.

Before, during, and after exposure to the source of fear, the child can begin to imagine controlling the environment and his own reactions in other ways. Creative visualization, for example, imagining a switch the child can use to control his fear when visiting the doctor or dentist, can sometimes be effective. A comforting ritual, a familiar object, or thoughts of a beloved person can be used as a good luck charm before embarking on a scary trip or performing a task such as speaking in class or sleeping alone. Relaxation techniques can also be taught to older children.

KEY TERMS

Agoraphobia—Abnormal anxiety regarding public places or situations from which the person may wish to flee or in which he or she would be helpless in the event of a panic attack.

Ailurophobia—Fear of cats.

Cognitive—The ability (or lack of) to think, learn, and memorize.

Didaskaleinophobia—Fear of going to school.

Entomophobia—Fear of insects.

Glossophobia—Fear of speaking.

Myctophobia—Fear of darkness.

Ophidiaphobia—Fear of snakes.

Pantophobia—Fear of everything.

Phobia—An intense and irrational fear of a specific object, activity, or situation that leads to avoidance.

Selective serotonin reuptake inhibitors (SSRIs)—A class of antidepressants that work by blocking the reabsorption of serotonin in the brain, thus raising the levels of serotonin. SSRIs include fluoxetine (Prozac), sertraline (Zoloft), and paroxetine (Paxil).

Triskaidekaphobia—Fear of the number thirteen.

Xenophobia—Fear of strangers or foreigners.

Zoophobia—Fear of animals.

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Febrile seizures

Definition

Febrile seizures are convulsions of sudden onset due to abnormal electrical activity in the brain that is caused by **fever**. Fever is a condition in which body temperature is elevated above normal (generally above 100.4°F [38°C]).

Description

Febrile seizures were first distinguished from epileptic seizures in the twentieth century. The National Institutes of Health defined febrile seizures in 1980 as "an event in infancy or childhood usually occurring between three months and five years of age, associated with fever, but without evidence of intracranial infection or defined cause."

There are three major subtypes of febrile seizures. The simple febrile seizure accounts for 70 to 75 percent of febrile seizures and is one in which the affected child is age six months to five years and has no history or evidence of neurological abnormalities, the seizure is generalized (affects multiple parts of the brain), and lasts less than 15 minutes, and the fever is not caused by brain illness such as **meningitis** or **encephalitis**. The complex febrile seizure shares similar characteristics with the exception that the seizure lasts longer than 15 minutes or is local (affects a localized part of the brain), or multiple seizures take place and accounts for about 20 to 25 percent of all febrile seizures. Lastly, about 5 percent of febrile seizures are diagnosed as symptomatic, in cases in which the child has a history or evidence of neurological abnormality.

The seizure activity itself is generally characterized as clonic (consisting of rhythmic jerking movements of the arms and/or legs), or tonic-clonic (commencing with a stiffening of the body followed by a clonic phase).

Demographics

Fever is the most common cause of seizures in children, occurring in 2 to 5 percent of children from six months to five years of age. First onset usually occurs by two years of age, with the risk decreasing after age three; most children stop having febrile seizures by the age of five or six. Male children have been shown to have a higher incidence of febrile seizures. The majority of children who experience a febrile seizure will only have one in their lifetime; approximately 33 percent will go on to have more than one.

Causes and symptoms

Under normal circumstances, information is transmitted in the brain by means of electrical discharges from brain cells. A seizure occurs when the normal electrical patterns of the brain become disrupted. A febrile seizure is caused by fever, most commonly a high fever that has risen quickly. The average fever temperature in which febrile seizures take place is 104°F (40°C). Conversely, a healthy person's body temperature fluctuates between 97°F (36.1°C) and 100°F (37.8°C).

Fevers are caused in most cases by viral or bacterial infections, such as **otitis media** (ear infection), upper respiratory infection, pharyngitis (throat infection), **pneumonia**, **chickenpox**, and urinary tract infection. Other conditions can induce a fever, including allergic reactions, ingestion of toxins, teething, autoimmune disease, trauma, **cancer**, excessive sun exposure, or certain drugs. In some cases no cause of the fever can be determined.

Febrile seizures generally last between one and ten minutes. A child experiencing a febrile seizure may exhibit some or all of the following behaviors:

- stiff body
- twitching or jerking of the extremities or face
- rolled-back eyes
- unconsciousness
- inability to talk
- problems breathing
- involuntary urination or defecation
- vomiting
- confusion, sleepiness, or irritability after the seizure

Approximately one third of children who have had a febrile seizure will experience recurrent seizures. Several risk factors are associated with recurrent febrile seizures; children who exhibit all four are at a 70 percent chance of developing recurrent seizures, while those who have none of the risk factors have only a 20 percent chance. The risk factors include:

- family history of febrile seizures
- young age of the child (i.e. less than 18 months of age)
- seizure occurs soon after or with onset of fever
- seizure-associated fever is relatively low

When to call the doctor

A healthcare provider should be contacted after a febrile seizure. A visit to the emergency room is warranted if the accompanying fever is greater than 103°F (39.4°C) in a child older than three months or 100.5°F (38°C) in an infant of three months or younger or if the seizure is the child's first. Emergency medical personnel (telephone 911) should be called if a febrile seizure lasts more than five minutes; if the child stops breathing; if the child's skin starts to turn blue; or if the fever is greater than 105.8°F (41°C), a condition called hyperpyrexia.

Diagnosis

A key focus of diagnostic tests will be to determine the underlying cause of the fever. A comprehensive medical history including the fever's duration and course, other symptoms the child is experiencing, prior or current medical conditions, recent vaccinations or exposure to communicable diseases, and the child's current behaviors may point to the fever's origin. A temperature below 100.4°F (38°C) suggests another cause for the seizure. The caregiver who was present with the child while he or she was having the seizure will be asked questions relating to the child's behaviors in an attempt to determine the type of seizure.

Physicians may administer tests to rule out conditions other than fever that could have caused the seizure, such as epilepsy, meningitis, or encephalitis. Children who suffer from recurrent febrile seizures are not diagnosed with epilepsy, a **seizure disorder** that is not caused by fever. In the case of children under 18 months of age, a lumbar puncture (spinal tap) may be recommended to rule out meningitis because symptoms are often lacking or subtle in children of that age. Because of the benign nature of the simple febrile seizure, tests such as **computed tomography** (CT) scans, **magnetic resonance imaging** (MRI), or **electroencephalogram** (EEG) are not usually recommended.

Treatment

During a seizure parents or caregivers need to remain calm and take steps to make sure the child remains safe. During the period after the seizure the child may be disoriented and/or sleepy (called the postictal state), but quick recovery from this state is normal, and medical treatment is not normally needed.

During a seizure

If a parent or caregiver observes a child having a seizure, there are a number of measures that should be taken to ensure the child's **safety**. These include:

- staying calm
- laying the child on his or her side or front to prevent vomited matter from being aspirated into the lungs

- loosening any tight clothing or items that could constrict breathing
- marking the start and end time of the seizure
- clearing the surrounding area of unsafe items
- attending to the child for the duration of the seizure
- clearing the child's airway if it becomes obstructed with vomited material or other objects

Parents or caregivers should not attempt to stop the seizure or slap or shake the child in attempt to wake him/her. The child may move around during the seizure, and parents should not try to hold the child down. If the child vomits, a suction bulb can be used to help clear the airway.

After a seizure

A healthcare professional should be called immediately after the seizure in the event that further treatment or tests are required. **Hospitalization** is not normally required unless the child is suffering from a serious infection or illness or the seizure itself was abnormally long. Parents or caregivers may be instructed to take certain measures at home to reduce the child's fever, such as administering fever-reducing drugs (called antipyretics) such as **acetaminophen** (Tylenol) or ibuprofen (Advil). There is, however, no evidence that shows fever-reducing therapies reduce the risk of another febrile seizure occurring. If the child is suffering from a bacterial infection that is the cause of the fever, he or she may be placed on **antibiotics**.

Treating the fever

The treatment of pediatric fever varies according to the age of the child and the fever's cause, if known. Physicians recommend that newborns less than four weeks of age with fever be admitted to the hospital and administered antibiotics until a complete workup can be done to rule out bacterial infection or other serious illness. The same is recommended for infants ages four to 12 weeks if they appear ill. Infants of this age who otherwise appear well can often be managed on an outpatient basis with antipyretics and antibiotics in the case of bacterial infection.

For children ages three months and older, the course of treatment depends on the extent and cause of the fever. Most fevers and associated conditions can be managed on an outpatient basis. Low-grade fevers often do not need to be treated in otherwise healthy children. Antipyretics may be suggested to lower a fever and make the child more comfortable but will not affect the course of an underlying infectious disease. Aspirin should not be given to a child or adolescent with a fever since this drug has been linked to an increased risk of the serious condition called **Reye's syndrome**. Antibiotics may be administered if the child has a known or suspected bacterial infection.

Alternative treatment

There are some outpatient treatments that parents or caregivers may administer to reduce their febrile child's discomfort, although there is no evidence that indicates such treatments reduce the risk of febrile seizures. These include dressing the child lightly, applying cold washcloths to the face and neck, providing plenty of fluids to avoid **dehydration**, and giving the child a lukewarm bath or sponging the child in lukewarm water.

Prognosis

The risk of complications associated with febrile seizures is very low. Some of the complications that may occur are:

- biting the tongue
- choking on items that were in the mouth at the start of the seizure
- injury from falling down
- aspirating fluid or vomit into the lungs
- developing recurrent febrile seizures
- developing recurrent seizures unrelated to fever (epilepsy)
- complications related the underlying cause of the fever

Children who have had a febrile seizure are at an increased risk of having another; approximately one third of febrile seizure cases become recurrent. The risk of recurrent seizures decreases with age: infants younger than 12 months have a 50 percent chance of having a second seizure, while children over the age of 12 months have a 30 percent chance. The risk of a child going on to develop epilepsy is slightly increased at approximately 2–5 percent, compared to 1 percent for the general population; such a risk is increased in children who have a history of neurological abnormalities such as **cerebral palsy** or developmental delays and in children whose seizures recur or are prolonged. Research has shown that febrile seizures do not affect a child's **intelligence** level or achievement in school.

Prevention

In some cases, a febrile seizure may be the first indication that a child is ill. Prevention is, therefore, not

KEY TERMS

Antipyretic drug—Medications, like aspirin or acetaminophen, that lower fever.

Autoimmune disorder—One of a group of disorders, like rheumatoid arthritis and systemic lupus erythematosus, in which the immune system is overactive and has lost the ability to distinguish between self and non-self. The body's immune cells turn on the body, attacking various tissues and organs.

Encephalitis—Inflammation of the brain, usually caused by a virus. The inflammation may interfere with normal brain function and may cause seizures, sleepiness, confusion, personality changes, weakness in one or more parts of the body, and even coma.

Hyperpyrexia—Fever greater than 105.8°F (41°C).

Meningitis—An infection or inflammation of the membranes that cover the brain and spinal cord. It is usually caused by bacteria or a virus.

always possible. While the use of anticonvulsants such as Phenobarbital or Valproate has been shown to prevent recurrent febrile seizures, these drugs are associated with significant side effects such as adverse behaviors, allergic reaction, and organ injury, and have not been shown to benefit simple febrile seizures. Only rarely is anticonvulsant therapy recommended for a child with febrile seizures because of the generally benign nature of the seizures and the risk of side effects from the drugs. In some cases oral diazepam (Valium) can be administered at the first sign of fever to reduce the risk of febrile seizures; about two-thirds of children who receive this drug experience side effects such as sleepiness and loss of coordination. The majority of children who have had a febrile seizure do not need drug therapy. Parents may be directed to administer over-the-counter antipyretics at the first sign of fever.

Parental concerns

A febrile seizure can be a frightening experience for both the child and his or her parents. It is important that parents be educated about the low risk of simple febrile seizures and the measures that can be taken to ensure their child's safety during and after a seizure.

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Fetal alcohol syndrome

Definition

Fetal alcohol syndrome (FAS) is a set of physical and mental birth defects that can result from a woman drinking alcohol during her pregnancy. The syndrome is characterized by brain damage, facial deformities, and growth deficits. Heart, liver, and kidney defects are also common, as well as vision and hearing problems. These

infants generally have difficulties with learning, attention, memory, and problem solving as they get older.

Description

Although there is a wide range of effects that result from in utero alcohol exposure, the diagnosis of FAS is recognized as the most severe birth defect that occurs. Fetal alcohol effect (FAE) is a term used to describe alcohol-exposed individuals whose condition does not meet the full criteria for an FAS diagnosis. The term alcohol-related neurodevelopmental disorders (ARND) is used for individuals with functional or cognitive impairments linked to prenatal alcohol exposure, including decreased head size at birth, structural brain abnormalities, and a pattern of behavioral and mental abnormalities. Alcohol-related birth defects (ARBD) describes the physical defects linked to prenatal alcohol exposure, including heart, skeletal, kidney, ear, and eye malformations.

FAS is the leading known preventable cause of **mental retardation** and birth defects. It affects one in 100 live births or as many as 40,000 infants born each year in the United States, and it is felt that the incidence is significantly under-reported. An individual with FAS can incur a lifetime health cost of over $800,000. In 2003, FAS cost the United States $3.9 billion in direct costs with indirect costs at approximately $1.5 billion. Children do not outgrow FAS. The physical and behavioral problems can last a lifetime. The syndrome is found in all racial and socio-economic groups. It is not a genetic disorder, so women with FAS or affected by FAS have healthy babies if they do not drink alcohol during their pregnancy.

Causes and symptoms

Alcohol is readily absorbed from the gastrointestinal tract into a pregnant woman's bloodstream and circulates to the fetus by crossing the placenta. Here it interferes with the ability of the fetus to receive sufficient oxygen and nourishment for normal cell development in the brain and other organs. The consumption of alcohol directly contributes to **malnutrition** because it contains no **vitamins** or **minerals**, and it uses up what the woman has for metabolism. Studies suggest that drinking a large amount of alcohol at any one time may be more dangerous to the fetus than drinking small amounts more frequently. The fetus is most vulnerable to various types of injuries depending on the stage of development in which alcohol is encountered. During the first eight weeks of pregnancy, organogenesis (the formation of organs) is taking place, which places the embryo at a higher risk of deformities when exposed to teratogens. Since a safe amount of alcohol intake during pregnancy has not been determined, twenty-first century authorities agree that women should not drink at all during pregnancy. A problem is that many women do not realize they are pregnant until the sixth to eight week. Therefore, women who are anticipating a pregnancy should abstain from all alcoholic beverages.

Unlike many birth defects which are identified at birth and then treated, FAS and FAE are usually overlooked at birth and treated later by mental health specialists, and often unknowingly. Possible FAS symptoms include:

- growth deficiencies: small body size and weight, slower than normal development, and failure to catch up
- skeletal deformities: deformed ribs and sternum; curved spine; hip dislocations; bent, fused, webbed, or missing fingers or toes; limited movement of joints; small head
- facial abnormalities: small eye openings; skin webbing between eyes and base of nose; drooping eyelids; nearsightedness; **strabismus**; failure of eyes to move in same direction; short upturned nose; sunken nasal bridge; flattened or absent groove between nose and upper lip; thin upper lip; **cleft palate** (opening in roof of mouth); small jaw; low-set or poorly formed ears
- organ deformities: heart defects, **heart murmurs**, genital malformations, kidney and urinary defects
- central nervous system handicaps: small brain; faulty arrangement of brain cells and connective tissue; mental retardation (usually mild to moderate but occasionally severe); learning disabilities; short attention span; irritability in infancy; hyperactivity in childhood; poor body, hand, and finger coordination

Since the primary birth defect in FAS and FAE involves central nervous system damage in utero, these newborns may have difficulties with feeding due to a poor suck, have irregular sleep-wake cycles, decreased or increased muscle tone, and seizures or tremors. Delays in achieving developmental milestones such as rolling over, **crawling**, walking, and talking may become apparent in infancy. Behavior and learning difficulties typical in the **preschool** or early school years include poor attention span, hyperactivity, poor motor skills, and slow **language development**. A common diagnosis that is associated with FAS is attention deficit-hyperactivity disorder. Learning disabilities or mental retardation may be diagnosed during this time. Arithmetic is often the most difficult subject for a child with FAS. During middle school and high school years, the behavioral

difficulties and learning difficulties can be significant. Memory problems, poor judgment, difficulties with daily living skills, difficulties with abstract reasoning skills, and poor social skills are often apparent by this time. It is important to note that animal and human studies have shown that neurologic and behavioral abnormalities can be present without characteristic facial features. These individuals may not be identified as having FAS but may fulfill criteria for alcohol-related diagnoses, as set forth by the Institute of Medicine.

In 1991, Streissguth and others reported some of the first long-term follow-up studies of adolescents and adults with FAS. In the approximate 60 individuals they studied, the average IQ was 68 (70 is the lower limit of the normal range). However, the range of IQ was quite large, as low as 20 (severely retarded) to as high as 105 (normal). The average achievement levels for reading, spelling, and arithmetic were fourth grade, third grade, and second grade, respectively. The Vineland Adaptive Behavior Scale was used to measure adaptive functioning in these individuals. The composite score for this group showed functioning at the level of a seven-year-old. Daily living skills were at a level of nine years, and social skills were at the level of a six-year-old.

In 1996, Streissguth and others published further data regarding the disabilities in children, adolescents, and adults with FAS. Secondary disabilities (those disabilities not present at birth and that might be preventable with proper diagnosis, treatment, and intervention) were described. These secondary disabilities include: mental health problems; disrupted school experiences; trouble with the law; incarceration for mental health problems, drug abuse, or a crime; inappropriate sexual behavior; alcohol and drug abuse; problems with employment; dependent living; and difficulties parenting their own children. In that study, only seven out of 90 adults were living and working independently and successfully. In addition to the studies by Streissguth, several other authors in different countries have as of the early 2000s reported on long term outcome of individuals diagnosed with FAS. In general, the neurologic, behavioral, and emotional disorders become the most problematic for individuals. The physical features change over time, sometimes making the correct diagnosis more difficult in older individuals, without old photographs and other historical data to review. Mental health problems, including attention deficit, depression, panic attacks, psychosis, **suicide** threats and attempts, were present in over 90 percent of the individuals studied by Streissguth. A 1996 study in Germany reported more than 70 percent of the adolescents they followed had persistent and severe developmental disabilities, and many had psychiatric disorders, the most common of which were emotional disorders, repetitive habits, **speech disorders**, and hyperactivity disorders. (Some of the above information derives from Ann Streissguth's book, *Fetal Alcohol Syndrome: A Guide for Families and Communities*, which appeared in 1997.)

Diagnosis

FAS is a clinical diagnosis, which means that there is no blood test, x ray, or psychological test that can be performed to confirm the suspected diagnosis. The diagnosis is made based on the history of maternal alcohol use and detailed physical examination for the characteristic major and minor birth defects and characteristic facial features. It is often helpful to examine siblings and parents of an individual suspected of having FAS, either in person or by photographs, to determine whether findings on the examination might be familial and if other siblings may also be affected. Individuals with **developmental delay** or birth defects may be referred to a clinical geneticist for genetic testing or to a developmental pediatrician or neurologist for evaluation and diagnosis of FAS. Psychoeducational testing to determine IQ and/or the presence of learning disabilities may also be part of the evaluation process.

Treatment

There is no treatment for FAS that will reverse or change the physical features or brain damage associated with maternal alcohol use during the pregnancy. Most of the physical birth defects associated with prenatal alcohol exposure are correctable with surgery. Children should have psychoeducational evaluation to help plan appropriate educational interventions. Commonly associated diagnoses as attention deficit-hyperactivity disorder, depression, or **anxiety** should be recognized and treated appropriately. The disabilities that present during childhood persist into adult life. However, some of the secondary disabilities already mentioned may be avoided or lessened by early diagnosis and intervention. Streissguth has describe a model in which an individual affected by FAS has one or more advocates to help provide guidance, structure, and support as the individual seeks to become independent, successful in school or employment, and develop satisfying social relationships.

Prognosis

The prognosis for FAS depends on the severity of birth defects and the brain damage present at birth.

KEY TERMS

Cleft palate—A congenital malformation in which there is an abnormal opening in the roof of the mouth that allows the nasal passages and the mouth to be improperly connected.

Congenital—Present at birth.

Intelligence quotient (IQ)—A measure of somebody's intelligence, obtained through a series of aptitude tests concentrating on different aspects of intellectual functioning.

Microcephaly—An abnormally small head.

Miscarriage—Loss of the embryo or fetus and other products of pregnancy before the twentieth week. Often, early in a pregnancy, if the condition of the baby and/or the mother's uterus are not compatible with sustaining life, the pregnancy stops, and the contents of the uterus are expelled. For this reason, miscarriage is also referred to as spontaneous abortion.

Organogenesis—The formation of organs during development.

Placenta—The organ that provides oxygen and nutrition from the mother to the unborn baby during pregnancy. The placenta is attached to the wall of the uterus and leads to the unborn baby via the umbilical cord.

Strabismus—A disorder in which the eyes do not point in the same direction.

Teratogen—Any drug, chemical, maternal disease, or exposure that can cause physical or functional defects in an exposed embryo or fetus.

Miscarriage, stillbirth, or death in the first few weeks of life may be outcomes in very severe cases. Major physical birth defects associated with FAS are usually treatable with surgery. Some of the factors that have been found to reduce the risk of secondary disabilities in FAS individuals include diagnosis before the age of six years, stable and nurturing home environments, never having experienced personal violence, and referral and eligibility for disability services. The long-term data help others understand the difficulties that individuals with FAS encounter throughout their lifetimes and can help families, caregivers, and professionals provide the care, supervision, education, and treatment geared toward their special needs.

A boy with fetal alcohol syndrome, a birth defect caused by his mother consuming alcohol during pregnancy. *(Photograph by David H. Wells. Corbis.)*

Parental concerns

Prevention of FAS is the key. Prevention efforts must include public education efforts aimed at the entire population, not just women of child bearing age, appropriate treatment for women with high-risk drinking habits, and increased recognition and knowledge about FAS by professionals, parents, and caregivers.

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March of Dimes Birth Defects Foundation. 1275 Mamaroneck Ave., White Plains, NY 10605. Web site: <www.modimes.org>.

National Institute on Alcohol Abuse and Alcoholism. 5635 Fishers Lane, MSC 9304, Bethesda, MD 20892–9304. Web site: <www.niaaa.nih.gov/>.

National Organization on Fetal Alcohol Syndrome (NOFAS). 900 17th Street, NW, Suite 910, Washington, DC 20006. Web site: <www.nofas.org>.

Linda K. Bennington

Fetal hemoglobin test

Definition

A fetal hemoglobin test (Hgb electrophoresis) measures the level of fetal hemoglobin (Hemoglobin F or HbF) in the blood of infants and children. It can also be measured in adults, though is more typically needed for diagnoses of congenital illnesses in children. Fetal hemoglobin, an alkali-resistant form of hemoglobin, is the major hemoglobin component in the bloodstream of the fetus. After birth, it decreases rapidly until only traces are found in healthy children and adults. Fetal hemoglobin is one of six types of hemoglobin measured in the clinical laboratory by a method called hemoglobin electrophoresis.

Purpose

The determination of fetal hemoglobin in the blood of infants and children identifies normal and abnormal levels, defining what percentage of total hemoglobin is made up of fetal hemoglobin. Knowing this level may help doctors evaluate low concentrations of normal hemoglobin in red blood cells (anemia), as well as higher-than-normal levels of fetal hemoglobin or its hereditary persistence. Fetal hemoglobin measurement helps diagnose a group of inherited disorders that affect hemoglobin production, among which are the thalassemias and **sickle cell anemia**. It may also be done to help doctors diagnose acquired illnesses such as acquired hemolytic anemia, leukemia, pernicious anemia, and certain types of **cancer**.

Description

Hemoglobin is the oxygen-carrying protein in red blood cells. It is also the pigment that gives red blood cells their color. Red blood cells deliver hemoglobin throughout the body, ensuring that all body tissues have the oxygen they need for life and proper function. Hemoglobin consists primarily of iron-bearing proteins called heme groups and moiety globin protein, which together give hemoglobin its ability to carry oxygen. The heme groups are molecular chains of different types and actually create six different hemoglobins that vary in their amino acid composition and also in the genes that control them. Among the six types of hemoglobin, HbA is the normal adult hemoglobin, and HbF is the major fetal hemoglobin. Abnormal types of hemoglobin include Hgb S and Hgb C. All types of hemoglobin are electrically charged, which enables them to be identified and quantified in the laboratory by hemoglobin electrophoresis techniques.

During fetal development, fetal hemoglobin composes about 90 percent of total hemoglobin. At birth, the newborn's blood is composed of about 70 percent fetal hemoglobin. As the infant's bone marrow begins to produce new red cells, fetal hemoglobin begins to decrease rapidly. Normally, only 2 percent or less of total hemoglobin is found as fetal hemoglobin after six months and throughout childhood; in adulthood, only traces (0.5% or less) are found in total hemoglobin.

In some diseases associated with abnormal hemoglobin production (hemoglobinopathy), fetal hemoglobin may persist in larger amounts. When this occurs, the increased amounts of fetal hemoglobin raise questions of possible underlying dysfunction or disease. For example, HbF can be found in higher levels in sickle cell anemia and other hereditary **anemias**. It has also been reported to be elevated in some other conditions such as leukemia, pregnancy, diabetes, thyroid disease, and sometimes as a side effect of anticonvulsant therapy. It may also reappear in adults when the bone marrow is overactive, as in disorders such as pernicious anemia, multiple myeloma, and invasive (metastatic) cancer affecting bone marrow. When HbF is elevated after age four, the cause is typically investigated. (Persistence of fetal hemoglobin in inherited hemolytic anemias can be associated with less severe disease symptoms.)

Defects in hemoglobin production may be either genetic or acquired. The genetic defects are subdivided

into errors of heme production (porphyria) and those of globin production, known collectively as the hemoglobinopathies. There are two categories of hemoglobinopathy: in one, abnormal globin chains give rise to abnormal hemoglobin molecules; in the other, normal hemoglobin chains are produced but in abnormal amounts. Sickle cell anemia, the inherited condition characterized by curved (sickle-shaped) red blood cells and chronic hemolytic anemia, is an example of the first category. Disorders in the second category are called the thalassemias, which are classified according to which amino acid chain, alpha or beta, is affected, and whether one defective gene (**thalassemia** minor) or two defective genes (thalassemia major) are responsible for the disorder. Testing for levels of fetal hemoglobin and other types of hemoglobin may be a first, important step in the investigation of possible hemoglobinopathies.

Levels of HbF are of interest in diagnosing several hemoglobinopathies, including the following:

- thalassemia minor or thalassemia trait (heterozygous thalassemia), in which HbF does not decrease normally after birth and may remain high in later life
- thalassemia major (homozygous thalassmia or Cooley's anemia), the hereditary persistence of HbF involving larger than normal amounts of HbF
- sickle cell anemia associated with the abnormal Hgb S, which occurs primarily in African-Americans; also Hgb C, another abnormal hemoglobin found in African-Americans, causing hemolytic anemia
- beta-chain hemoglobinopathies with increased HbF during childhood
- disease-related hematologic stress as in hemolytic anemias, leukemia, and aplastic anemia

Precautions

Blood transfusions received prior to testing may alter results.

Preparation

Parents may wish to explain the blood-drawing procedure to older children to help prepare them for the slight discomfort they will experience.

Testing for fetal hemoglobin requires that a blood sample be drawn from the child. No preparation is needed before performing fetal hemoglobin tests, and fasting (nothing to eat or drink for a period of hours before the test) is not required. Proper identification and careful handling of the child are important when a blood sample is being obtained for testing. A site, usually the heel on an infant and a finger on an older child, is chosen by the phlebotomist who will draw the blood. A baby's foot may be wrapped in a warm cloth for a few minutes to bring blood to the surface and allow it to flow more easily. The baby's heel or child's finger is then wiped with alcohol and/or an antibacterial solution such as betadine to sterilize the surface. Puncture is performed quickly with a lancet, avoiding the center of an infant's heel to prevent inflammation of the bone. The blood sample is drawn into tiny capillary tubes, properly labeled, and taken to the laboratory for testing. In rare instances, a phlebotomist will not be able to draw sufficient blood from an infant's heel puncture, and a physician may draw venous blood from a femoral vein in the groin area, which is larger than veins in an infant's arms. Older children may also have venous blood drawn, particularly if other blood tests are being done.

Aftercare

Pressure is applied to the blood-drawing site for a few minutes to prevent bleeding. The site of heel stick, finger stick, or venipuncture must be kept clean and dry and observed for any undue bleeding or bruising. A small bandage can be used to cover the site. Any unusual conditions or reactions should be reported to the pediatrician.

Risks

Risks for this test are minimal but may include slight bleeding from the blood-drawing site or blood may accumulate under the puncture site (hematoma). Fainting, **nausea**, or feeling lightheaded after venipuncture may occur in some children.

Normal results

Reference values vary from laboratory to laboratory but results of hemoglobin electrophoresis are generally reported within the following ranges:

- Newborn to six months: HbF may be up to 70 percent of total hemoglobin.
- Six months to adult: HbF may be up to 2 percent of total hemoglobin.

Levels of HbF greater than 2 percent of total hemoglobin is abnormal after the age of six months. Some hemoglobinopathies have elevated levels of other types of hemoglobin and normal levels of HbF as shown below.

Typical results for certain hemoglobinopathies include:

KEY TERMS

Anemia—A condition in which there is an abnormally low number of red blood cells in the bloodstream. It may be due to loss of blood, an increase in red blood cell destruction, or a decrease in red blood cell production. Major symptoms are paleness, shortness of breath, unusually fast or strong heart beats, and tiredness.

Hemoglobinopathy—A disorder of hemoglobin, which can be either the presence of abnormal types of hemoglobin or abnormal levels of specific types of hemoglobin.

Hemolytic anemia—A form of anemia characterized by chronic premature destruction of red cells in the bloodstream. Hemolytic anemias are classified as either inherited or acquired.

Heterozygote/Heterozygous—Having two different versions of the same gene.

Homozygote/Homozygous—Having two identical copies of a gene.

Trait—A distinguishing feature of an individual.

- Sickle cell disease: Hemoglobin S 80–100%, Hgb A 0%, HbF 2%.
- Sickle cell trait: Hemoglobin S 20–40%, Hgb A 60–80%, HbF 2%.
- Hgb C disease: Hgb C 90–100%, Hgb A 0%, and Hb F 2%.
- Thalassemia major: HbF 65–100%, Hgb A 5–25%.
- Thalassemia minor: HbF 1–3%, Hgb A 50–90%.

Parental concerns

Children generally respond well to blood-drawing procedures if they are prepared for the slight discomfort. If parents are concerned about the possibility of inherited blood disorders, it may be helpful to remember that abnormal levels of fetal hemoglobin may be caused by a variety of conditions, not all of which are inherited or serious, and early recognition of a blood condition usually leads to early treatment and effective management of the condition.

See also Sickle cell anemia; Thalassemia.

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Fetal monitoring, electronic *see* **Electronic fetal monitoring**

Fever

Definition

A fever is any body temperature elevation over 100.4°F (38°C).

Description

A healthy person's body temperature fluctuates between 97°F (36.1°C) and 100°F (37.8°C), with the average being 98.6°F (37°C). The body maintains stability within this range by balancing the heat produced by the metabolism with the heat lost to the environment. The "thermostat" that controls this process is located in the hypothalamus, a small structure located deep within the brain. The nervous system constantly relays information about the body's temperature to the thermostat, which in turn activates different physical responses designed to cool or warm the body, depending on the circumstances. These responses include: decreasing or increasing the flow of blood from the body's core, where it is warmed, to the surface, where it is cooled; slowing down or speeding up the rate at which the body turns food into energy (metabolic rate); inducing shivering, which generates heat through muscle contraction; and inducing sweating, which cools the body through evaporation.

A fever occurs when the thermostat resets at a higher temperature, primarily in response to an infection. To reach the higher temperature, the body moves blood to the warmer interior, increases the metabolic rate, and induces shivering. The chills that often accompany a fever are caused by the movement of blood to the body's core, leaving the surface and extremities cold. Once the higher temperature is achieved, the shivering and chills stop. When the infection has been overcome or drugs such as aspirin or **acetaminophen** have been taken, the thermostat resets to normal and the body's cooling mechanisms switch on: the blood moves to the surface and sweating occurs.

Fever is an important component of the immune response, though its role is not completely understood. Physicians believe that an elevated body temperature has several effects. The immune system chemicals that react with the fever-inducing agent and trigger the resetting of the thermostat also increase the production of cells that fight off the invading bacteria or viruses. Higher temperatures also inhibit the growth of some bacteria, while at the same time speeding up the chemical reactions that help the body's cells repair themselves. In addition, the increased heart rate that may accompany the changes in blood circulation also speeds the arrival of white blood cells to the sites of infection.

Demographics

Fevers are components of many disease entities. Virtually all persons experience fevers at some time in their lives. Elevations in temperature are not reportable events. Thus, accurate data regarding the prevalence of fevers are not available.

Causes and symptoms

Fevers are primarily caused by viral or bacterial infections, such as **pneumonia** or **influenza**. However, other conditions can induce a fever, including allergic reactions; autoimmune diseases; trauma, such as breaking a bone; **cancer**; excessive exposure to the sun; intense **exercise**; hormonal imbalances; certain drugs; and damage to the hypothalamus. When an infection occurs, fever-inducing agents called pyrogens are released, either by the body's immune system or by the invading cells themselves that trigger the resetting of the thermostat. In other circumstances, the immune system may overreact (allergic reactions) or become damaged (autoimmune diseases), causing the uncontrolled release of pyrogens. A **stroke** or tumor can damage the hypothalamus, causing the body's thermostat to malfunction. Excessive exposure to the sun or intense exercise in hot weather can result in heat stroke, a condition in which the body's cooling mechanisms fail. Malignant hyperthermia is a rare, inherited condition in which a person develops a very high fever when given certain anesthetics or muscle relaxants in preparation for surgery.

How long a fever lasts and how high it may go depends on several factors, including its cause, the age of the person, and his or her overall health. Most fevers caused by infections are acute, appearing suddenly and then dissipating as the immune system defeats the infectious agent. An infectious fever may also rise and fall throughout the day, reaching its peek in the late afternoon or early evening. A low-grade fever that lasts for several weeks is associated with autoimmune diseases such as lupus or with some cancers, particularly leukemia and lymphoma.

When to call the doctor

A doctor or other healthcare provider should be called when a fever does not resolve after two to three days. Anyone with a fever over 104°F (40.0°C) should seek immediate medical treatment.

A doctor should be called when an infant's temperature rises above 100°F (37.8°C) and cannot be brought down within a few minutes. Infants whose temperatures exceed 102°F (38.9°C) should be immersed in warm or tepid water to help reduce temperature slowly, while waiting for emergency help to arrive.

Diagnosis

A fever is usually diagnosed using a thermometer. A variety of different thermometers are available. Glass thermometer should not be used since they can break and release mercury, which is toxic. Digital thermometers can and should be used in place of glass thermometers rectally, orally, and under the arm in all age groups. Electronic thermometers can be inserted in the ear to quickly register the body's temperature.

As important as registering a person's temperature is determining the underlying cause of the fever. The presence or absence of accompanying symptoms, a person's medical history, and information about what he or she may have ingested, any recent trips taken, or possible exposures to illness all help the physician make a diagnosis. Blood tests can aid in identifying an infectious agent by detecting the presence of antibodies against it or providing samples for growth of the organism in a culture. Blood tests can provide the doctor with white blood cell counts. Ultrasound tests, **magnetic resonance imaging** (MRI) tests, or **computed tomography** (CT) scans may

KEY TERMS

Antipyretic drug—Medications, like aspirin or acetaminophen, that lower fever.

Autoimmune disorder—One of a group of disorders, like rheumatoid arthritis and systemic lupus erythematosus, in which the immune system is overactive and has lost the ability to distinguish between self and non-self. The body's immune cells turn on the body, attacking various tissues and organs.

Febrile seizure—Convulsions brought on by fever.

Hyperthermia—Body temperature that is much higher than normal (i.e. higher than 98.6°F).

Malignant hyperthermia—A type of reaction (probably with a genetic origin) that can occur during general anesthesia and in which the patient experiences a high fever, muscle rigidity, and irregular heart rate and blood pressure.

Meningitis—An infection or inflammation of the membranes that cover the brain and spinal cord. It is usually caused by bacteria or a virus.

Metabolism—The sum of all chemical reactions that occur in the body resulting in growth, transformation of foodstuffs into energy, waste elimination, and other bodily functions. These include processes that break down substances to yield energy and processes that build up other substances necessary for life.

Pyrogen—A chemical circulating in the blood that causes a rise in body temperature.

Reye's syndrome—A serious, life-threatening illness in children, usually developing after a bout of flu or chickenpox, and often associated with the use of aspirin. Symptoms include uncontrollable vomiting, often with lethargy, memory loss, disorientation, or delirium. Swelling of the brain may cause seizures, coma, and in severe cases, death.

be ordered if the doctor cannot readily determine the cause of a fever.

Treatment

Physicians agree that the most effective treatment for a fever is to address its underlying cause, such as through the administration of **antibiotics**. Also, because a fever helps the immune system fight infection, it usually should be allowed to run its course. Drugs to lower fever (antipyretics) can be given if a person (particularly a child) is uncomfortable. These include acetaminophen and ibuprofen. Aspirin, however, should not be given to a child or adolescent with a fever since this drug has been linked to an increased risk of **Reye's syndrome**. Bathing a person in tepid water can also help alleviate a high fever.

An ear thermometer is used to measure body temperature through the ear canal. *(© LWA-Stephen Welstead/Corbis.)*

A fever requires emergency treatment under the following circumstances:

- newborn (three months or younger) with a fever over 100.5°F (38°C)
- infant or child with a fever over 103°F (39.4°C)
- fever accompanied by severe **headache**, neck stiffness, mental confusion, or severe swelling of the throat

A very high fever in a small child can trigger seizures (**febrile seizures**) and, therefore, should be treated immediately. A fever accompanied by the above symptoms can indicate the presence of a serious infection, such as **meningitis**, and should be brought to the immediate attention of a physician.

Prognosis

Most fevers caused by infection end as soon as the immune system rids the body of the pathogen, and these fevers do not produce lasting effects. The prognosis for fevers associated with more chronic conditions, such as autoimmune disease, depends upon the overall outcome of the disorder.

Prevention

Fevers may be prevented by avoiding the various diseases that cause them.

Nutritional concerns

Adequate **nutrition** via a well-balanced diet and sufficient intake of liquid help to reduce many fevers. Adequate intake of electrolytes such as sodium, chloride, potassium, phosphate, and bicarbonate helps to prevent **dehydration** that often accompanies a fever.

Parental concerns

Parents should carefully monitor their infants and young children for symptoms of fever. Any fever that exceeds 103°F (39.4°C) for more than a few minutes should be promptly treated.

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American Academy of Emergency Medicine. 611 East Wells St., Milwaukee, WI 53202. Web site: <www.aaem.org/>.

American Academy of Pediatrics. 141 Northwest Point Boulevard, Elk Grove Village, IL 60007–1098. Web site: <www.aap.org/>.

American Association of Naturopathic Physicians. 8201 Greensboro Drive, Suite 300, McLean, VA 22102. Web site: <naturopathic.org/>.

American Society of Clinical Pathologists. 2100 West Harrison Street, Chicago IL 60612. Web site: <www.ascp.org/index.asp>.

College of American Pathologists. 325 Waukegan Road, Northfield, IL 60093. Web site: <www.cap.org/>.

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Fever blister *see* **Cold sore**

Fever of unknown origin

Definition

Fever of unknown origin (FUO) refers to the presence of a documented elevation in body temperature for a specified time, for which a cause has not been found after basic medical evaluation. FUO is categorized as classic, hospital acquired FUO; FUO associated with low white blood cell counts (immunosuppression); and HIV-associated (AIDS-related) FUO.

Description

Fever, an elevation of normal body temperature, is a natural response of the body that helps fight off foreign substances, such as microorganisms (bacteria and viruses), parasites, fungi, and toxins. Body temperature is set by the thermoregulatory center, located in an area in the brain called the hypothalamus. Body temperature is not constant all day, but actually is lowest at 6 a.m. and highest around 4 to 6 p.m. Temperature also varies in different regions of the body; for example, rectal and urine temperatures are about one degree Fahrenheit higher than oral temperature, and rectal temperature is higher than urine. Certain normal conditions can also effect body temperature, such as food ingestion, age, pregnancy, and certain hormonal changes.

Substances that cause fever are known as pyrogens, which can be either exogenous (originate outside the body, such as bacterial toxins) or endogenous (formed by the body's own cells in response to an outside stimulus, such as a bacterial toxin). Researchers have discovered that there are several endogenous pyrogens, each made up of small groups of amino acids, the building blocks of proteins. When these natural pyrogens, called cytokines, are injected into humans, fever and chills develop within an hour. Interferon, tumor necrosis factor, and various interleukins are the major fever-producing cytokines.

In the complex process that produces fever, cytokines cause the thermoregulatory center in the hypothalamus to reset the normal temperature level. The body's initial response is to conserve heat by vasoconstriction, a process in which blood vessels narrow and prevent heat loss from the skin and elsewhere. This process alone raises temperature by two to three degrees. Certain behavioral activities also occur, such as adding more clothes and seeking a warmer environment. If the hypothalamus requires more heat, shivering occurs.

In children, the definition of FUO is applied when fever has been present for 14 days with no apparent cause, even though physical examinations have been made and laboratory tests performed. Doctors pay special attention to the ears, nose, throat, sinuses, and chest as sites of infection, since most childhood infections are respiratory in nature. The majority of children with FUO are eventually found to have one of several infectious diseases or an autoimmune disease. In many cases the disease is common, and in some cases an allergic response is causing the fever. Fever increases the body's metabolic rate and oxygen consumption, which can have a devastating effect on individuals with poor circulation. In addition, fever can lead to seizures in the very young. Some possible infectious causes shown in studies of children with FUO are as follows:

- acute **otitis media** (middle ear infection)
- bacterial meningitis
- blastomycosis
- brucellosis
- cystic fibrosis
- ehrlichiosis
- endocarditis
- enteric infection
- herpes infection
- HIV infection
- infectious mononucleosis
- lower respiratory tract infection
- malaria
- osteomyelitis
- Rocky Mountain spotted fever
- strep infection (streptococcus infection)
- systemic viral syndrome

- tonsillophyaryngitis or tonsillitis
- **tuberculosis**
- tularemia
- urinary tract infection
- viral meningoencephalitis

Transmission

It is possible for a child with FUO to spread infection or illness to other individuals, particularly if an infectious organism is the underlying cause. If a child has FUO, it is best to reduce contact with other young children or immune compromised **family** members until the cause of the fever has been identified.

Demographics

Fever of unknown origin can occur in anyone, male or female, of any age at any time depending upon exposure to infectious organisms such as bacteria or viruses or to other causes of illness such as fungi, parasites, or toxins or to underlying autoimmune or allergic conditions. Because the underlying cause of the fever is usually recorded as the diagnosis, accurate statistics for those presenting with FUO are not available.

Causes and symptoms

There are many possible causes of FUO; generally though, a diagnosis can be found. The most frequent cause of FUO is still infection, though the percentage has decreased in the early 2000s. Tuberculosis remains an important cause, especially when it occurs outside the lungs. The decrease in infections as a cause of FUO is due in part to improved culture techniques that allow more precise identification of organisms and, therefore, more appropriate treatment. In addition, advances in diagnostic technologies have made it easier to identify non-infectious causes. For example, tumors and autoimmune diseases were as of 2004 easier to diagnose. An autoimmune disease is one that arises when the body's immune system attacks its own tissue as if it were foreign. This happens when the immune system does not recognize protein markers (antigens) on its own cells. In some cases, reactions to medications can also cause prolonged fever.

In about 10 percent of cases, no definite cause is found. In another 10 percent, ''factitious fevers'' (either self induced or no fever at all) are identified.

General constitutional symptoms tend to occur along with fever, including muscle aches and pains (myalgias), chills, and **headache**. Sometimes symptoms such as a rash suggest an allergic reaction.

When to call the doctor

An infant under three months should be seen as soon by a pediatrician as possible if a fever develops. If a toddler or older child has a fever for more than a day or two (48 to 72 hours), with or without other symptoms, the pediatrician should be consulted so that an early diagnosis can be made and treatment begun. It is especially important to watch for signs of **dehydration**, particularly if the child is not drinking liquids or seems too sick to drink. A crying child with fever may have **pain** associated with a specific condition and should be seen by the pediatrician as soon as possible.

Diagnosis

Few symptoms in medicine present such a diagnostic challenge as fever. Nonetheless, if a careful, logical, and thorough evaluation is performed, the underlying cause generally can be diagnosed. The child's medical history is first reviewed along with travel, social, and family history, which can reveal important clues.

The first step medically is to search for an infectious cause. Skin and other screening tests for diseases such as tuberculosis and examination of blood, urine, and stool are generally indicated. Antibodies to a number of infectious agents can be measured; if antibody levels are rising, they may point to an active infection. In some cases, a febrile agglutination test can be performed to detect the presence in blood of certain infectious organisms that may stimulate the immune system to produce antibodies known as febrile agglutinins. The test helps diagnose or confirm certain febrile diseases that are known to be associated with febrile agglutinins. These may include:

- brucellosis, a type of infection caused by bacteria belonging to the genus *Brucella* and characterized by intermittent fever, sweating, chills, aches, and mental depression
- rickettsial infections, a group of diseases caused by the bacteria *Rickettsia*
- salmonellosis, caused by *Salmonella* bacteria and marked by **nausea** and severe diarrhea
- tularemia, also called rabbit fever, a bacterial infection characterized by a high fever and swollen lymph nodes

Various x-ray studies are of value and may be performed, particularly if organisms are identified that may indicate involvement of abdominal organs. Imaging techniques such as ultrasound, **computed tomography** (CT scan), and **magnetic resonance imaging** (MRI)

may be performed. These enable physicians to examine areas that were once accessible only through surgery. Furthermore, new studies using radioactive materials (nuclear medicine) can detect areas of infection and inflammation previously almost impossible to find, even with surgery.

The removal and microscopic examination of tiny bits of tissue (biopsy) from any suspicious areas found on an x-ray exam can be performed by either traditional or newer surgical techniques. Material obtained by biopsy is then examined by a pathologist in order to look for clues as to the cause of the fever. Evidence of infection, tumor, or other diseases can be found in this way. Portions of the biopsy are also sent to the laboratory for culture in an attempt to grow and identify an infectious organism.

Fever in an individual with HIV, primary immune deficiency, recent transplant, on **chemotherapy**, or anyone else who is immunocompromised constitutes an especially difficult problem, as these patients often suffer from many unusual infections. HIV itself is a potential cause of fever.

Treatment

Most children who undergo evaluation for FUO do not receive treatment until a clear-cut cause is found. **Antibiotics** or medications designed to suppress a fever such as antipyretics (**acetaminophen**) or non-steroidal anti-inflammatory drugs (NSAIDs) will only hide the true cause. Once physicians are satisfied that there is no infectious cause, they may recommend medications such as acetaminophen, NSAIDs, or corticosteroids to decrease inflammation and reduce constitutional symptoms. Parents are advised not to give children aspirin for fever because of a side effect called Reye syndrome, which may cause liver failure. Fluids are replenished by having the child consume clear liquids. A child too sick to drink may be hospitalized and given intravenous fluids.

The development of FUO in certain settings, such as when it is acquired by patients in the hospital or in immunosuppressed individuals with a low white blood count, often needs rapid treatment to avoid serious complications. Therefore, in these instances antibiotics may be given after a minimal number of diagnostic studies. Once test results are known and causative organisms identified, treatment can be adjusted appropriately.

Alternative treatment

Practitioners of complementary medicine recommend herbs to treat fever. A tea made with catnip, lobelia, and dandelion will reduce fever or catnip tea enemas administered daily. Rest is recommended and large quantities of clear liquids to flush out toxins and help prevent dehydration. Some practitioners recommend letting a fever run its course or cooling the body with cool sponge baths.

Supporting the immune system is one way to help avoid infection by exposure to bacteria, viruses, and toxins as a potential source of fever. Green drinks made with young barley are believed to cleanse the blood and supply chlorophyll and nutrients for maintaining healthy tissue. Because stress is known to produce biochemicals that reduce white blood cell functioning, it is important to get sufficient **sleep** and reduce stress to help keep the immune system functioning well. Therapeutic massage, **yoga**, and other types of stress reduction programs are available in most communities.

Prognosis

The outlook for children with FUO depends on the cause of the fever. If the basic illness is easily treatable and can be found rather quickly, the potential for a cure is quite good. Some children may continue to have an elevated temperature steadily or intermittently for six months or more. If no serious disease is found, medications such as NSAIDs are used to decrease the effects of the fever. Careful follow-up and reevaluation is recommended in these cases.

Prevention

Although FUO cannot actually be prevented because the sources are unknown, the immune system can be strengthened to help avoid infection from bacteria, viruses, and toxins. Several nutritional supplements are reported to help build the immune system. These include garlic (contains the essential trace element germanium), essential fatty acids (found in flax seed oil, evening primrose oil, and fish oils), sea vegetables such as kelp, acidophilus to supply natural bacteria in the digestive tract, and **vitamins** A and C, both powerful antioxidants that improve immune function and increase resistance to infection. Zinc is another nutrient essential to immune system functioning.

Nutritional concerns

Immune system function requires ingesting certain essential nutrients and avoiding others that depress immunity. A diet that improves immune system functioning includes fresh fruits and vegetables, as many eaten raw as possible to provide necessary enzymes; whole grain cereals, brown rice, and whole grain pasta

KEY TERMS

Acquired immune deficiency syndrome (AIDS)—A disease associated with infection by the human immunodeficiency virus (HIV) that attacks the immune system.

Antibiotics—Drugs that are designed to kill or inhibit the growth of the bacteria that cause infections.

Antibody—A special protein made by the body's immune system as a defense against foreign material (bacteria, viruses, etc.) that enters the body. It is uniquely designed to attack and neutralize the specific antigen that triggered the immune response.

Antigen—A substance (usually a protein) identified as foreign by the body's immune system, triggering the release of antibodies as part of the body's immune response.

Computed tomography (CT)—An imaging technique in which cross-sectional x rays of the body are compiled to create a three-dimensional image of the body's internal structures; also called computed axial tomography.

Culture—A test in which a sample of body fluid is placed on materials specially formulated to grow microorganisms. A culture is used to learn what type of bacterium is causing infection.

Immunosuppression—Techniques used to prevent transplant graft rejection by the recipient's immune system.

Magnetic resonance imaging (MRI)—An imaging technique that uses a large circular magnet and radio waves to generate signals from atoms in the body. These signals are used to construct detailed images of internal body structures and organs, including the brain.

Nonsteroidal anti-inflammatory drugs (NSAIDs)—A group of drugs, including aspirin, ibuprofen, and naproxen, that are taken to reduce fever and inflammation and to relieve pain. They work primarily by interfering with the formation of prostaglandins, enzymes implicated in pain and inflammation.

Toxin—A poisonous substance usually produced by a microorganism or plant.

Ultrasonography—A medical test in which sound waves are directed against internal structures in the body. As sound waves bounce off the internal structure, they create an image on a video screen. Ultrasonography is often used to diagnose fetal abnormalities, gallstones, heart defects, and tumors. Also called ultrasound imaging.

for essential vitamins, **minerals**, and fiber; and non-meat sources of protein such as nuts, seeds, tofu, legumes (beans), and eggs. Fish, fowl, and lean meats can be consumed in small amounts. Sweets, especially if sweetened with refined sugars, should be reduced or avoided altogether. A diet high in fats and processed foods made with refined flours and sugars can actually suppress the immune system. Alcohol and **caffeine** should be avoided.

Parental concerns

When FUO is present, parents may be concerned that effective treatment will be delayed by waiting for a diagnosis, which may depend on waiting for the results of diagnostic tests. Depending on the extent of the fever, keeping the child quiet and in bed is probably recommended until all results are available and a definitive diagnosis is made. The doctor will undoubtedly recommend giving clear liquids as often as possible to avoid dehydration from the high body temperature. After the tests have been performed, the physician may recommend fever-reducing medications such as acetaminophen or ibuprofen. If the fever is high enough for concern, physicians may prescribe a broad spectrum antibiotic as initial treatment rather than waiting for the results of all diagnostic tests. When results are available, the physician will likely prescribe a new medication most appropriate for the diagnosis.

See also HIV infection and AIDS; Rheumatic fever.

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Fever seizures
see **Febrile seizures**

Fifth disease

Definition

Fifth disease is a mild childhood illness caused by the human parvovirus B19 that causes flu-like symptoms and a rash. It is called fifth disease because it was fifth on a list of common childhood illnesses that are accompanied by a rash, including **measles**, **rubella** (or German measles), **scarlet fever** (or scarlatina), and scarlatinella, a variant of scarlet fever.

Description

The Latin name for fifth disease is *erythema infectiosum*, meaning infectious redness. It is also called the "slapped cheek disease" because, when the bright red rash first appears on the cheeks, it looks as if the face has been slapped. Anyone can get the disease, but it occurs more frequently in school-aged children. The disease is usually mild, and both children and adults usually recover quickly without complications. Some individuals exhibit no symptoms and never even feel ill.

Transmission

The virus that causes fifth disease lives in the nose, mouth, and throat of an infected person; therefore, the virus can be spread through the air by coughing and sneezing. It can also be spread through shared drinking glasses and eating utensils.

Demographics

Fifth disease is very common in children between the ages of five and 15. Studies show that although 40 percent to 60 percent of adults worldwide have laboratory evidence of a past parvovirus B19 infection, most of these adults cannot remember having had symptoms of fifth disease. This fact leads medical experts to believe that most people with parvovirus B19 infection have either very mild symptoms or no symptoms at all.

Fifth disease occurs everywhere in the world. Outbreaks of parvovirus tend to occur in the late winter and early spring, but there may also be sporadic cases of the disease any time throughout the year.

In households where a child has fifth disease, another **family** member who has not previously had fifth disease has about a 50 percent chance of also getting the infection, while classmates of a child with fifth disease have about a 60 percent chance of getting the disease.

Causes and symptoms

Fifth disease is caused by the human parvovirus B19, a member of the Parvoviridae family of viruses, that lives in the nose, mouth, and throat of an infected person. Because the virus needs a rapidly dividing cell in order to multiply, it attacks the red blood cells of the body. Once infected, a person is believed to be immune to reinfection.

Symptoms may appear four to 28 days after being exposed to the virus, with the average time to onset ranging from 16 to 17 days. Initial symptoms are flu-like and include **headache**, stuffy or runny nose, body ache, **sore throat**, a mild fever of 101°F (38.3°C), and chills. It is at this time, prior to the development of the rash, that individuals are contagious. These symptoms last for two to three days. Other symptoms that sometimes occur with fifth disease include swollen glands, red eyes, and **diarrhea**. In older teens and adults, fifth disease may be followed by joint swelling or **pain** in the hands, wrists, knees, and ankles, lasting from a few months to several years.

In children, especially those under the age of ten, a bright red rash that looks like a slap mark develops suddenly on the cheeks a few days after the original symptoms were experienced. The rash may be flat or raised and may or may not be itchy. Sometimes, the rash spreads to the arms, legs, and trunk, where it has a lace-like or net-like appearance as the centers of the blotches fade. By the time the rash appears, individuals are no longer infectious. On average, the rash lasts for ten to 11 days but may last for as long as five to six weeks. The rash may fade away and then reappear upon exposure to sunlight, hot baths, emotional distress, or vigorous **exercise**.

The virus causes the destruction of red blood cells and, therefore, a deficiency in the oxygen-carrying capacity of the blood (anemia) can result. In healthy children, the anemia is mild and only lasts a short while. In children with weakened immune systems, however, either because they have a chronic disease like **AIDS** or **cancer** (immunocompromised) or are receiving medication to suppress the immune system (immunosuppressed), such as organ transplant recipients, this anemia can be severe and last long after the infection has subsided. Symptoms of anemia include fatigue, lack of color, lack of energy,

KEY TERMS

Anemia—A condition in which there is an abnormally low number of red blood cells in the bloodstream. It may be due to loss of blood, an increase in red blood cell destruction, or a decrease in red blood cell production. Major symptoms are paleness, shortness of breath, unusually fast or strong heart beats, and tiredness.

Antibody—A special protein made by the body's immune system as a defense against foreign material (bacteria, viruses, etc.) that enters the body. It is uniquely designed to attack and neutralize the specific antigen that triggered the immune response.

Immunocompromised—A state in which the immune system is suppressed or not functioning properly.

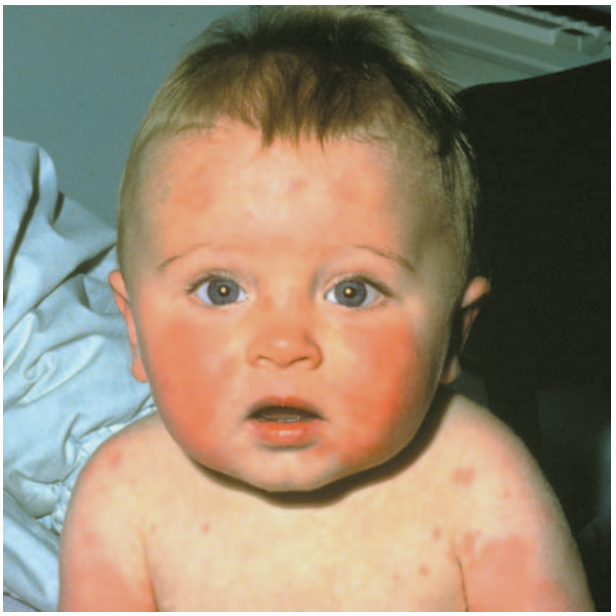
Immunosuppressed—A state in which the immune system is suppressed by medications during the treatment of other disorders, like cancer, or following an organ transplantation.

Reye's syndrome—A serious, life-threatening illness in children, usually developing after a bout of flu or chickenpox, and often associated with the use of aspirin. Symptoms include uncontrollable vomiting, often with lethargy, memory loss, disorientation, or delirium. Swelling of the brain may cause seizures, coma, and in severe cases, death.

Sickle cell anemia—An inherited disorder in which red blood cells contain an abnormal form of hemoglobin, a protein that carries oxygen. The abnormal form of hemoglobin causes the red cells to become sickle-shaped. The misshapen cells may clog blood vessels, preventing oxygen from reaching tissues and leading to pain, blood clots and other problems.

and shortness of breath. Some individuals with **sickle cell anemia**, iron deficiency, a number of different hereditary blood disorders, and those who have received bone marrow transplantations may be susceptible to developing a potentially life-threatening complication called a transient aplastic crisis in which the body is temporarily unable to form new red blood cells.

In very rare instances, the virus can cause inflammation of different areas of the body, including the brain (**encephalitis**), the covering of the brain and spinal cord (**meningitis**), the lungs (pneumonitis), the liver (hepatitis), and the heart muscle (myocarditis). The virus can also aggravate symptoms for people with an autoimmune disease called systemic lupus erythematosus.



Infant with a rash caused by fifth disease, or erythema infectiosum. *(Custom Medical Stock Photo, Inc.)*

Although no association with an increased number of birth defects has been demonstrated, there is concern that infection during the first three months of pregnancy may lead to a slight increase in the number of miscarriages due to the fetus developing severe anemia. There is also some concern that infection later in pregnancy may involve a very small risk of premature delivery or stillbirth. As a result, women who get fifth disease while they are pregnant should be monitored closely by a physician.

When to call the doctor

Parents should call their child's doctor if their child develops a rash, especially if the rash is widespread over the child's body or if the rash is accompanied by other symptoms.

Diagnosis

Fifth disease is usually suspected based on a patient's symptoms, including the typical appearance of the bright red rash on the cheeks, patient history, age, and the time of year. The physician will also exclude other potential causes for the symptoms and rash, including rubella, **infectious mononucleosis**, bacterial infections such as **Lyme disease**, allergic reactions, and lupus.

In addition, there is a blood test for fifth disease, but it is generally used only for pregnant women and for people who have weakened immune systems or who suffer from blood disorders, such as sickle cell anemia. The test involves measuring for a particular antibody or protein that the body produces in response to infection with the human parvovirus B19. The test is 92–97 percent specific for this disease.

Because fifth disease can pose problems for an unborn fetus exposed to the disease through the mother, testing for the disease may be conducted while a fetus is still in the uterus. This test uses fluid collected from the sac around the fetus (amniotic fluid) instead of blood to detect the viral DNA.

Prognosis

Generally, fifth disease is mild, and patients tend to improve without any complications. In cases where the patient is either immunocompromised or immunosuppressed, a life-threatening aplastic crisis can occur. With prompt treatment, however, the prognosis is good. Mothers who develop the infection while pregnant can pass the infection on to their fetus and thus stand an increased risk of miscarriage and stillbirth. There are tests and treatments, however, that can be performed on the fetus while still in the uterus that can reduce the risk of anemia or other complications.

Prevention

As of 2004, there was no vaccine against fifth disease. Avoiding contact with persons who exhibit symptoms of a cold and maintaining good personal hygiene by regularly washing hands may minimize the chances of an infection. Pregnant women should avoid exposure to persons infected with the disease and notify their obstetricians immediately if they are exposed so that they can be tested and monitored closely.

Parental concerns

Generally parents should not be concerned if their child contracts fifth disease. However, if the mother is pregnant, she should contact her doctor.

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Fighting *see* **Aggressive behavior**

Fine motor skills

Definition

Fine motor skills generally refer to the small movements of the hands, wrists, fingers, feet, toes, lips, and tongue.

Description

Motor skills are actions that involve the movement of muscles in the body. They are divided into two groups: **gross motor skills**, which include the larger movements of arms, legs, feet, or the entire body (**crawling**, running, and jumping); and fine motor skills, which are smaller actions, such as grasping an object between the thumb and a finger or using the lips and tongue to taste objects. Both types of motor skills usually develop together, because many activities depend on the coordination of gross and fine motor skills.

Infancy

The hands of newborn infants are closed most of the time and, like the rest of their bodies, they have little control over them. If their palms are touched, they will make a very tight fist, but this is an unconscious reflex action called the Darwinian reflex, and it disappears within two to three months. Similarly, infants will grasp at an object placed in their hands, but without any awareness that they are doing so. At some point their hand muscles relax, and they drop the object, equally unaware that they have let it fall. Babies may begin flailing at

objects that interest them by two weeks of age but cannot grasp them. By eight weeks, they begin to discover and **play** with their hands, at first solely by touch, and then, at about three months, by sight as well. At this age, however, the deliberate grasp remains largely undeveloped.

Hand-eye coordination begins to develop between the ages of two and four months, inaugurating a period of trial-and-error practice at sighting objects and grabbing at them. At four or five months, most infants can grasp an object that is within reach, looking only at the object and not at their hands. Referred to as ''top-level reaching,'' this achievement is considered an important milestone in fine motor development. At the age of six months, infants can typically hold on to a small block briefly, and many have started banging objects. Although their grasp is still clumsy, they have acquired a fascination with grabbing small objects and trying to put them in their mouths. At first, babies will indiscriminately try to grasp things that cannot be grasped, such as pictures in a book, as well as those that can, such as a rattle or ball. During the latter half of the first year, they begin exploring and testing objects before grabbing, touching them with an entire hand and, eventually, poking them with an index finger.

One of the most significant fine motor accomplishments is the pincer grip, which typically appears at about 12 months. Initially, infants can only hold an object, such as a rattle, in their palm, wrapping their fingers (including the thumb) around it from one side. This awkward position is called the palmar grasp, which makes it difficult to hold on to and manipulate the object. By the age of eight to 10 months, a finger grasp begins, but objects can only be gripped with all four fingers pushing against the thumb, which still makes it awkward to grab small objects. The development of the pincer grip—the ability to hold objects between the thumb and index finger—gives the infant a more sophisticated ability to grasp and manipulate objects and also to deliberately drop them. By about the age of one, an infant can drop an object into a receptacle, compare objects held in both hands, stack objects, and nest them within each other.

Toddlerhood

Toddlers develop the ability to manipulate objects with increasing sophistication, including using their fingers to twist dials, pull strings, push levers, turn book pages, and use crayons to produce crude scribbles. Dominance of either the right or left hand usually emerges during this period as well. Toddlers also add a new dimension to touching and manipulating objects by simultaneously being able to name them. Instead of only random scribbles, their **drawings** include patterns, such as circles. Their play with blocks is more elaborate and purposeful than that of infants, and they can stack as many as six blocks. They are also able to fold a sheet of paper in half (with supervision), string large beads, manipulate snap **toys**, play with clay, unwrap small objects, and pound pegs.

Preschool

The more delicate tasks facing **preschool** children, such as handling silverware or tying shoelaces, represent more challenge than most of the gross motor activities learned during this period of development. The central nervous system is still in the process of maturing sufficiently for complex messages from the brain to get to the child's fingers. In addition, small muscles tire more easily than large ones, and the short, stubby fingers of preschoolers make delicate or complicated tasks more difficult. Finally, gross motor skills call for energy, which is boundless in preschoolers, while fine motor skills require patience, which is in shorter supply. Thus, there is considerable variation in fine motor development among this age group.

School age

By the age of five, most children have clearly advanced beyond the fine motor skill development of the preschool age. They can draw recognizably human figures with facial features and legs connected to a distinct trunk. Besides drawing, five-year-olds can also cut, paste, and trace shapes. They can fasten visible buttons (as opposed to those at the back of clothing), and many can tie bows, including shoelace bows. Their right- or left-handedness is well established, and they use the preferred hand for writing and drawing.

School-age children six to 12 years old should have mastered hand and eye coordination. Early school age children should be able to use eating utensils and other tools, be able to help with household chores, such as sweeping, mopping, and dusting; care for pets; draw, paint, and engage in making crafts; and begin developing writing skills. Children will continue to fine-tune their fine motor skills through **adolescence** with such activities as **sports**, crafts, hobbies, learning musical instruments, computer use, and even **video games**.

Helping a child succeed in fine motor tasks requires planning, time, and a variety of play materials. Fine motor development can be encouraged by activities that youngsters enjoy, including crafts, puzzles, and playing with building blocks. Helping parents with everyday domestic activities, such as baking, can be fun for the child in addition to helping the child develop fine motor skills. For example, stirring batter provides a good work-

KEY TERMS

Beery-Buktenica Test—A test that identifies problems with visual perception, fine motor skills (especially hand control), and hand-eye coordination.

Darwinian reflex—An unconscious action in infants in which if a palm is touched, the infant makes a very tight fist. This instinct disappears within two to three months.

Developmental coordination disorder—A disorder of motor skills.

Gross motor skills—The abilities required to control the large muscles of the body for walking, running, sitting, crawling, and other activities. The muscles required to perform gross motor skills are generally found in the arms, legs, back, abdomen, and torso.

Hand-eye coordination—The ability to grasp or touch an object while looking at it.

Lincoln-Oseretsky Motor Development Scale—A test that assesses the development of motor skills.

Palmar grasp—A young infant's primitive ability to hold an object in the palm by wrapping fingers and thumb around it from one side.

Pincer grip—The ability to hold objects between thumb and index finger, which typically develops in infants between 12 and 15 months of age.

Top-level reaching—The ability of an infant to grasp an object that is within reach, looking only at the object and not at the hands. Typically develops between four and five months of age.

out for the hand and arm muscles, and cutting and spooning out cookie dough requires hand-eye coordination. Even a computer keyboard and mouse can provide practice in finger, hand, and hand-eye coordination. Because the development of fine motor skills plays a crucial role in school readiness and **cognitive development**, it is considered an important part of the preschool curriculum.

Fine motor skills

Age	Skill
One to three months	Reflexively grasps finger or toy placed in hand.
Three months	Grasping reflex gone. Briefly holds small toy voluntarily when it is placed in the hand.
Four months	Holds and shakes rattle. Brings hands together to play with them. Reaches for objects but frequently misses them.
Five months	Grasps objects deliberately. Splashes water. Crumples paper.
Six months	Holds bottle. Grasps at own feet. May bring toes to mouth.
Seven months	Transfers toy from hand to hand. Bangs objects on table. Puts everything into the mouth. Loves playing with paper.
Nine months	Able to grasp small objects between thumb and forefinger.
Ten months	Points at objects with index finger. Lets go of objects deliberately.
Eleven months	Places object into another's hand when requested, but does not release.
Twelve months	Places and releases object into another's hand when requested. Rolls ball on floor. Starts to hold crayon and mark paper with it.
Fifteen months	Builds tower of two blocks. Repeatedly throws objects on floor. Starts to be able to take off clothing, starting with shoes.
Eighteen months	Builds tower of three blocks. Starts to feed self well with spoon. Turns book pages two or three at a time. Scribbles on paper.
Two years	Builds tower of six or seven blocks. Turns book pages one at a time. Turns door knobs and unscrews jar lids. Washes and dries hands. Uses spoon and fork well.
Two and a half years	Builds tower of eight blocks. Holds pencil between fingers instead of grasping with fist.
Three years	Builds tower of nine or ten blocks. Puts on shoes and socks. Can button and unbutton. Carries containers with little spilling or dropping.
Four years	Dresses self except for tying. Cuts with scissors, but not well. Washes and dries face.
Five years	Dresses without help. Ties shoes. Prints simple letters.

SOURCE: *Miller-Keane Encyclopedia and Dictionary of Medicine, Nursing, and Allied Health, 5th ed.* and Child Development Institute, http://www.childdevelopmentinfo.com.

(Table by GGS Information Services.)

Common problems

Fine motor skills can become impaired in a variety of ways, including injury, illness, **stroke**, and congenital deformities. An infant or child up to age five who is not developing new fine motor skills for that age may have a developmental disability. These problems can include major health conditions including **cerebral palsy**, **mental retardation**, blindness, deafness, and diabetes. Children with delays in fine motor skills development have difficulty controlling their coordinated body movements, especially with the face, hands, and fingers. Signs of fine motor skills delays include a failure to develop midline

A toddler demonstrates his fine motor skills by grasping and munipulating building blocks. *(© S. Villeger/Explorer/ Photo Researchers, Inc.)*

orientation by four months, reaching by five months, transferring objects from hand to hand by six months, a raking grasp by eight months, a mature pincer grip by one year, and index finger isolation by one year.

Developmental coordination disorder is a disorder of motor skills. A person with this disorder has a hard time with things like riding a bike, holding a pencil, and throwing a ball. People with this disorder are often called clumsy. Their movements are slow and awkward. People with developmental coordination disorder may also have a hard time completing tasks that involve movement of muscle groups in sequence. For example, such a person might be unable to do the following in order: open a closet door, get out a jacket, and put it on. It is thought that up to 6 percent of children may have developmental coordination disorder, according to the 2002 issue of the annual journal *Clinical Reference Systems*. The symptoms usually go unnoticed until the early years of elementary school. It is usually diagnosed in children who are between five and 11 years old.

Parental concerns

Parents, teachers, and primary caregivers need to have a clear understanding of how young children develop fine motor skills and the timetable for development of the skills.

Fine motor skills development tests

The Lincoln-Oseretsky Motor Development Scale is an individually administered test that assesses the development of motor skills in children and adults. Areas covered include fine and gross motor skills, finger dexterity and speed, and hand-eye coordination. The test consists of 36 tasks arranged in order of increasing difficulty. These include walking backwards, standing on one foot, touching one's nose, jumping over a rope, throwing and catching a ball, putting coins in a box, jumping and clapping, balancing on tiptoe while opening and closing one's hands, and balancing a rod vertically. Norms have been established for each part of the test for children aged six to 14.

The **Beery-Buktenica Test**, also known as VMI or Developmental Test of Visual-Motor Integration, is designed for individuals two years of age through adult. The text identifies problems with visual perception, fine motor skills (especially hand control), and hand-eye coordination. It is usually administered individually but can also be given in groups. The child is given a booklet containing increasingly complex geometric figures and asked to copy them without any erasures and without rotating the booklet in any direction. The test is given in two versions: the Short Test Form, containing 15 figures, is used for ages three through eight; the Long Test Form, with 24 figures, is used for older children, adolescents, and adults with **developmental delay**. A raw score based on the number of correct copies is converted based on norms for each age group, and results are reported as converted scores and percentiles. The test is not timed but usually takes 10 to 15 minutes to administer.

When to call the doctor

Some symptoms of impaired fine motor skills may appear up to age two. These symptoms include having a difficult time sitting up or raising the head, being unable to stand without help or having a very hard time standing without help, being unable to crawl or having a very hard time crawling, and walking very late or having a hard

time walking. Other symptoms usually appear during the preschool or grade school years. These may include the child having difficulty holding a pencil or drawing, throwing a ball, riding a bicycle, playing sports, having a hard time with clothes that have buttons or zippers, having poor handwriting, and being clumsy.

Children with any one or combination of these symptoms should be seen by a pediatrician who specializes in motor skills development delays. Children who lose previously acquired motor skills should also be seen by a doctor. There are many ways to address fine motor skills impairment, such as physical therapy. This type of therapy can include treating the underlying cause, strengthening muscles, and learning how to compensate for impaired movements.

See also Gross motor skills.

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Ken R. Wells

Fingertip injuries

Definition

Fingertip injuries include any **wounds** to the area at tip of the finger. They range from a simple bruise or scrape to having the fingertip taken off. Fingertip injuries occur frequently in infants and children because hands are used to explore surroundings and **play**.

Description

Fingers each have three bones (phalanges); the thumb has two. The fingertip consists of the uppermost phalanx with surrounding muscle, tissue, nerves, and nail. A fingertip is a highly complex structure, with many specialized features, one of which is a rich network of sensory nerves. The fingernail is called the nail plate. Underneath the nail plate is the nail bed, the mostly pink tissue seen under the nail. The pulp is the area of skin opposite the fingernail and is usually very vulnerable to injury. Fingertip injuries are extremely common and varied. Blunt or crush injuries can cause bleeding under the nail plate (subungual hematomas), which can be very painful. Nails can also be torn off (nail avulsions), and the fingertip bone can be broken (fracture). Sharp or shearing injuries from knives and glass result in cuts (lacerations) and punctures. Occasionally, the end of the fingertip is torn off (amputated). When portions of the fingertip are missing, the injury is described as a partial

amputation. When the finger is cut more than halfway through, the injury is described as a subtotal amputation. **Burns** and **frostbite** also commonly injure fingertips.

Demographics

In the United States, fingertip injuries account for approximately two-thirds of hand injuries in children. Damage to the nail bed is reported to occur in 15–24 percent of fingertip injuries. A Florida study of hand injuries in children conducted in 2002 showed that the most frequent hand injury setting was outdoors (47%). The most frequent injuries were lacerations (30%), followed by **fractures** (16%). The fingers were the most commonly injured part of the hand, particularly the thumb (19%), and fingertips were involved in 21 percent of cases. Children younger than two years suffered fingertip injuries mostly inside the home.

Causes and symptoms

In children, fingertip injuries are the result of accidents occurring at play or in the home. They involve cuts, by glass, knives, or other sharp objects; or crushing injuries, as when the fingertip gets caught in a door or window or is hit by a hammer or rock. Symptoms depend on the nature of the fingertip injury and may include some of the following:

- **pain**
- bleeding
- swelling
- tissue loss
- movement restriction
- amputation

When to call the doctor

Parents should always see a doctor right away if their child injures the tip of a finger or thumb. Fingertips contain many nerves and are extremely sensitive. Without prompt and proper care, a fingertip injury can disrupt the complex function of the hand, resulting in permanent deformity and disability. Bleeding from minor fingertip cuts often stop on its own with direct pressure applied to the wound with a clean cloth. If continuous pressure does not slow or stop the bleeding after 15 minutes, an emergency room visit is indicated.

Diagnosis

The treating physician begins a diagnosis by carefully evaluating the fingertip injury. Bones and joints are examined for motion and tenderness. Nerves are examined for sensory (feeling sensations) and motor (movement) function. As part of injury diagnosis, the treating physician also considers the following factors:

- nature of injury (crush or sharp)
- nail or nail bed involvement
- bone involvement
- viability the tip
- presence of foreign bodies

X rays may be required either to assess alignment of fingertip phalanx fractures or to detect presence of foreign bodies.

Treatment

Doctors provide individualized treatment for fingertip injuries based on the nature and extent of the injury. The treating physician usually gives an injection (digital block anesthesia) to stop pain in the affected finger. Then he or she may rinse the wound (irrigate) with a saline solution, inspect it for exposed bone, soft tissue loss, and nail or nail bed injury. Infected or dead tissue or foreign materials are also removed (debridement) to reduce risk of infection. If blood has accumulated under the nail (subungual hematoma), the doctor may drain it by piercing through the fingernail. **Antibiotics** and a **tetanus** shot may also be prescribed. The goal of treatment is a painless fingertip that has durable and feeling skin. A normal fingertip has sensation without pain, stable pulp padding, and an acceptable appearance. The hand should be able to pinch, grip, and perform other normal functions. In cases of severe injury and whenever possible, the doctor will try to maintain the fingertip's length and appearance and preserve its fingernail.

Surgery

Fingertip injuries often require surgical treatment, usually performed with local anesthesia. Fingertip repair surgery includes the following:

- Sutures: Laceration wounds are stitched (sutured) after application of a digital block.
- Nail bed surgery: If the nail bed is injured, the nail plate may be removed, the bed carefully debrided and repaired. The nail plate is then reinserted and the injury dressed. A finger splint may be applied.
- Fingertip amputation: Various methods are used for amputation injuries including simple amputation of the fingertip, full or partial skin grafts, and skin flaps.

KEY TERMS

Avulsion—The forcible separation of a piece from the entire structure.

Debridement—The surgical removal of dead tissue and/or foreign bodies from a wound or cut.

Laceration—A cut or separation of skin or other tissue by a tremendous force, producing irregular edges. Also called a tear.

Nail bed—The layer of tissue underneath the nail.

Phalanx—Plural, plananges. Any of the digital bones of the hand or foot. Humans have three phalanges to each finger and toe with the exception of the thumb and big toe which have only two each.

Sensory nerves—Sensory or afferent nerves carry impulses of sensation from the periphery or outward parts of the body to the brain and spinal cord.

Subungual hematoma—Accumulation of blood under a nail.

Tetanus—A potentially fatal infection caused by a toxin produced by the bacterium *Clostridium tetani*. The bacteria usually enter the body through a wound and the toxin they produce affects the central nervous system causing painful and often violent muscular contractions. Commonly called lockjaw.

- Reconstructive flap surgery: If a fingertip injury exposes bone, and there is not enough tissue available on the fingertip to close it, the surgeon may need to shorten the bone and transfer a piece of skin and underlying fat and blood vessels from a healthy part of the patient's body to the injury site. A bulky dressing and splint supports the hand after surgery, with uninjured fingers left free to **exercise**. A second operation may be necessary after a few weeks to detach the flap from its origin.
- Fractured phalanx: In case of fingertip bone fracture, the bone is straightened and put in a splint or cast. A temporary metal pin may also be inserted into the bone to hold it in place until bone growth occurs. If bone is partly missing, the finger may be shortened or the surgeon may use a bone graft.

Prognosis

The outcome depends on the extent of traumatic damage to the fingertip and whether surgery is required. Small wounds to a fingertip's skin and pulp usually close on their own with complete healing within three to five weeks. In 60 percent of patients with subungual hematomas that involve more than 50 percent of the nail surface, laceration is repairable. This prognosis increases to over 95 percent when an associated fracture of the phalanx is present. Nail lacerations that are not treated may cause nail deformities. The average healing time for fingertip amputation is 21–27 days. In many cases, fingertip repair surgery gives back a large degree of feeling and function. However, infection, poor healing, loss of feeling or motion, blood clots, and adverse reactions to anesthesia are all possible complications of surgery. Mild to severe pain and sensitivity to cold following treatment for a fingertip amputation sometimes occur.

Prevention

Children should at all times be supervised in their activities. Parents should not allow children to use sharp tools and knives without supervision. They should teach them to always wash and thoroughly dry any tool or knife before use. Children should also be taught how to use knives properly, to always cut away from themselves, and to cut in small, controlled strokes.

Parental concerns

Parents should be aware that recovery from a serious fingertip injury may take months and require hand therapy. This may include hand exercises to improve movement and strength, heat and **massage therapy**, electrical nerve stimulation, splinting, traction, and special wrappings to control swelling.

See also Frostbite and frostnip.

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Monique Laberge, Ph.D.

5p minus syndrome *see* **Cri du chat syndrome**

Flu *see* **Influenza**

Flu vaccine

Definition

The flu vaccine protects a person against getting **influenza**, caused by the influenza virus. It is administered either by injection or by inhalation.

Description

There are two types of flu vaccine. Live attenuated influenza vaccine (LAIV) was first approved for use in 2003. It contains live, but weakened, influenza virus and is administered as a nasal spray. Inactivated influenza vaccine contains killed viruses and is given by intramuscular injection.

Because influenza changes from year to year, a flu shot, unlike other types of vaccinations, is required every year. Each year, the United States Centers for Communicable Disease Control predicts the strains of influenza that are likely to appear in the coming year. Vaccine manufacturers then make products to protect against these types of flu. These vaccines only protect against the type of influenza viruses from which they are made. They are usually of no value against other types of influenza virus.

There are three types of influenza virus: A, B, and C. They differ by the proteins on their outer surface. Type A viruses are found in many different animals, including ducks, chickens, pigs, whales, horses, and seals. Influenza B viruses circulate widely, but only among humans. Type C viruses, also only among humans, cause a very mild infection, and flu vaccines do not include protection against type C influenza.

General use

Influenza is a contagious illness. Every year, 10 to 20 percent of the U.S. population gets the flu, and over 100,000 people are hospitalized because of influenza. Symptoms of flu include **fever**, **headache**, extreme tiredness, dry **cough**, **sore throat**, runny or stuffy nose, and muscle aches. When children get the flu, they often complain of **nausea** and have **vomiting** and **diarrhea**, although these problems are less common in older children and adults. While influenza virus vaccines cannot give complete protection against flu, they greatly reduce the risk of flu-like infections, reduce the risk of **hospitalization**, and shorten the duration of these infections.

All healthy children between the ages of six months and 23 months should receive influenza vaccine because these young children do not yet have fully developed immune systems and are at increased risk of getting the flu and requiring hospital treatment. Children over the age of six months should also receive the vaccine if they have certain medical risk factors:

- asthma or other lung disease
- congenital heart disease with defects that require medications or surgery or other heart disease
- glomerulonephritis, kidney failure, or other kidney disease
- diabetes or other metabolic disease
- sickle cell disease or other anemia
- immune system problems or other anemia
- juvenile rheumatoid arthritis or any other disease needing aspirin therapy

KEY TERMS

Attenuated—A live but weakened microorganism that can no longer produce disease.

Guillain-Barré syndrome—Progressive and usually reversible paralysis or weakness of multiple muscles usually starting in the lower extremities and often ascending to the muscles involved in respiration. The syndrome is due to inflammation and loss of the myelin covering of the nerve fibers, often associated with an acute infection. Also called acute idiopathic polyneuritis.

Influenza—An infectious disease caused by a virus that affects the respiratory system, causing fever, congestion, muscle aches, and headaches.

Vaccine—A substance prepared from a weakened or killed microorganism which, when injected, helps the body to form antibodies that will prevent infection by the natural microorganism.

Otherwise healthy children above the age of 23 months may have a flu shot simply to reduce the risk of influenza.

Side effects

Most people have no adverse effects from a flu shot other than soreness at the injection site that lasts a few days. The greatest risk is an allergic reaction, which can be serious, but this is very rare. A low fever occasionally occurs.

Interactions

Vaccines should not be given to patients taking antibiotic drugs. While both flu vaccines may be administered at the same time as other vaccines, if two vaccines are not given at exactly the same time, they should be spaced four weeks apart.

Flu vaccine should not be given to the following groups:

- children with a severe allergic reaction to chickens or egg protein
- children who have exhibited a moderate to severe reaction after a previous influenza shot
- children who have ever been paralyzed due to Guillain-Barré syndrome
- children who are sick with anything beyond a slight cold

Parental concerns

Unlike the swine flu vaccine used in 1976, flu vaccines in the last decades of the twentieth century and early 2000s have shown no association with Guillain-Barré syndrome (GBS) in children.

Both types of influenza vaccine may be administered to **family** members of immunosuppressed patients, as long as the patients do not require a protected environment, although the killed virus vaccine is preferred for this purpose. Family members of patients who do require a protected environment, such as people who have had a stem cell transplant, should only receive the killed virus vaccine (injection).

Parents, siblings, and other people who will be in close contact with a sick child or adult should have a flu shot whether or not they need it for their own protection.

The risk of a local reaction to a flu injection increases with age. Children under the age of 13 years rarely have reactions to the flu vaccine. Over the age of 13 years, about 10 percent of children have a reaction to the shot.

Since flu season is usually in late fall or early winter, a flu shot should be given in the autumn, so that full immunity can be built up. However, it is never too late to get a flu shot.

See also Influenza; Vaccination.

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Fluoridation

Definition

Fluoridation is the addition of fluoride to water supplies to help prevent **tooth decay**.

Description

The element fluorine is the seventeenth most abundant element in the earth's crust. It occurs as fluoride ion in combination with other elements such as sodium. Most water supplies naturally contain low levels of fluoride. In much of the United States, as well as in other parts of the world, fluoride is added to community water systems to bring fluoride levels up to the recommended amount for preventing teeth decay: 0.7–1.2 parts of fluoride to 1 million parts of water (parts per million or ppm). The levels of naturally occurring fluoride in fresh water range from less than 0.1 ppm to more than 13 ppm. Seawater contains about 1.5 ppm. As of 2000, about 162 million Americans—two-thirds of the population—were served by fluoridated water systems.

Mode of action

Systemic fluorides, including fluoridated water and prescription fluoride supplements supplied as tablets, drops, or lozenges, can be incorporated into the enamel of children's developing teeth. The enamel that covers the crown, the part of the tooth that is above the gum, is made of a substance called hydroxyapatite. When enough fluoride from water, supplements, food, or other sources enters the bloodstream and reaches the teeth while the enamel is forming, the fluoride can replace a piece of the hydroxyapatite molecule to form fluorapatite. Thus fluoride becomes part of the tooth enamel. Fluoride makes the tooth more resistant to acids produced by the bacteria that cause tooth decay. These acids dissolve the enamel, causing cavities. Fluoride in the enamel appears to do the following:

- make the enamel less susceptible to bacterial acids
- reduce the bacteria's ability to produce acid
- reduce the number of bacteria in plaque deposits

It is unlikely that sufficient fluoride will be incorporated into the enamel throughout the years of crown formation. With optimally fluoridated water (1 ppm) as the primary source of fluoride, a child would have to drink two quarts of water every day for 12 to 14 years to incorporate fluoride into all of the baby and adult teeth as they form. The child would be ingesting about 2 mg. of fluoride daily.

Topical fluorides are applied directly to the surfaces of fully-formed teeth. Fluoridated water acts as a topical—as well as a systemic—source of fluoride. Other topical fluorides include fluoridated toothpastes and mouthwashes and fluoride gels that are applied to children's teeth at dental examinations. Topical fluoride is the most effective mineral for renewing or remineralizing the surface layers of enamel and dentin as they wear out and are eaten away by acids from food and bacteria. Fluoride remineralization makes the tooth surface more resistant to decay and reverses early decay processes. Thus topical fluorides help prevent decay in both children and adults. Systemic fluorides also can provide topical protection because they are incorporated into the saliva that bathes the teeth.

Sources of fluoride

Fluoridated water is a major source of fluoride. Most bottled water contains only trace amounts of fluoride. Filtered water and well water vary greatly in their fluoride content. Children who drink water that is low in fluoride may be given fluoride supplements.

Fluoride occurs in many different foods and is also added to some foods. Fruits and vegetables may contain more than 0.2 mg of fluoride per serving, depending on where they were grown and whether fluoridated water was used for irrigation and processing. Most seafood is high in fluoride. The amount of fluoride in beverages depends on the amount of fluoride in the water used to make them. Many **vitamins** and medicines also contain fluoride. Most baby food is made with nonfluoridated water.

Fluoridated toothpastes and mouthwashes contain high amounts of fluoride. A tube of fluoridated toothpaste may contain as much as 1 to 2 gm of fluoride. Nonprescription mouthwashes can contain up to 120 mg of fluoride. Children between the ages of two and six swallow about 33 percent of the toothpaste they use; children between seven and 15 swallow about 20 percent. The average child using the typical amount of fluoridated toothpaste will swallow or absorb 0.5 to 1.0 mg. of fluoride per brushing. Much of this fluoride is excreted.

It is believed that fluoridated water is between 20 and 60 percent effective in preventing cavities in children and adults. Early studies suggested that water fluoridation was eliminating tooth decay in children. However, other factors are recognized in the early 2000s as having contributed to the decline in dental cavities. The widespread use of fluoridated toothpastes and mouthwashes has increased children's sources of fluoride significantly. Furthermore, both children's and adults' knowledge about dental care and dental hygiene has improved in the last quarter of the twentieth century.

General use

History

In the early twentieth century a young dentist in Colorado Springs, Colorado, named Frederick McKay, noticed that many local residents had brown stains on their permanent teeth and that their teeth were surprisingly resistant to decay. McKay eventually discovered that this "mottling"—as he called it—resulted from high levels of naturally occurring fluoride in the drinking water.

The first fluoridation of a public water system took place in Grand Rapids, Michigan, in 1945. By the 1950s and 1960s increasing numbers of communities were fluoridating their water using by-products from the phosphate fertilizer industry. The practice became mired in controversy, and it remains so in the first decade of the twenty-first century. Since the decision to fluoridate usually is made at the local level, by public officials or a vote of the people, fluoridation has become a political as well as a scientific controversy.

Proponents

Proponents of water fluoridation argue the following:

- It significantly reduces tooth decay, both before and after tooth enamel has formed.
- The fluoride levels used are completely safe.
- The children of parents who are poorly informed about dental hygiene and cavity prevention or who cannot afford dental treatment are still protected against tooth decay.

Most government agencies and scientific and professional organizations agree that water fluoridation is safe and effective in preventing tooth decay and cost-effective in that it reduces the need for expensive dental treatment. Among the organizations that endorse fluoridation of water supplies are the following:

- American Academy of Pediatric Dentistry
- American Dental Association
- American Medical Association
- National Institute of Dental and Craniofacial Research
- U.S. Centers for Disease Control and Prevention (CDC)
- U.S. Public Health Service
- World Health Organization (WHO)

Opponents

Opponents of water fluoridation often use one or more of the following arguments:

- Any fluoride above the naturally occurring (usually trace) amounts is unnecessary and possibly toxic.
- An individual dose of fluoride cannot be controlled because it depends on the amount of fluoridated water that a child ingests each day.
- Fluoridation of public water systems deprives people of freedom-of-choice as to what they ingest.
- People can choose from a variety of fluoride-containing products that are just as effective as fluoridated water.
- Where the water is not fluoridated, schools often provide fluoridation programs, and parents can choose whether their children participate.

- Although fluoride may help prevent decay, good diet, good **oral hygiene**, and regular dental cleanings can be just as effective.
- Fluoride can be toxic and even fatal at higher doses.
- The difference between the amount of fluoride that is beneficial and the amount that can cause mottling is only two to four-fold.
- People vary in their susceptibility to the effects of fluoride.
- It is impossible to determine how much fluoride a child is ingesting because of the numerous sources of fluoride in food and products; a child may regularly drink water from sources with different fluoride levels.
- Fluoride is ineffective against gum disease, the major destroyer of teeth.

Communities throughout the United States, as well as many countries, have chosen not to fluoridate their water. In the early 2000s a number of countries have discontinued fluoridation because of ongoing concerns about possible health effects.

Fluoride dosages

There is some disagreement as to whether fluoride is an essential mineral in humans. Relatively low levels of fluoride (20–80 mg) are considered toxic. Less than 1 gm of fluoride can be fatal to a small child. The Food and Nutrition Board of the Institute of Medicine of the U.S. National Institutes of Health has determined an adequate daily intake of fluoride and a maximal safe daily intake, based on a child's weight:

- infants up to six months of age or about 16 lb (7 kg): 0.01 mg is adequate and 0.7 mg is the maximum safe intake
- infants between six and 12 months or about 20 lb (9 kg): 0.5 mg and 0.9 mg
- children one to three years of age or about 29 lb (13 kg): 0.7 mg and 1.3 mg
- children aged four to seven or about 48 lb (22 kg): 1.0 mg and 2.0 mg
- children aged nine to 13 or about 88 lb (40 kg): 2.0 mg and 10 mg
- children aged 14 to 19 or about 125–166 lb (57–76 kg): 3.0 mg and 10 mg

Fluoride supplements often are prescribed for children who drink nonfluorinated water and do not use fluoride toothpaste. Fluoride supplements should not be used if the drinking water contains more than 0.6 ppm of fluoride. One ppm of fluoride is equivalent to about 1 mg per quart (or liter) of water. Fluoride supplements should not be given to babies under six months of age regardless of the fluoride content of the water. Babies get adequate fluoride from breast milk or infant formula. Powdered or concentrated infant formula should be mixed with low-fluoride or fluoride-free water.

If the water supply contains 0.0 to 0.3 ppm fluoride, the recommended daily dosage of fluoride supplement is:

- 0.25 mg for children aged six months to three years
- 0.50 mg for children aged three to six years
- 1 mg for children aged six to 16 years

If the water supply contains 0.3 to 0.6 ppm fluoride, the recommended daily dosage of fluoride supplement is:

- 0.0 mg for babies aged six months to three years
- 0.25 mg for children aged three to six years
- 0.50 mg for children aged six to 16

Fluoride supplements usually come in the form of sodium fluoride: 2.2 mg of sodium fluoride supplies 1 mg of fluoride ion.

Precautions

A child easily can swallow enough fluoridated toothpaste to exceed the recommended daily amount of fluoride by four-fold. A medium-sized toothpaste tube contains enough fluoride to make a child seriously ill or even cause death should the child eat it all. The flavorings added to toothpaste to encourage children to brush also can entice them into eating it. Toothpaste always should be stored out of the reach of children.

Side effects

As little as four to eight mg of fluoride ingested daily while the tooth enamel is forming can cause mottling—often called fluorosis—in children under age eight. Fluorosis only affects children whose teeth are still developing within the gums. Symptoms of fluorosis include:

- teeth discoloration
- white or brown chalky spots
- brown enamel
- pitting of teeth
- excessive wear on the enamel
- structural damage to the enamel

KEY TERMS

Dentin—The middle layer of a tooth, which makes up most of the tooth's mass.

Enamel—The hard, outermost surface of a tooth.

Fluorapatite—Fluoride-substituted hydroxyapatite.

Fluorosis—Mottled discoloration of tooth enamel due to excessive systemic ingestion of fluoride during tooth development.

Mottling—Fluorosis; spotting on the teeth due to excess fluoride as the tooth enamel is forming.

Plaque—A deposit, usually of fatty material, on the inside wall of a blood vessel. Also refers to a small, round demyelinated area that develops in the brain and spinal cord of an individual with multiple sclerosis.

Remineralization—Recalcification; the process by which minerals from saliva and food are added to the surface of the enamel or to the dentin.

Systemic absorption—Any substance topical, inhaled, or ingested that is absorbed into the blood-stream and distributed throughout the body.

Topical—Not ingested; applied to the outside of the body, for example to the skin, eye, or mouth.

- brittle teeth in which the enamel breaks easily

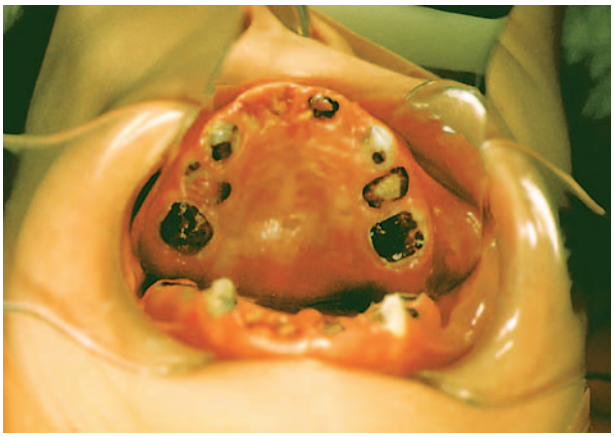
The extent of mottling depends on the following:

- when the excess fluoride is ingested
- how much is ingested
- over how long of a period it is ingested
- how much of the fluoride reaches the enamel

Most cases of fluorosis are very mild. Very mild to mild fluorosis has no effect on tooth function and may help prevent decay. Even severe fluorosis is not harmful. Fluorosis in children appears as of 2004 to be increasing; however, it is not known whether this is from water fluoridation, the excessive use of fluoride-containing products, or both.

Prevention

No type of fluoridation can replace good dental care and hygiene, which are necessary for preventing gum disease as well as tooth decay. Weekly rinsing with a fluoride mouthwash can reduce decay in children by 20–40 percent. Fluoride supplements can reduce decay in children by 40 percent, if administered at least 150 days of every year that enamel is forming.



Lack of fluoridation caused this four-year old child's tooth decay. *(AP/Wide World Photos.)*

Topical fluoride treatments given in a dentist's office have been proven to be 40 percent or more effective in preventing decay. These treatments include fluoride gels and foams and fluoride varnishes. Advantages of fluoride varnishes include the following:

- They are more concentrated than other fluoride treatments and so are less likely to be ingested.
- They are fast and easy to apply.
- They continue to provide fluoride to the enamel for about 24 hours after application.

Varnishes may be particularly appropriate for young children and those with special needs since varnishes do not require the use of a fluoride tray.

Parental concerns

It is the parents' responsibility to monitor their child's fluoride intake. Pregnant and nursing mothers should pay close attention to how much fluoride they ingest. Children should:

- be over two years of age before using a fluoridated toothpaste; younger children are likely to swallow most of their toothpaste
- use a pea-sized amount of toothpaste or less
- not use fluorinated mouthwashes until the age of six
- be prevented from swallowing fluoridated toothpaste or mouthwash

See also Dental development.

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Margaret Alic, PhD

Folate *see* **Folic acid**

Folic acid

Definition

Folic acid is a water-soluble vitamin belonging to the B-complex group of **vitamins**. These vitamins help the body break down complex carbohydrates into simple sugars to be used for energy. Excess B vitamins are excreted from the body rather than stored for later use. This is why sufficient daily intake of folic acid is necessary.

Description

Folic acid is also known as folate or folacin. It is one of the nutrients most often found to be deficient in the Western diet. There is evidence that folate deficiency is a worldwide problem. Folic acid is found in leafy green vegetables, beans, peas and lentils, liver, beets, brussel sprouts, poultry, nutritional yeast, tuna, wheat germ, mushrooms, oranges, asparagus, broccoli, spinach, bananas, strawberries, and cantaloupes. In 1998, the U.S. Food and Drug Administration (FDA) required food manufacturers to add folic acid to enriched bread and grain products to boost intake. Pregnant women whose diets are deficient in folic acid have a greater chance of having a baby with neural tube defects (NTD), such as **spina bifida**.

General use

Folic acid works together with vitamin B_{12} and vitamin C to metabolize protein in the body. It is important for the formation of red and white blood cells. It is also necessary for the proper differentiation and growth of cells in fetal development. It is also used to form the nucleic acid of DNA and RNA. It increases the appetite and stimulates the production of stomach acid for digestion, and it aids in maintaining a healthy liver. A deficiency of folic acid may lead to anemia, in which there is

KEY TERMS

Homocysteine—A sulfur-containing amino acid.

Preeclampsia—A condition that develops after the twentieth week of pregnancy and results in high blood pressure, fluid retention that doesn't go away, and large amounts of protein in the urine. Without treatment, it can progress to a dangerous condition called eclampsia, in which a woman goes into convulsions.

Recommended Dietary Allowance (RDA)—The Recommended Dietary Allowances (RDAs) are quantities of nutrients in the diet that are required to maintain good health in people. RDAs are established by the Food and Nutrition Board of the National Academy of Sciences, and may be revised every few years. A separate RDA value exists for each nutrient. The RDA values refer to the amount of nutrient expected to maintain good health in people. The actual amounts of each nutrient required to maintain good health in specific individuals differ from person to person.

Water-soluble vitamins—Vitamins that are not stored in the body and are easily excreted. They must, therefore, be consumed regularly as foods or supplements to maintain health.

decreased production of red blood cells. This situation reduces the amounts of oxygen and nutrients that are able to get to the tissues. Symptoms may include fatigue, reduced secretion of digestive acids, confusion, and forgetfulness. During pregnancy, a folic acid deficiency may lead to preeclampsia, premature birth, and increased bleeding after birth.

Pregnant women have an increased need for folic acid, both for themselves and their unborn child. Folic acid is necessary for the proper growth and development of the fetus. Adequate intake of folic acid is vital for the prevention of several types of birth defects, particularly NTDs. The neural tube of the embryo develops into the brain, spinal cord, spinal column, and the skull. If this tube forms incompletely during the first few months of pregnancy, a serious, and often fatal, defect results in spina bifida or anencephaly (formation of the head without the brain). Folic acid, taken from one year to one month before conception through the first four months of pregnancy, can reduce the risk of NTDs by 50 to 70 percent. It also helps prevent **cleft lip and palate**.

Research shows that folic acid can be used to successfully treat cervical dysplasia. This condition is considered to be a possible precursor to cervical **cancer** and is diagnosed as an abnormal Pap smear. Daily consumption of 1,000 mcg of folic acid for three or more months has resulted in improved cervical cells upon repeat Pap smears.

Precautions

Folic acid is not stable. It is easily destroyed by exposure to light, air, water, and cooking. Therefore, the supplement should be stored in a dark container in a cold, dry place, such as a refrigerator. Many medications interfere with the body's absorption and use of folic acid. This includes sulfa drugs, sleeping pills, estrogen, anticonvulsants, birth control pills, antacids, quinine, and some **antibiotics**. Using large amounts of folic acid (e.g., over 5,000 mcg per day) can mask a vitamin B_{12} deficiency and thereby risk of irreversible nerve damage.

Side effects

At levels of 5,000 mcg or less, folic acid is generally safe for use. Side effects are uncommon. However, large doses may cause **nausea**, decreased appetite, bloating, gas, decreased ability to concentrate, and insomnia. Large doses may also decrease the effects of phenytoin (Dilantin), a seizure medication.

Parental concerns

Pregnant women or those thinking of becoming pregnant should ensure that that they get the recommended amount of folic acid daily. As with all B-complex vitamins, it is best to take folic acid with the other B vitamins. Vitamin C is important to the absorption and functioning of folic acid in the body.

To correct a folic acid deficiency, supplements are taken in addition to food. Since the functioning of the B vitamins is interrelated, it is generally recommended that the appropriate dose of B-complex vitamins be taken in place of single B vitamin supplements. The Recommended Dietary Allowances (RDA) for folate is 400 mcg per day for adults, 600 mcg per day for pregnant women, and 500 mcg for nursing women. Medicinal dosages of up to 1,000–2,000 mcg per day may be prescribed. Nearly all multivitamin formulations for women include the RDA for folic acid.

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Food allergies and sensitivities

Definition

A food allergy or sensitivity is a person's immune system reaction to eating a particular food.

Description

The word allergy comes from two Greek words: alos, meaning "other" and argon, meaning "action." When one has an allergy, he or she has a reaction other than the one expected.

Food **allergies** and sensitivities are the body's reaction to a specific food. In a food allergy or sensitivity, when the child eats a particular food, (such as eggs, for example) usually by the time the eggs reach the stomach or the intestines, the body reads the presence of eggs as an allergen (something harmful). It sends out immunoglobulin E (IgE), an antibody, to destroy the eggs and protect the body, releasing histamines. The body remembers and produces histamines every time the food is eaten. These histamines trigger allergic symptoms that affect many areas of the body, particularly the skin, respiratory system, nervous system, and digestive system. Digestive disorders after eating specific foods are not always allergies. These reactions can be food sensitivities or intolerances. They can also be symptoms of other, more serious digestive diseases and malfunctions.

In the United states, 90 percent of all food allergies are caused by wheat, peanuts, nuts, milk, eggs, shellfish, soy, and fish. Many other foods can be at the root of food allergies or sensitivities, especially berries and other fruits, tomatoes, corn, and some meats like pork. Migraine headaches have been associated with sensitivities

to chemicals contained in red wine, deli meats, aged cheeses, and the tannins in tea.

Usually, when a child is allergic to one food in a food family, he or she will most likely react to other foods in that food family. For example, if a child is sensitive to one type of fish, he or she also may be sensitive to other types of fish. This is called cross-reactivity.

Demographics

Nearly three million children in the United States have been diagnosed with food allergies. Nearly 600,000 of them have severe allergies to peanuts and possibly twice as many have severe shellfish allergies. Each year about 200 adults and children in the United States die from food-related **anaphylaxis**, an extreme reaction that causes swelling of the throat and bronchial passages, shock, and a severe drop in blood pressure. Nevertheless, food allergies tend to be under-diagnosed by doctors.

Genetics seems to play a part in food allergies. If one parent has a food allergy, the child's risk of having a food allergy is doubled. If both parents have food allergies, the risk is even higher. The child, however, may be allergic to a completely different food from the one to which the parent has demonstrated sensitivity. There also is increased risk when there are other kinds of allergy-related diseases in the family, such as hay fever or **asthma**.

Causes and symptoms

Causes

Allergies are caused by the immune system's reaction to a particular food. Usually, a child will have had a prior exposure before IgE or specific histamines are produced.

Food intolerance is often put into the same category as food allergy, even though there may be an entirely different mechanism involved. In these cases, the digestive tract reacts to a specific part of the food; for example, the protein or the sugar in a specific food. The digestive system rebels, resulting in gas, bloating, upset stomach, **diarrhea**, **nausea**, or **vomiting**. Many times, these responses are due to eating food contaminated with bacteria, rather than a true food allergy. In other cases, the child's reaction is due to an underlying digestive disorder such as **irritable bowel syndrome**, which is a chronic condition that is often triggered by specific types of food.

Gluten intolerance is not an allergy. It is a disease called **celiac disease**, or gluten-sensitive enteropathy. The body cannot process gluten found in wheat and other grains. Though the immune system is involved, celiac disease does not behave as a true allergy. Its treatment is like many food allergies, namely avoidance of the offending substance, which in this case is gluten.

Some children may lack a specific enzyme needed to metabolize certain foods. About 10 percent of all adults and older children have **lactose intolerance**. There are two forms of lactose intolerance: inherited and acquired. The inherited form (autosomal recessive) is extremely rare and severe. The acquired type is very common, and occurs in older children (not infants) and adults. It is distressing, but not life-threatening, and occurs with increased frequency in African Americans. Sometimes infants, as well as older children and adults, have a transient lactose deficiency after an episode of diarrhea.

Children with lactose intolerance have a lactase deficiency that keeps them from processing milk and milk products. These children can often drink milk that has had this enzyme introduced into the product. Some children can drink milk that has acidophilus bacteria put into it. This bacteria breaks down the lactose, or milk sugar, in the milk so that the child can tolerate it. Some children with lactose intolerance cannot drink whole milk, but can eat cheese or drink low-fat buttermilk in small quantities. This is different from a true milk allergy where even a small amount of any dairy product will produce a reaction.

Some children may also be intolerant of food colorings, additives, and preservatives. Among these are yellow dye number 5, which can cause **hives**; and monosodium glutamate, which produces flushing, headaches, and chest **pain**. Sulfites, another additive, have been found to cause asthmatic reactions and even anaphylactoid reactions. Sulfites are preservatives used in wines, maraschino cherries, seafood, and soft drinks. They are sometimes put on fresh fruits and lettuce to maintain their fresh appearance, on red meats to prevent brown discoloration, and even in prepared deli foods like crab salad. Sulfites appear on food labels as sodium sulfite, sodium bisulfite, potassium bisulfite, sulfur dioxide, and potassium metabisulfite. The U.S. Food and Drug Administration (FDA) has banned the use of sulfites as a preservative for fruits and vegetables, but they are still in use in some foods.

Symptoms

Food allergies and sensitivities can produce a wide range of symptoms involving the skin, respiratory system, and nervous system. Children may have watery eyes, runny noses, and sneezing.

Skin **rashes** or hives can range from measles-like rashes to itchy welts. The rashes or welts can appear on a specific part of the body or can be widespread. Some children have swelling of the eyes, lips, and/or tongue.

Symptoms vary among children, even those who are sensitive to the same food. One child's specific reaction to an offending food does not mean that all children react the same. Nut allergies and shellfish, however, seem to be the most documented triggers for anaphylaxis. Nevertheless, anaphylaxis is not limited to those foods. IgE-mediated allergic reactions can progress to other allergic symptoms. For example, a child who has had hives is at risk for angioedema (swelling of the blood vessels) and anaphylaxis.

Symptoms also vary in intensity and by the amount eaten. One child may have a mild rash on the forearms when eating half a dozen strawberries. Another may be covered with a rash after eating only one. This variation is individualized and is a factor in the body's sensitivity to the target food.

Although the time between ingestion and symptoms is somewhat variable for allergic reactions (IgE-mediated), the vast majority occur within minutes. Nearly all occur within two hours. Reactions due to intolerances, like lactose, may occur somewhat later. Symptoms occurring days after a food is ingested are not likely related to the food.

When to call the doctor

Anaphylaxis is an extreme reaction to a food, usually peanuts or nuts. It causes swelling of the throat and bronchial passages, a drop in blood pressure, shock, and even death. A child with anaphylaxis should be taken to the emergency room immediately. If an emergency epinephrine pen is available, it should be administered immediately.

If a child experiences any type of allergy symptoms after eating, the child should be evaluated. Of particular concern are digestive symptoms that keep the child from eating properly or cause the child to lose weight. Equally important are neurological symptoms, especially headaches. Digestive and neurological symptoms could also be an indication of other underlying disorders. Therefore, the child should be seen by a doctor.

Diagnosis

Having a child evaluated as soon as possible will identify the offending food and allow parents to eliminate it from the child's diet. Many allergists, or doctors who specialize in allergies, will do a skin-prick test followed by a blood test. The skin-prick test is a series of pricks on the child's skin with a plastic applicator that contains a single food in concentrated form. A food allergy has been identified if the child's skin reacts by welting or becoming red. The skin-prick test for foods (not for aeroallergens) has a high incidence of false positives; that is, the test may be positive but the child is not truly allergic, or does not have symptoms from the food. This test is not used on a child with severe anaphylactic reactions or on children with widespread eczema, a skin disorder.

The allergist may also do a food challenge in the doctor's office. The child is fed the suspected food in increasing amounts to see what kind of reaction occurs.

One of the tests allergists use is called the RAST (Radio-Allergo-Sorbent Test). It measures the amount of IgE antibody in the blood that is produced for certain known food allergens. Like the skin-prick test, RAST and other antibody tests have a high rate of false positives.

Some doctors will put the child on an **elimination diet** for one week to 10 days. The basic elimination diet is a series of foods that have proven not to be allergy triggers. This diet consists of foods such as lamb, poultry, rice, vegetables, and all fruits, except citrus and berries. One new food is introduced each week. Parents record the child's reaction to each food. If the child has no reaction, the food is considered safe and can remain in the diet. If there is a reaction, it is noted and the food is removed. The child continues the elimination diet for a few more days, at which time another food is introduced. The elimination diet is often done after skin testing, so there is a logical guide for what to eliminate.

Treatment

The only treatment for IgE-mediated reactions to foods is avoidance. These reactions, as well as intolerances, are not responsive to desensitization. An epi-pen should be kept in the home for all IgE mediated food allergies and all inadvertent reactions should be treated.

It is not unusual for children to crave the very foods to which they are allergic. When the child is placed on an elimination diet, often the body will rebel at not being given the foods that cause it to react and will produce cravings for those foods.

Some doctors will prescribe **antihistamines** to help manage symptoms. These, however, are for use after an episode and not for an extended period.

Nutritional concerns

Eliminating one food or even one food family from a child's diet will not interfere with his or her nutritional

KEY TERMS

Allergen—A foreign substance that provokes an immune reaction or allergic response in some sensitive people but not in most others.

Allergists—Doctors who specialize in allergies.

Antibody—A special protein made by the body's immune system as a defense against foreign material (bacteria, viruses, etc.) that enters the body. It is uniquely designed to attack and neutralize the specific antigen that triggered the immune response.

Cross-reaction—A reaction that occurs in blood testing when a disease agent reacts to the specific antibody for another disease agent.

Histamine—A substance released by immune system cells in response to the presence of an allergen. It stimulates widening of blood vessels and increased porousness of blood vessel walls so that fluid and protein leak out from the blood into the surrounding tissue, causing localised inflammation of the tissue.

Immunoglobulin G (IgG)—Immunoglobulin type gamma, the most common type found in the blood and tissue fluids.

Lactose—A sugar found in milk and milk products.

needs, nor will it keep the child from growing properly. There is enough variety in the foods available in the American diet to meet any child's needs. Some foods, however, may be difficult to find sufficient replacements for if the child wants substitutes. Wheat is particularly difficult to replace, although bread, pasta, and pastry products made with oat and rice flours are good substitutes. However, they do not taste or look exactly like risen wheat bread or regular pasta.

Prognosis

Children are known to outgrow milk allergies in most cases, but—for safety purposes—reintroduction in a medical setting is advised. Egg allergy disappearance is not as high as it is for milk allergy. Sensitivities to wheat and soy are also outgrown. Allergies to peanuts, shellfish, and other foods that can produce anaphylaxis usually remain with the child throughout life.

Foods such as whole-wheat flour, tuna, soy sauce, eggs, peanut butter, almonds, and milk are common causes of food allergies. *(© Erik Freeland/Corbis.)*

Prevention

If both parents have food allergies, precautions should be taken to minimize the risk of the child having a food allergy, too. Before birth and while breastfeeding, the mother can limit the baby's exposure to allergens by not bingeing on foods known to cause allergies. Breastfeeding delays the onset of allergies, but does not avoid them. The secretory IgA in breast milk fights infection but is not shown to avoid absorption of allergies.

Solid foods are slowly introduced at four to six months of age. The first solid foods should be those that have shown the potential for not producing an allergic reaction, such as fruits (except citrus fruits and berries), vegetables, and rice. Early introduction of highly allergenic foods may predispose a child to reactions, but this is controversial. It is recommended that parents avoid feeding the child highly allergenic foods until three years of age, if possible. The list of highly allergenic foods includes nuts, peanuts, fish, shellfish, and eggs. Whole cow's milk—not cow's milk formula—should be avoided during the first year. Having the child eat a variety of foods will also keep the child from too much exposure to any one particular food family.

Parents should make sure that the baby's first foods and those during the first few years of life are pure, unprocessed foods. Packaged and prepared foods (soups, stews, and toddler dinners, for example) contain many foods mixed together, along with fillers, usually wheat products, and possibly flavorings, sugar, and salt. By feeding a toddler a piece of boneless chicken, some green peas, a few cooked carrots, and a bit of potato instead of a can of chicken stew, parents can identify exactly what foods the child is eating and in what

quantities. Therefore, if there is an allergic reaction, it is easier to identify what triggered it.

Parental concerns

Because children can come into contact with food allergens at school and during **extracurricular activities**, parents should meet with school officials to discuss procedures for keeping their children safe. Parents and school personnel should develop an action plan for dealing with allergens in the school and handling an emergency. Not only should the cafeteria staff be notified about the food allergy, but parents should also ask about snack time, birthdays and holiday celebrations, field trips, and arts and crafts projects. Arrangements should be made to keep medications to treat accidental exposure at the school, and personnel should be trained in their use.

Due to the seriousness of nut allergies and other allergies that can cause anaphylaxis, some school districts have created policies that forbid nuts on school premises and do not allow students to trade food at lunch. Some parents have lobbied school boards for such restrictions.

Avoiding the trigger food may be very problematic, even at home. Parents need to become proficient label readers, especially if the allergen is a nut or other food that may cause anaphylaxis. Understanding what the ingredient names mean is critical to total food avoidance. For example, dairy products can be listed as milk, casein, whey, and sodium caseinate. If a child is allergic to corn, it can be found not only as corn and corn syrup, but also cornstarch, which is a binding agent in a number of medications, including **acetaminophen** (Tylenol). Consultation with a dietitian can help parents understand food labels.

Peanut allergies in the United States doubled between 1997 and 2002. A controversial British study, reported in 2003, found a peanut/soy link, which is clinically rare. The study reported a link between early use of soy formula and peanut oil baby lotion in the later diagnosis of peanut food allergy. Soy formula may sensitize an infant to legumes, and therefore to peanuts. Peanut oil, known by doctors and nurses as arachis oil, is found in baby lotion and creams, especially those used to treat **diaper rash**, eczema, and dry skin.

Children who have severe reactions to a trigger food should wear a medical alert bracelet. Parents should also have on hand an emergency epinephrine-filled syringe like those found in bee-sting kits, or an epinephrine pen.

Parents should also notify day-care providers, Girl Scout and Boy Scout leaders, religious education teachers, **sports** coaches, and parents of their child's friends. They should explain what foods their child is allergic to, how the child reacts to the food, and how adults can help, either by making sure these foods are not served as snacks or by giving emergency care or support during an allergic reaction. In addition, parents can teach their child how to ask for help.

See also Lactose intolerance.

Resources

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Janie Franz

Food poisoning

Definition

Food poisoning refers to illness arising from eating contaminated food. Food may be contaminated by bacteria, viruses, environmental toxins, or toxins present

within the food itself, such as the poisons in some mushrooms or seafood. Symptoms of food poisoning are usually gastrointestinal, such as **nausea**, abdominal **pain**, **vomiting**, and/or **diarrhea**. Some food-borne toxins can affect the nervous system. Food poisoning is sometimes called bacterial **gastroenteritis** or infectious diarrhea and is sometimes incorrectly called ptomaine poisoning.

Description

Every year millions of people of all ages suffer from bouts of vomiting and diarrhea blamed correctly on something they ate. According to the Centers for Disease Control and Prevention (CDC), up to 33 million cases of food poisoning are reported in the United States each year. Many cases are mild and pass so rapidly that they are never diagnosed. Occasionally a severe outbreak affects many people at once, creating a newsworthy public health hazard. Although the food supply in the United States is probably one of the safest in the world, anyone can get food poisoning. Outbreaks have occurred in schools and colleges (up to 25 incidents reported annually in the United States), among restaurant clientele, in institutions such as long-term care facilities, and in other settings serving the public. Serious outbreaks are rare. When they occur, the very young, the very old, and those with weakened immune systems are subject to the most severe and life-threatening cases.

A variety of bacteria cause food poisoning, including *Salmonella*, *Staphylococcus aureus*, *Campylobacter jejuni*, *Escherichia coli*, *Shigella*, and *Clostridium botulinum*. Each type of bacteria has a different incubation period and duration, and all except the botulinum toxin cause inflammation of the intestines and diarrhea. Food and water can also be contaminated by viruses such as the Norwalk and hepatitis viruses. Environmental toxins (heavy metals) in foods or water, and poisonous substances in certain foods such mushrooms and shellfish are other causes of food poisoning.

Careless food handling between farm and table may create conditions for the growth of bacteria. Vegetables eaten raw, such as lettuce, may be contaminated by bacteria in soil, water, and dust during washing and packing. Home canned and commercially canned food may be improperly processed at too low a temperature or for too short a time to kill bacteria.

Raw meats carry many bacterial contaminants. The United States Food and Drug Administration (FDA) estimates that 60 percent or more of raw poultry sold at retail carries some disease-causing bacteria. Other raw meat products and eggs are contaminated to a lesser degree. Although thorough cooking kills bacteria and makes the food harmless, recontamination can occur in properly cooked food if it comes into contact with cutting boards, countertops, or utensils that were used with raw meat and not cleaned and sanitized after use. Food can also become contaminated by environmental contaminants and by food handlers carrying bacteria on their hands while preparing foods for the public.

It is estimated that 50 percent of healthy people have staphylococcus organisms in their nasal passages and throats and on their skin and hair. Rubbing a runny nose and then touching food can introduce the bacteria into cooked food. Bacteria flourish at room temperature and grow rapidly in quantities capable of causing illness. To prevent this growth, food must be kept hot or cold but never just warm or at room temperature.

Travel to countries where less attention is paid to sanitation, water purification, and good food-handling practices may expose individuals to bacterial contaminants. Institutional food preparation also increases the risk of food poisoning, especially if food is allowed to stand on warming trays, under warming lights, or at room temperature before being served.

Transmission

Food poisoning is not spread from one individual to another but through direct contact with the causative bacteria, viruses, or other toxins in consumed food.

Demographics

The Centers for Disease Control and Prevention (CDC) estimates that there are from six to 33 million cases of food poisoning in the United States annually, affecting men, women, and children. Food poisoning by *E. coli* occurs in three out of every 10,000 people. One out of every 1,000 people are reported to have food poisoning caused by *Salmonella*; two-thirds are young people under age 20, and the majority are children under age nine. Although camplyobacter infections can occur in anyone, children under age five and young adults between ages 15 and 29 are more frequently infected.

Causes and symptoms

Classic food poisoning cases are caused by a variety of bacteria. The most common are the following:

- *Salmonella*
- *Staphylococcus aureus*
- *Campylobacter jejuni*
- *Escherichia coli*

- *Shigella*
- *Clostridium botulinum*

Food poisoning symptoms occur when food-borne bacteria release toxins or poisons as a byproduct of their growth in the body. These toxins (except those from *C. botulinum*) cause inflammation of the stomach lining and the small and/or large intestines, resulting in abdominal **muscle cramps**, vomiting, diarrhea, and **fever**. The severity of symptoms depends on the type of bacteria, the amount consumed, and the individual's general health and sensitivity to the toxin. **Dehydration** can result from loss of fluids through persistent vomiting and diarrhea; it is one of the most frequent and serious complications of food poisoning. When more fluids are being lost than are replaced, dehydration may occur in the very young and in the elderly, as well as in individuals who take diuretics.

Salmonella

A 2001 CDC report states that culture-confirmed cases of salmonella poisoning affected almost 50,000 people in the United States. However, it is believed that between 2 and 4 million unconfirmed cases actually occur each year. Salmonella is found in egg yolks from infected chickens, raw and undercooked poultry and other meats, dairy products, fish, shrimp, and many other foods. The CDC estimates that one of every 50 consumers is exposed to a contaminated egg yolk each year, although thorough cooking kills the bacteria, making the food harmless. Salmonella is also found in feces of pet reptiles such as turtles, lizards, and snakes. Most cases of salmonella poisoning occur in the warm months between July and October.

Symptoms of food poisoning, such as abdominal pain, diarrhea, vomiting, and fever, begin eight to 72 hours after eating food contaminated with salmonella. Symptoms generally last one to five days. Dehydration can be a complication of severe cases with persistent vomiting and/or diarrhea. People generally recover without antibiotic treatment, although they may feel tired for a week or so after the active symptoms subside.

Staphylococcus aureus

Staph organisms are found on humans and in the environment in dust, air, and sewage. The bacteria are spread primarily by food handlers using poor sanitary practices. Almost any food can be contaminated, but salad dressings, milk products, cream pastries, and any food kept at room temperature, rather than hot or cold, are likely candidates. It is difficult to estimate the number of annual cases of *Staphylococcus* food poisoning because its symptoms are so similar to those caused by other food-borne bacteria. Many cases are mild. Victims may miss a day of school or work but never see a doctor for confirmation of food poisoning. Symptoms appear rapidly, usually one to six hours after the contaminated food is eaten. Acute symptoms of vomiting and severe abdominal cramps without fever usually last three to six hours and rarely more than 24 hours. Most people recover without medical assistance. Deaths are rare.

Escherichia coli (E. coli)

The many strains of *E. coli* are not all harmful. Nonpathogenic E. coli are, in fact, a major part of normal gut flora. The strain that causes the most severe food poisoning, however, is *E. coli O157:H7*, which affects three people in every 10,000. The food-borne organisms are found and transmitted mainly in food derived from cows, such as raw milk and raw or rare ground beef. Fruit or vegetables can also be contaminated.

Symptoms of *E. coli* poisoning are slower to appear than those caused by other food-borne bacteria. Because *E. coli* toxins are produced in the large intestine rather than higher up in the digestive system, symptoms typically occur from one to three days after eating contaminated food. Those affected have severe abdominal cramps and watery diarrhea that usually becomes bloody within 24 hours, a condition that can last from one to eight days. There is little or no fever and vomiting occurs only rarely.

Campylobacter jejuni

Campylobacter is the leading cause of bacterial diarrhea worldwide, responsible for more cases (2 million or more) of bacterial diarrhea in the United States than *Shigella* and *Salmonella* combined. *Campylobacter* is carried by healthy cattle, chickens, birds, and flies. It is also found in ponds and stream water and has been found in bottled water and on salad vegetables washed with water. Although eating chicken is a known risk factor, drinking water and eating salads have not been considered significant risks until studies of causes released in 2003 showed possible association with *Campylobacter* diarrheal infections. It is not known whether contamination occurs at the site of production or in the home or institution after contact with other contaminated foods, surfaces, or utensils. The ingestion of only a few hundred *Campylobacter* bacteria can cause food poisoning symptoms, which may begin two to five days after eating contaminated food. Symptoms will typically include fever, abdominal pain, nausea, **headache**, muscle pain, and diarrhea. The diarrhea can be watery or sticky and may contain blood. Symptoms last from seven to ten days and relapses occur in about one-fourth of infected

individuals. Dehydration is a common complication. Other complications, such as arthritis-like joint pain and hemolytic-uremic syndrome (HUS), occur in rare cases.

Shigella

Shigella is a common cause of diarrhea in travelers to developing countries. It is associated with contaminated food and water, crowded living conditions, and poor sanitation. The bacterial toxins affect the small intestine. Symptoms of *Shigella* infection appear about 36–72 hours after eating contaminated food. In addition to the familiar watery diarrhea, nausea, vomiting, and abdominal cramps, the individual may also have chills, fever, and neurological symptoms. The diarrhea may be quite severe with cramping and progresses to classic dysentery. Up to 40 percent of children with severe infections show neurological symptoms. These include confusion, headache, lethargy, a stiff neck, and possible seizures. The symptoms of food poisoning by *Shigella* organisms may resemble **meningitis** and a differential diagnosis must be made by isolating the causative bacteria.

The disease runs its course usually in two to three days but may last longer. Dehydration is a common complication. Most people recover on their own, although they may feel exhausted. Children who are malnourished or have weakened immune systems may be severely affected and death can result.

Clostridium botulinum

C. botulinum causes both adult and infant **botulism** and differs significantly from other contaminants in its sources and symptoms. *C. botulinum*'s common food-borne form is an anaerobic bacterium that can only live and reproduce in the absence of oxygen. Exposure to the botulinum toxin usually occurs while eating contaminated food stored in an airless environment, as in home-canned or commercially canned or vacuum-packed food. Also, botulinum toxin is a neurotoxin that blocks the ability of motor nerves to release acetylcholine, the neurotransmitter that relays nerve signals to muscles. This neurological process can result in unresponsive muscles, a condition known as flaccid paralysis. Breathing may be severely compromised in progressive botulism because of failure of the muscles that control the airway and breathing. In infants, botulism may be caused by specific types of clostridia obtained from soil, inhaled spores, or honey containing the spores. Contamination from any of the sources results in growth of the bacteria in the infant's intestine and production of the neurotoxin.

Infant botulism is a form of botulism first recognized in 1976 that differs from food-borne botulism. Infant botulism occurs when a child younger than one year ingests the spores of *C. botulinum*. Although these spores are commonly found in soil, honey is a more frequent source of spores causing infant botulism by lodging in the baby's intestinal tract and producing the neurotoxin. Onset of symptoms is gradual. Initially, the baby is constipated, followed by poor feeding, lethargy, weakness, drooling, and a distinctive wailing cry. Eventually, the baby loses the ability to control its head muscles. From there the paralysis progresses to the rest of the body. Immediate treatment is required to avoid neurological complications and death. Infant botulism is much more likely to be fatal than other food poisoning infections. Infant botulism is a special form of food poisoning not related to the food-borne toxins that cause adult botulism.

Adult botulism outbreaks are usually associated with toxins found in home-canned food, although poisoning occasionally results from eating commercially canned or vacuum-packed foods. *C. botulinum* grows well in non-acidic, oxygen-free environments, meaning that if the cooking temperatures are too low or the cooking time too brief the bacteria in the food are not killed. Instead, bacteria may reproduce inside the can or jar, releasing the deadly neurotoxin. Heating canned food to boiling for ten minutes can render the toxin harmless. However, consuming even a very small amount of the toxin can result in serious illness or death because of lethal neurological complications.

Symptoms of adult botulism appear about 18–36 hours after the contaminated food is eaten, although times of onset have been documented ranging from four hours to eight days. Initially a person suffering from botulism feels weak and dizzy and later experiences double vision. Symptoms progress to difficulty speaking and swallowing. Paralysis moves down the body, and when the respiratory muscles are paralyzed, death can result from asphyxiation. Individuals with any signs of botulism poisoning must receive immediate emergency medical care to increase their chance of survival.

When to call the doctor

Any unexplained abdominal pain accompanied by persistent vomiting or diarrhea, whether or not a food source is suspected, should be reported to the doctor. A child having difficulty swallowing, speaking, holding the head up, or maintaining an upright posture should receive emergency medical attention. Signs of confusion, lethargy, headache, stiff neck, or seizures also require immediate medical attention.

Diagnosis

One important part of diagnosing food poisoning is the need for doctors and community health professionals

to determine if a number of people have eaten the same food and show the same symptoms. If this can be proven, food poisoning is strongly suspected. The diagnosis is confirmed when the suspected bacteria is identified in the culture of a stool sample or a fecal smear from the affected individual. In some cases, the suspected bacteria, virus, or toxin can be identified in the actual food source.

Laboratory tests are used to make a definitive diagnosis, but treatment of symptoms may be started immediately without waiting for test results, which may take up to two days. Diagnostic tests focus on identifying the organism causing the illness. This process may involve performing a culture on contaminated material from the suspect food, a stool sample, or swabs of the nose or throat of the affected individual if inhaled spores are a possibility. Culture results are available from the microbiology laboratory as soon as bacteria grow in a special plate incubated at temperatures at or above body temperature. The growth of specific bacteria confirms the diagnosis. The microbiology laboratory may use samples of the bacteria grown to perform other special techniques to help identify the causative organism.

In infant botulism, the infant's stool may be cultured to isolate the organism; this test may be performed by the state health department or the Centers for Disease Control (CDC). Early diagnosis of botulism is critical so that treatment can begin in time to avoid neurological involvement. Although the definitive diagnosis comes from laboratory tests, it can usually be diagnosed by recognizing the distinctive neurological symptoms typical of contamination with *C. botulinum*.

While waiting for diagnostic test results, the doctor performs a physical examination and may ask about recently consumed food, possible open sores, recent activities and behavior, and other information that may help to rule out other disease possibilities. Imaging studies or additional diagnostic tests may be done to rule out other diseases or conditions with similar symptoms.

Many cases of food poisoning go undiagnosed, since a definite diagnosis is not necessary to effectively treat the symptoms. Because it takes time for symptoms to develop, the most recent food one has eaten may not be the cause of the symptoms.

Treatment

Treatment of food poisoning, except for botulism, focuses on preventing or correcting dehydration by replacing critical fluids and electrolytes lost through vomiting and diarrhea. Electrolytes are mineral salts that form electrically charged particles (ions) in body fluids; they help control body fluid balance and participate in many essential body functions. Pharmacists can recommend effective, pleasant-tasting, electrolyte replacement fluids that are available without a prescription. To prevent dehydration, a doctor may decide to give fluids intravenously. In very serious cases of food poisoning, medications may be given to stop abdominal cramping and vomiting. Antidiarrheal medications are not usually given. Stopping the diarrhea actually maintains toxin levels in the body for longer periods and may prolong the infection. Severe bacterial food poisonings are sometimes treated with intravenous **antibiotics**.

Modifying the diet while recovering from food poisoning is usually recommended. During a period of active vomiting and diarrhea, solid food should be avoided and only small quantities of clear liquids should be consumed as frequently as possible. Once active symptoms stop, bland, soft, easy-to-digest foods should be consumed for two to three days. One example is the BRAT diet of bananas, rice, applesauce, and toast, all of which are easy to digest. Milk products, spicy food, and fresh fruit should be avoided for a few days, although babies should continue to breastfeed. These modifications are often the only treatment that is necessary.

Botulism is treated in an entirely different way. Older children and adults can be treated with injections of a specific antitoxin for botulism if it can be administered within 72 hours after symptoms are first observed. If given later, it provides little or no benefit. Infants, however, cannot receive this antitoxin and are usually treated instead with injections of human botulism immune globulin (BIG), an antiserum that neutralizes the botulinum toxin. This antiserum is available in the United States through the Infant Botulism Treatment and Prevention Program in Berkeley, California. Both infants and adults may require **hospitalization**, often in the intensive care unit. Mechanical ventilators may be used for those whose ability to breathe is impaired and intravenous **nutrition** may be provided until any paralysis is corrected.

Alternative treatment

Alternative practitioners offer the same advice as traditional practitioners concerning diet modification, treatment of diarrhea and vomiting, and prevention of dehydration. Charcoal tablets, *Lactobacillus acidophilus*, *Lactobacillus bulgaricus*, and citrus seed extract can be taken to help normalize the digestive system. An electrolyte replacement fluid can be made at home by adding one teaspoon of salt and four teaspoons of sugar to one quart of water. For food poisoning other than botulism, two homeopathic remedies, either *Arsenicum*

album or *Nux vomica*, are recommended to help reduce symptoms.

Prognosis

Most cases of food poisoning (except botulism) clear up on their own within one week without medical assistance. As symptoms subside, the individual may continue to feel tired or weak for a few days. If dehydration has been effectively corrected or prevented, few complications can be expected. Deaths are rare and usually occur in the very young, the very old, and people whose immune systems are already weakened.

Complications of **salmonella food poisoning** may include arthritis-like symptoms that occur three to four weeks after infection. Although deaths from salmonella infection are rare, they do occur. Most deaths reported have occurred among elderly adults in long-term care.

Adults usually recover from *E. coli* poisoning without medical intervention, but many children require hospitalization for contamination with this organism. Toxins may be absorbed into the blood stream where they destroy red blood cells and platelets, tiny cells important in blood clotting. About 5 percent of victims develop hemolytic-uremic syndrome (HUS), which can result in sudden kidney failure that requires a medical procedure (dialysis) to perform the kidney's task of filtering the body's waste products.

Botulism is the deadliest of the bacterial food-borne illnesses. With prompt medical care, the death rate is less than 10 percent in children and adults.

Prevention

Food poisoning is almost entirely preventable by practicing good sanitation and good food handling techniques. These include the following measures:

- Keep hot foods hot and cold foods cold.
- Cook meat to the recommended internal temperature.
- Use a meat thermometer to check meat and cooking eggs until they are no longer runny.
- Refrigerate leftovers promptly, not letting food stand at room temperature.
- Before preparing other foods, carefully clean surfaces (cutting boards and counters, knives and other utensils) contaminated with the juices of uncooked meats.
- Do not refreeze meat once it has been thawed.
- Wash fruits and vegetables before using.
- Consume only pasteurized dairy products and fruit juices.

KEY TERMS

Antitoxin—An antibody against an exotoxin, usually derived from horse serum.

Culture—A test in which a sample of body fluid is placed on materials specially formulated to grow microorganisms. A culture is used to learn what type of bacterium is causing infection.

Diuretics—A group of drugs that helps remove excess water from the body by increasing the amount lost by urination.

Electrolytes—Salts and minerals that produce electrically charged particles (ions) in body fluids. Common human electrolytes are sodium chloride, potassium, calcium, and sodium bicarbonate. Electrolytes control the fluid balance of the body and are important in muscle contraction, energy generation, and almost all major biochemical reactions in the body.

Hemolysis—The process of breaking down red blood cells. As the cells are destroyed, hemoglobin, the component of red blood cells which carries the oxygen, is liberated.

Lactobacillus acidophilus—Commonly known as acidophilus, a bacteria found in yogurt that changes the balance of the bacteria in the intestine in a beneficial way.

Neurological—Relating to the brain and central nervous system.

Neurotoxin—A poison that acts directly on the central nervous system.

Platelet—A cell-like particle in the blood that plays an important role in blood clotting. Platelets are activated when an injury causes a blood vessel to break. They change shape from round to spiny, "sticking" to the broken vessel wall and to each other to begin the clotting process. In addition to physically plugging breaks in blood vessel walls, platelets also release chemicals that promote clotting.

Spore—A dormant form assumed by some bacteria, such as anthrax, that enable the bacterium to survive high temperatures, dryness, and lack of nourishment for long periods of time. Under proper conditions, the spore may revert to the actively multiplying form of the bacteria. Also refers to the small, thick-walled reproductive structure of a fungus.

Toxin—A poisonous substance usually produced by a microorganism or plant.

- Discard bulging or leaking cans or any food that smells spoiled.
- Wash hands well before and during food preparation and after using the bathroom.
- Sanitize food preparation surfaces regularly.

It is especially important to discard any food that seems spoiled and not to eat food that has been stored at room temperature or above for more than a few hours. Home canners must be diligent about using sterile equipment and following U.S. Department of Agriculture canning guidelines.

Infant botulism is perhaps the most difficult poisoning to prevent, because what goes into an infant's mouth is often beyond control. One important preventative measure, however, is to avoid feeding honey to infants younger than 12 months since it is a known source of botulism spores. As infants begin eating solid foods, the same food precautions should be followed as for older children and adults.

Parental concerns

Symptoms of food poisoning can appear as early as an hour after consuming the contaminated food or up to several days later. Parents may be concerned about possible contamination from unknown sources and that symptoms may occur suddenly, without warning. Practicing good sanitation and good food handling techniques is the best way parents can prevent contamination. Normal watchfulness of the parents is sufficient to notice symptoms, paying attention to any change in eating, unusual crying, increases or decreases in bowel movements, the presence of vomiting or a lack of normal responses such as turning of the head and body movements. An early report of symptoms, even if no particular food is suspected of causing illness, helps get early treatment and avoid complications.

See also Botulism; Gastroenteritis.

Resources

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Foreign objects

Definition

Foreign bodies can enter the human body by swallowing, insertion, or traumatic force, either accidentally or on purpose. The word "foreign" in this context means "originating elsewhere" or simply "outside the body."

Description

Children and adolescents may experience health problems caused by foreign objects getting stuck in their bodies. Young children in particular are naturally curious and may intentionally put such shiny objects as coins or button batteries into their mouths. They also like to stick small items in their ears and up their noses. Older children and teenagers may accidentally swallow a nonfood object or ingest a foreign body that gets stuck in the throat, like a fish bone or toothpick. Airborne particles can lodge in the eyes of people at any age. In addition, foreign bodies may be driven into the face or other parts of the body by the force of a collision or explosion.

Foreign bodies may be found in hollow organs (like swallowed batteries in the stomach) or in tissues (like bullets). They can be inert or irritating. If they irritate the surrounding tissue, they cause inflammation and scarring. Foreign objects can bring infection with them or acquire it and protect it from the body's immune defenses. They can obstruct passageways in the body either by their size or by the scarring they cause. Some foreign objects, particularly lead shot or other small objects containing lead, are toxic.

Demographics

Swallowing of foreign bodies is a fairly common pediatric emergency; about 80,000 cases involving per-

sons 19 years old or younger are reported each year to the 67 poison control centers in the United States. In a recent survey of the parents of 1,500 children, 4 percent reported that their children had swallowed a foreign object of some kind. The highest incidence of swallowed foreign bodies is in children between the ages of six months and four years.

The type of object most frequently swallowed varies somewhat across different historical periods and cultures. A recent study comparing the Jackson collection of foreign bodies removed from children between 1920 and 1932 with data collected from North American children's hospitals between 1988 and 2000 found that coins have replaced safety pins as the objects most commonly swallowed by American children. In Asia, on the other hand, fish bones are a frequent offender because fish is a dietary staple in most countries of the Far East.

In younger children, boys are at slightly greater risk than girls (53% to 47%) of swallowing foreign objects. Among teenagers, however, males are at a much higher risk than females of swallowing foreign bodies or inserting them into the rectum.

Younger children usually swallow or insert foreign objects into their bodies accidentally, usually as a result of **play** or exploring their environment. Adolescents, however, are more likely to swallow or insert foreign bodies intentionally as a risk-taking behavior, a bid for attention, or under the influence of drugs or alcohol. Adolescent girls with eating disorders have been reported to swallow toothbrushes. A small minority of teenagers who harm themselves by swallowing or inserting foreign bodies suffer from **schizophrenia** or another psychotic disorder.

Causes and symptoms

Causes

The causes of foreign body ingestion or insertion range from traumatic accidents or casual exploration and play to intentional risk-taking, desire for sexual stimulation, an eating or personality disorder, or psychotic behavior. Cases of repeated swallowing of foreign objects by small children may indicate neglect or a dysfunctional home environment.

Symptoms

The symptoms of foreign body ingestion or insertion depend in part on the organ or part of the body affected.

EYES Dust, dirt, sand, or other airborne material can lodge in the eyes as a result of high wind or an explosion, causing minor irritation and redness. More serious damage can be caused by hard or sharp objects that penetrate the surface of the eye and become embedded in the cornea or conjunctivae (the mucous membranes lining the inner surface of the eyelids). Swelling, redness, bleeding from the surface blood vessels, sensitivity to light, and sudden vision problems are all symptoms of foreign matter in the eyes.

EARS AND NOSE Toddlers sometimes put small objects into their noses, ears, and other openings. Beans, dried peas, popcorn kernels, hearing-aid batteries, raisins, and beads are just a few of the many items that have been found in these bodily cavities. On occasion, insects may also fly into a child's ears or nose. **Pain**, hearing loss, and a feeling of fullness in the ear are symptoms of foreign bodies in the ears. A smelly or bloody discharge from one nostril is a symptom of foreign bodies in the nose.

AIRWAYS AND STOMACH At a certain age children will eat almost anything. A very partial list of items recovered from young stomachs includes the following: coins, chicken bones, fish bones, beads, pebbles, plastic **toys**, pins, keys, buckshot, round stones, marbles, nails, rings, batteries, ball bearings, screws, staples, washers, a heart pendant, a clothespin spring, and a toy soldier. Some of these items will pass right on through the digestive tract and leave the body through the feces. The progress of metal objects has been successfully followed with a metal detector or **x rays**. Other objects, like needles, broken poultry bones, or razor blades, can get stuck at various points in the digestive tract and cause trouble.

Most complications of swallowed foreign bodies occur in the esophagus at one of three points: the thoracic inlet at collarbone level (70%); the mid-esophagus (15%); and the sphincter at the lower end of the esophagus where the esophagus joins the stomach (15%). If a swallowed object passes into the stomach, it is unlikely to cause complications unless it is either sharp and pointed in shape or made of a toxic material.

Some foreign objects lodge in the airway. Although children eat small objects and stick things into their bodily openings of their own volition, they inhale them unwittingly while **choking**. Probably the most commonly inhaled item is a peanut. Items as unusual as crayons and cockroaches have also been found in children's windpipes. These items always cause symptoms (difficulty swallowing and spitting up saliva, for instance) and may elude detection for some time while the child is being treated for **asthma** or recurring **pneumonia**.

RECTUM Sometimes a foreign object will successfully pass through the throat and stomach only to get stuck at the juncture between the rectum and the anal canal. Items may also be self-introduced to enhance sex-

ual stimulation and then get stuck in the rectum. Sudden sharp pain during elimination may signify that an object is lodged in the rectum. Other symptoms vary depending upon the size of the object, its location, how long it has been in place, and whether or not infection has set in.

When to call the doctor

The specific symptoms of foreign body ingestion vary somewhat depending on the item and its location in the body. Parents or caregivers may observe the child swallowing the object, or the child may report doing so. In general, parents should take the child to the doctor or emergency room in any of the following situations occurs:

- foreign bodies in the eyes or skin that are the result of an automobile accident, explosion, gunshot injury, or similar trauma
- foreign body appears to have caused an infection in the surrounding tissue
- foreign body is made of lead or contains corrosive chemicals (most commonly batteries)
- foreign body is pointed or has sharp edges (needles, pins, broken glass, toothpicks, razor blades, pop-off tabs from soda cans, etc.)
- child complains of pain on swallowing, pain in the chest, abdominal pain, or severe pain on defecation
- child drools heavily
- child coughs up, vomits, or defecates blood
- child loses consciousness or becomes delirious as a result of esophageal or airway blockage
- child is known to have Crohn's disease, Meckel's diverticulum, or other chronic disorder of the digestive tract (These disorders increase the risk of complications from swallowed foreign bodies.)

Diagnosis

In most cases the doctor needs only a brief history to determine what type of foreign object is involved and where it may be lodged in the child's body. Objects in the ear, nose, or eye can usually be seen on visual examination. In the case of swallowed objects, the doctor examine the inside of the child's mouth and throat to look for signs of tissue damage and bleeding. The doctor may perform a digital examination to locate objects lodged in the rectum.

In general, the doctor may use an endoscope to look for a foreign object in the body as well as to remove it. He or she may order an x ray of the neck, chest, and/or abdomen to locate a foreign body in the esophagus, airway, or lower digestive tract. Most foreign bodies swallowed by small children are radiopaque, which means that they show up on a standard x ray. Metal detectors can successfully identify the location of soda can tops and other aluminum objects that will not show up on an x ray.

Blood tests are not usually necessary unless the doctor suspects that the foreign body has caused an infection or bleeding.

Treatment

Eyes

Small particles like sand may be removable without medical help, but if the object is not visible or cannot be retrieved, prompt emergency treatment is necessary. Trauma to the eyes can lead to loss of vision and should never be ignored. Before an adult attempts any treatment, he or she should move the child to a well-lighted area where the object can be more easily spotted. Hands should be washed and only clean, preferably sterile, materials should make contact with the eyes. If the particle is small, it can be dislodged by blinking or pulling the upper lid over the lower lid and flushing out the speck. A clean cloth can also be used to pick out the offending particle. Afterwards, the eye should be rinsed with clean, lukewarm water or an ophthalmic wash.

If the foreign object cannot be removed at home, the eye should be lightly covered with sterile gauze to discourage rubbing. A physician will use a strong light and possibly special eye drops to locate the object. Surgical tweezers can effectively remove many objects. An antibiotic sterile ointment and a patch may be prescribed. If the foreign body has penetrated the deeper layers of the eye, an eye surgeon will be consulted for emergency treatment.

Ears and nose

A number of ingenious extraction methods have been devised for removing foreign objects from the nose and ears. A bead in a nostril, for example, can be popped out by blowing into the mouth while holding the other nostril closed. Skilled practitioners have removed peas from children's ears by tiny improvised corkscrews and marbles by cotton-tipped applicators with super glue. Tweezers often work well, too. Insects can be floated out of the ear by pouring warm (not hot) mineral oil, olive oil, or baby oil into the ear canal. Metal objects can be removed from the nose or ears with the help of a magnet. Items that are lodged deep in the ear canal are more difficult to remove because of the possibility of damaging the eardrum. These require emergency treatment from a qualified physician.

Airways and stomach

Mechanical obstruction of the airways, which commonly occurs when food gets lodged in the throat, can be treated by applying the **Heimlich maneuver**. If the object is lodged lower in the airway, a bronchoscope (a special instrument to view the airway and remove obstructions) can be inserted. On other occasions, as when the object is blocking the entrance to the stomach, a fiberoptic endoscope (an illuminated instrument that views the interior of a body cavity) may be used. The physician typically administers a sedative and anesthetizes the child's throat. The foreign object then is either pulled out or pushed into the stomach, depending on whether the physician thinks it will pass through the digestive tract on its own. Objects in the digestive tract that are not irritating, sharp, or large may be followed as they continue on through. Sterile objects that are causing no symptoms may be left in place. Surgical removal of the offending object is necessary, however, if it contains a toxic substance; is likely to penetrate the stomach wall; or is longer than 2.36 inches (6 cm) or wider than 0.8 inches (2 cm).

Rectum

A rectal retractor can remove objects that a physician can feel during a digital examination of the rectum. In most cases the doctor will inject a local anesthetic before extracting the object. Surgery under general anesthesia may be required for objects deeply lodged within the body, as in the case of a 14-year-old Dutch adolescent who had inserted a soda can into his rectum.

Treatment of any health problem related to a foreign body may include a psychiatric consultation if the doctor suspects that the swallowing or insertion of the foreign body is related to **autism** or **mental retardation** (in small children) or an eating or personality disorder (in adolescents).

Prognosis

The prognosis of foreign body ingestion or insertion varies according to the nature of the object and its location in the body but is quite good in most cases. With regard to foreign bodies in the digestive tract, between 80 percent and 90 percent pass through without incident; 10–20 percent can be removed with an endoscope; and fewer than 1 percent require surgical removal.

Prevention

Using common sense and following safety precautions are the best ways to prevent foreign objects from

KEY TERMS

Bronchoscope—A lighted instrument that is inserted into the windpipe to view the bronchi and bronchioles, to remove obstructions, or to withdraw specimens for testing.

Conjunctiva—Plural, conjunctivae. The mucous membrane that covers the white part of the eyes (sclera) and lines the eyelids.

Cornea—The clear, dome-shaped outer covering of the eye that lies in front of the iris and pupil. The cornea lets light into the eye.

Crohn's disease—A chronic, inflammatory disease, primarily involving the small and large intestine, but which can affect other parts of the digestive system as well.

Endoscopy—Visual examination of an organ or body cavity using an endoscope, a thin, tubular instrument containing a camera and light source. Many endoscopes also allow the retrieval of a small sample (biopsy) of the area being examined, in order to more closely view the tissue under a microscope.

Heimlich maneuver—An emergency procedure for removing a foreign object lodged in the airway that is preventing the person from breathing. To perform the Heimlich maneuver on a conscious adult, the rescuer stands behind the victim and encircles his waist. The rescuer makes a fist with one hand and places the other hand on top, positioned below the rib cage and above the waist. The rescuer then applies pressure by a series of upward and inward thrusts to force the foreign object back up the victim's trachea.

Meckel's diverticulum—A congenital abnormality of the digestive tract consisting of a small pouch off the wall of the small bowel that was not reabsorbed before birth. A Meckel's diverticulum increases the risk that a foreign object in the digestive tract will get trapped or stuck in the small intestine and cause problems.

Radiopaque—Not penetrable by x rays. A radiopaque object will look white or light when the x-ray film is developed. Most objects that children swallow can be detected by an x-ray study because they are radiopaque.

Sphincter—A circular band of muscle that surrounds and encloses an opening to the body or to one of its hollow organs. These muscles can open or close the opening by relaxing or contracting.

entering the body. For instance, parents and grandparents should toddler-proof their homes, storing batteries in a locked cabinet and properly disposing of used batteries, so they are not in a location where curious preschoolers can retrieve them from a wastebasket. Sewing kits, razor blades, and other potentially dangerous items should also be stored in childproof locations. To minimize the chance of youngsters inhaling food, parents should not allow children to eat while walking or playing. Fish should be carefully boned before it is served to younger children. Many eye injuries can be prevented by wearing safety glasses while using tools or participating in certain **sports**.

Parental concerns

Parental concerns in younger children should be directed toward the prevention of accidental swallowing or ingestion of foreign bodies. In most cases, these accidents can be successfully treated when they do occur, and they are unlikely to cause long-term damage to the child's health. In addition, small children are not likely to repeat behaviors that result in a trip to the doctor's office or hospital emergency room.

Ingestion or insertion of foreign bodies in older children and adolescents is a matter of greater concern to parents, however, because it is much more likely to be intentional, to reflect the presence of an eating disorder or other psychiatric problem, to be a repeated behavior, and to result in serious bodily harm.

See also Heavy metal poisoning; Lead poisoning.

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Foster care

Definition

Foster care is full-time substitute care of children outside their own home by people other than their biological or adoptive parents or legal guardians.

Description

Children who are removed from their biological or adoptive parents, or other legal guardians, are placed in foster care in a variety of settings. They may be placed in the care of relatives other than the **family** members involved in the neglect or abuse (kin placement), non-relatives, therapeutic or treatment foster care, or in an institution or group home.

Children come to foster care for a number of reasons. In many cases, they have suffered physical or sexual abuse, or neglect at home, and are placed in a safe environment. A small percentage of children are in foster care because their parents feel unable to control them, and their behavior may have led to delinquency or **fear** of harm to others. Some children have been neglected by their parents or legal guardians, or have parents or legal guardians who are unable to take care of them because of substance abuse, incarceration, or mental health problems. These children are placed into custodial care while the parents or guardians receive treatment or counseling, or fulfill their sentences.

In all foster care cases, the child's biological or adoptive parents, or other legal guardians, temporarily give up legal custody of the child. (The guardian gives up custody, but not necessarily legal guardianship.) A child may be placed in foster care with the parents' consent. In a clear case of abuse or neglect, a court can order a child into foster care without the parents' or guardians' consent. Foster care does not necessarily mean care by strangers. If a government agency decides a child must be removed from the home, the child may be placed with relatives or with a family friend. Children may also be placed in a group home, where several foster children live together with a staff of caregivers. Therapeutic or treatment foster care can be in a group home or foster home with a specific structure and treatment focus. Foster homes are the most well-known option. The child temporarily becomes a part of another family, either with other foster children, the family's biological or adoptive children, or alone. State or county social service agencies oversee foster care decisions, although they may also work with private foundations.

Foster parents must be licensed by the agency that handles a specific region's foster care. The foster home must pass an inspection for health and **safety** and, in most states, the parents must attend training sessions covering issues of foster care and how to deal with problems. When a child is placed, the foster family takes responsibility for feeding and clothing the child, getting the child to school and to appointments, and doing any of the usual things a child's parents or legal guardians might be called to do. The foster parents might also need to meet with the foster child's therapist and will meet regularly with the child's caseworker as well. The foster parent aims to help the foster child develop normally in a safe, family environment.

Foster parents usually receive money for taking in foster children. They are expected to use the money to buy the child's food, clothing, school supplies, and other incidentals. Most of the foster parent's responsibilities toward the foster child are clearly defined in a legal contract. Foster parents do not become the guardians of foster children; legal guardianship remains with the state agency.

Foster placements may last for a single day or several weeks; some continue for years. If the parents give up their rights permanently, or their rights to their child are severed by the court, the foster family may adopt the foster child or the child may be placed for **adoption** by strangers. Foster parenting is meant to be an in-between stage, while a permanent placement for the child is settled. As such, it is stressful and uncertain, but for many families very necessary.

Federal money supports most foster care programs, and federal law governs foster care policy. The Adoption Assistance and Child Welfare Reform Act of 1980 emphasizes two aims of foster care. One is to preserve the child's family, if at all possible. Children are placed in foster care only after other options have failed, and social service agencies work with the family to resolve its problems so that children can return to their homes. The second aim of the Child Welfare Reform Act is to support the so-called "permanency planning." This means that if a child must be removed from the home, the social service agency handling the case can decide quickly whether or not the child will ever be returned. If it seems likely that parents will not be able to care for their children again, their parental rights may be terminated so that the child is free to be adopted. This policy is articulated in this law in order to prevent children from living too long in an unstable and uncertain situation.

The goal of foster care is the care of the child within the child welfare system, but also is to place all appropriate and available services at the disposal of the parents so that they can create a safe, fit home environment for their children when they are reunited. Children in the child welfare system are also overseen by a multitude of agencies. The caseworker from the state or county social services agency oversees the child's placement and makes regular reports to the court. Others involved in the child's case are private service providers (including foster homes and group homes), welfare agencies, mental health counselors, substance abuse treatment centers (for the child or the parent), and Medicaid (federal medical insurance for seniors and children at risk).

Demographics

In 1980, about 300,000 children in the United States spent some time in foster care placement. By 2001, there were nearly 800,000 children in foster care, with 540,000 children in the system at any given time. The majority of these children were the victims of abuse. The emergence of widespread homelessness, substance abuse (especially crack and methamphetamines), unemployment, increased incarceration rates, street violence, and HIV/AIDS have all impacted poor communities. Children from families with multiple problems flooded the child welfare system. Young children with physical handicaps, mental delays or mental illness, and complex medical conditions have become the fastest-growing foster care population.

The foster care population is quite young. About one-fourth of all children entering foster care for the first time are infants. Sixty percent of foster children are under four years old. Teenagers comprise one-third of the foster care population. Minority children comprise most of the foster care population, with the largest groups being African American and American Indian children.

Poor children are more likely to be in foster care than middle-class children because their families have fewer resources. Illness or loss of a job may be devastating to a poor family with no savings and no relatives who can afford to assist them. These children are also more likely to stay in foster care longer or to have been in foster care since infancy. Also, children of alcoholics or drug abusers are at high risk for neglect or abuse, and comprise 75 percent of all placements.

More than half (57 percent) of all children in foster care are returned to their original homes; however, reunification rates have declined in the 1990s and early twenty-first century. Children also spend more time in the system. The average length of stay for a child in foster care is 33 months. However, some spend a very short time in a foster home, and others are there for their entire childhoods, ‘‘aging out’’ at 18 when they become legal adults.

Instead of reunification, more children are being adopted from foster care. Most states doubled, and some tripled, the number of foster care adoptions since 1997. This steady increase is a response to the Adoption and Safe Families Act (ASFA) of 1997 that recommends termination of parental rights and encourages adoption if a child has been in foster care for 15 out of the previous 22 months. This can be waived by the court if the parents are making substantive progress or the caseworker believes that legal guardianship, but not adoption, is in the child’s best interests.

Half of all children in foster care live with nonrelative foster caregivers; about one-fourth live with relatives, and this number is growing. ASFA also recognized kinship caregivers as legitimate placements. It was customary for many poor families to take in a child informally when the child’s parents or legal guardians were incarcerated, in treatment, or had died, but ASFA allowed relatives to take care of a child legally and receive financial help, and also opened the doors to a number of agencies and services the relatives could not afford.

Common problems

In most cases, children placed in foster care have been subjected to some form of abuse or neglect, and being removed from familiar surroundings is, in itself, usually highly traumatic. Children in foster care may have **nightmares**, problems sleeping or eating, and may be depressed, angry, and confused. Many young children in foster care are unable to understand why they have been taken from their parents. Even if a child is in some sense relieved to be out of a home that was dangerous, the child may still miss the parents or legal guardians, and may imagine that there is something he or she must do to get back to them. There is evidence that children from abusive and neglectful homes start to feel better in foster care; however, separation is almost always difficult for children, regardless of the circumstances.

Half of all foster children spend as much as two years in foster care and are moved from placement to placement at least three times. This leads not only to uncertainty and lack of stability in the child’s life, but some of these placements may be inappropriate for the child’s specific circumstances. This often is due to the lack of qualified, licensed foster caregivers, but it can also occur as a result of inexperienced or overloaded caseworkers trying to get through their caseloads.

Foster care can be difficult for foster parents as well. A child who has been neglected or abused suffers psychological damage that may make him or her withdrawn, immature, aggressive, or otherwise difficult to reach. Children with severe medical and mental problems can tax caregivers. Foster placements sometimes fail because these surrogate parents simply cannot handle the demands of a troubled foster child.

Unfortunately, the number of foster caregivers has been declining since the mid-1980s as the demand for placements has increased. States have responded by licensing responsible adults who were not married (even divorced men and women) and reaching out to seniors and children’s relatives. In some areas, single mothers make up a large proportion of foster parents.

KEY TERMS

Adoption—The legal process that creates a parent and child relationship between two individuals who are not biologically related at birth.

Age out—Become a legal adult at age 18 and move out of foster care.

Medicaid—A program jointly funded by state and federal governments that reimburses hospitals and physicians for the care of individuals who cannot pay for their own medical expenses. These individuals may be in low-income households or may have chronic disabilities.

In 2002, about 405,000 children were placed in court-appointed kinship care. Caseworkers placed almost 140,000 more in the care of relatives, without court intervention. Many of these kinship caregivers are grandparents or elderly aunts and uncles. Kinship caregivers offer family support and stability, and more frequent contact with parents or legal guardians, and siblings. They also are more apt to get children to talk to them about their problems, and the presence of relatives can help ease the trauma of separation from parents.

Nevertheless, kinship caregivers, especially grandparents, face a number of challenges. Most of the formal and informal kinship caregivers experience economic hardship as they take in one or more of their relative's children. Nearly two-thirds of these placements are with financially strapped families who may not have essentials such as a car seat, crib, or **toys**. They also may not have adequate medical insurance; however, Medicaid often will cover the foster child in a formal kinship arrangement. Grandparents may not know how to raise a child in today's world, with the amount of freedoms or lack of them that children experience today. They may not be able to help their foster children with homework. Many social service agencies offer counseling, homework help, and even home tutoring for both the child and caregiver.

One other problem inherent in the child welfare system is the teenager who "ages out," or turns 18 and moves out of foster care to live independently. Many teenagers mark time within the system, without adequate preparation for the transition to adulthood. Less than one-fourth of social service agencies provide employment services for teenagers. Only 17 percent provide employment and career assessments, and 16 percent provide job-training. One-fourth offer vocational training. Without help, these teenagers often never go on to college, do not find good jobs if they find jobs at all, and become prey to bad influences on the street. If they have children of their own, these offspring fall back into the child welfare system just as they did. Adequate training, counseling, and preparation can break this cycle.

In addition, children in all types of foster care face more challenges financially, emotionally, and developmentally. A study by the Child Welfare League of America in 2004 showed that children in foster care experienced more health and developmental problems than children who had similar economic circumstances but lived with their parents or legal guardians. Foster children also have more neglect, abuse, family dysfunction, poverty, and emotional problems. This may be a direct result of the reasons for their initial placement, but these conditions continue throughout foster care. Another reason for these results may be that foster children are given more frequent and thorough medical and psychological care than their counterparts.

Parental concerns

Other foster care placements are made by families who cannot afford medical or psychological services for their children. These children may have multiple disabilities or severe social or mental disorders that have depleted the family's financial and emotional resources. Convinced by social workers that this option is the only one available to them, they give up their parental rights in order to get their children into proper treatment. According to the U.S General Accounting Office, 12,700 children were placed into the child welfare system or the juvenile justice system to receive mental health services in 2001. Despite the noble reasons for placing these children in foster care, the parents' names are placed on state registries as child abusers, and they have to petition the court and prove their fitness to get their children returned to their homes.

See also Child abuse.

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Foster Care Children. 507 North Sullivan Road Suite A-6. Spokane Valley, WA 99037. (509) 924-3175. Web site: <www.fostercarechildren.com>.

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Fractures

Definition

A fracture is a complete or incomplete break in a bone resulting from the application of excessive force.

Description

A fracture usually results from traumatic injury to a bone, causing the continuity of bone tissues or bony cartilage to be disrupted or broken. Fracture classifications include simple or compound and incomplete or complete. Simple fractures (often called "closed") are not obvious as the skin has not been ruptured and remains intact. Compound fractures (commonly called "open") break the skin, exposing bone and causing additional soft tissue injury and possible infection. A single fracture means that one fracture has occurred, and multiple fractures refer to more than one fracture occurring in the same bone. Fractures are termed complete if the break is completely through the bone and described as incomplete or "greenstick" if the fracture occurs partly across a bone shaft. This latter type of fracture is often the result of bending or crushing forces applied to a bone.

Fractures are also named according to the specific part of the bone involved and the nature of the break. Identification of a fracture line can further classify fractures. Types include linear, oblique, transverse, longitudinal, and spiral fractures. Fractures can be further subdivided by the positions of bony fragments and are described as comminuted, non-displaced, impacted, overriding, angulated, displaced, avulsed, and segmental. Additionally, an injury may be classified as a fracture-dislocation when a fracture involves the bony structures of any joint with associated dislocation of the same joint.

Fractures line identification

Linear fractures have a break that runs parallel to the bone's main axis or in the direction of the bone's shaft. For example, a linear fracture of the arm bone could extend the entire length of the bone. Oblique and transverse fractures differ in that an oblique fracture crosses a bone at approximately a 45° angle to the bone's axis. In contrast, a transverse fracture crosses a bone's axis at a 90° angle. A longitudinal fracture is similar to a linear fracture. Its fracture line extends along the shaft but is more irregular in shape and does not run parallel to the bone's axis. Spiral fractures are described as crossing a bone at an oblique angle, creating a spiral pattern. This break usually occurs in the long bones of the body such as the upper arm bone (humerus) or the thigh bone (femur).

Bony fragment position identification

Comminuted fractures have two or more fragments broken into small pieces, in addition to the upper and lower halves of a fractured bone. Fragments of bone that maintain their normal alignment following a fracture are described as being non-displaced. An impacted fracture is characterized as a bone fragment forced into or onto another fragment resulting from a compressive force. Overriding is a term used to describe bony fragments that overlap and shorten the total length of a bone. Angulated fragments result in pieces of bone being at angles to each other. A displaced bony fragment occurs from disruption of normal bone alignment with deformity of these segments separate from one another. An avulsed fragment occurs when bone fragments are pulled from their normal position by forceful muscle contractions or resistance from ligaments. Segmental fragmented positioning occurs if fractures in two adjacent areas occur, leaving an isolated central segment. An example of segmental alignment occurs when the arm bone fractures in two separate places, with displacement of the middle section of bone.

Demographics

The exact number of fractures sustained in the United States each year is not known as many are not treated. Experts estimate the number of fractures at between 10 and 20 million. People of all ages and races experience fractures. Broken bones are slightly more common among children due to their increased level of activity

and among older people due to their lack of **exercise** and inadequate intake of calcium.

Causes and symptoms

Individuals with high activity levels appear to be at greater risk for fractures. This group includes children and athletes participating in contact **sports**. Because of an increase in bone brittleness with aging, elderly persons are also included in this high-risk population. Up to the age of 50, more men suffer from fractures than women due to occupational hazards. However, after the age of 50, women are more prone to fractures than men. Specific diseases causing an increased risk for fractures include Paget's disease, rickets, **osteogenesis imperfecta**, osteoporosis, bone **cancer** and tumors, and prolonged disuse of a nonfunctional body part such as after a **stroke**.

Symptoms of fractures usually begin with **pain** that increases with attempted movement or use of the area and swelling at the involved site. The skin in the area may be pale and an obvious deformity may be present. In more severe cases, there may be a loss of pulse below the fracture site, such as in the extremities, accompanied by **numbness**, **tingling**, or paralysis below the fracture. An open or compound fracture is often accompanied by bleeding or bruising. If the lower limbs or pelvis are fractured, pain and resistance to movement usually accompany the injury causing difficulty with weight bearing.

When to call the doctor

A physician should be called when a child complains of bone pain. This is a deep pain that may be exquisitely tender to the touch.

Diagnosis

Diagnosis begins immediately with an individual's own observation of symptoms. A thorough medical history and physical exam by a physician often reveals the presence of a fracture. An x ray of the injured area is the most common test used to determine the presence of a bone fracture. Any x-ray series performed involves at least two views of the area to confirm the presence of the fracture because not all fractures are apparent on a single x ray. Some fractures are often difficult to see and may require several views at different angles to see clear fracture lines. In some cases, CT, MRI, or other imaging tests are required to demonstrate fracture. Sometimes, especially with children, the initial x ray may not show any fractures, but if it is repeated seven to 14 days later, the x ray may show changes in the bone(s) of the affected area. If a fracture is open and occurs in conjunction with soft tissue injury, further laboratory studies are often conducted to determine if blood loss has occurred.

In the event of exercise-related stress fractures (micro-fractures due to excessive stress), a tuning fork can provide a simple, inexpensive test. The tuning fork is a metal instrument with a stem and two prongs that vibrate when struck. If an individual has increased pain when the tuning fork is placed on a bone, such as the tibia or shinbone, the likelihood of a stress fracture is high. Bone scans also are helpful in detecting stress fractures. In this diagnostic procedure, a radioactive tracer is injected into the bloodstream and images are taken of specific areas or the entire skeleton by CT or MRI.

Treatment

Treatment depends on the type of fracture, its severity, the individual's age, and the person's general health. The first priority in treating any fracture is to address the entire medical status of the patient. Medical personnel are trained not to allow a painful, deformed limb to distract them from potentially life-threatening injury elsewhere or shock. If an open fracture is accompanied by serious soft tissue injury, it may be necessary to control bleeding and the shock that can accompany loss of blood.

First aid is the appropriate initial treatment in emergency situations. It includes proper splinting, control of blood loss, and monitoring vital signs such as breathing and circulation.

Immobilization

Immobilization of a fracture site can be done internally or externally. The primary goal of immobilization is to maintain the realignment of a bone long enough for healing to start and progress. Immobilization by external fixation uses splints, casts, or braces. This may be the primary and only procedure for fracture treatment. Splinting to immobilize a fracture can be done with or without traction. In emergency situations if the injured individual must be moved by someone other than a trained medical person, splinting is a useful form of fracture management. It should be done without causing additional pain and without moving the bone segments. In a clinical environment, plaster of Paris casts are used for immobilization. Braces are useful as they often allow movement above and below a fracture site. Treatments for stress fractures include rest and decreasing or stopping any activity that causes or increases pain.

KEY TERMS

Avulsion fracture—A fracture caused by the tearing away of a fragment of bone where a strong ligament or tendon attachment forcibly pulls the fragment away from the bone tissue.

Axis—A line that passes through the center of the body or body part.

Comminuted fracture—A fracture where there are several breaks in a bone creating numerous fragments.

Compartment syndrome—A condition in which the blood supply to a muscle is cut off because the muscle swells but is constricted by the connective tissue around it.

Contrast hydrotherapy—A series of hot and cold water applications. A hot compress (as hot as an individual can tolerate) is applied for three minutes followed by an ice cold compress for 30 seconds. These applications are repeated three times each and ending with the cold compress.

Osteogenesis imperfecta—An inherited disorder of the connective tissues that involves multiple symptoms, including weakened bones that break easily.

Osteoporosis—Literally meaning "porous bones," this condition occurs when bones lose an excessive amount of their protein and mineral content, particularly calcium. Over time, bone mass and strength are reduced leading to increased risk of fractures.

Paget's disease—A chronic disorder of unknown cause usually affecting middle aged and elderly people and characterized by enlarged and deformed bones. Changes in the normal mechanism of bone formation occur in Paget's disease and can cause bones to weaken, resulting in bone pain, arthritis, deformities, and fractures. Also known as osteitis deformans.

Reduction—The restoration of a body part to its original position after displacement, such as the reduction of a fractured bone by bringing ends or fragments back into original alignment. The use of local or general anesthesia usually accompanies a fracture reduction. If performed by outside manipulation only, the reduction is described as closed; if surgery is necessary, it is described as open. Also describes a chemical reaction in which one or more electrons are added to an atom or molecule.

Rickets—A condition caused by the dietary deficiency of vitamin D, calcium, and usually phosphorus, seen primarily in infancy and childhood, and characterized by abnormal bone formation.

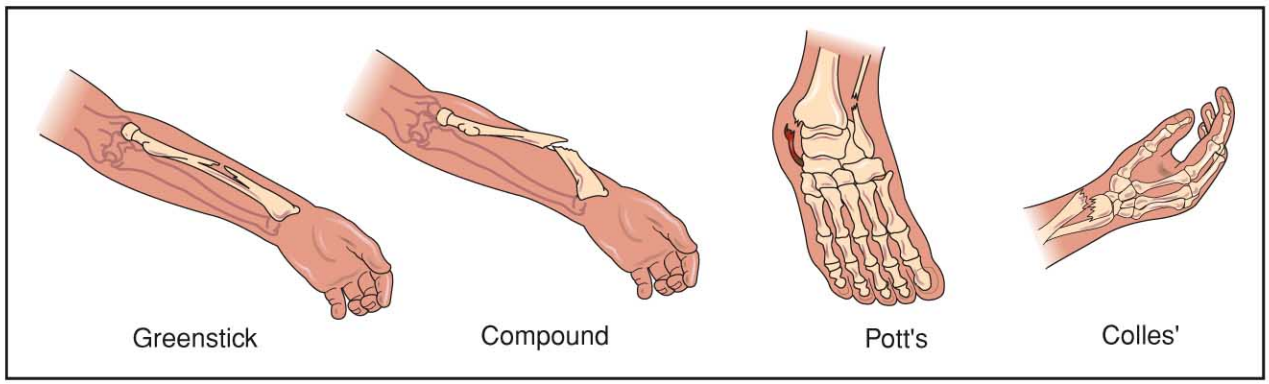
Traction—The process of placing a bone, limb, or group of muscles under tension by applying weights and pulleys. The goal is to realign or immobilize the part or to relieve pressure on that particular area to promote healing and restore function.

Fracture reduction

Fracture reduction is the procedure by which a fractured bone is realigned in normal position. It can be either closed or open. Closed reduction refers to realigning bones without breaking the skin. It is performed with manual manipulation and/or traction and is commonly done with some kind of anesthetic. Open reduction primarily refers to surgery that is performed to realign bones or fragments. Fractures with little or no displacement may not require any form of reduction.

Traction is used to help reposition a broken bone. It works by applying pressure to restore proper alignment. The traction device immobilizes the area and maintains realignment as the bone heals. A fractured bone is immobilized by applying opposing force at both ends of the injured area, using an equal amount of traction and countertraction. Weights provide the traction pull needed or the pull is achieved by positioning the individual's body weight appropriately. Traction is a form of closed reduction and is sometimes used as an alternative to surgery. Since it restricts movement of the affected limb or body part, it may confine a person to bed rest for an extended period of time.

A person may need open reduction if there is an open, severe, or comminuted fracture. This procedure allows a physician to examine and surgically correct associated soft tissue damage while reducing the fracture and, if necessary, applying internal or external devices. Internal fixation involves the use of metallic devices inserted into or through bone to hold the fracture in a set position and alignment while it heals. Devices include plates, nails, screws, and rods. When healing is complete, the surgeon may or may not remove these devices. Virtually any hip fracture requires open reduction and internal fixation so that the bone will be able to support the patient's weight.



Fractures usually result from a traumatic injury to a bone where the continuity of bone tissues or bony cartilage is disrupted or broken. The illustrations above feature common sites where fractures occur. *(Illustration by Electronic Illustrators Group.)*

Alternative treatment

In addition to the importance of calcium for strong bones, many alternative treatment approaches recommend use of mineral supplements to help build and maintain a healthy, resilient skeleton. Some physical therapists use electro-stimulation over a fractured site to promote and expedite healing. Chinese traditional medicine may be helpful by working to reconnect chi (life energy) through the meridian lines along the line of a fracture. Homeopathy can enhance the body's healing process. Two particularly useful homeopathic remedies are arnica (*Arnica montana*) and symphytum (*Symphytum officinalis*). If possible, applying contrast hydrotherapy to an extremity (e.g., a hand or foot) of a fractured area can assist healing by enhancing circulation.

Prognosis

Fractures involving joint surfaces almost always lead to some degree of arthritis of the joint. Fractures can normally be cured with proper first aid and appropriate aftercare. If determined necessary by a physician, the fractured site should be manipulated, realigned, and immobilized as soon as possible. Realignment has been shown to be much more difficult after six hours. Healing time varies from person to person with the elderly generally needing more time to heal completely. A non-union fracture may result when a fracture does not heal, such as in the case of an elderly person or an individual with medical complications. Recovery is complete when there is no bone motion at the fracture site, and **x rays** indicate complete healing. Open fractures may lead to bone infections, which delay the healing process. Another possible complication is compartment syndrome, a painful condition resulting from the expansion of enclosed tissue and that may occur when a body part is immobilized in a cast.

Prevention

Fractures can be prevented if **safety** measures are taken seriously. These measures include using seat belts in cars and encouraging children to wear protective sports gear. Weight-bearing exercise also helps to strengthen bones.

Nutritional concerns

Persons who consume diets that are rich in calcium are less likely to experience a fracture than those who have diets that are deficient in calcium. Good dietary sources of calcium are milk, cheese, and other dairy products.

Parental concerns

Parents should ensure that their children drink milk to provide an adequate intake of calcium. Children should also participate in regular physical exercise.

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American Academy of Family Physicians. 11400 Tomahawk Creek Parkway, Leawood, KS 66211–2672. Web site: <www.aafp.org/>.

American Academy of Orthopedic Surgeons. 6300 North River Road, Rosemont, Illinois 60018–4262. Web site: <www.aaos.org/>.

American Academy of Pediatrics. 141 Northwest Point Boulevard, Elk Grove Village, IL 60007–1098. Web site: <www.aap.org/default.htm>.

American Academy of Physical Medicine and Rehabilitation. One IBM Plaza, Suite 2500, Chicago, IL 60611–3604. Web site: <www.aapmr.org/>.

American College of Foot and Ankle Surgeons. 515 Busse Highway, Park Ridge, Illinois 60068–3150. Web site: <www.acfas.org/index.html>.

American College of Sports Medicine. 401 W. Michigan St., Indianapolis, IN 46202–3233. Web site: <www.acsm.org/>.

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Fragile X syndrome

Definition

Fragile X syndrome, a genetic condition involving changes in the long arm of the X chromosome, is the most common form of inherited **mental retardation**. Individuals with this condition have **developmental delay**, variable levels of mental retardation, and behavioral and emotional difficulties. They may also have characteristic physical traits. Generally, males are affected with moderate mental retardation (since they only have one X chromosome) and females with mild mental retardation.

Description

Fragile X syndrome is the most common form of inherited mental retardation in the United States. Fragile X syndrome is caused by a mutation in the FMR-1 gene, located on the X chromosome. The FMR-1 gene is thought to play an important role in the development of the brain, but the exact way that the gene acts in the body is not fully understood. Language delays, behavioral problems, **autism** or autistic-like behavior (including poor eye contact and hand-flapping), enlarged genitalia (macroorchidism), large or prominent ears, hyperactivity, delayed motor development, and/or poor sensory skills are among the wide range of characteristics associated with this disorder.

Fragile X syndrome is also known as Martin-Bell syndrome, Marker X syndrome, and FRAXA syndrome.

Demographics

Fragile X syndrome affects males and females of all ethnic groups. A summary of existing research conducted by the Centers for Disease Control and Prevention in 2001 estimated that approximately one in 3,500–8,900 males is affected by the full mutation of the FMR-1 gene and that one in 1,000 males has the premutation form of the FMR-1 gene. This study also estimated that one in 250–500 females in the general

population has the premutation. Another study estimated that one in 4,000 females is affected by the full mutation.

Causes and symptoms

For reasons not fully understood, the CGG sequence in the FMR-1 gene can expand through succeeding generations to contain between 54 and 230 repeats. This stage of expansion is called a premutation. People who carry a premutation do not usually have symptoms of fragile X syndrome, although there have been reports of individuals with a premutation who have subtle intellectual or behavioral symptoms. Individuals who carry a fragile X premutation are at risk for having children or grandchildren with the premutation. Female premutation carriers may also be at increased risk for earlier onset of menopause.

Premutation carriers may exist through several generations of a **family** though no symptoms of fragile X syndrome appear. However, the size of the premutation can expand over succeeding generations. When a man carries a premutation on his X chromosome, it tends to be stable and usually will not expand if he passes it on to his daughters (he passes his Y chromosome to his sons). Thus, all of his daughters will be premutation carriers like he is. When a woman carries a premutation, it is unstable and can expand as she passes it on to her children; therefore, a man's grandchildren are at greater risk of developing the syndrome. There is a 50 percent risk for a premutation carrier female for transmitting an abnormal mutation with each pregnancy. The likelihood for the premutation to expand is related to the number of repeats present; the higher the number of repeats, the greater the chance that the premutation will expand to a full mutation in the next generation. All mothers of a child with a full mutation are carriers of an FMR-1 gene expansion.

Once the size of the premutation exceeds 230 repeats, it becomes a full mutation, and the FMR-1 gene is disabled. Individuals who carry the full mutation may have fragile X syndrome. Since the FMR-1 gene is located on the X chromosome, males are more likely to develop symptoms than females. This greater inclination occurs because males have only one copy of the X chromosome. Males who inherit the full mutation are expected to have mental impairment. A female's normal X chromosome may compensate for her chromosome with the fragile X gene mutation. Females who inherit the full mutation have an approximately 30–50 percent risk of mental impairment, ranging from mild learning disability to mental retardation and behavioral problems.

Another feature of fragile X syndrome is that mosaicism is present in 15 to 20 percent of those affected by the condition. Mosaicism refers to the presence of cells of two different genetic materials in the same individual.

Individuals with fragile X syndrome appear normal at birth, but their development is delayed. Most boys with fragile X syndrome have mental impairment. The severity of mental impairment ranges from learning disabilities to severe mental retardation. Behavioral problems include attention deficit and hyperactivity at a young age. Some may show **aggressive behavior** in adulthood. Short attention span, poor eye contact, delayed and disordered speech and language, emotional instability, and unusual hand mannerisms (hand flapping or hand biting) are also seen frequently. Other behavioral characteristics include whirling, spinning, and occasionally autism or autistic-like behavior.

Characteristic physical traits appear later in childhood. These traits include a long and narrow face, prominent jaw, large ears, and enlarged testes. In females who carry a full mutation, the physical and behavioral features and mental retardation tend to be less severe. About 50 percent of females who have a full mutation are mentally retarded.

Children with fragile X syndrome often have frequent ear and sinus infections. Nearsightedness and lazy eye are also common. Many babies with fragile X syndrome may have trouble with sucking, and some experience digestive disorders that cause frequent gagging and **vomiting**. A small percentage of children with fragile X syndrome may experience seizures. Children with fragile X syndrome also tend to have loose joints, which may result in joint dislocations. Some children develop a curvature in the spine, flat feet, and a heart condition known as mitral valve prolapse.

When to call the doctor

If a child exhibits delayed development and mental impairment and has other symptoms typical of fragile X syndrome, the doctor should be consulted to determine the cause of the problems.

Diagnosis

A birth, there may be few outward signs of fragile X syndrome in the newborn infant. However, fragile X symptoms may include a large head circumference and oversized testes in males. An experienced geneticist may recognize subtle differences in facial characteristics.

However, any child with signs of developmental delay of speech, language, or motor development with

KEY TERMS

Amniocentesis—A procedure performed at 16-18 weeks of pregnancy in which a needle is inserted through a woman's abdomen into her uterus to draw out a small sample of the amniotic fluid from around the baby for analysis. Either the fluid itself or cells from the fluid can be used for a variety of tests to obtain information about genetic disorders and other medical conditions in the fetus.

CGG or CGG sequence—Shorthand for the DNA sequence: cytosine-guanine-guanine. Cytosine and guanine are two of the four molecules, called nucleic acids, that make up DNA.

Chorionic villus sampling—A procedure performed at 10 to 12 weeks of pregnancy in which a needle is inserted either through the mother's vagina or abdominal wall into the placenta to withdraw a small amount of chorionic membrane from around the early embryo. The chorionic membrane can be examined for signs of chromosome abnormalities or other genetic diseases.

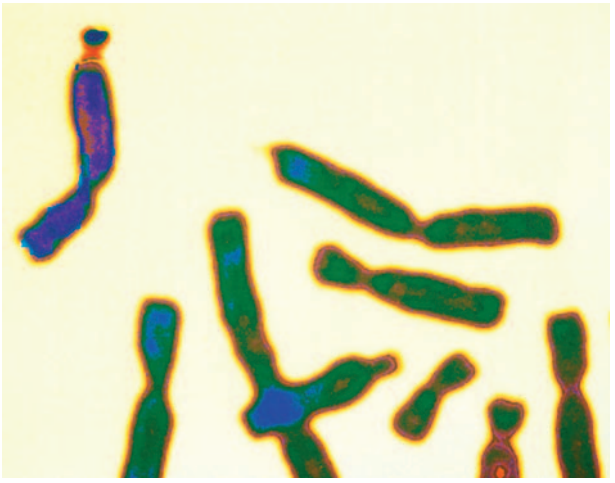
Chromosome—A microscopic thread-like structure found within each cell of the human body and consisting of a complex of proteins and DNA. Humans have 46 chromosomes arranged into 23 pairs. Chromosomes contain the genetic information necessary to direct the development and functioning of all cells and systems in the body. They pass on hereditary traits from parents to child (like eye color) and determine whether the child will be male or female.

FMR-1 gene—A gene found on the X chromosome. Its exact purpose is unknown, but it is suspected that the gene plays a role in brain development.

Mitral valve prolapse—A heart defect in which the mitral valve of the heart (which normally controls blood flow from the left atrium to the left ventricle) becomes floppy. Mitral valve prolapse may be detected as a heart murmur but there are usually no symptoms.

Premutation—A change in a gene that precedes a mutation; this change does not alter the function of the gene.

X chromosome—One of the two sex chromosomes (the other is Y) that determine a person's gender. Normal males have both an X and a Y chromosome, and normal females have two X chromosomes.



Fragile X chromosome, shaded in purple at upper left corner, is shown among other chromosomes. *(© Siebert/Custom Medical Stock Photo, Inc)*

no known cause should be considered for fragile X testing, especially if there is a family history of the condition. Behavioral and developmental problems may indicate fragile X syndrome, particularly if there is a family history of mental retardation. Definitive identification of the fragile X syndrome is made by means of a genetic test to assess the number of CGG sequence repeats in the FMR-1 gene. Individuals with the premutation or full mutation may be identified through genetic testing. Genetic testing for and detection of the fragile X mutation can be performed on the developing baby before birth through **amniocentesis**, chorionic villus sampling (CVS), and percutaneous umbilical blood sampling. Prenatal testing is recommended after the fragile X carrier status of the parents has been confirmed, and the couple has been counseled regarding the risks of recurrence.

Prognosis

Early diagnosis and intensive intervention offer the best prognosis for individuals with fragile X syndrome. Adults with fragile X syndrome may benefit from vocational training and may need to live in a supervised setting. About 50 percent of affected individuals develop mitral valve prolapse, a heart condition, as adults. However, life span is typically normal.

Prevention

Neither the fragile X premutation nor mutation is preventable as of 2004. Genetic counseling may help prospective parents with a family history of fragile X

syndrome. Genetic testing can help determine the level of risk in the family.

Parental concerns

A child with fragile X syndrome requires many services, so parents must be prepared to invest significant time and resources to ensure the child receives the help that he or she needs.

Families may wish to seek counseling regarding the effects of the syndrome on relationships within the family. Many people respond with guilt, **fear**, or blame when a genetic disorder is diagnosed in the family, or they may overprotect the affected member. Support groups are often good sources of information about fragile X syndrome; they can offer helpful suggestions about living with it as well as emotional support.

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ORGANIZATIONS

Arc of the United States (formerly Association for Retarded Citizens of the United States). 500 East Border St., Suite 300, Arlington, TX 76010. Web site: <http://thearc.org>.

FRAXA Research Foundation. 45 Pleasant Street, Newburyport, MA 01950. Web site: <www.fraxa.org>.

National Fragile X Foundation. PO Box 190488, San Francisco, CA 94119–0988. Web site: <www.FragileX.org>.

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Friedreich's ataxia

Definition

Friedreich's ataxia (FA) is an inherited, progressive nervous system disorder causing loss of balance and coordination, speech problems, and heart disease.

Description

FA is an inherited disease marked by impaired coordination that is a result of degeneration of the structures in the cerebellum and the spinal cord, which are responsible for coordination, muscle movement, and some sensory functions, including color vision and hearing. The intellect of a child with FA is normal.

FA is an autosomal recessive disease, which means that two defective gene copies, one from each parent, must be inherited to develop symptoms. A person with only one defective gene copy will not show signs of FA, but may pass along the gene to offspring. Couples with one child affected by FA have a 25 percent chance in each pregnancy of conceiving another affected child.

FA is also referred to as spinocerebellar degeneration.

Demographics

Friedreich's ataxia is the most common inherited ataxia, affecting one in 50,000 people in the United States. Females and males are affected equally.

Causes and symptoms

Causes

The gene for FA codes for a protein called frataxin. Normal frataxin is found in the cellular energy structures known as mitochondria, where it is thought to be involved in regulating the transport of iron. In FA, the frataxin gene on chromosome 9 is expanded when a particular sequence of bases in the DNA is repeated too many times. Ordinarily, there are seven to 22 repeats of the frataxin gene; in FA, this sequence may be repeated between 800 to 1,000 times. This extra DNA interferes with normal production of frataxin, thereby impairing iron transport. The triplet repeat expansion seems to interfere with the normal assembly of amino acids into proteins, significantly reducing the amount of frataxin that is made. Without a normal level of frataxin, some of the body's cells—especially those of the brain, spinal cord, and muscle—cannot handle the normal amounts of "oxidative stress," which the mitochondria produce. When excess iron in the cells (as a result of the

deficiency of frataxin) reacts with oxygen, free radicals are produced. Free radicals are necessary molecules in the body's metabolism, but in excess they can also destroy cells and harm the body.

The types of symptoms and severity of FA seems to be associated with the number of repetitions. Children with more copies have more severe symptomatology, with symptoms starting at a younger age.

The nerve cells most affected by FA are those in the spinal cord involved in relaying information between muscles and the brain. Control of movement requires complex feedback between the muscles promoting a movement, those restraining it, and the brain. Without this control, movements become uncoordinated, jerky, and inappropriate to the desired action.

Symptoms

Symptoms of FA usually first appear between the ages of five and 15 years, although onset as early as 18 months or as late as age 30 years is possible. The first symptom is usually gait incoordination. A child with FA may graze doorways when passing through, for instance, or trip over low obstacles. Unsteadiness when standing still and deterioration of position sense is common. Children with FA may develop foot deformities such as **clubfoot**, hammertoe, and high arches. Walking up off the heels often results from uneven muscle weakness in the legs. **Muscle spasms and cramps** may occur, especially at night. Other early symptoms include changes in speech, swallowing difficulties, loss of reflexes, and jerky eye movements (**nystagmus**).

Ataxia in the arms follows, usually within several years, leading to decreased **hand-eye coordination**. Arm weakness does not usually occur until much later. There is often a gradual loss of sensation in the extremities, which may spread to other parts of the body. In about 10 percent of children with FA, **diabetes mellitus** may develop in the later stages of the disease. Some loss of visual acuity may be noted. Hearing loss occurs in about 10 percent of children with FA, and about 20 percent develop **carbohydrate intolerance**. A side-to-side curvature of the spine (**scoliosis**) occurs in many cases, and may become severe. About 50 percent of people develop problems with control of their urge to urinate (urinary urgency), or become incontinent.

Various forms of heart disease often accompany FA, including cardiomyopathy (enlargement of the heart), myocardial fibrosis (formation of fiber-like materials in heart muscles), and cardiac failure. Symptoms of heart involvement include chest **pain**, shortness of breath, and heart palpitations. Heartbeat abnormalities such as tachycardia (rapid heart rate) and heart block (impaired conduction of the heart's cardiac impulses) are common occurrences.

When to call the doctor

Any time a child with FA reports unusual heart symptoms, such as shortness of breath on exertion, **dizziness**, fainting, chest pain or discomfort, or abnormal heart rhythms, the doctor or cardiologist should be called, or the child should be taken immediately to a hospital emergency room.

Diagnosis

Diagnosis of FA involves a careful medical history and thorough neurological exam. Laboratory tests include electromyography (a measurement of the electrical activity of muscle cells) and nerve conduction velocity tests, which measure the speed that nerves transmit impulses. An electrocardiogram and echocardiogram may be performed to diagnose heart disease. Imaging studies are conducted to provide pictures of the brain and spinal cord. A spinal tap is performed to evaluate the cerebrospinal fluid. Blood and urine samples are tested for elevated glucose levels, to determine whether the child has diabetes.

Direct DNA testing is available, allowing FA to be more easily distinguished from other types of ataxia. The same test may be used to determine the presence of the genetic defect in unaffected individuals, such as siblings.

Treatment

There is no cure for FA, nor is there any treatment that can slow its progress. Therefore, the goal of treatment is to control symptoms and maintain general health. Amantadine may provide some limited improvement in ataxic symptoms, but is not recommended in children with cardiac abnormalities. Physical therapy and activity are used to maintain range of motion in weakened muscles and to compensate for loss of coordination and strength. Some children find that using weights on the arms can help dampen the worst of the uncoordinated arm movements. Scoliosis and foot deformities can be treated with braces or surgery.

Safety is an important consideration in this disease since the child will eventually experience loss of balance and sensation. Occupational therapy is recommended to select adaptive techniques and devices such as safety railings, walkers, or other safety appliances. If the child loses feelings in various body parts, injuries can be avoided by testing bath water to prevent **burns**, inspect-

KEY TERMS

Ataxia—A condition marked by impaired muscular coordination, most frequently resulting from disorders in the brain or spinal cord.

Oxidative stress—A condition where the body is producing an excess of oxygen-free radicals.

Scoliosis—An abnormal, side-to-side curvature of the spine.

ing the body visually for injuries, and using protective shoes and helmets.

Diabetes is treated with insulin and dietary changes. Some of the heart problems can be treated with medications.

Since the disease may be associated with damage to cells caused by free radicals, antioxidants such as vitamin E and coenzyme Q10 are often prescribed for children with FA.

Prognosis

The rate of progression of FA is highly variable. Most children lose the ability to walk within 15 to 20 years after the onset of symptoms, and will require aids for walking such as scooters, walkers, or wheelchairs. In later stages of the disease, people become incapacitated.

Reduction in lifespan from FA complications is also quite variable. Average age at death is in the mid-thirties, but may be as late as the mid-sixties in persons with less severe symptoms. Many persons with FA will develop untreatable heart disease, which may shorten life expectancy.

Prevention

There is no way to prevent development of FA in a person carrying two defective gene copies. Genetic counseling and testing are recommended for prospective parents with a **family** history of FA.

Parental concerns

Coping with the challenges of raising a child with a chronic serious disease is difficult for parents and other family members. Psychological counseling and support groups are invaluable tools to help families meet the challenges and provide the child with needed support.

A child with Friedreich's ataxia is entitled to an Individual Education Plan (IEP) through the Individuals with Disabilities Education Act (IDEA). An IEP team consisting of parents, administrators, teachers, and sometimes the student and outside experts, will collaborate to develop the child's IEP.

Children with FA are considered high-risk for flu and **pneumonia**. The children and family members should receive a flu shot every year, unless there are other health considerations that would indicate that there is a reason not to receive the vaccine. The children should also periodically receive pneumococcal pneumonia shots as recommended by their doctors.

See also Movement disorders.

Resources

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ORGANIZATIONS

Friedreich's Ataxia Research Alliance. 2001 Jefferson Davis Hwy, Suite 209, Arlington, VA 22202. (703)413-4468. Fax: (703) 413-4467. Web site: <www.frda.org>.

Muscular Dystrophy Association. 3300 East Sunrise Drive, Tucson, AZ 85718-3208. (520) 529-2000 or (800) 572-1717. Fax: 520-529-5300. Web site: <www.mdausa.org>.

National Ataxia Foundation. 2600 Fernbrook Lane, Suite 119, Minneapolis, MN 55447-4752. (763) 553-0020. Web site: <www.ataxia.org>.

National Organization for Rare Disorders (NORD). P.O. Box 1968, 55 Kenosia Avenue, Danbury, CT 06813-1968. (203) 744-0100 or (800) 999-6673. Fax: (203) 798-2291. Web site: <www.rarediseases.org>.

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Friedreich's Ataxia Fact Sheet. Available online at: <www.ninds.nih.gov/health_and_medical/pubs/friedreich_ataxia.htm>.

Friedreich's Ataxia Parents Group (FAPG). Available online at: <www.fortnet.org/fapg/>.

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Frostbite and frostnip

Definition

Frostbite is damage to the skin and other tissues caused by freezing. Frostnip is a mild form of this cold injury.

Description

Skin exposed to temperatures a little below 32°F (0°C) can take hours to freeze, but very cold skin can freeze in minutes or seconds. Nevertheless, under extreme conditions, even warm skin exposed to subzero temperatures and high wind chill factors can freeze rapidly. Air temperature, wind speed, humidity, and altitude all affect how cold the skin becomes. A strong wind can lower skin temperature considerably by dispersing the thin protective layer of warm air that surrounds the body. Wet clothing readily draws heat away from the skin because water is a potent conductor of heat. The evaporation of moisture on the skin also produces cooling. For these reasons, wet skin or clothing on a windy day can lead to frostbite even if the air temperature is above freezing.

The extent of permanent injury, however, is determined not by how cold the skin and the underlying tissues become but by how long they remain frozen. When skin is exposed to freezing temperatures, three things happen. The skin begins to freeze, causing ice crystal formation, damage to capillaries (the tiny blood vessels that connect the arteries and veins), and other changes that damage and eventually kill cells. Much of this harm occurs because the ice produces pressure changes that force water (crucial for cell survival) out of the cells.

Tissue hypoxia, or oxygen deficiency, occurs next as a survival mechanism in the body kicks in, causing the blood vessels in the hands, feet, and other extremities to narrow in response to cold. Among its many tasks, blood transfers body heat to the skin, which then dissipates the heat into the environment. Blood vessel narrowing is the body's way of protecting vital internal organs at the expense of the extremities by reducing heat flow away from the center of the body. However, blood also carries life-sustaining oxygen to the skin and other tissues, and narrowed vessels result in oxygen starvation. Narrowing also causes acidosis (an increase in tissue acidity) and increases blood viscosity (thickness). Ultimately, blood stops flowing through the capillaries) and blood clots form in the arterioles and venules (the smallest arteries and veins). Damage also occurs to the endothelial cells that line the blood vessels.

Hypoxia, blood clots, and endothelial damage lead, in turn, to the release of inflammatory mediators. These are substances that act as links in the inflammatory process, which promote further blood vessel damage, hypoxia, and cell destruction. Tissue damage is greatest when skin is exposed to freezing slowly or over a long period of time. More damage can occur when rewarming is slow or the affected area is warmed and refrozen.

Demographics

In North America, frostbite frequently occurs in Alaska, Canada, and the northern states, which have extremely cold winter temperatures. Frostbite, however, can occur almost anywhere, given the right conditions. Though there has been in the early 2000s a substantial decline in the number of frostbite cases in the United States, due to better winter clothing and footwear and greater public understanding of how to avoid cold-weather dangers, these cases are rising among the homeless who do not have adequate clothing or shelter. Frostbite has thus become an urban as well as a rural public health concern. The growing popularity of outdoor winter activities has also expanded the at-risk population.

Children are at a higher risk of experiencing frostbite and frostnip than adults because they experience heat loss from their skin more rapidly. Those children with disorders that affect circulation, such as diabetes, may be even more susceptible to frostbite and frostnip. Children who have had a recent injury, surgery, or blood loss are at risk, as well as teenagers who might be **smoking**, drinking alcohol, or taking beta-blockers for high blood pressure or a heart condition. Also, children who have had a frostbite injury in the past are more prone to having a recurrence in the same location. In addition, children from tropical climates may not be able to withstand cold temperatures as well as their cold-climate counterparts, making them more susceptible to frostbite and frostnip at higher temperatures.

Causes and symptoms

Causes

Skin damage from frostbite and frostnip occurs because of freezing, either by extremely cold weather,

wet clothing in cold temperatures, or through chemical exposures, such as dry ice or highly compressed gases. Most children encounter frostbite when they participate in outdoor **sports**, camp in winter, get wet and cannot change their clothing immediately, or do not dress according to the weather conditions. Frostnip and frostbite are associated with ice crystal formation in the tissues.

Symptoms

In frostnip, no tissue destruction occurs and the ice crystals dissolve as soon as the skin is warmed. Frostnip affects areas such as the earlobes, cheeks, nose, fingers, and toes. The skin turns pale, and the person experiences **numbness** or **tingling** in the affected part until warming begins.

Frostbite, by contrast, has a range of severity. Most injuries affect the hands and feet, but about 10 percent of all frostbite cases affect the nose, cheeks, ears, and even the penis. Frostbite is classified by degree of injury (first, second, third, or fourth), or simply divided into two types, superficial (corresponding to first- or second-degree injury) and deep (corresponding to third- or fourth-degree injury). Frostnip is sometimes labeled a first-degree frostbite case.

Once frostbite sets in, the affected part begins to feel cold and, usually, numb. This condition is followed by a feeling of clumsiness. The skin turns white or yellowish. Many patients experience severe **pain** in the affected part during rewarming treatment and an intense throbbing pain that arises two or three days later and can last days or weeks. As the skin begins to thaw during treatment, edema (excess tissue fluid) often accumulates, causing swelling. In frostbite injuries of second-degree or higher, blisters appear. Third-degree cases produce deep, blood-filled blisters and a hard black eschar (scab). Fourth-degree frostbite penetrates below the skin to the muscles, tendons, nerves, and bones. Septicemia or blood poisoning and infection may also be present, as well as the possible need for amputation (the surgical removal of appendages such as fingers, toes, foot, or leg).

When to call the doctor

If a child's clothing has been wet for a long period of time or the child has been exposed to freezing temperatures, shows skin discoloration, and complains of feeling numb, the child should be seen by a doctor. In most cases, the child will be hospitalized to monitor the rewarming process and to do the necessary tests needed to determine the extent of the frostbite. Prolonged exposure to extreme temperatures can also produce hypothermia (lowered body temperature), which can be life threatening.

Diagnosis

Initial diagnosis is usually made based on the environmental conditions. Physical examination of the skin reveals that the skin is extremely cold and may have white, red, blue, or black areas on it. The patient may report feeling numb or a tingling sensation.

Frostbite diagnosis may also include conventional radiography (x rays), angiography (x-ray examination of the blood vessels using an injected dye to provide contrast), thermography (use of a heat-sensitive device for measuring blood flow), and other techniques for predicting the course of injury and identifying tissue that requires surgical removal. During the initial treatment period, however, a physician cannot judge how a case may progress. Diagnostic tests only become useful three to five days after rewarming, once the blood vessels have stabilized.

Treatment

Frostnip

Frostnipped fingers are helped by blowing warm air on them or holding them under one's armpits. Other frostnipped areas can be covered with warm hands. The injured areas should never be rubbed.

Frostbite

Emergency medical help should always be summoned whenever frostbite is suspected. While waiting for help to arrive, one should, if possible, remove wet or tight clothing and put on dry, loose clothing or cover with a blanket. Rubbing the area with snow or anything else is dangerous because it can cause tissue damage. The key to prehospital treatment is to avoid partial thawing and refreezing, which releases more inflammatory mediators and makes the injury substantially worse. For this reason, the affected part must be kept away from heat sources such as campfires and car heaters. Experts advise rewarming in the field only when emergency help will take more than two hours to arrive and refreezing can be prevented.

Because the outcome of a frostbite injury cannot be predicted at first, all hospital treatment follows the same routine. Treatment begins by rewarming the affected part for 15 to 30 minutes in water at a temperature of 104–108°F (40–42°C). This rapid rewarming halts ice crystal formation and dilates narrowed blood vessels. Aloe vera (which acts against inflammatory mediators) is applied

KEY TERMS

Acidosis—A disturbance of the balance of acid to base in the body causing an accumulation of acid or loss of alkali (base). Blood plasma normally has a pH of 7.35-7.45. Alkaline blood has a pH value greater than pH 7.45. When the blood pH value is less than 7.35, the patient is in acidosis.There are two types of acidosis: metabolic and respiratory. One of the most common causes of metabolic acidosis is an overdose of aspirin. Respiratory acidosis is caused by impaired breathing caused by conditions such as severe chronic bronchitis, bronchial asthma, or airway obstruction.

Amputation—Surgical removal of any portion of the body.

Angiography—Radiographic examination of blood vessels after injection with a radiopaque contrast substance or dye.

Arteriole—The smallest type of artery.

Capillaries—The tiniest blood vessels with the smallest diameter. These vessels receive blood from the arterioles and deliver blood to the venules. In the lungs, capillaries are located next to the alveoli so that they can pick up oxygen from inhaled air.

Hypothermia—A serious condition in which body temperature falls below 95°F (35 °C). It is usually caused by prolonged exposure to the cold.

Hypoxia—A condition characterized by insufficient oxygen in the cells of the body

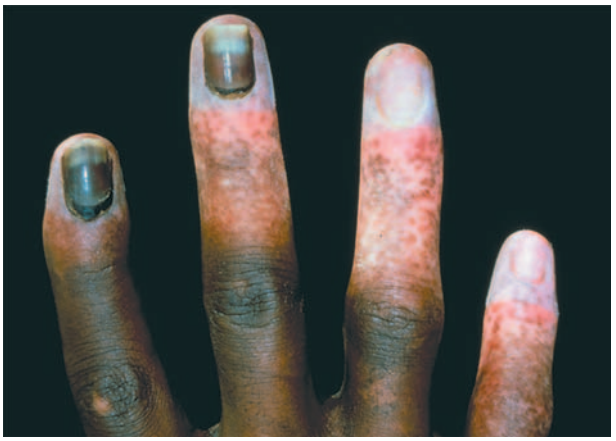
Radiography—Examination of any part of the body through the use of x rays. The process produces an image of shadows and contrasts on film.

Thermography—Use of a heat-sensitive device for measuring blood flow.

Venules—The smallest veins.

Viscosity—Thickness of a liquid.

to the affected part, which is then splinted, elevated, and wrapped in a dressing. Depending on the extent of injury, blisters may be debrided (cleaned by removing foreign material) or simply covered with aloe vera. A **tetanus** shot and, possibly, penicillin, are used to prevent infection, and the patient is given ibuprofen to combat inflammation. Narcotics are needed in most cases to reduce the excruciating pain that occurs as sensation returns during rewarming. Except when injury is minimal, treatment generally requires a hospital stay of several days, during



Hand with effects of frostbite. *(Photograph by SIU. National Audubon Society Collection/Photo Researchers, Inc.)*

which hydrotherapy and physical therapy are used to restore the affected part to health. Experts recommend a cautious approach to tissue removal and advise that 22 to 45 days must pass before a decision on amputation can safely be made.

Alternative treatment

Alternative practitioners suggest several kinds of treatment to speed recovery from frostbite after a person leaves the hospital. Bathing the affected part in warm water or using contrast hydrotherapy can help enhance circulation. Contrast hydrotherapy involves a series of hot and cold water applications. A hot compress (as hot as the patient can stand) is applied to the affected area for three minutes followed by an ice-cold compress for 30 seconds. These applications are repeated three times each, ending with the cold compress.

Nutritional therapy to promote tissue growth in damaged areas may also be helpful. Homeopathic and botanical therapies may also assist recovery from frostbite. Homeopathic *Hypericum* (*Hypericum perforatum*) is recommended when nerve endings are affected (especially in the fingers and toes) and *Arnica* (*Arnica montana*) is prescribed for shock. Cayenne pepper (*Capsicum frutescens*) can enhance circulation and relieve pain. Drinking hot ginger (*Zingiber officinale*) tea also aids circulation. Other possible approaches include acupuncture to avoid permanent nerve damage and oxygen therapy.

Prognosis

The rapid rewarming approach to frostbite treatment, pioneered in the 1980s, has proved to be much

more effective than older methods in preventing tissue loss and amputation. A study of 56 first-, second-, and third-degree frostbite patients treated with rapid rewarming between 1982 and 1985 found that 68 percent recovered without tissue loss, 25 percent experienced some tissue loss, and 7 percent needed amputation. In a comparison group of 98 patients, treatment using older methods resulted in a tissue loss rate of nearly 35 percent and an amputation rate of nearly 33 percent. Although the comparison group included a higher proportion of second- and third-degree cases, the difference in treatment results was determined to be statistically significant.

The extreme throbbing pain that many frostbite sufferers endure for days or weeks after rewarming is not the only prolonged symptom of frostbite. During the first weeks or months, people often experience tingling, a burning sensation, or a sensation resembling shocks from an electric current. Other possible consequences of frostbite include skin-color changes, nail deformation or loss, joint stiffness and pain, **hyperhidrosis** (excessive sweating), and heightened sensitivity to cold. For everyone, a degree of sensory loss lasting at least four years, and sometimes a lifetime, is inevitable.

Prevention

With the appropriate knowledge and precautions, frostbite can be prevented even in the coldest and most challenging environments. Appropriate clothing and footwear are essential. To prevent heat loss and keep the blood circulating properly, clothing should be worn loosely and in layers. Covering the hands, feet, and head is also crucial for preventing heat loss. Children especially should wear hats that cover their heads and ears, mittens, and coats that are wind and water resistant. Wet clothing and footwear must be removed as quickly as possible and replaced with dry clothing and shoes.

Alcohol and drugs should be avoided because of their harmful effects on judgment and reasoning. Experts also warn against alcohol use and smoking in the cold because of the circulatory changes they produce.

Parental concerns

Parents should pay close attention to weather reports before sending children out to **play** or to take part in long-exposure outdoor activities such as sledding, skiing, and winter camping. Listening to winter driving warnings and road reports is also important before taking trips in the winter or in the mountains.

In addition, parents should keep a close eye on their children when the children play outdoors in winter around lakes, streams, and other water sources. Even older children and teenagers can slip on ice or snow and fall in. The risk of hypothermia and frostbite is too great to ignore. Sometimes, even a child's getting his or her shoes wet and then continuing to play in the cold can produce serious frostbite.

See also X rays.

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Fructose intolerance *see* **Hereditary fructose intolerance**

Fugue disorder *see* **Dissociative disorders**

Fused fingers and toes *see* **Polydactyly and syndactyly**

G

Galactosemia

Definition

Galactosemia is an inherited disease in which the body is unable to metabolize the simple sugar galactose, which is found primarily in dairy products but is also produced by the body. If left unaddressed, galactose can increase to toxic levels in the body and may lead to damage of the liver, central nervous system, and various other body systems.

Description

Galactosemia is a rare but potentially life-threatening disease resulting from the body's inability to metabolize galactose. Galactose makes up half of lactose, the sugar that is found in milk. Persons with galactosemia either have very low levels of or are entirely lack the enzyme that assists the body in breaking down galactose. This enzyme is called galactose-1-phosphate uridyl transferase (GALT). The GALT enzyme enables the body to break down galactose into glucose for energy. The severity of the disease may vary from person to person, because some individuals with galactosemia have higher levels of this enzyme than do others.

The two main types of galactosemia are called classic and Duarte variant. Individuals with the classic type of galactosemia lack the enzyme activity necessary to metabolize galactose. Individuals with the Duarte variant have approximately 5–20 percent of the enzyme activity necessary to metabolize this sugar and often do not have signs or symptoms of galactosemia.

Transmission

Galactosemia is an hereditary disease. In order to understand this disorder, it is necessary to have a very basic knowledge of genetics. Genes, the basic components of hereditary material, contain the "blueprint" that directs the development and functioning of every cell and tissue in the human body. Genes are situated on larger structures called chromosomes that contain several thousand genes each. Genes and chromosomes exist in pairs. Every cell in the body has 23 pairs of chromosomes containing two copies of every gene. Human beings receive one copy of every gene from their mother, and another copy from their father. Most of the time, genes function normally. However, in some cases, a change or mutation in a gene can cause it to not function. These mutations can cause inherited or genetic disorders.

In galactosemia, the gene mutation which occurs is inherited in what is known as an autosomal recessive pattern. This means that a non-working copy of the gene must be inherited from both parents for a child to be affected with the disease. The parents of children are called "carriers" of the disorder, because though they themselves do not have galactosemia, they may have children who do.

Unfortunately, as of 2004, parents had no way of knowing if they carry the mutated gene that causes galactosemia until they have a child diagnosed with the disease. The chance that two parents each of whom has the defective gene will produce a child with a recessive disorder is one in four or 25 percent with each pregnancy. The chances that their child will be a carrier, like themselves, is one in two, or 50 percent, with each pregnancy.

Demographics

Galactosemia is an inherited disorder that occurs in approximately one out of 30,000 live births. The incidence for the Duarte variant type of galactosemia is estimated to be one in 16,000 live births. Although galactosemia occurs in all ethnic groups worldwide, some mutations cause a less severe type of disease and are more commonly seen in specific ethnic groups, such as African-Americans. In Japan, classic galactosemia is not diagnosed as frequently as it is in Caucasian populations in the United States.

Causes and symptoms

Galactosemia is an inherited disorder. People with the disease are unable to fully break down galactose. If an infant with galactosemia is given milk, byproducts of galactose will build up in the baby's body, causing damage to the liver, kidneys, brain, and eyes. Characteristically, a newborn with galactosemia who is fed milk products will have **jaundice**, **vomiting**, lethargy, irritability, and convulsions. Continued feeding of milk products to the infant will lead to cirrhosis, cataracts, kidney failure, and **mental retardation**.

When to call the doctor

Parents should notify their doctor if their child displays any of the symptoms of galactosemia. Couples should consult their physician if there is a **family** history of galactosemia, and they are considering having a child.

Diagnosis

As of late 2004, all 50 states had mandatory screening of newborns for galactosemia. If parents receive a call from a healthcare provider saying the screening test indicates possible galactosemia, they should promptly stop milk products and have a blood test done for galactosemia through their doctor. The physician may also perform enzyme studies on or look for the presence of "reducing substances" in the child's urine, look for ketones in the urine, and measure enzyme activity in the red blood cells.

Treatment

Galactosemia is treated by removing foods that contain galactose from the diet. Foods containing lactose and, therefore, galactose should be avoided. Because milk and milk products are the most common food source of galactose, persons with galactosemia should avoid ingesting these foods. It is recommended that persons with galactosemia avoid eating foods with galactose throughout their entire lives.

Nutritional concerns

The goal of dietary treatment is to minimize galactose intake. It is impossible to have a galactose-free diet. However, all persons with galactosemia should limit galactose intake as much as possible. The galactose-1-phosphate levels of the individual will establish the level of dietary restriction necessary. Infants can be fed soy, meat-based, or other lactose-free formulas. Abstinence from milk and milk products must continue throughout life. Parents need to find some of the many listings available that identify the galactose content in foods. Since the primary source of calcium is usually milk or milk-based foods, calcium must be supplemented. Parents of a child with galactosemia should note that lactose is often used as filler in medicines. This very frequently is not listed on the package. Parents should always check with the pharmacist before administering any new medications.

KEY TERMS

Galactose—One of the two simple sugars (glucose is the other one) that makes up the protein, lactose, found in milk. Galactose can be toxic in high levels.

Recessive trait—An inherited trait or characteristic that is outwardly obvious only when two copies of the gene for that trait are present.

Prognosis

Approximately 75 percent of the babies with galactosemia who are not diagnosed and treated die within the first two weeks of life. By contrast, if an early diagnosis is made and if milk products are strictly avoided, most children lead a relatively normal life. With appropriate treatment, liver and kidney problems do not develop, and early mental development progresses in a normal manner. However, even with proper treatment, children with galactosemia often have a lower **intelligence** quotient (IQ) than their siblings, and they frequently have speech problems. Girls often have ovaries that do not function, and only a few are able to conceive naturally. Boys, however, have normal testicular function.

Prevention

Since galactosemia is a recessive genetic disease, it is usually first detected on a newborn screening test, as most people are not aware that they are carriers of a gene mutation causing the disease. If there is a family history of galactosemia, genetic counseling is recommended for prospective parents as they make decisions regarding pregnancy and prenatal testing. Once one child in a family is diagnosed with galactosemia, it is recommended that other members of the family receive genetic counseling as well.

Parental concerns

One of the most important things parents of a child with galactosemia can do is educate themselves thoroughly on an appropriate diet. All other caregivers or teachers involved with the child need to be notified of the child's dietary restrictions, and the potential consequences if they are not maintained.

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Gangs

Definition

Youth gangs are variously defined in the social science and criminal justice literature. They are commonly understood to be a loosely-organized association of socially excluded, alienated, or bigoted individuals acting together within a fluid structure with informal leadership. Youth gangs are bound by a common ethnicity, race, social class, or other determinant and employ distinctive symbols, including style and color of dress, hand signs, **tattoos**, and graffiti. Loyal gang members follow a gang-defined system of rules, rituals, and codes of behavior. Gangs serve some individuals as a substitute **family** structure. Membership imparts a sense of empowerment as members act together to defend territory and provide mutual protection. Youth gangs typically engage in delinquent, criminal, and violent activities, often for financial gain.

Description

Gangs have been a part of U.S. culture since the early 19th century. Immigrant youth organized themselves into street gangs, often as a means of economic survival. Social scientists have been studying and reporting on gang membership and attributes since early in the 20th century. Gangs have been seen as a normal adolescent peer activity that occurs "within a continuum of behaviors, from conventional to wild," as suggested by the classic 1927 research of Frederic Thrasher, a social scientist who studied 1,313 Chicago gangs. A more recent view by the U.S. Department of Justice in 1998 holds that "a group must be involved in a pattern of criminal acts to be considered a youth gang." This criterion is also used by the Federal Bureau of Investigation who contend that it is "participation in criminal activity" that separates a community group or social club from a gang.

Gangs are more prevalent in neighborhoods where the community network is weak, with few ties among individual residents or between residents and conventional community institutions. Among adolescent males, the best predictor of gang membership is the absence of a positive male role model. Most girls who participate in gang activity have run away from home at least once due to family problems including the drug **addiction** and/or arrest of a parent.

Gang violence has reached a crisis level in the United States. A 1998 study revealed that gang members possess significantly more guns than other at-risk youth. The ready availability of such deadly weapons has led to an increase in violence such as drive-by shootings and a loss of life among gang members and others caught in the crossfire. Research reported in 1991 found that gang access to firearms "led to lethal violence in circumstances that might otherwise have been settled with less-than lethal means." Gang culture increasingly involves its youth membership in the use of weapons, drugs, and criminal activity.

Risk factors

According to Lonnie Jackson, author of the book *Gangbusters: Strategies for Prevention and Intervention,* many factors contribute to the likelihood of youth gang involvement. Some of the factors he cites include:

- frequent exposure to crime and violence during formative years
- few positive role models, particularly of their own ethnicity
- unstable family life, with little parental control
- lack of economic opportunities conducive to lawful self-sufficiency
- inadequate constructive social and recreational activities for youths
- hopelessness engendered by minimal employment opportunity
- inadequate skills, education, or employment qualifications
- lure of power and money, particularly through the drug trade
- cultural environment that highly values immediate gratification
- unmet needs for **safety**, a sense of belonging, and secure emotional relationships
- low **self-esteem** and feelings of insignificance and powerlessness

Demographics

"Gang activity is notably prevalent in the biggest cities (over 100,000 population) in the United States," according to research reported by the National Youth Gang Center. Between 1996 and 2001, more than 90 percent of the largest U.S. cities reported gang activity. However, between 1998 and 1999, the research shows an increase in gang membership by 27 percent in suburban areas and by 29 percent in rural areas. Gang membership is no longer limited to ethnic minorities in America's inner cities, but is found in all ethnic groups, economic classes, and in rural, urban, and suburban settings.

Researchers studying gang life focused first on the behavior of male gangs. Later research, however, has revealed a growing number of girl gangs, with estimates as high as 10 percent of all youth gangs. However, the incidence of female gangs may be much higher than reports indicate. Female gang activity is less violent than that of their male counterparts and is underreported by law enforcement agencies.

Gang membership remains predominantly the province of male adolescents and young adults from 12 to 24 years of age. When young women become involved in gangs, it is usually through relationships with boyfriends or brothers, according to research by A. Campbell reviewed in the *Journal of Criminal Justice*. Girl gang members experience more long-term, harmful effects from gang membership than their male counterparts, and some research finds that "gang membership itself opened up young women to additional victimization risk."

The proportion of gang members of particular race or ethnicity reflects the demographics of the community where they live. "Nearly half (49 percent) of all gang members are Hispanic/Latino, 34 percent are African American/black, 10 percent are Caucasian/white, 6 percent are Asian, and the remainder are of some other race/ethnicity," according to respondents to the 2001 National Youth Gang Survey. The Survey estimated that "youth gangs were active in over 2,300 cities with populations over 2,500 in 2002."

Causes

Research studies throughout the 1980s and 1990s, during a period of growing gang involvement among North American youth, cite complex social problems as the root cause of the persistence and proliferation of youth gangs. Dysfunctional families, often with an absent father, low socio-economic circumstances, poor educational opportunities, unemployment, indigence, deteriorated neighborhoods with high crime rates, racism, and limited opportunities for bringing about a change in circumstances, are among the serious factors that put youth at high risk for gang involvement.

Though there is no conclusive evidence, many critics of popular media cite youth exposure to violent films and song lyrics, particularly rap music, as a negative influence glamorizing gang life and encouraging at-risk youth to join gangs or to participate in gang-related crime as a means of gaining a sense of belonging and empowerment.

When to intervene

Early intervention is the most effective means of diverting at-risk youth into pro-social activities and associations before they seek affiliation with youth gangs. Children as young as eight years old are attracted by the lure of gang membership. Parents, teachers, and concerned others should seek the help of culturally-sensitive and well-trained counselors who can intervene with information and alternatives that address unmet needs for safety, and provide a feeling of belonging, and a sense of power and purpose.

Indicators

Concerned and attentive parents and school counselors should be on the alert for indications of possible

gang membership in at-risk youth. Some indicators are poor academic achievement and frequent **truancy**, antisocial and delinquent behaviors, adoption of gang dress in style and color, appearance of tattoos, use of hand signals, and other gang-related signs, preference for music with gang themes, and the presence of gang activity in the community.

Treatment

Effective treatment must be culturally sensitive, diverse, and experienced as relevant to the lives of the gang-involved youth. Treatment plans must address the myriad and serious underlying personal and social problems that lead to gang involvement. Young people need information about alternatives to street gangs that can realistically meet their needs in pro-social ways. Treatment for drug addiction, sexual abuse, and other physical and emotional traumas are a prerequisite to providing lasting help. Mental health treatment must address delayed stress issues from repeated exposure to trauma, violence, and economic hardship. Education and training in skills of nonviolent conflict resolution are also important components of a successful treatment plan. Counselors must be skilled, knowledgeable, and trustworthy and able to help the gang-involved youth to examine choices in ways that encourage clear thinking and provide a broader view of potential and possibilities outside gang life.

Prognosis

Early intervention with at-risk youth to relieve some of the personal and environmental stressors that lead to gang involvement has the best prognosis. Youth who have already joined a gang usually also have well-developed manipulative skills. They exhibit a fierce loyalty to other gang members and are highly resistant to change, even after arrest and detention for gang-related crimes.

Prevention

Community intervention at the grassroots, neighborhood level, can be an effective first step in a multifaceted approach to prevention of gang involvement. Eliminating underlying social problems that lead to development of youth gangs and strengthening community ties can reduce the influence of gangs and deter gang crime that thrives when neighborhoods fail to work together. Parental involvement with teachers can head off many problems of truancy, and community education on gang culture will help parents and teachers to identify early signs of gang involvement. Strong after-school programs that assist working parents meet children's needs for supervision and provide structured, pro-social activities to young children may reduce attraction to gang-related activities. Former gang members who are willing to speak about the negative side of gang life, and adults who are willing to serve as mentors and tutors can provide critical positive role models for at-risk youth, an indispensable component to a successful prevention strategy. Job skills training and meaningful employment opportunities will divert many youth from the path to gang membership.

Parental concerns

The prevalence of youth gangs throughout the United States, and the increase in violence associated with gang membership are serious issues of concern for any parent. Delinquent and antisocial behaviors in young children, particularly those who live in environments where poverty, unemployment, and drug addiction are common, are early danger signs. Seeking help from concerned and qualified school counselors, church, and community leaders can alleviate many parental concerns and provide opportunity for early intervention.

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Gastroenteritis

Definition

Gastroenteritis is an inflammation of the digestive tract, particularly the stomach, and large and small intestines. Viral and bacterial gastroenteritis are intestinal infections associated with symptoms of **diarrhea**, abdominal cramps, **nausea**, and **vomiting**.

Description

Gastroenteritis is an uncomfortable and inconvenient ailment, but is rarely life-threatening in the United States and other developed nations. Viral gastroenteritis is frequently referred to as the stomach or intestinal flu, although the **influenza** virus is not associated with this illness.

Demographics

Viral gastroenteritis is one of the most common acute (sudden-onset) illnesses in the United States, with millions of cases reported annually. Each year, an estimated 220,000 children younger than age five are hospitalized with gastroenteritis symptoms. Of these children, 300 die as a result of severe diarrhea and **dehydration**. In developing nations, diarrheal illnesses are a major source of mortality.

Causes and symptoms

Causes

Gastroenteritis is caused by the ingestion of viruses, certain bacteria, or parasites. Food that has spoiled may also cause illness. Young children may develop signs and symptoms of gastroenteritis as a reaction to a new food.

VIRAL INFECTION Viral infection is the most common cause of gastroenteritis. Viral gastroenteritis is highly contagious and can be spread through close contact with an infected person. Exposure also can occur through the fecal-oral route, such as by consuming foods or beverages contaminated by fecal material related to poor sanitation or poor hygiene, or by touching contaminated surfaces and then touching the mouth and ingesting the germs. The four types of viruses that cause most viral gastroenteritis include rotavirus, adenovirus, calicivirus, and astrovirus.

Typically, children ages three to 15 months are more vulnerable to rotaviruses, the most significant cause of acute watery diarrhea. Outbreaks of diarrhea caused by rotaviruses are common during the winter and early spring months, especially in child care centers. Symptoms in children last for three to eight days, and occur one to two days after exposure to the virus. Worldwide, rotaviruses are estimated to cause 800,000 deaths annually in children under five years of age. For this reason, much research has gone into developing a vaccine to protect children from this virus. Adults can be infected with rotaviruses, but these infections typically have minimal or no symptoms.

Children under age two are more susceptible to adenovirus serotypes 40 and 41. Vomiting and diarrhea symptoms occur about one week after exposure to the virus.

Calciviruses cause infection in people of all ages. This family of viruses includes the noroviruses (such as the Norwalk virus) and the sapoviruses (such as the sapporo virus). Calciviruses are transmitted from person-to-person contact, as well as through contaminated water or food. These viruses are the most likely to produce vomiting as a major symptom. Muscle aches also are common symptoms. The symptoms usually appear within one to three days after exposure to the virus.

Astrovirus primarily infects infants, young children, and the elderly. This virus is most active during the winter months. Symptoms of vomiting and diarrhea appear within one to three days after exposure to the virus.

BACTERIAL AND PARASITIC INFECTIONS Bacterial gastroenteritis is frequently a result of poor sanitation,

the lack of safe drinking water, or contaminated food (conditions common in developing nations). Natural or man-made disasters can worsen underlying problems in sanitation and food safety.

In developed nations, including the United States, bacterial gastroenteritis may result from contaminated water supplies, improperly processed or preserved foods, or person-to-person contact in places such as child-care centers. The modern food production system potentially exposes millions of people to disease-causing bacteria through its intensive production and distribution methods. Common types of bacterial gastroenteritis can be linked to *Salmonella* and *Campylobacter bacteria*. However, *Escherichia coli (E. coli)* 0157:H7 and *Listeria monocytogenes*, bacterial causes of food borne illnesses, have caused increased concern in developed nations.

Cholera and *Shigella* remain two diseases of great concern in developing countries, and research to develop long-term vaccines against them is underway. *Shigella* bacteria are dangerous because they attack the intestinal wall and cause bleeding ulcers.

Parasitic infections that cause gastroenteritis are most commonly caused by *Giardia*, which is easily spread through contaminated water and human contact. *Cryptosporidium* is another common parasitic organism that causes the symptoms of gastroenteritis.

Symptoms

Gastroenteritis symptoms include **nausea and vomiting**, watery diarrhea, and abdominal **pain** and cramps. These symptoms are sometimes accompanied by bloating, low **fever**, chills, **headache**, and overall tiredness or weakness. Gastroenteritis symptoms typically last two to three days, but some viruses may last up to a week.

Infants, young children, the elderly, and anyone with an underlying disease are more vulnerable to complications of gastroenteritis. The greatest danger presented by gastroenteritis is dehydration. The loss of fluids through diarrhea and vomiting can upset the body's electrolyte balance, leading to potentially life-threatening problems such as heart beat abnormalities (arrhythmia). The risk of dehydration increases as symptoms become prolonged. Untreated, severe dehydration can be life threatening. Dehydration should be suspected if symptoms of a dry mouth, increased or excessive thirst, or decreased urination are experienced.

When to call the doctor

If symptoms do not resolve within one week, an infection or disorder more serious than gastroenteritis may be involved. Prompt medical attention is required if the child has any of these symptoms:

- a high fever of 102°F (38.9°C) or above
- blood or mucus in the diarrhea
- blood in the vomit
- bloody stools or black stools
- confusion
- severe abdominal pain or swelling
- inability to keep liquids down

If a child has the following symptoms, the parent should contact the child's pediatrician:

- diarrhea or vomiting that wakes the child during the night
- persistent or severe diarrhea or vomiting
- dehydration symptoms, including dry mouth, increased or excessive thirst, few or no tears when crying, decreased urination, dark yellow urine, irritability, low energy, lightheadedness or fainting, severe weakness, and sunken abdomen, eyes, and cheeks
- no improvement in symptoms after 36 hours

Diagnosis

A usual bout of gastroenteritis should not require a visit to the doctor. However, medical treatment is essential if symptoms worsen or if the child has any symptoms of dehydration.

A physician makes the diagnosis of gastroenteritis based on the presence of symptoms and after performing a medical examination. Unless there is an outbreak affecting several people or complications are encountered in a particular case, identifying the specific cause of the illness is not a priority. However, if identification of the infectious agent is required, a stool sample will be collected and analyzed for the presence of rotavirus, disease-causing (pathogenic) bacteria, or parasites.

When symptoms continue even after treatment or to rule out the presence of other illnesses with similar symptoms, the diagnostic evaluation may include blood tests, a hydrogen breath test, or an x ray of the bowel, called a barium enema. Endoscopic tests such as a colonoscopy or sigmoidoscopy may be performed. An endoscopic test is an internal examination of the colon using a flexible instrument (sigmoidoscope or colonoscope) inserted through the anus. When symptoms persist, a nutritional **assessment**, performed by a registered dietitian, may be included in the child's diagnostic evaluation.

Treatment

Gastroenteritis is a self-limiting illness that will resolve by itself. **Acetaminophen** (such as Tylenol) or ibuprofen (such as Advil or Motrin) should be used sparingly for relief of discomfort. Parents should ask the child's doctor for specific guidelines. Should pathogenic bacteria or parasites be identified in the patient's stool sample, medications such as **antibiotics** will be prescribed. Over-the-counter antidiarrheal medications such as Imodium should not be given to the child unless advised by the child's doctor, as these drugs may make it more difficult for the child's body to eliminate the virus.

An adequate intake of liquids and oral rehydrating solutions may be enough to treat mild dehydration. More severe dehydration requires medical treatment with intravenous (IV) fluids and may require **hospitalization**. IV therapy can be followed with oral rehydration as the patient's condition improves. Once normal hydration is achieved and symptoms have cleared, the patient can resume a regular diet.

Nutritional concerns

It is important for the child to stay hydrated and nourished during a bout of gastroenteritis. Formula feeding and breastfeeding should continue as normal. If dehydration is absent, drinking generous amounts of fluids, such as water or juice, is adequate. **Caffeine** should be avoided since it increases urine output and can contribute to or worsen dehydration. Dairy products, sugary beverages and foods, highly seasoned foods, and fatty or fried foods should be avoided until symptoms have cleared.

When diarrhea and vomiting symptoms have subsided, plain foods can be given. The traditional BRAT diet—bananas, rice, applesauce, and toast—is tolerated by the tender gastrointestinal system. Other foods can be gradually reintroduced into the diet once the child is symptom-free.

Minimal to moderate dehydration can be treated by giving the child generous amounts of fluids, including water, clear liquids, and oral rehydrating solutions containing glucose and electrolytes. Oral rehydrating solutions—including brands such as Pedialyte, Infalyte, Ceralyte, and Oralyte—are available at most grocery and drug stores. They are essential for replacing fluids, **minerals**, and salts lost from diarrhea or vomiting, and should be given when diarrhea or vomiting first occur.

Small sips of water, clear liquids, or ice chips are usually tolerated better than a large glass of liquid given all at once.

If the water supply is thought to be contaminated because of a recent storm or other reason, the water should be boiled or bottled water should be given.

The Centers for Disease Control and Prevention (CDC) recommends that families with infants and young children keep a supply of oral rehydration solution (two bottles or packages) at home at all times. However, it is important to make sure that the product has not expired before giving it to the child. Parents and caregivers should follow usage directions on the package.

Oral rehydrating solutions are formulated based on physiological properties. Fluids that are not based on these properties—such as cola, apple juice, broth, and sports beverages—are not recommended to treat dehydration.

Alternative treatment

Alternative and complementary therapies include approaches that are considered to be outside the mainstream of traditional health care. Symptoms of uncomplicated gastroenteritis can be relieved with adjustments in diet and homeopathy.

Probiotics, bacteria that are beneficial to a person's health, are recommended during the recovery phase of gastroenteritis. Specifically, live cultures of *Lactobacillus acidophilus* are said to be effective in soothing the digestive tract and returning the intestinal flora to normal. *L. acidophilus* is found in live-culture yogurt, as well as in capsule or powder form at health food stores. The use of probiotics has some support in the medical literature. Castor oil packs applied to the abdomen can reduce inflammation and also lessen spasms or discomfort.

Before using any alternative remedy, it is important for the parent/caregiver and child to learn about the therapy, its safety and effectiveness, and potential side effects. Although some remedies are beneficial, others may be harmful to certain patients. Dietary supplements should not be used as a substitute for medical therapies prescribed by a doctor. Parents should discuss these alternative treatments with the child's doctor to determine the techniques and remedies that may be beneficial for the child.

Prognosis

For most people, gastroenteritis is not a serious illness. It typically resolves within two to three days and there are usually no long-term effects. If dehydration occurs, recovery is extended by a few days. Gastroenteritis is not an anatomical or structural defect, nor is it an identifiable physical or chemical disorder.

KEY TERMS

Barium enema—An x ray of the bowel using a liquid called barium to enhance the image of the bowel. This test is also called a lower GI (gastrointestinal) series.

Colonoscopy—An examination of the lining of the colon performed with a colonoscope.

Constipation—Difficult bowel movements caused by the infrequent production of hard stools.

Defecation—The act of having a bowel movement or the passage of feces through the anus.

Dehydration—An excessive loss of water from the body. It may follow vomiting, prolonged diarrhea, or excessive sweating.

Diarrhea—A loose, watery stool.

Electrolytes—Salts and minerals that produce electrically charged particles (ions) in body fluids. Common human electrolytes are sodium chloride, potassium, calcium, and sodium bicarbonate. Electrolytes control the fluid balance of the body and are important in muscle contraction, energy generation, and almost all major biochemical reactions in the body.

Endoscopy—Visual examination of an organ or body cavity using an endoscope, a thin, tubular instrument containing a camera and light source. Many endoscopes also allow the retrieval of a small sample (biopsy) of the area being examined, in order to more closely view the tissue under a microscope.

Feces—The solid waste, also called stool, that is left after food is digested. Feces form in the intestines and pass out of the body through the anus.

Gastroenterologist—A physician who specializes in diseases of the digestive system.

Glucose—A simple sugar that serves as the body's main source of energy.

Hydrogen breath test—A test used to determine if a person is lactose intolerant or if abnormal bacteria are present in the colon.

Influenza—An infectious disease caused by a virus that affects the respiratory system, causing fever, congestion, muscle aches, and headaches.

Intravenous (IV) therapy—Administration of fluids or medications through a vein, usually in the hand or arm.

Lactose—A sugar found in milk and milk products.

Microflora—The bacterial population in the intestine.

Pathogenic bacteria—Bacteria that produce illness.

Probiotics—Bacteria that are beneficial to a person's health, either through protecting the body against pathogenic bacteria or assisting in recovery from an illness.

Sigmoidoscopy—A procedure in which a thin, flexible, lighted instrument, called a sigmoidoscope, is used to visually examine the lower part of the large intestine. Colonoscopy examines the entire large intestine using the same techniques.

Prevention

A few steps can be taken to avoid gastroenteritis. Thorough hand washing is the most effective way to prevent the fecal-oral transmission of certain viruses, especially rotaviruses. People should wash their hands frequently, especially after using the bathroom and before eating. Child-care providers and caregivers should wash their hands after diapering a child and before preparing, serving, or eating, food. The child's hands also should be washed after every diaper change. Separate towels or disposable paper towels should be used to dry hands. Clean bathroom surfaces, disinfected **toys**, and prompt washing of soiled clothes in hot water also help prevent the spread of infectious germs.

Ensuring that food is prepared safely well-cooked and unspoiled can prevent bacterial gastroenteritis, but may not be effective against viral gastroenteritis. All kitchen utensils, counters, or cutting boards that come in contact with raw meat, especially poultry, should be washed with hot water and a chlorine bleach-based cleaner to prevent the spread of harmful bacteria. Meats should be refrigerated as soon as possible after bringing them home from the grocery store, and cooked leftovers should be refrigerated as soon as possible after a meal to prevent spoilage.

Consuming contaminated food or water can cause gastroenteritis when traveling to other countries. To reduce the risk, travelers should use bottled water for drinking and brushing teeth, and avoid ice (it may be made with contaminated water) and raw foods, including peeled fruit, raw vegetables, and salads.

Research is underway involving vaccines that will decrease the risk of rotavirus infection, especially among infants and young children.

Parental concerns

Parents should reinforce with the child that gastroenteritis is not a serious condition and that symptoms usually subside in a few days. It is most important to prevent dehydration by following the recommendations listed previously. Parents should assure that the child gets adequate rest; the child should be kept home from school or **day care** until the symptoms have cleared. The child may be contagious before the onset of diarrhea and a few days after the diarrhea has ended. To prevent the spread of infection among family members, soiled clothing or bedding should be washed in hot water immediately, hands must be washed frequently, there should be no sharing of utensils or cups used by the child, and toys and bathroom surfaces should be cleaned with a chlorine-based cleaner.

See also Food poisoning.

Resources

PERIODICALS

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ORGANIZATIONS

American College of Gastroenterology (ACG). P.O. Box 3099, Alexandria, VA 22302. (703) 820-7400. Web site: <www.acg.gi.org/patientinfo>.

American Gastroenterological Association. 4930 Del Ray Ave., Bethesda, MD 20814. (301) 654-2055. Patient Information Resources. Web site: <www.gastro.org/generalPublic.html>.

Centers for Disease Control and Prevention. 1600 Clifton Rd., Atlanta, GA 30333. (800) 311-3435 or (404) 639-3534. Web site: <www.cdc.gov>.

National Institute of Diabetes and Digestive and Kidney Diseases (NIDDK). 2 Information Way, Bethesda, MD 20892-3570. (800) 891- 5389. Web site: <www.niddk.nih.gov>.

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Gastroesophageal reflux disease

Definition

Gastroesophageal reflux disease (GERD) is a gastric disorder which causes stomach acids to back up into the esophagus, the tube leading from the mouth to the stomach. This action causes **pain**, which is often called heartburn. GERD can disrupt **sleep** and make eating difficult. It can lead to respiratory infections, ulcers, and even **cancer**.

Description

The reflux action of gastroesophageal reflux disease is a function of the weakening of the lower esophageal sphincter (LES). The LES is a muscle located at the bottom of the esophagus and acts as a doorkeeper to the stomach. When food is eaten, it passes through the esophagus and the LES and into the stomach. The LES closes after food enters the stomach and usually keeps the stomach contents from returning up the esophagus.

In an infant, the LES may not be well formed, which causes the baby to spit up or vomit. In an older child or adolescent, the LES weakens and acids from the stomach come into the esophagus, causing the characteristic burning in the middle of the chest, known as heartburn.

Everyone has experienced this reflux occasionally, and it is not a concern. It is when the reflux occurs often that the condition should be evaluated. Infants and children who do not vomit or complain of heartburn or stomachache may have this condition. When the stomach contents moves into the esophagus, there is the possibility that this material will be aspirated into the windpipe, which can cause **asthma**, **pneumonia**, and possibly suffocation or sudden death. GERD was thought to be implicated in **sudden infant death syndrome** (SIDS); however, subsequent studies concluded it was not.

Some children and adults have few episodes of heartburn over their lifetimes, but they have frequent bouts of

ear infection, **sinusitis**, **bronchitis**, and even asthma. Some children and adults only experience a vague indigestion. They come to the doctor because they are having trouble eating. They feel that there is something in their throats or that their food keeps getting stuck when they eat. This may be a serious condition called dysphagia, which develops from long-term GERD. The stricture of the esophagus is caused by a thickening of the lining of the esophagus in response to acids from the stomach. Sometimes, when swallowing hurts, the condition is called odynophagia. This type of GERD is often referred to as silent reflux.

Constant irritation by stomach acids in the esophagus can cause a condition called esophagitis, in which the esophagus becomes red and irritated. Because the lining of the esophagus is thinner and not as acid-proof as the stomach or the intestines, undiagnosed GERD over many years can cause ulcers along the esophagus. These can bleed and can, in turn, result in anemia. Scar tissue can also build up.

Sometimes, the body tries to protect the esophagus by growing a thicker lining, made up of cells like those in the stomach and intestine. This is known as Barrett's esophagus and is a pre-cancerous condition that usually leads to cancer of the esophagus.

Demographics

One-third of the adult population (95 million) have GERD symptoms once a month, while 15 million have symptoms every day. Though half of people who have GERD are between the ages of 45 and 64, infants, children, and teenagers also have GERD.

GERD affects 50 percent of all healthy, full-term newborns. It is the primary reason for most **vomiting** in infants during the first four months, at which time the vomiting should stop. Less than 5 percent of infants with GERD continue the problem into adulthood. However, this figure may be revised upward as more and more young children experience GERD symptoms and are diagnosed with this condition.

Some children seem to be more at risk for having GERD than others, particularly children who have hiatal **hernia**, **cystic fibrosis**, neurological impairment or delay, or an immature esophagus and LES.

Causes and symptoms

Causes

GERD is caused by a weakened or immature LES. It can also be caused by a hiatal hernia that traps the stomach contents. Having too much acid in the stomach can also weaken the LES.

Heredity plays a small part in whether a child has GERD. GERD seems to be more prone to occur in some families than others.

Other factors that seem to weaken the LES are **allergies** and neurological disorders that affect specific muscles in the body. Diabetes and rapid weight gain can also be factors in causing GERD.

Some medications also can weaken the LES. They include calcium channel blockers used to treat high blood pressure, theophyline used to treat asthma, and **antihistamines**. Nitrates in medications and foods can also trigger GERD.

In infants, it may simply be a matter of having an immature digestive system. Once the body begins to mature, the GERD goes away. For adolescents, the hormones of **puberty** seem to trigger acid reflux.

Certain foods have been known to affect the muscle tone of the LES and increase stomach acids. Chocolate, peppermint, and high fat foods can allow the LES to relax and stay open more often. Citrus foods, tomatoes, and tomato products increase acid production in the stomach.

Lifestyle habits can also trigger episodes of acid reflux. Using **caffeine** and alcohol, **smoking**, eating large meals, and having poor posture can produce GERD.

Symptoms

Though heartburn is the characteristic symptom of GERD in adolescents and adults, GERD in children and infants is not so easy to recognize. Frequent vomiting or spitting up is the usual indicator for GERD in children. However, vomiting can be a symptom of many other childhood disorders, including stomach flu, allergy, or a related symptom to almost any illness. Frequent vomiting that continues after the first four months of life or is excessive at any time usually indicates the presence of GERD. Constant crying with back arching usually accompanies the frequent vomiting.

Children with GERD who are **preschool** age and older often have gas and abdominal pain above the navel. They only have intermittent vomiting. They can also experience chest pain or true heartburn symptoms, which can last up to two hours and get worse after eating. Bending over or lying down makes the heartburn worse.

Children with GERD exhibit difference symptoms. They can either gain or lose weight. One group of children

will eat more because they are uncomfortable and a full stomach seems to make them feel better temporarily. Another group of children are often very picky about what they eat, refusing specific foods. These children will only eat a few bites even though they might be very hungry. A third group of children report having trouble swallowing; they choke or gag whenever they eat, no matter what foods are served. A fourth group of children will drink liquids constantly because doing so soothes the burning feeling in their esophagus.

Respiratory symptoms are twice as likely to occur in children with GERD as those who do not have it. Children often have frequent sore throats when they wake up in the morning, sinus infections, bronchitis, and dry coughs. These children have a constantly runny nose or a hoarse, deep voice. They can also experience wheezing or other asthma symptoms. Some children aspirate the stomach contents, which can cause pneumonia or even sudden death.

Sleep is often disturbed. Children often wake up with a nighttime **cough** or choke when they lie down. Some children experience sleep apnea (interrupted breathing).

Other children have frequent ear infections or drool a lot. Some infants and toddlers will insist on being held upright and not laid down, often falling asleep over a parent's shoulder or in a parent's arms. In some extreme cases, when there is a lot of stomach acid regurgitation, the child's teeth will show enamel erosion.

Children with GERD may also have hiccups or belch a lot. They can also have bad breath and complain of having a sour taste in their mouths.

Some children with GERD have anemia. This condition usually develops because there is an ulcer in the lining of the esophagus that has begun to bleed.

When to call the doctor

It is important to call the doctor if GERD symptoms occur frequently or get worse. If symptoms disturb the child's sleep and interfere with school and **play**, a doctor should be consulted to determine a course of treatment. Also, if a child is not eating or gaining weight or has breathing difficulty, parents should seek medical advice as soon as possible. For a child of any age, if blood is present in vomit, a doctor should be called. If a child over two complains of swallowing difficulty, a serious condition could exist and a doctor should be called.

Diagnosis

In some cases, the doctor will diagnose GERD after taking a thorough medical history, listening carefully for GERD symptoms, and doing a physical exam. Many doctors will also order a series of tests to gauge the extent of damage done by GERD. Sometimes, chest **x rays** are ordered to check for pneumonia or lung damage due to aspiration of stomach contents.

The most common tests, however, are the upper GI (gastrointestinal) series and the upper GI endoscopy. The upper GI series looks at the esophagus, the stomach, and the duodenum, or the first section of the small intestine. The child is asks to drink a cup of liquid that coats the digestive track. Because this liquid has usually been barium, a metallic, chalky substance, the upper GI series is sometimes called a barium swallow.

X rays or images are then taken as the barium flows down the esophagus, into the stomach, and into the duodenum. The child may be asked to turn on his or her side so that the technician can gently massage the stomach to move the barium into the duodenum. Images are often sent to a video monitor where the doctors and technicians observe the behavior of the upper digestive tract and snap still images from the monitor.

The upper GI series is particularly important in diagnosing infants. It can tell if there are anatomical changes in the esophagus, such as a hiatal hernia, a condition where the stomach bulges above the diaphragm. It can also assess damage to the esophagus and can determine if there are stomach ulcers or ulcers in the duodenum.

The upper GI endoscopy, also called the esophagogastroduodenoscopy (EGD), by contrast, is a more sensitive test and offers a more complete picture of what is happening in the upper digestive tract. As of 2004, it was the test of choice for many gastroenterologists (doctors specializing in diseases of the digestive system).

For the endoscopy, the patient receives a mild sedative, then a small, flexible tube is inserted into the esophagus. The tube has a light and a tiny camera attached to its end. There also is a small instrument to take tissue samples if the doctor needs to do so. The camera broadcasts live images from the esophagus and stomach to a video monitor. Using these tools, the doctor can capture still images for further diagnosis and hospital records, and the doctor can examine suspicious areas more closely with the camera or by taking tissue samples.

The EGD allows the doctor to determine the extent of damage to the esophagus and to rule out serious complications like Barrett's esophagus. Mild GERD may show no damage to the esophagus at all. The GED is a good tool for determining esophagitis.

Another test the doctor may order is esophageal manometry. It measures how well the LES and motor

function of the esophagus are. A thin tube is inserted through the nose and down the throat. Coupled with the 24-hour pH probe study, the test becomes the best determinant of GERD because it actually monitors how often the patient has reflux into the esophagus during a full day. One episode of acid reflux is considered having a pH of less than 4 for at least 15 to 30 seconds. This test can see if there is a correlation between episodes of acid reflux and other symptoms, such as chronic cough, wheezing, or sleep apnea.

The doctor may also order a gastric emptying study. For this test, the child is asked to drink milk mixed with a radioactive chemical. Then, the child is monitored, using a special camera. Episodes of reflux can be seen with this test.

Though esophagitis may have been found in one of these tests, the doctor will need to determine whether it was caused by GERD or by milk allergy, which does not respond to acid suppressant therapy.

Treatment

There are two main treatment methods for GERD. The first is lifestyle change. This usually means that patients should not eat within three hours of going to bed. This lets the stomach empty and the acid decrease. Lying down will cause the stomach contents to come back up. Elevating the bed about six inches will also keep the acid within the stomach. Eating smaller meals more frequently will control the amount of acid in the stomach. Patients should also avoid fatty foods, caffeine, mints and mint-flavoring, spicy foods, citrus fruits, and anything with tomatoes. Carbonated beverages can also irritate the already sensitive lining of the esophagus. Alcohol and smoking should be avoided. Improved posture, with no slumping, will reduce pressure on the stomach, as will losing excess weight.

For an infant, lifestyle changes are simple. Holding a baby upright for about a half hour after breastfeeding or bottle feeding will help keep reflux to a minimum. Feeding a baby on formula smaller portions more frequently can also help manage spitting up. Some doctors recommend thickening the baby's formula with rice cereal or using pre-thickened formulas such as Enfamil. This will decrease the amount of spit up or vomit, but it does nothing for reflux. It does fill up the child on a smaller amount of food and can also make the baby sleep and thus stop crying. Placing a baby in a semi-prone position as in an infant car seat only makes GERD worse. Babies with GERD should sleep on their backs in a crib or bed that has the head of the bed elevated to a 30 degree angle.

Medication is the second main way to treat GERD. The doctor may first recommend non-prescription medications, such as antacids and histamine-2 receptor blockers (H2 blockers). Antacids, such as Gaviscon, Maalox, Mylanta, and Tums, help neutralize acid already in the stomach or esophagus. Some have a foaming agent, which also helps prevent acid from backing up into the esophagus. Antacids can be used every day for three weeks. If taken longer, they can produce **diarrhea**, interfere with calcium absorption in the body, and build up magnesium, which can damage the kidneys. The doctor will determine if they can be taken longer. Infants are only given antacids in limited doses because of the risk of aluminum toxicity.

Common H2 blockers are nizatidine (Axid), ranitidine (Zantac), famotidine (Pepcid), and cimetidine (Tagamet). These should be taken one hour before meals. They block acid formation but have no effect on acid already present in the stomach.

If these remedies do not work or the patient's GERD is very serious, the doctor will usually move onto the more powerful proton-pump inhibitors (PPIs). These include omeprazole (Prilosec), esomeprazole (Nexium), lansoprazole (Prevacid), rabeprazole (Aciphex), and pantoprazole (Protonix). These medications block the production of an enzyme that aids in the production of acid. PPIs stop acid production better than H2 blockers.

In addition to PPIs, the doctor may prescribe coating agents, such as sucralfate (Carafe), to cover the sores and mucous membranes of the esophagus and stomach. This acts as a protective barrier to stomach acids.

Some doctors also use promotility agents to tighten the LES and promote faster emptying of the stomach. These include metoclopramide (Reglan) and bethanechol (Duvoid). However, many doctors are reluctant to use these drugs because they have serious side effects. For example, cisapride (Propulsid) was pulled from the market because of safety concerns about lethal drug interactions.

One last option that doctors have to treat GERD is surgery. Because lifestyle changes and medications work for most children and adolescents with GERD, the election of surgery is only used for a small number of people for whom all the other options did not work.

Fundoplication is a surgical procedure that puts pressure on the LES to keep acid from backing up. During the surgery, the doctor wraps a part of the stomach around the esophagus and sews it down. This produces a one-way valve. This procedure can be done laparoscopically, a less invasive surgery where the doctor makes

small cuts into the abdomen to insert a camera and the surgical instruments. This surgery produces very little scarring and has a faster recovery rate. Fundoplication is not always successful and can have complications. The surgery also comes undone in about 20 percent to 30 percent of cases.

Prognosis

Many babies outgrow infantile GERD, but some keep having symptoms well into adulthood. In most cases, GERD is easily managed. For 60 percent of children and adolescents with mild to moderate GERD, lifestyle changes and H2 blockers are very effective. For those with severe symptoms, including esophagitis, PPI therapy works well. For relapses, long-term therapy or surgery may be necessary.

Prevention

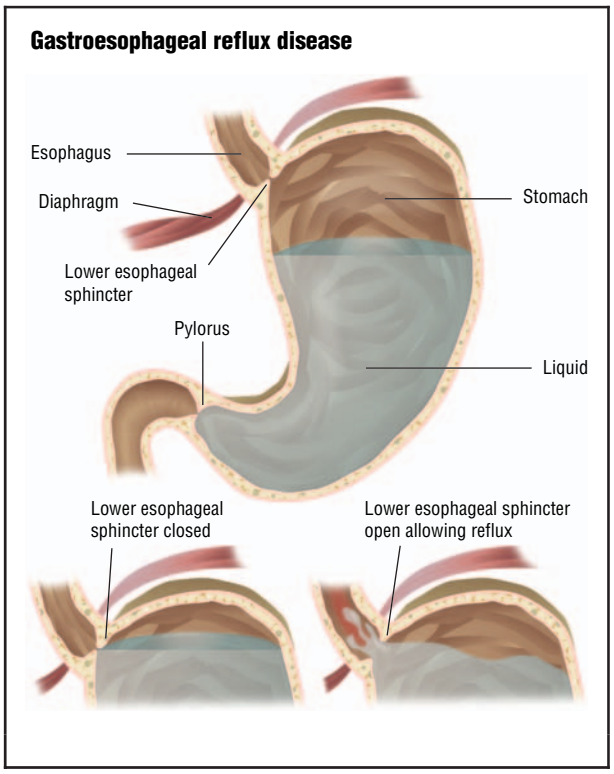
GERD can be prevented by maintaining a healthy body weight, avoiding alcohol and smoking, eating smaller meals, limiting fatty foods, and eliminating trigger foods.

Parental concerns

GERD diagnosis is a recent phenomenon. Though it may have existed in the distant past, only since the 1990s have doctors begun to recognize GERD as an individual disease. Diagnosing GERD in children is sometimes controversial. Some doctors have recognized GERD as a temporary condition in infants but do not recognize GERD in children or in adolescents. Many doctors are, as of 2004, beginning to understand that GERD, like many other digestive disorders, can occur at any age. The North American Society for Pediatric Gastroenterology and Nutrition drafted guidelines for treating children and adolescents with GERD in 2001. Being educated about the disease can help parents discuss their child's GERD symptoms and treatment options with their child's doctor.

Parents should help children understand that they need to take their medications regularly and that they need to make lifestyle changes. It can be hard to explain to a child that chocolate and candy canes are off limits, but like **food allergies**, children will learn to modify their food choices because of their special sensitivities.

Parents can also help children cope emotionally with this disease. For some children, it is just a matter of eating right and taking medication once a day. For others, it is a lifelong struggle with food and their digestive tract. They will have good days and bad days. It may be difficult for children to communicate their condition to their friends or their teachers who might not always understand that on one day they are fine and on the next they are not. Parental support will help children and teenagers cope with GERD.



Normally, the lower esophageal sphincter keeps the stomach contents contained with the stomach (top). However, with gastroesophageal reflux disease, the sphincter opens, allowing the acidic contents to flow up the esophagus. *(Illustration by GGS Information Services.)*

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KEY TERMS

Dysphagia—Difficulty in swallowing.

Esophagogastroduodenoscopy (EGD)—An imaging test that involves visually examining the lining of the esophagus, stomach, and upper duodenum with a flexible fiberoptic endoscope.

Esophagus—The muscular tube that leads from the back of the throat to the entrance of the stomach. It is coated with mucus and surrounded by muscles, and pushes food to the stomach by sequential waves of contraction. It functions to transport food from the throat to the stomach and to keep the contents of the stomach in the stomach.

Fundoplication—A surgical procedure that increases pressure on the lower esophageal sphincter by stretching and wrapping the upper part of the stomach around the sphincter.

Gastroenterologist—A physician who specializes in diseases of the digestive system.

H2RAs—Medications used to treat some GERD symptoms, for example, Tagamet, Pepcid, Axid.

Heartburn—A burning sensation in the chest that can extend to the neck, throat, and face. It is the primary symptom of gastroesophageal reflux (the movement of stomach acid into the esophagus).

Hiatal hernia—A condition in which part of the stomach protrudes through the diaphragm into the chest cavity.

Laparoscopic surgery—Minimally invasive surgery in which a camera and surgical instruments are inserted through a small incision.

Odynophagia—Pain in swallowing.

Lower esophageal sphincter (LES)—A muscle located at the base of the esophagus which keeps the stomach contents from coming back into the esophagus.

Proton pump—A structure in the body that produces and pumps acid into the stomach.

Silent reflux—An acid reflux problem that does not cause vomiting but can cause chronic, recurrent respiratory symptoms much like asthma.

Regurgitation—The flow of material back up the esophagus and into the throat or lungs. Also refers to the backward flow of blood through a partly closed heart valve.

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Janie Franz

Gastroschisis *see* **Abdominal wall defects**

Gaucher's disease *see* **Lipidoses**

Gay issues *see* **Homosexuality and bisexuality**

Gender constancy

Definition

A child's realization that gender is fixed and does not change over time.

Description

The concept of gender constancy, influenced by the **cognitive development** theory of French psychologist Jean Piaget (1896–1980), was introduced by Lawrence Kohlberg (1927–1987). In 1966, Kohlberg presented a revolutionary new view of early gender role development. Parting with previous views generally held by psychologists, Kohlberg emphasized that children actively self-construct their gender through a conceptual pattern in the mind called a schema. Gender schema models stress the roles of children's selective attention to gender and their internal motivation to conform to societal standards and stereotypes of gender roles. These cognitive-developmental models of the development of gender roles are perhaps best known, however, for the introduction of the construct of gender constancy. Gender con-

stancy has been defined as children's understanding of the irreversibility of their sex, which develops in stages between about the ages of two and seven years. Kohlberg acknowledged that some awareness of gender roles emerges in children before full attainment of an understanding of gender constancy, although he argued that once children attain full understanding of gender constancy, they become increasingly motivated to observe, incorporate, and respect gender roles.

Addressing the formation of **gender identity** in terms of cognitive development, Kohlberg advanced the idea that the development of sex roles depends in large part on a child's understanding that gender remains constant throughout a person's lifetime. Children realize that they are male or female and are aware of the gender of others by the age of three. However, at these ages they still do not understand that people cannot change genders the way they can change their clothes, names, or behavior. Kohlberg theorized that children do not learn to behave in gender-appropriate ways until they understand that gender is permanent, which occurs at about the age of seven. At this point they start modeling the behavior of members of their own sex.

Kohlberg's idea of the prerequisite significance of an understanding of gender constancy for gender typing has been controversial, and empirical support for the notion has been mixed. Kohlberg's exact claims about the prerequisite importance of gender constancy understanding for gender-typing to take place have added to the controversy. Although it has been supported by some research studies, Kohlberg's theory has also been criticized on the grounds that children do show certain types of gender-associated behavior, such as toy and playmate selection, by the ages of two or three. This observable pattern points to the fact that there are others factors, such as parental reinforcement, that influence the adoption of sex-typed behavior. The significance of gender constancy understanding on early gender-typing remains unclear, both theoretically and empirically.

A gender constancy interview is often used by psychologists to determine a child's level of gender constancy. Although questions used in the interview can vary, the interview generally consists of 13 questions and counterquestions. Previous research indicated that, based on their responses to questions in the interview, children may reliably be assigned to one of four levels of gender constancy understanding. Children who failed to express any understanding of the 13 questions in levels two through four are classified at level one and are considered pre-gender constant. Level two has four questions and examines children's understanding of their own sex and the sex of a pictured person. Level three has two questions and verbally measures children's understanding that sex is permanent over time despite changes in appearance, desires, or activities. Level four contains seven questions that measures children's understanding that pictured people do not change sex through changes in hair length, clothing, or both.

Infancy and toddlerhood

There is a growing amount of scientific research that suggests gender identity develops at a very early age. Several studies show that infants can discriminate between male and female faces and associate faces and voices according to gender by the time they reach one year old. However, gender-labeling tasks, such as toy identification, do not occur until about age two. Gender identity and awareness of sex differences generally emerge in the first three to four years of a child's life. However, children begin to demonstrate a preference for their own sex starting at about age two.

Gender identification is often associated with the choice and use of **toys** in this age group, according to a number of studies done in the 1970s, 1980s, and 1990s. Sex differences in toy **play** have been found in children as young as one year of age. By age two, children begin to spontaneously choose their types of toys based on gender. Several of these studies show that by age one, boys display a more assertive reaction than girls to toy disputes. By age two, the reaction of boys is more aggressive.

Between the ages of 18 and 24 months, children know whether they are boys or girls and can identify adults as males or females. However, they do not develop a sense of gender constancy for several or a few more years. This means that they do not fully understand that they will be a boy or girl for the rest of their lives. At this age, children are unsure about whether gender remains constant from childhood to adulthood.

Preschool

By age three, most children know that men have a penis and women have breasts. Also at age three, children begin to apply gender labels and stereotypes, identifying gentle, empathic characteristics with females and strong and aggressive characteristics with males. Even in the twenty-first century, most young children develop stereotypes regarding gender roles, associating nurses, teachers, and secretaries as females and police officers, firefighters, and construction workers as males.

School age

By the first or second grade (ages six or seven), children's thinking becomes more logical, and they come to realize and understand that they will either be a boy or girl for the rest of their lives. They will draw upon what

they have learned and what they see in the world around them as they continue to refine their ideas about what it means to be either a girl or a boy.

Common problems

While most children follow a predictable pattern in the acquisition of gender constancy, some develop a gender identity inconsistent with their biological sex, a condition variously known as gender confusion, gender identity disorder, or transsexualism, which affects about one in 20,000 males and one in 50,000 females. Researchers have found that both early socialization and hormonal factors may play a role in the development of gender identity disorder. Children with gender identity disorder usually feel from their earliest years that they are trapped in the wrong body and begin to show signs of gender confusion between the ages of two and four. They prefer playmates of the opposite sex at an age when most children prefer to spend time in the company of same-sex peers. They also show a preference for the clothing and typical activities of the opposite sex: transsexual boys like to play house and play with dolls. Girls with gender identity disorder are bored by ordinary female pastimes and prefer the rougher types of play typically associated with boys, such as contact **sports**.

Both male and female transsexuals believe and repeatedly insist that they actually are, or will grow up to be, members of the opposite sex. Girls cut their hair short, favor boys' clothing, and have negative feelings about maturing physically as they near **adolescence**. In childhood, girls with gender identity disorder experience less overall social rejection than boys, as it is more socially acceptable for a girl to be a tomboy than for a boy to be perceived as a "sissy." About five times more boys than girls are referred to therapists for this condition. Teenagers with gender identity disorder suffer social isolation and are vulnerable to depression and **suicide**. They have difficulty developing peer relationships with members of their own sex as well as romantic relationships with the opposite sex. They may also become alienated from their parents.

Parental concerns

Children with gender identity disorder refuse to dress and act in sex-stereotypical ways. It is important to remember that many emotionally healthy children experience fantasies about being a member of the opposite sex. The distinction between these children and gender identity disordered children is that the latter experience significant interference in functioning because of their cross-gender identification. They may become severely depressed, anxious, or socially withdrawn. Most children eventually outgrow gender identity disorder. About 75 percent of boys with gender identity disorder develop a homosexual or bisexual orientation by late adolescence or adulthood, but without continued feelings of transsexuality. Most of the remaining 25 percent become heterosexuals (also without transsexuality). Those individuals in whom gender identity disorder persists into adulthood retain the desire to live as members of the opposite sex, sometimes manifesting this desire by cross-dressing, either privately or in public. In some cases, adult transsexuals (both male and female) have their primary and secondary sexual characteristics altered through a sex change operation, consisting of surgery followed by hormone treatments.

When to call the doctor

Gender identity disorder (GID) is generally diagnosed when children display any four of the following symptoms:

- They repeatedly state a strong desire to be, or insist that they are, of the opposite sex.
- They show a preference for cross-dressing.
- They display a strong and long-term preference for fantasies and role-play that allows them to see themselves as members of the opposite sex.
- They participate in or want to play stereotypical games of the opposite sex.
- They show a strong preference for friends and playmates of the opposite sex.

GID is typically diagnosed by a psychiatrist or psychologist, who conducts an interview with the patient and takes a detailed social history. **Family** members may also be interviewed during the **assessment** process. Most children diagnosed with GID eventually grow out of it but some psychiatrists try to speed up the process, usually using psychotherapy. This treatment itself is controversial and has received much criticism within both the psychiatric and gay, lesbian, bisexual, and transgendered communities. For children, a clear diagnosis may not be possible until the teenage years since most children grow out of GID problems. Some psychiatrists are critical of gender identity disorder being classified as a psychiatric condition at all, saying it is more a social stigma. To prove their case, some psychiatrists point to the fact that boys are up to six times more likely to be diagnosed with GID and singled out for treatment than girls. This is not because the disorder is more common in boys, but because most parents tend to worry more if a son starts wearing dresses than if their daughter starts playing with toy trucks.

See also Gender identity.

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National Academy of Child & Adolescent Psychiatry. 3615 Wisconsin Ave. NW, Washington, DC 20016. Web site: <www.aacap.org>.

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Gender identity

Definition

Gender identity is a person's sense of identification with either the male or female sex, as manifested in appearance, behavior, and other aspects of a person's life.

Description

Psychologists believe human sexual identities are made up of three separate components. The first shows the direction of a child's sexual orientation, whether he or she is heterosexual (straight), homosexual (gay), or bisexual. The second is the child's style of behavior, whether a female is a "tomboy" or homemaker-type and a male is a "macho guy" or a "sensitive boy." The third component is what psychologists call the core gender identity. According to an article in the May 12, 2001 issue of *New Scientist*, it is the most difficult to ascertain but is essentially the deep inner feeling a child has about whether he or she is a male or female.

In most people, the three components point in the same direction but in some people, the components are more mixed. For example, a gay woman (lesbian) might look and act either feminine or masculine (butch), but she still deeply feels she is a female. Scientists are uncertain about where the inner feeling of maleness or femaleness comes from. Some believe it is physical, from the body, while others believe it is mental, from the hypothalamus region of the brain. There is also debate on whether the determination is shaped by hormones, particularly testosterone and estrogen, or by genes assigned at conception.

Gender identity emerges by the age of two or three and is influenced by a combination of biological and sociological factors reinforced at **puberty**. Once established, it is generally fixed for life.

Aside from sex differences, other biological contrasts between males and females are already evident in childhood. Girls mature faster than boys, are physically healthier, and are more advanced in developing oral and written linguistic skills. Boys are generally more advanced at envisioning and manipulating objects. They are more aggressive and more physically active, preferring noisy, boisterous forms of **play** that require larger groups and more space than the play of girls the same age.

In spite of conscious attempts to reduce sex role stereotyping in the final decades of the twentieth century and in the early 2000s, boys and girls are still treated differently by adults from the time they are born. The way adults play with infants has been found to differ based on gender. Girls are treated more gently and approached more verbally than boys. As children grow older, many parents, teachers, and other authority figures still tend to encourage independence, competition, aggressiveness, and exploration more in boys and expression, nurturance, motherhood and childrearing, and obedience more in girls.

Infancy and toddlerhood

There is a growing amount of scientific research that suggests gender identity develops at a very early age.

Several studies show that infants can discriminate between male and female faces and associate faces and voices according to gender by the time they reach one year old. However, gender-labeling tasks, such as toy identification, do not occur until about age two. Gender identity and awareness of sex differences generally emerge in the first three to four years of a child's life. However, children begin to demonstrate a preference for their own sex starting at about age two.

Gender identification is often associated with the choice and use of **toys** in this age group, according to a number of studies done in the 1970s, 1980s, and 1990s. Sex differences in toy play have been found in children as young as one year old. By age two, children begin to spontaneously choose their types of toys based on gender. Several of these studies show that by age one, boys display a more assertive reaction than girls to toy disputes. By age two, the reaction of boys is more aggressive.

Most two-year-olds know whether they are boys or girls and can identify adults as males or females. By age three, most children know that men have a penis and women have breasts. Also at age three, children begin to apply gender labels and stereotypes, identifying gentle, empathic characteristics with females and strong, aggressive characteristics with males. Even in the twenty-first century, most young children develop stereotypes regarding gender roles, associating nurses, teachers, and secretaries as females and police officers, firefighters, and construction workers as males.

Preschool

Preschoolers develop an increasing sense of self-awareness about their bodies and gender differences. Fears about the body and body mutilation, especially of the genitals, are often major sources of **fear** in preschoolers. As children become more aware of gender differences, preschoolers often develop intense feelings of vulnerability and **anxiety** regarding their bodies.

School age

By the age of six years, children are spending about 11 times as much time with members of their own sex as with children of the opposite sex. This pattern begins to change as the child approaches puberty, however.

By the teenage years, most children have an established sexual orientation of heterosexual, homosexual, or bisexual. They have also established their style of behavior and core sexual identity. However, a very small fraction have not.

While most children follow a predictable pattern in the acquisition of gender identity, some develop a gender identity inconsistent with their biological sex, a condition variously known as gender confusion, gender identity disorder, or transsexualism, which affects about one in 20,000 males and one in 50,000 females. Researchers have found that both early socialization and hormonal factors may play a role in the development of gender identity disorder. Children with gender identity disorder usually feel from their earliest years that they are trapped in the wrong body and begin to show signs of gender confusion between the ages of two and four. They prefer playmates of the opposite sex at an age when most children prefer to spend time in the company of same-sex peers. They also show a preference for the clothing and typical activities of the opposite sex; transsexual boys like to play house and play with dolls. Girls with gender identity disorder are bored by ordinary female pastimes and prefer the rougher types of play typically associated with boys, such as contact **sports**.

Both male and female transsexuals believe and repeatedly insist that they actually are, or will grow up to be, members of the opposite sex. Girls cut their hair short, favor boys' clothing, and have negative feelings about maturing physically as they near **adolescence**. In childhood, girls with gender identity disorder experience less overall social rejection than boys, as it is more socially acceptable for a girl to be a tomboy than for a boy to be perceived as feminine. About five times more boys than girls are referred to therapists for this condition. Teenagers with gender identity disorder suffer social isolation and are vulnerable to depression and **suicide**. They have difficulty developing peer relationships with members of their own sex as well as romantic relationships with the opposite sex. They may also become alienated from their parents.

Common problems

The psychological diagnosis of gender identity disorder (GID), commonly called transsexualism, is used to describe a male or female who feels a strong identification with the opposite sex and experiences considerable distress because of their actual sex. Children with gender identity disorder have strong cross-gender identification. They believe that they are, or should be, the opposite sex. They are uncomfortable with their sexual role and organs and may express a desire to alter their bodies.

While not all persons with GID are labeled as transsexuals, there are those who are determined to undergo sex change procedures or have done so, and, therefore, are classified as transsexual. They often attempt to pass socially as the opposite sex. Transsexuals alter their physical appearance cosmetically and hormonally and may eventually undergo a sex-change operation.

Most children eventually outgrow gender identity disorder. About 75 percent of boys with gender identity disorder develop a homosexual or bisexual orientation by late adolescence or adulthood, but without continued feelings of transsexuality. Most of the remaining 25 percent become heterosexuals (also without transsexuality). Those individuals in whom gender identity disorder persists into adulthood retain the desire to live as members of the opposite sex, sometimes manifesting this desire by cross-dressing, either privately or in public. In some cases, adult transsexuals (both male and female) have their primary and secondary sexual characteristics altered through a sex change operation, consisting of surgery and hormone treatments.

Parental concerns

Children with gender identity disorder refuse to dress and act in sex-stereotypical ways. It is important to remember that many emotionally healthy children experience fantasies about being a member of the opposite sex. The distinction between these children and gender identity disordered children is that the latter experience significant interference in functioning because of their cross-gender identification. They may become severely depressed, anxious, or socially withdrawn.

According to an article in the January 2003 issue of *The Brown University Child and Adolescent Behavior Letter*, psychiatrists offer these suggestions for parents of children diagnosed with GID:

- Create an atmosphere of acceptance so the child feels safe within the **family** to express his or her interests. Identify and praise the child's talents.
- Use gender-neutral language in discussing romantic attachments.
- Watch television programs and movies and read books that have gay themes or characters.
- Encourage the child to find activities that respect his or her interests, yet help the child "fit in" to society.
- Insist on classroom discussions about diversity and tolerance. Ensure the child's school has anti-discrimination policies that include gender identity and that the policies are enforced.
- If indicated, take the child to a psychotherapist with expertise and tolerance for issues related to gender identity and sexual orientation.

When to call the doctor

Gender identity disorder is generally diagnosed when children display any four of the following symptoms:

- They repeatedly state a strong desire to be, or insist that they are, of the opposite sex.
- They show a marked preference for cross-dressing.
- They display a strong and long-term preference for fantasies and role-play as members of the opposite sex.
- They participate in or want to play stereotypical games of the opposite sex.
- They show a strong preference for friends and playmates of the opposite sex.

KEY TERMS

Core gender identity—The deep inner feeling a child has about whether he or she is a male or female.

Estrogen—Female hormone produced mainly by the ovaries and released by the follicles as they mature. Responsible for female sexual characteristics, estrogen stimulates and triggers a response from at least 300 tissues. After menopause, the production of the hormone gradually stops.

Gender identity disorder (GID)—A strong and lasting cross-gender identification and persistent discomfort with one's biological gender (sex) role. This discomfort must cause a significant amount of distress or impairment in the functioning of the individual.

Hypothalamus—A part of the forebrain that controls heartbeat, body temperature, thirst, hunger, body temperature and pressure, blood sugar levels, and other functions.

Psychotherapy—Psychological counseling that seeks to determine the underlying causes of a patient's depression. The form of this counseling may be cognitive/behavioral, interpersonal, or psychodynamic.

Testosterone—Male hormone produced by the testes and (in small amounts) in the ovaries. Testosterone is responsible for some masculine secondary sex characteristics such as growth of body hair and deepening voice. It also is sometimes given as part of hormone replacement therapy to women whose ovaries have been removed.

Transsexualism—A term used to describe a male or female that feels a strong identification with the opposite sex and experiences considerable distress because of their actual sex. Also called gender identity disorder.

GID is typically diagnosed by a psychiatrist or psychologist, who conducts an interview with the patient and takes a detailed social history. Family members may also be interviewed during the **assessment** process. Most children diagnosed with GID eventually grow out of it, but some psychiatrists try to speed up the process, usually using psychotherapy. This treatment itself is controversial and has received much criticism within both the psychiatric and gay, lesbian, bisexual, and transgendered communities. For children, a clear diagnosis may not be possible until the teenage years, since most children grow out of GID problems.

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GERD *see* **Gastroesophageal reflux disease**

German measles *see* **Rubella**

Gingivitis *see* **Periodontal disease**

Gluten sensitive enteropathy *see* **Celiac disease**

Gonadal dysgenesis *see* **Turner syndrome**

Granular conjuctivitis *see* **Trachoma**

Grieving *see* **Death and mourning**

Gross motor skills

Definition

Gross motor skills are the abilities required in order to control the large muscles of the body for walking, running, sitting, **crawling**, and other activities.

Description

Motor skills are actions that involve the movement of muscles in the body. They are divided into two groups: gross motor skills, which are the larger movements of arms, legs, feet, or the entire body (crawling, running, and jumping); and **fine motor skills**, which are smaller actions, such as grasping an object between the thumb and a finger or using the lips and tongue to taste

objects. Motor skills usually develop together since many activities depend on the coordination of gross and fine motor skills. Gross motor skills develop over a relatively short period of time. Most development occurs during childhood. However, soldiers, some athletes, and others who engage in activities requiring high degrees of endurance may spend years improving their level of muscle and body coordination and gross motor skills.

Gross motor skills development is governed by two principles that also control physical growth. Head to toe development refers to the way the upper parts of the body develop, beginning with the head, before the lower ones. The second principle of development is trunk to extremities. Head control is gained first, followed by the shoulders, upper arms, and hands. Upper body control is developed next, followed by the hips, pelvis, and legs.

Encouraging gross motor skills requires a safe, open **play** space, peers to interact with, and some adult supervision. Promoting the development of gross motor abilities is considerably less complicated than developing fine motor skills. Helping a child succeed in gross motor tasks requires patience and opportunities for a child to practice desired skills. Parents and other persons must understand the child's level of development before helping him or her master gross motor skills. Children reach developmental milestones at different rates. Pushing a child to perform a task that is impossible due to development status promotes frustration and disappointment. Children should be allowed to acquire motor skills at their own paces.

There are a number of activities parents can have children do to help develop gross motor skills. These include:

- playing hopscotch and jumping rope; activities that help children learn balance
- hitting, catching, kicking, or throwing a ball, such as a baseball, football, or soccer ball; activities that help develop hand-eye or foot-eye coordination
- kangaroo hop, in which children hold something, such as a small ball or orange, between their knees and then jump with their feet together frontward, backwards, and sideways
- playing wheelbarrow, in which someone holds the children's legs while they walk on their hands along a specific route
- walking on a narrow bar or curb, while holding a bulky object in one hand, then the other hand, and then repeating the activity walking backwards and sideways
- toss and catch, in which children toss an object, such as a baseball, in the air and then catch it, while sitting or lying down and also while using alternate hands

Infancy

The first gross motor skill infants learn usually is to lift their heads and shoulders before they can sit up, which, in turn, precedes standing and walking. Lifting the head is usually followed by head control. Although they are born with virtually no head or neck control, most infants can lift their heads to a 45-degree angle by the age of four to six weeks, and they can lift both their head and chest at an average age of eight weeks. Most infants can turn their heads to both sides within 16 to 20 weeks and lift their heads while lying on their backs within 24 to 28 weeks. By about nine to 10 months, most infants can sit up unassisted for substantial periods of time with both hands free for playing.

One of the major tasks in gross motor development is locomotion, the ability to move from one place to another. Infants progress gradually from rolling (eight to 10 weeks) to creeping on their stomachs and dragging their legs behind them (six to nine months) to actual crawling (seven to 12 months). While infants are learning these temporary means of locomotion, they are gradually becoming able to support increasing amounts of weight while in a standing position. In the second half-year of life, babies begin pulling themselves up on furniture and other stationary objects. By the ages of 28 to 54 weeks, on average, they begin navigating a room in an upright position by holding on to the furniture to keep their balance. Eventually, they are able to walk while holding on to an adult with both hands and then with only one. They usually take their first uncertain steps alone between the ages of 36 and 64 weeks and are competent walkers by the ages of 12 to 18 months.

Toddlerhood

Toddlers are usually very active physically. By the age of two years, children have begun to develop a variety of gross motor skills. They can run fairly well and negotiate stairs holding on to a banister with one hand and putting both feet on each step before going on to the next one. Most infants this age climb (some very actively) and have a rudimentary ability to kick and throw a ball. By the age of three, children walk with good posture and without watching their feet. They can also walk backwards and run with enough control for sudden stops or changes of direction. They can hop, stand on one foot, and negotiate the rungs of a jungle gym. They can walk up stairs alternating feet but usually still walk down putting both feet on each step. Other achievements include riding a tricycle and throwing a ball, although they have trouble catching it because they hold their arms out in front of their bodies no matter what direction the ball comes from.

Preschool

Four-year-olds can typically balance or hop on one foot, jump forward and backward over objects, and climb and descend stairs alternating feet. They can bounce and catch balls and throw accurately. Some four-year-olds can also skip. Children this age have gained an increased degree of self-consciousness about their motor activities that leads to increased feelings of pride and success when they master a new skill. However, it can also create feelings of inadequacy when they think they have failed. This concern with success can also lead them to try daring activities beyond their abilities, so they need to be monitored especially carefully.

School age

School-age children, who are not going through the rapid, unsettling growth spurts of early childhood or **adolescence**, are quite skilled at controlling their bodies and are generally good at a wide variety of physical activities, although the ability varies according to the level of maturation and the physique of a child. Motor skills are mostly equal in boys and girls at this stage, except that boys have more forearm strength and girls have greater flexibility. Five-year-olds can skip, jump rope, catch a bounced ball, walk on their tiptoes, balance on one foot for over eight seconds, and engage in beginning acrobatics. Many can even ride a small two-wheel bicycle. Eight- and nine-year-olds typically can ride a bicycle, swim, roller skate, ice skate, jump rope, scale fences, use a saw, hammer, and garden tools, and play a variety of **sports**. However, many of the sports prized by adults, often scaled down for play by children, require higher levels of distance judgment and **hand-eye coordination**, as well as quicker reaction times, than are reasonable for middle childhood. Games that are well suited to the motor skills of elementary school-age children include kick ball, dodge ball, and team relay races.

In adolescence, children develop increasing coordination and motor ability. They also gain greater physical strength and prolonged endurance. Adolescents are able to develop better distance judgment and hand-eye coordination than their younger counterparts. With practice, they can master the skills necessary for adult sports.

Gross motor skills

Age	Skill
One month	May hold up head momentarily.
Two months	Lifts head when placed on stomach. Holds up head briefly when held in a seated or standing position.
Three months	Holds head and shoulders up when placed on stomach. Puts weight on forearms.
Four months	Holds head up well in sitting position. Can lift head to a 90-degree angle when placed stomach. May start to roll over.
Five months	Has full head control. When pulled by hands to a sitting position, the head stays in line with body.
Six months	Rolls over (front to back first). Bears a large. percentage of body weight when held in a standing position.
Seven months	Can stand with support. May sit without support for short periods. Pushes upper part of body up while on stomach.
Eight months	Stands while holding onto furniture. Sits well unsupported. Gets up on hands and knees; may start to crawl backwards.
Nine months	Crawls first by pulling body forward with hands. May move around a room by rolling.
Ten months	Pulls up to standing. Is very steady while sitting; moves from sitting to crawling position and back. Crawls well.
Eleven months	"Cruises," walking while hanging onto furniture. Walks with two hands held.
Twelve months	Walks with one hand held. May walk with hands and feet. Stands unsupported for longer periods of time.
Fifteen months	Walks without help. Crawls up stairs. Gets into a standing position without support.
Eighteen months	Seldom falls while walking. Can walk and pull toy. Runs. Climbs stairs holding railing. May walk backward.
Two years	Kicks a ball. Walks up and down stairs, two feet per step.
Two and a half years	Jumps with both feet. Jumps off step. Can walk on tiptoe.
Three years	Goes upstairs one foot per step. Stands on one foot briefly. Rides tricycle. Runs well.
Four years	Skips on one foot. Throws ball well overhand. Jumps a short distance from standing position.
Five years	Hops and skips. Good balance. Can skate or ride scooter.

SOURCE: *Miller-Keane Encyclopedia and Dictionary of Medicine, Nursing, and Allied Health, 5th ed.* and Child Development Institute, http://www.childdevelopmentinfo.com.

(Table by GGS Information Services.)

Common problems

There are a range of diseases and disorders that affect gross motor skill development and skills. Among young persons, developmental problems such as genetic disorders, **muscular dystrophy**, **cerebral palsy**, and some neurological conditions adversely impact gross motor skill development.

Gross motor skills can become impaired in a variety of ways, including injury, illness, **stroke**, and congenital deformities. Developmental coordination disorder affects motor skills. A person with this disorder has a hard time with skills such as riding a bike, holding a

Girls using their gross motor skills, large muscle movements, to play soccer. *(Photograph by Tony Freeman. PhotoEdit.)*

pencil, and throwing a ball. People with this disorder are often called clumsy. Their movements are slow and awkward. People with developmental coordination disorder may also have a hard time completing tasks that involve movement of muscle groups in sequence. For example, such a person might be unable to do the following in order: open a closet door, get out a jacket, and put it on. It is thought that up to 6 percent of children may have developmental coordination disorder, according to the 2002 issue of the annual journal *Clinical Reference Systems*. The symptoms usually go unnoticed until the early years of elementary school; the disorder is usually diagnosed in children who are between five and 11 years old.

Children with any one or combination of developmental coordination disorder symptoms should be seen by a pediatrician who specializes in motor skills development delays. There are many ways to address gross motor skills impairment, such as physical therapy. This type of therapy can include treating the underlying cause, strengthening muscles, and teaching ways to compensate for impaired movements.

Parental concerns

Parents, teachers, and primary caregivers need to have a clear understanding of how young children develop gross motor skills and the timetable for development of the skills. The Lincoln-Oseretsky Motor Development Scale is an individually administered test that assesses the development of motor skills in children and adults. Areas covered include fine and gross motor skills, finger dexterity and speed, and hand-eye coordination. The test consists of 36 tasks arranged in order of increasing difficulty. These include walking backwards, standing on one foot, touching one's nose, jumping over a rope, throwing and catching a ball, putting coins in a box, jumping and clapping, balancing on tiptoe while opening and closing one's hands, and balancing a rod

KEY TERMS

Cerebral palsy—A nonprogressive movement disability caused by abnormal development of or damage to motor control centers of the brain.

Congenital malformation—A deformity present at birth.

Developmental coordination disorder—A disorder of motor skills.

Fine motor skill—The abilities required to control the smaller muscles of the body for writing, playing an instrument, artistic expression and craft work. The muscles required to perform fine motor skills are generally found in the hands, feet and head.

Lincoln-Oseretsky Motor Development Scale—A test that assesses the development of motor skills.

Locomotion—The ability to move from one place to another.

Muscular dystrophy—A group of inherited diseases characterized by progressive wasting of the muscles.

vertically. Norms have been established for each part of the test for children aged six to 14.

When to call the doctor

Parents, who suspect that their child has a delay in developments should follow their instincts in having that child evaluated. The earliest intervention possible offers the highest response and success rate among children with special needs. Parents should call the doctor any time they have a concern about their child's motor skill development. Parents should keep in mind that children develop at different rates and try to focus on the skills their children have mastered instead of those they may have yet to master. Still, there are certain signs that may point to a problem, and these should be discussed with a pediatrician or physician. These signs include not walking by 15 months of age, not walking maturely (heel to toe) after walking for several months, walking only on the toes, and not being able to push a toy on wheels by age two. Toddlers may begin to prefer one hand to the other, the first sign of right- or left-handedness, or to use both hands equally. This preference should be allowed to develop naturally. Parents should call a doctor if the child does not seem to use one hand at all or has a strong hand preference before he or she is one year old.

See also Fine motor skills.

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Growth *see* **Skeletal development**

Growth hormone tests

Definition

Growth hormone tests measure the levels of specific hormones that regulate human growth. These hormone levels are measured in blood serum samples obtained by venipuncture. To study growth hormone function under specific conditions, certain medications may be administered before blood is taken and hormone levels are measured. Human growth hormone (hGH) (somatotropin) is produced by somatotropes in the anterior pituitary gland. Its role in normal body growth and development is to stimulate protein production in muscle cells and trigger energy release from the breakdown of fats. Diagnostic tests for growth hormones include the somatotropin hormone test, somatomedin C test, growth hormone stimulation test (also known as the arginine test or insulin tolerance test), and growth hormone suppression test (glucose loading test).

Purpose

Growth hormone tests are ordered by physicians to determine whether levels of hGH and other related hormones in the blood are normal, increased, or decreased, and to help diagnose conditions that may result from abnormal hormone levels or pituitary gland dysfunction. Some of the common reasons for testing are:

- to identify growth abnormalities that may cause delayed **puberty** and small stature in adolescents
- to aid in the diagnosis of hyperpituitarism, which can cause **gigantism** or **acromegaly**
- to screen for pituitary gland dysfunction
- to assist in the diagnosis of pituitary tumors or tumors related to the hypothalamus, an area of the brain
- to monitor the effects of hGH therapy administered for certain conditions

Description

Human growth hormones play an important role in normal human growth and development. The major human growth hormone is a protein made up of 191 amino acids, the building blocks of proteins. The production of this protein is controlled by two other hormones secreted by the hypothalamus: growth hormone releasing hormone (GHRH), which controls secretion of hGH; and growth hormone-inhibiting hormone (GHIH), which inhibits secretion of hGH. All healthy individuals have measurable levels of hGH throughout life, but there are two notable growth spurts, one at birth and the other at puberty, and hGH plays a vital role at each time. The most obvious effect of hGH is on linear skeletal growth (height), but metabolic effects of hGH (the results of hGH activity in the body) on muscle, the liver, and fat cells are a critical part of its function. When any question arises about growth or development, pediatricians may investigate the levels of the major growth hormone hGH, its receptors and stimulants, the glands that produce the hormones, and the complex hormone interactions that control normal development.

Somatotropin (hGH) is secreted by somatotropes in the anterior pituitary gland. It is typically secreted during **sleep**, with peak release occurring around 10 p.m., midnight, and 2 a.m. Most of the effects of hGH are mediated by other hormones, including the somatomedins, IGH-I (somatomedin C) and IGH-II, which are insulin-like growth hormones that also influence linear growth, and the two hypothalamic hormones (GHRH and GHIH) that regulate hGH by responding to changes in the individual's blood sugar (glucose) and protein levels. When blood glucose levels fall, GHRH triggers the secretion of stored hGH. As blood glucose levels rise, hGH secretion is turned off by GHIH activity. Increases in blood protein levels trigger a similar response. This feedback loop, along with the effects of eating and **exercise**, is responsible for the fluctuating levels of hGH throughout the day. In addition, blood glucose and amino acid availability for growth is also regulated by the hormones adrenaline, glucagon, and insulin. All of these growth factors may be evaluated in order to understand hormone deficiencies or gland dysfunction when growth deficiencies are suspected.

A number of hormonal conditions can lead to excessive or diminished growth. Because of its critical role in producing hGH and other hormones, a dysfunctional pituitary gland will often lead to altered growth. **Dwarfism** (very small stature) can be due to underproduction of hGH, lack of IGH-I, or a flaw in target tissue response to either of these growth hormones. Overproduction of hGH or IGH-I, or an exaggerated response to these hormones, can lead to gigantism or acromegaly, both of which are characterized by a very large stature.

Gigantism is the result of hGH overproduction in early childhood leading to a skeletal height up to 8 feet (2.5 m) or more. Acromegaly results when hGH is overproduced after the onset of puberty. In this condition, the epiphyseal plates of the long bones of the body do not close, and they remain responsive to additional stimulated growth by hGH. This disorder is characterized by an enlarged skull, hands and feet, nose, neck, and tongue.

Somatrotropin

Somatrotropin (hGH) is measured in the clinical laboratory to identify hGH deficiency in adolescents with short stature, delayed sexual maturity, and other growth or development abnormalities. The somatotropin test also aids in documenting the excess hGH production responsible for gigantism or acromegaly, and confirms underactivity or overproduction of the pituitary gland (hypopituitarism or hyperpituitarism, respectively). However, due to variable secretion of hGH, as well as hGH production in response to stress, exercise, or other factors, random assays are not an adequate determination of hGH deficiency. To obtain more accurate readings, a blood sample can be drawn one to one-and-a-half hours after sleep (hGH levels increase during sleep), or strenuous exercise can be performed for 30 minutes before blood is drawn. (A person's hGH levels increase after exercise.) The hGH levels at the end of an exercise period are expected to be maximal.

Somatomedin C

The somatomedin C test is usually ordered to help detect pituitary abnormalities, hGH deficiency, and acromegaly. Also called insulin-like growth factor (IGF-1), somatomedin C is considered a more accurate reflection of the blood concentration of hGH because such variables as time of day, activity levels, or diet do not influence test results. Somatomedin C is part of a group of peptides, called somatomedins, through which hGH exerts its effects. Because it circulates in the bloodstream bound to long-lasting proteins, it is more stable than hGH. Levels of somatomedin C do depend on hGH levels, however, and typically somatomedin C levels will be low when hGH levels are deficient. Abnormally low levels of somatomedin C will require further investigation, so doctors may perform the hGH stimulation test to diagnose hGH deficiency. Nonpituitary causes of reduced somatomedin C include **malnutrition**, severe chronic illness, severe liver disease, **hypothyroidism**, and Laron's dwarfism.

Growth hormone stimulation test

The hGH stimulation test, also called hGH provocation test, insulin tolerance, or arginine test, is performed to test the body's ability to produce human growth hormone and to confirm suspected hGH deficiency. A normal patient can have low hGH levels, but if hGH is still low after stimulation, a more definitive diagnosis can be made. The test involves creating a condition of insulin-induced **hypoglycemia** (via intravenous injection of insulin) to stimulate production of hGH and corticotropin secretion as well. If such stimulation is unsuccessful, a malfunction of the anterior pituitary gland is likely. It may be necessary to obtain blood samples following an energetic exercise session lasting 20 minutes.

A substance called hGH-releasing factor has also been used for hGH stimulation. This approach is believed to be more accurate and specific for hGH deficiency caused by the pituitary. Growth hormone deficiency is also suspected when x-ray determination of bone age indicates retarded growth in comparison to chronological age. As of 2004, the best method to identify hGH-deficient patients was a positive stimulation test followed by a positive response to a therapeutic trial of hGH.

Growth hormone suppression test

This procedure, also called the glucose loading test, is performed to evaluate excessive baseline levels of hGH and to confirm diagnosis of gigantism in children (and acromegaly in adults). The procedure requires drawing two different blood samples, one before the child ingests 100 grams of glucose by mouth and a second sample two hours after glucose ingestion. Normally, a glucose load such as this will suppress hGH secretion. In a child with excessive hGH levels, failure of suppression indicates anterior pituitary dysfunction and confirms a diagnosis of gigantism (or acromegaly).

Precautions

Taking certain drugs such as amphetamines, dopamine, corticosteroids, and phenothiazines may increase or decrease growth hormone secretion. A pediatrician may discontinue certain medications prior to the performance of growth hormone tests. Other factors that may influence hGH secretion include stress, exercise, diet, and abnormal glucose levels. The pediatrician may make recommendations for the child's activity prior to testing. Growth hormone tests should not be done within a week after any radioactive scan such as an x ray, MRI, or CT scan.

Preparation

The hGH or somatotropin test requires that a fasting blood sample be drawn from a vein, usually in the arm. The child should have nothing to eat or drink from midnight the night before the test until after the blood sample is drawn. Since stress and exercise increase hGH levels, the child must be at complete rest for 30 minutes before the blood sample is drawn. If the physician has requested two samples, they should be drawn on consecutive days at approximately the same time, preferably between 6 a.m. and 8 a.m.

The somatomedin C test also requires a fasting blood sample. The patient should have nothing to eat or drink from midnight the night before until after the blood sample is drawn.

Growth hormone stimulation testing requires intravenous administration of arginine and/or insulin. Venous blood samples will be drawn at 0, 60, and 90 minutes after the injection.

Growth hormone suppression testing requires two fasting blood samples, one before the test and another two hours after the child is given a glucose solution by mouth. The child should have nothing to eat or drink from midnight the night before until after the blood samples are drawn, and physical activity should be limited for at least 10 to 12 hours before the test.

Aftercare

Usually there will be no effects from hormone testing and normal activities can be resumed. A bandage may be applied to keep the site of venipuncture or intravenous administration of medications clean and to stop any bleeding that may occur. Unusual bleeding or bruising of the site should be reported to the pediatrician. The child should be observed closely after the more extensive growth hormone stimulation test and growth hormone suppression test. A pediatrician may limit activities for the immediate pre-test period.

Risks

Growth hormone tests do not have significant risks. Minor discomfort may be experienced during and after the growth hormone stimulation test because of the intravenous line for delivery of insulin. A low blood sugar (hypoglycemia) will result from the insulin injected into the child's system, which may make some children lightheaded or lethargic. Some children may experience sleepiness, sweating, and/or nervousness, all of which can be corrected after the test by ingestion of juice or a glucose infusion, as recommended by the pediatrician. Severe cases of hypoglycemia may cause ketosis (excessive amounts of fatty acid byproducts in the body), acidosis (a disturbance of the body's acid-base balance), or shock. Medical personnel provide close observation during the test to help prevent or react to these unlikely reactions. Growth hormone suppression tests can cause some children to feel nauseous after the administration of glucose. Ice chips can help alleviate this symptom.

Normal results

Results are reported in nanograms per milliliter (ng/ml). Normal results may vary from laboratory to laboratory depending upon the method used for measurement, but results are usually within the following ranges.

Somatotropin (hGH):

- men: 5 ng/ml
- women: less than 10 ng/ml
- children: 0–10 ng/ml
- newborn: 10–40 ng/ml

Somatomedin C:

- adult: 42–110 ng/ml
- child: 0–8 years; girls 7–110 ng/ml; boys 4–87 ng/ml
- 9–10 years: girls 39–186 ng/ml; boys 26–98 ng/ml
- 11–13 years: girls 66–215 ng/ml; boys 44–207 ng/ml
- 14–16 years: girls 96–256 ng/ml; boys 48–255 ng/ml

Growth hormone stimulation: greater than 10 ng/ml

Regarding growth hormone suppression, normally, glucose suppresses hGH to levels ranging from undetectable to 3 ng/ml within 30 minutes to two hours. In children, rebound stimulation may occur after two to five hours.

KEY TERMS

Acromegaly—A rare disease resulting from excessive growth hormone caused by a benign tumor. If such a tumor develops within the first ten years of life, the result is gigantism (in which growth is accelerated) and not acromegaly. Symptoms include coarsening of the facial features, enlargement of the hands, feet, ears, and nose, jutting of the jaw, and a long face.

Dwarfism, pituitary—Short stature. When caused by hGH deficiency, as opposed to late growth spurt or genetics, abnormally slow growth and short stature with normal proportions may be seen.

Gigantism—Excessive growth, especially in height, resulting from overproduction of growth hormone during childhood or adolescence by a pituitary tumor. Untreated, the tumor eventually destroys the pituitary gland, resulting in death during early adulthood. If the tumor develops after growth has stopped, the result is acromegaly, not gigantism.

Pituitary gland—The most important of the endocrine glands (glands that release hormones directly into the bloodstream), the pituitary is located at the base of the brain. Sometimes referred to as the "master gland," it regulates and controls the activities of other endocrine glands and many body processes including growth and reproductive function. Also called the hypophysis.

Parental concerns

Parents may be concerned about the child's response to venipuncture or reaction to intravenous administration of medications prior to testing. The parents can play a calming role by reassuring the child and explaining the procedure beforehand. Medical personnel and the pediatrician may provide instruction before, during, and after the tests, as well as close observation to minimize any risks. Parents can be prepared for post-testing sleepiness or lightheadedness by having juice handy and ice chips to help relieve any feelings of **nausea**.

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Gum disease *see* **Periodontal disease**

H

Haemophilus influenzae infections *see* **Hemophilus infections**

Haemophilus influenzae type B vaccine *see* **Hib vaccine**

Hair loss *see* **Alopecia**

Handedness

Definition

Handedness is the preferred use of the right hand, the left hand, or one or the other depending on the task.

Description

Handedness is defined and categorized in different ways. Most people define handedness as the hand that one uses for writing. Within the scientific community some researchers define handedness as the hand that is faster and more precise for manual tasks. Others define it as the preferred hand, regardless of its abilities. Whereas some people always use their right hand or their left hand for most activities, others use one hand or the other depending on the activity. Still other people can use either hand for most functions.

Lefthanders usually prefer using their left hand for delicate tasks; however, there is no good method for predicting which hand a lefthander will choose for a specific task. Although left-handed children usually are more flexible in their hand usage than right-handers, this may be because they are forced to function in a world designed for right-handers.

There is no standard measure for determining degrees of handedness. Some scientists believe that there are only two types of handedness: right and non-right. These researchers believe that true left-handedness is rare and that most lefthanders are really mixed-handed. Others believe that ambidexterity—the equal use of both hands—is a third type of handedness, and some think that there are two types of ambidexterity. Other scientists believe that handedness should be measured on a continuum from completely right-handed to completely left-handed.

Demographics

It is commonly estimated that about 10 percent of the human population is left-handed or ambidextrous. Boys are about 1.5 times more likely than girls to be left-handed. Archeological evidence indicates that the proportion of left-handed to right-handed people was about the same 30,000 years ago as it is in the early 2000s.

In the past there were many social and cultural biases against left-handed children. In particular, left-handed children often were forced by parents or teachers to use their right hand for eating and writing. In the early 2000s the frequency of left-handedness appears to be on the increase. This may be due to the increased acceptance of children determining their own hand preferences. Left-handedness appears to be rarer in restrictive societies as compared with more permissive societies.

Basis of handedness

The physical basis of handedness is not well-understood. Through the centuries left-handedness has been attributed to numerous physical, psychological, and supernatural causes.

Each hemisphere of the brain has some specialized functions, a poorly-understood phenomenon called brain lateralization. In the late nineteenth century Paul Broca, a French neurosurgeon, identified an area of the left hemisphere that has a major role in the production of speech. Carl Wernicke, a German neurologist, identified another region in the left hemisphere that was responsible for language comprehension. Broca suggested that people's handedness was the opposite of their language-specialized hemisphere, so that a person with

left-hemisphere language specialization would be right-handed. Thus until the 1960s, handedness was believed to be indicative of brain lateralization. Between 70 and 90 percent of humans have language specialization in their left hemispheres. The remainder may have right-hemisphere specialization or no real distinction between the two hemispheres in language specialization. However, among lefthanders, about 50 percent process language on both sides of their brains, 10 percent process language primarily in their right brains, and the remainder process language primarily in their left brains.

The 1987 Geschwind-Behan-Galaburda (GBG) Theory of Left-Handedness suggested that left-handedness was a result of some brain injury or trauma or chemical variations in the fetal environment, such as high levels of the male hormone testosterone.

For decades during the twentieth century scientists argued about whether there is a genetic basis for handedness. Children of left-handed parents have a 50 percent chance of being right-handed and 18 percent of identical **twins** differ in their handedness. Furthermore, right-handed twins are equally as likely as their left-handed twins to have left-handed children. A 2003 study appeared to identify a single gene that controls both handedness and the direction that hair spins on the scalp. An individual possessing at least one copy of the dominant form of this gene is both right-handed and has a clockwise hair spiral. However, when an individual has two copies of the recessive form of the gene—one copy from the mother and one copy from the father—the gene does not determine handedness. Thus 50 percent of these individuals are right-handed and 50 percent are non-right-handed. Furthermore, these individuals have a separate 50 percent likelihood of hair that spins clockwise or counterclockwise.

Handedness determines few if any lateralized behaviors other than fine finger dexterity. However, one study showed that right-handers preferred turning to their left side and non-right-handers preferred turning to their right side. Turning to the right or left is strongly correlated with turning toward the side of the brain that has less dopamine, an important brain hormone.

Infancy

In his pioneering work on child behavior, the American developmental psychologist Arnold Gesell claimed that infants as young as four weeks display signs of handedness and that right-handedness is clearly established by age one. However, it was as of 2004 commonly believed that babies are born ambidextrous. Although a hand preference may seem apparent towards the end of the first year, this is not necessarily due to right- or left-handedness and may change several times over the subsequent few years.

Toddlerhood

Toddlers usually go through phases of using one hand for some activities and the other hand for other activities. Although many children exhibit clear left- or right-handedness from the age of two—and handedness usually is determined during the third year—it is not unusual for a child to repeatedly switch hand preferences well into their **preschool** years. Early hand preference may be due to a pathological problem (e.g. **stroke**).

Common problems

It may be hard for right-handers to appreciate the daily problems confronting non-right-handers. Although most of these difficulties are simply annoying or frustrating, others can cause physical injury or serious life-long problems. Most systems and tools are designed for right-handers and so have an intrinsic bias. Many items, such as screws and light bulbs, require a left-to-right turning that is easier for a right-hander. Items designed specifically for right-handers include:

- cooking utensils
- can openers
- scissors
- telephones
- calculators
- computer keyboards
- sports equipment
- musical instruments, especially stringed instruments

Non-right-handed children must either learn to use tools with their right hand, which can be awkward, inefficient, and frustrating, or to use tools backwards with their left hand. The latter can be dangerous for a child.

In the past left-handed children often were forced to write with their right hand. In the early 2000s, a non-right-handed child may still feel pressure to conform to a right-handed world. Most parents and teachers as of 2004 probably accept that it is wrong to attempt to suppress or change a child's handedness. Nevertheless, lefthanders may still suffer at school. A teacher may label a lefthander's writing as "sloppy" because of an unconscious reaction to handwriting that looks different. Left-handed children may hook their wrists while writing in order to see the paper. However, hand or wrist twisting can reduce legibility and writing fluency. This problem is avoidable with correct positioning of the paper for the

lefthander. In art and science lefthanders may struggle with tools, instruments, and equipment designed for the right-handed majority. Some left-handed children may seem clumsy as they try to adapt to a right-handed world.

At various times in the past, left-handedness has been wrongly associated with numerous physical, mental, emotional, and behavioral disorders. However, there is some evidence that left-handed people may be more at risk for **schizophrenia**, **bipolar disorder**, or language-processing disorders, including **dyslexia** and **stuttering**.

Parental concerns

Many children repeatedly switch hand preferences until at least the age of three. This is normal unless it seems to interfere with the child's **fine motor skills**. By the age of two most children should be able to do the following:

- hold a fork and spoon well enough to feed themselves
- handle small objects well
- hold the paper in place while drawing

Once a child's handedness becomes apparent, parents or caregivers should never try to change it. Parents can assist left-handed children by the following:

- placing their table settings according to their handedness
- providing left-handed scissors
- helping them find the easiest ways to handle paper and pencil
- assuring that teachers and caregivers treat their lefthanders appropriately
- not dwelling on their child's non-right-handedness

Left-handed children can become very frustrated when they are trying to imitate a right-handed parent or sibling, particularly with activities such as shoe-tying. In these cases the parent or sibling should sit across from the child—rather than next to or behind the child—so as to be the child's mirror.

If handedness is not apparent by the time a child enters school, the teacher must determine which hand the child should learn to write with. Observing which hand the child consistently uses for various activities—or whether the child switches hands when repeating the activity—can help the teacher make this determination. Example of such activities include:

- holding a spoon
- cutting with scissors

KEY TERMS

Ambidextrous—Equally competent with either hand.

Brain lateralization—A function that is dominated by either the left or the right hemisphere of the brain.

Intrinsic bias—An assumed bias that favors one group over another; as in systems and hand implements that assume that all people are right-handed.

- playing with puppets
- using a lock and key
- hammering nails
- screwing lids on jars
- throwing a ball

Teachers should help lefthanders to hold a pencil and place the paper in ways that are appropriate for left-handed writing.

When to call the doctor

A child who exhibits a strong preference for one hand at about one year of age—too early to clearly express handedness—may have a weakness or neuromuscular problem in the other arm or hand.

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Margaret Alic, PhD

Hand-eye coordination

Definition

Hand-eye coordination is the ability of the vision system to coordinate the information received through the eyes to control, guide, and direct the hands in the accomplishment of a given task, such as handwriting or catching a ball. Hand-eye coordination uses the eyes to direct attention and the hands to execute a task.

Description

Vision is the process of understanding what is seen by the eyes. It involves more than simple visual acuity (ability to distinguish fine details). Vision also involves fixation and eye movement abilities, accommodation (focusing), convergence (eye aiming), binocularity (eye teaming), and the control of hand-eye coordination. Most hand movements require visual input to be carried out effectively. For example, when children are learning to draw, they follow the position of the hand holding the pencil visually as they make lines on the paper. Between four and 14 months of age, infants explore their world and develop hand-eye coordination, in conjunction with **fine motor skills**. Fine motor skills are involved in the control of small muscle movements, such as when an infant starts to use fingers with a purpose and in coordination with the eyes.

Infants are eager to move their eyes, their mouths, and their bodies toward the people and objects that comfort and interest them. They practice skills that let them move closer to desired objects and also move desired objects closer to themselves. By six months of age, many infants begin reaching for objects quickly, without jerkiness, and may be able to feed themselves a cracker or similar food. Infants of this age try to get objects within their reach and objects out of their reach. Many infants are also able to look from hand to object, to hold one object while looking for a second object, and to follow the movements of their hands with their eyes. At this age, most infants begin to poke at objects with their index fingers. After six months, infants are usually able to manipulate a cup and hold it by the handle. Many infants at this age also begin to reach for objects with one arm instead of both. At about eight months of age, as dexterity improves, many infants can use a pincher movement to grasp small objects, and they can also clap and wave their hands. They also begin to transfer objects from hand to hand, and bang objects together. Hand-eye coordination development milestones are as follows.

Birth to three years

Between birth and three years of age, infants can accomplish the following skills:

- start to develop vision that allows them to follow slowly moving objects with their eyes
- begin to develop basic hand-eye skills, such as reaching, grasping objects, feeding, dressing
- begin to recognize concepts of place and direction, such as up, down, in
- develop the ability to manipulate objects with fine motor skills

Three to five years

Between three and five years of age, little children develop or continue to develop the following skills:

- continue to develop hand-eye coordination skills and a preference for left or right **handedness**
- continue to understand and use concepts of place and direction, such as up, down, under, beside
- develop the ability to climb, balance, run, gallop, jump, push and pull, and take stairs one at a time
- develop eye/hand/body coordination, eye teaming, and depth perception

Five to seven years

Children between five and seven years old develop or continue to develop the following skills:

- improve fine motor skills, such as handling writing tools, using scissors
- continue to develop climbing, balancing, running, galloping, and jumping abilities
- continue to improve hand-eye coordination and handedness preference
- learn to focus vision on school work for hours every day

Common problems

Hand-eye coordination problems are usually first noted as a lack of skill in drawing or writing. Drawing shows poor orientation on the page and the child is unable to stay "within the lines" when using a coloring book. Often the child continues to depend on his or her hand for inspection and exploration of **toys** or other objects. Poor hand-eye coordination can have a wide variety of causes, but the main two conditions responsible for inadequate hand-eye coordination are vision problems and **movement disorders**.

Vision impairment

Vision impairment is a loss of vision that makes it hard or impossible to perform daily tasks without specialized adaptations. Vision impairment may be caused by a loss of visual acuity, in which the eye does not see objects as clearly as usual. It may also be caused by a loss of visual field, in which the eye cannot see as wide an area as usual without moving the eyes or turning the head. Vision impairment changes how a child understands and functions in the world. Impaired vision necessarily affects a child's hand-eye coordination, as well as cognitive, emotional, neurological, and physical development by limiting the range of experiences and the kinds of information to which the child is exposed.

Movement disorders

Movement disorders are characterized by impaired body movements. They have a variety of causes. An example is ataxia, characterized by a lack of coordination while performing voluntary movements. The problem may appear as clumsiness, inaccuracy, or instability. Movements are not smooth and may appear disjointed or jerky. Another example is hypertonia, a condition marked by an abnormal increase in muscle tension and a reduced ability of a muscle to stretch. Whatever their origin, movement disorders almost always prevent the normal development of hand-eye coordination.

Good hand-eye coordination is needed to play baseball. *(Photograph by Henry Horenstein. Harcourt.)*

Parental concerns

When a child is between one and two years old, parents should start encouraging activities that allow the child to learn how to use hands to manipulate objects and learn how different things fit together. These activities are extremely important for the development of hand-eye coordination, which is itself crucial for the overall physical development of the child. Some toys are designed to assist hand-eye coordination development. They usually involve "fitting things together," but parents can also use simple kitchen cups or bowls. Letting children **play** with these objects teaches them how to fit objects together. Jigsaw puzzles are also good hand-eye coordination developers, as are blocks or tower toys that encourage playing and coordination. These toys help children learn what items can fit on top of each other and which ones will stack easily. By the time children reach two or three years of age, they are ready to start throwing things and wanting to catch them. These play activities are a great way to improve hand-eye coordination. Most child development professionals believe that the best thing parents can do when trying to improve hand-eye coordination in their children is simply to let them play with a variety of toys on their own.

When to call the doctor

Parents of **preschool** children should be alert for signs of abnormal visual development or coordination

KEY TERMS

Ataxia—A condition marked by impaired muscular coordination, most frequently resulting from disorders in the brain or spinal cord.

Binocular vision—Using both eyes at the same time to see an image.

Convergence—The natural movement of the eyes inward to view objects close-up.

Fine motor skill—The abilities required to control the smaller muscles of the body for writing, playing an instrument, artistic expression and craft work. The muscles required to perform fine motor skills are generally found in the hands, feet and head.

Gross motor skills—The abilities required to control the large muscles of the body for walking, running, sitting, crawling, and other activities. The muscles required to perform gross motor skills are generally found in the arms, legs, back, abdomen and torso.

Hypertonia—Having excessive muscular tone or strength.

Motor skills—Controlled movements of muscle groups. Fine motor skills involve tasks that require dexterity of small muscles, such as buttoning a shirt. Tasks such as walking or throwing a ball involve the use of gross motor skills.

Visual acuity—Sharpness or clearness of vision.

problems. If a child is reluctant to engage in activities that require hand-eye coordination (does not like Legos, tinker toys) or has very irregular handwriting with excessive erasures, they should consider having the child evaluated.

See also Bayley Scales of Infant Development.

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Hand-foot-mouth disease

Definition

Hand-foot-mouth disease is an infection of young children in which characteristic fluid-filled blisters appear on the hands, feet, and inside the mouth.

Description

Coxsackie viruses belong to a family of viruses called enteroviruses. These viruses live in the gastrointestinal tract and are, therefore, present in feces. They can be spread easily from one person to another when poor hygiene allows the virus within the feces to be passed from person to person. After exposure to the virus, development of symptoms takes only four to six days. Hand-foot-mouth disease can occur year-round, although the largest number of cases are in summer and fall months.

An outbreak of hand-foot-mouth disease occurred in Singapore in 2000, with more than 1,000 diagnosed cases, all in children, resulting in four deaths. A smaller outbreak occurred in Malaysia in 2000. In 1998, a serious outbreak of enterovirus in Taiwan resulted in more than 1 million cases of hand-foot-and-mouth disease. Of these, there were 405 severe cases and 78 deaths, 71 of which were children younger than five years of age.

Hand-foot-mouth should not be confused with foot and mouth disease, which infects cattle but is extremely rare in humans. An outbreak of foot and mouth disease swept through Great Britain and into other parts of Europe and South America in 2001.

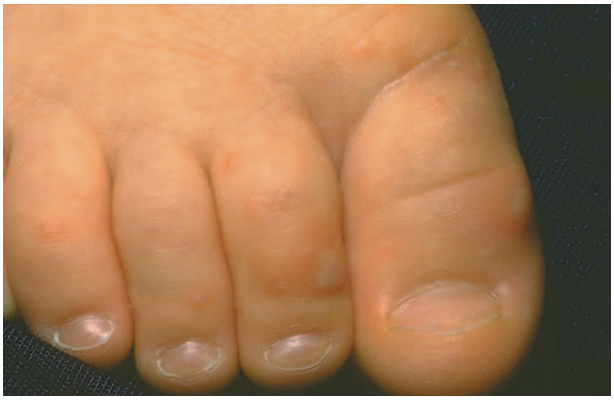
Demographics

Hand-foot-mouth disease is very common among young children and often occurs in clusters of children who are in daycare together.

Causes and symptoms

Hand-foot-mouth disease is spread when poor hand washing after a diaper change or contact with saliva allows the virus to be passed from one child to another.

Within about four to six days of acquiring the virus, an infected child may develop a relatively low-grade **fever**, ranging from 99 to 102°F (37.2–38.9°C). Other symptoms include fatigue, loss of energy, decreased appetite, and a sore sensation in the mouth that may interfere with feeding. After one to two days, fluid-filled bumps (vesicles) appear on the inside of the mouth, along the surface of the tongue, on the roof of the mouth, and on the insides of the cheeks. These are tiny blisters, about 3–7 mm in diameter. Eventually, they may appear on the palms of the hands and on the soles of the feet. Occasionally, these vesicles may occur in the diaper region.



Blisters shown on the foot a child with hand-foot-mouth disease. *(Custom Medical Stock Photo, Inc.)*

The vesicles in the mouth cause the majority of discomfort, and the child may refuse to eat or drink due to **pain**. This phase usually lasts for an average of a week. As long as the bumps have clear fluid within them, the disease is at its most contagious. The fluid within the vesicles contains large quantities of the causative viruses. Extra care should be taken to avoid contact with this fluid.

Diagnosis

Diagnosis is made by most practitioners solely on the basis of the unique appearance of blisters of the mouth, hands, and feet, in a child not appearing very ill.

Treatment

As of 2004, there were no treatments available to cure or decrease the duration of the disease. Medications like **acetaminophen** or ibuprofen may be helpful for decreasing pain and helping the child to eat and drink. It is important to try to encourage the child to take in adequate amounts of fluids, in the form of ice chips or Popsicles if other foods or liquids are too uncomfortable. There is a risk of developing **dehydration**.

KEY TERMS

Enterovirus—Any of a group of viruses that primarily affect the gastrointestinal tract. The coxsackievirus and the poliovirus are both enteroviruses.

Vesicle—A bump on the skin filled with fluid.

Prognosis

The prognosis for a child with hand-foot-mouth disease is excellent. The child is usually completely recovered within about a week of the start of the illness.

Prevention

Prevention involves careful attention to hygiene. Thorough, consistent hand-washing practices and discouraging the sharing of clothes, towels, and stuffed **toys** are all helpful. Virus continues to be passed in the feces for several weeks after infection, so good hygiene should be practiced long after all signs of infection have passed.

Prevention

Parents should be aware of the characteristic rash of hand-foot-mouth disease and monitor their children, especially if they are in a child care setting. Good hygiene practices should be strictly followed to prevent the spread of the disease.

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Happy puppet syndrome *see* **Angelman's syndrome**
Harelip *see* **Cleft lip and palate**
Hashimoto's thyroiditis *see* **Hypothyroidism**
Haverhill fever *see* **Rat-bite fever**
Hay fever *see* **Allergic rhinitis**
HBF test *see* **Fetal hemoglobin test**
HBV *see* **Hepatitis B vaccine**

Head injury

Definition

Head injury is an injury to the scalp, skull, or brain. The most important consequence of head trauma is traumatic brain injury. Head injury may occur either as a closed head injury, such as the head hitting a car's windshield; or as a penetrating head injury, as when a bullet pierces the skull. Both may cause damage that ranges from mild to profound. Very severe injury can be fatal because of profound brain damage.

Description

External trauma to the head is capable of damaging the brain, even if there is no external evidence of damage. More serious injuries can cause skull fracture, blood clots between the skull and the brain, or bruising and tearing of the brain tissue itself.

Injuries to the head can be caused by traffic accidents, **sports injuries**, falls, workplace accidents, assaults, or bullets. Most people have had some type of head injury at least once in their lives, but rarely do they require a hospital visit.

Demographics

Each year about two million people suffer from a more serious head injury, and up to 750,000 of those are severe enough to require **hospitalization**. Brain injury is most likely to occur in males between ages 15 and 24, usually as a result of car and motorcycle accidents. About 70 percent of all accidental deaths are due to head injuries, as are most of the disabilities that occur after trauma. Among children and infants, head injury is the most common cause of death and disability. The most

common cause of head injury in children under age two is **child abuse**.

Causes and symptoms

A head injury may cause damage both from the direct physical injury to the brain and from secondary factors, such as lack of oxygen, brain swelling, and disturbance of blood flow. Both closed and penetrating head injuries can cause swirling movements throughout the brain, tearing nerve fibers and causing widespread bleeding or a blood clot in or around the brain. Swelling may raise pressure within the skull (intracranial pressure) and may block the flow of oxygen to the brain.

Head trauma may cause a **concussion**, in which there is a brief loss of consciousness without visible structural damage to the brain. In addition to loss of consciousness, initial symptoms of brain injury may include:

- memory loss and confusion
- vomiting
- **dizziness**
- partial paralysis or numbness
- shock
- anxiety

After a head injury, there may be a period of impaired consciousness followed by a period of confusion and impaired memory with disorientation and a breakdown in the ability to store and retrieve new information. Others experience temporary amnesia following head injury that begins with memory loss over a period of weeks, months, or years before the injury (retrograde amnesia). As a person recovers, memory slowly returns. Post-traumatic amnesia refers to loss of memory for events during and after the accident.

Epilepsy occurs in 2–5 percent of those who have had a head injury; it is much more common in people who have had severe or penetrating injuries. Most cases of epilepsy appear right after the accident or within the first year and become less likely with increased time following the accident.

Closed head injury

Closed head injury refers to brain injury without any penetrating injury to the brain. It may be the result of a direct blow to the head; of the moving head being rapidly stopped, such as when a person's head hits a windshield in a car accident; or by the sudden deceleration of the head without its striking another object. The kind of injury the brain receives in a closed head injury is determined by whether the head was unrestrained upon impact and the direction, force, and velocity of the blow. If the head is resting on impact, the maximum damage will be found at the impact site. A moving head will cause a contrecoup injury where the brain damage occurs on the side opposite the point of impact, as a result of the brain slamming into that side of the skull. A closed head injury also may occur without the head being struck, such as when a person experiences whiplash. This type of injury occurs because the brain is of a different density than the skull and can be injured when delicate brain tissues hit against the rough, jagged inner surface of the skull.

Penetrating head injury

If the skull is fractured, bone fragments may be driven into the brain. Any object that penetrates the skull may implant foreign material and dirt into the brain, leading to an infection.

Skull fracture

A skull fracture is a medical emergency that must be treated promptly to prevent possible brain damage. Such an injury may be obvious if blood or bone fragments are visible, but it is possible for a fracture to have occurred without any apparent damage. A skull fracture should be suspected if there is:

- blood or clear fluid leaking from the nose or ears
- unequal pupil size
- bruises or discoloration around the eyes or behind the ears
- swelling or depression of part of the head

Intracranial hemorrhage

Bleeding (hemorrhage) inside the skull may accompany a head injury and cause additional damage to the brain. A blood clot (hematoma) may occur if a blood vessel between the skull and the brain ruptures; when the blood leaks out and forms a clot, it can press against brain tissue, causing symptoms from a few hours to a few weeks after the injury. If the clot is located between the bones of the skull and the covering of the brain (dura), it is called an epidural hematoma. If the clot is between the dura and the brain tissue itself, the condition is called a **subdural hematoma**. In other cases, bleeding may occur deeper inside the brain. This condition is called intracerebral hemorrhage or intracerebral contusion (from the word for bruising).

In any case, if the blood flow is not stopped, it can lead to unconsciousness and death. The symptoms of bleeding within the skull include:

- nausea and vomiting
- **headache**
- loss of consciousness
- unequal pupil size
- lethargy

Postconcussion syndrome

If the head injury is mild, there may be no symptoms other than a slight headache. There also may be confusion, dizziness, and blurred vision. While the head injury may seem to have been quite mild, in many cases symptoms persist for days or weeks. Up to 60 percent of persons who sustain a mild brain injury continue to experience a range of symptoms called postconcussion syndrome as long as six months or a year after the injury.

The symptoms of postconcussion syndrome can result in a puzzling interplay of behavioral, cognitive, and emotional complaints that can be difficult to diagnose, including the following:

- headache
- dizziness
- mental confusion
- behavior changes
- memory loss
- cognitive deficits
- depression
- emotional outbursts

When to call the doctor

A parent of a child who has had a head injury and who is experiencing any the following symptoms should seek medical care immediately:

- serious bleeding from the head or face
- loss of consciousness, however brief
- confusion and lethargy
- lack of pulse or breathing
- clear fluid drainage from the nose or ear

Diagnosis

The extent of damage in a severe head injury can be assessed with **computed tomography** (CT) scan, **magnetic resonance imaging** (MRI), positron emission tomography (PET) scans, electroencephalograms (EEG), and routine neurological and neuropsychological evaluations.

A three-dimensional computed tomography (CT) scan of a human skull showing a depressed skull fracture above the right eye. *(Custom Medical Stock Photo, Inc.)*

Doctors use the Glasgow Coma Scale to evaluate the extent of brain damage based on observing a person's ability to open his or her eyes, respond verbally, and respond to stimulation by moving (motor response). People can score from three to 15 points on this scale. People who score below eight when they are admitted usually have suffered a severe brain injury and will need rehabilitative therapy as they recover. In general, higher scores on the Glasgow Coma Scale indicate less severe brain injury and a better prognosis for recovery.

Individuals with a mild head injury who experience symptoms are advised to seek out the care of a specialist; unless a **family** physician is thoroughly familiar with medical literature in this area, experts warn that there is a good chance that people's complaints after a mild head injury will be downplayed or dismissed. In the case of mild head injury or postconcussion syndrome, CT and MRI scans, electroencephalograms (EEG), and routine

neurological evaluations all may be normal because the damage is so subtle. In many cases, these tests cannot detect the microscopic damage that occurs when fibers are stretched in a mild, diffuse injury. In this type of injury, the axons lose some of their covering and become less efficient. This mild injury to the white matter reduces the quality of communication between different parts or the brain. A PET scan, which evaluates cerebral blood flow and brain metabolism, may be of help in diagnosing mild head injury.

Persons with continuing symptoms after a mild head injury should call a local chapter of a head-injury foundation that can refer people to the best nearby expert.

Treatment

If a concussion, bleeding inside the skull, or skull fracture is suspected, the person should be kept quiet in a darkened room, with head and shoulders raised slightly on pillow or blanket.

After initial emergency treatment, a team of specialists may be needed to evaluate and treat the problems that result. A penetrating wound may require surgery. Those with severe injuries or with a deteriorating level of consciousness may be kept hospitalized for observation. If there is bleeding inside the skull, the blood may need to be surgically drained; if a clot has formed, it may need to be removed. Severe skull **fractures** also require surgery. Supportive care and specific treatments may be required if the person experiences further complications. People who experience seizures, for example, may be given anticonvulsant drugs, and people who develop fluid on the brain (**hydrocephalus**) may have a shunt inserted to drain the fluid.

In the event of long-term disability as a result of head injury, there are a variety of treatment programs available, including long-term rehabilitation, coma treatment centers, transitional living programs, behavior management programs, life-long residential or day treatment programs and independent living programs.

Prognosis

Prompt, proper diagnosis and treatment can help alleviate some of the problems that may develop after a head injury. However, it usually is difficult to predict the outcome of a brain injury in the first few hours or days; a person's prognosis may not be known for many months or even years.

The outlook for someone with a minor head injury generally is good, although recovery may be delayed, and symptoms such as headache, dizziness, and cognitive problems can persist for up to a year or longer after an accident. This can limit a person's ability to work and cause strain in personal relationships.

Serious head injuries can be devastating, producing permanent mental and physical disability. Epileptic seizures may occur after a severe head injury, especially a penetrating brain injury, a severe skull fracture, or a serious brain hemorrhage. Recovery from a severe head injury can be very slow, and it may take five years or longer to heal completely. Risk factors associated with an increased likelihood of memory problems or seizures after head injury include age, length and depth of coma, duration of post-traumatic and retrograde amnesia, presence of focal brain injuries, and initial Glasgow Coma Scale score.

As researchers learn more about the long-term effects of head injuries, they uncover links to later conditions. A 2003 report found that mild brain injury during childhood could speed up expression of **schizophrenia** in those who were already likely to get the disorder

KEY TERMS

Computed tomography (CT)—An imaging technique in which cross-sectional x rays of the body are compiled to create a three-dimensional image of the body's internal structures; also called computed axial tomography.

Electroencephalogram (EEG)—A record of the tiny electrical impulses produced by the brain's activity picked up by electrodes placed on the scalp. By measuring characteristic wave patterns, the EEG can help diagnose certain conditions of the brain.

Magnetic resonance imaging (MRI)—An imaging technique that uses a large circular magnet and radio waves to generate signals from atoms in the body. These signals are used to construct detailed images of internal body structures and organs, including the brain.

Positron emission tomography (PET)—A computerized diagnostic technique that uses radioactive substances to examine structures of the body. When used to assess the brain, it produces a three-dimensional image that shows anatomy and function, including such information as blood flow, oxygen consumption, glucose metabolism, and concentrations of various molecules in brain tissue.

because of genetics. Those with a history of a childhood brain injury, even a minor one, were more likely to get familial schizophrenia than a sibling and to have earlier onset. Another study in 2003 found that people who had a history of a severe head injury were four times more likely to develop Parkinson's disease than the average population. Those requiring hospitalization for their head injuries were 11 times as likely. The risk did not increase for people receiving mild head injuries.

Prevention

Many severe head injuries could be prevented by wearing protective helmets during certain **sports** and when riding a bike or motorcycle. Seat belts and airbags can prevent many head injuries that result from car accidents. Appropriate protective headgear always should be worn on the job where head injuries are a possibility.

Parental concerns

Parents should insist that their children always use a seat belt when riding in a car. They should also insist that appropriate protective headgear always be worn when children engage in activities such as bicycling or roller-blading during which a head injury is possible. If a parent suspects a caregiver of abusing their child, prompt intervention is required.

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American Academy of Neurology. 1080 Montreal Avenue, St. Paul, MN 55116. Web site: <www.aan.com/>

American Academy of Pediatrics. 141 Northwest Point Blvd., Elk Grove Village, IL 60007–1098. Web site: <www.aap.org/>.

American College of Emergency Physicians. PO Box 619911, Dallas, TX 75261–9911. Web site: <www.acep.org/>.

American College of Sports Medicine. 401 W. Michigan St., Indianapolis, IN 46202–3233. Web site: <www.acsm.org/>.

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Head lice *see* **Lice infestation**

Head Start programs

Definition

Head Start is a federally funded **preschool** program that provides comprehensives services to both low-income children and their families.

Description

Head Start is a federal program for preschool children three to five years of age in low-income families. Its aim is to prepare children for success in school through an early learning program. The Head Start program is managed by local nonprofit organizations in almost every county in the country. Children who attend Head Start engage in various education activities. They also receive free medical and dental care, have healthy meals and snacks, and enjoy playing indoors and outdoors in a safe setting.

Head Start helps all children succeed, even those with disabilities. Services are also available to infants and toddlers in selected sites.

Head Start began in 1965 as part of the War on Poverty program launched by president Lyndon B. Johnson. Nearly half the nation's poor people were children under age 12, and Head Start was developed to respond as early as possible to the needs of poor children. A few privately funded preschool programs for poor children in inner cities and rural areas showed marked success in raising children's intellectual skills. Many low-income children also had unrecognized health problems and had not been immunized. Head Start was imagined as a comprehensive program that would provide health and nutritional services to poor children, while also developing their cognitive skills. The program aimed to involve parents as well. Many parents of children in the program were employed as teachers' aides so they would understand what their children were learning and help carry on that learning at home.

The program was political from its beginning. Head Start was launched with much fanfare by Lady Bird Johnson, Lyndon Johnson's wife. Measuring the program's success is not a simple matter, however. Head Start saves taxpayers' money, because children who attend Head Start are more likely to graduate high school and get a job than their peers who do not attend. While the savings long-term that result from this program cannot be estimated in dollar value, some sources have suggested that $6 are probably saved for every $1 invested in the Head Start program. Other studies merely suggest that Head Start graduates are more likely than their peers to stay in the proper grade level for their age in elementary school.

In the early 2000s, Head Start serves nearly 700,000 children across the nation. Most programs are half-day and include lunch. The curriculum is not the same in every program, but in most programs school readiness is stressed. Children may be taught the alphabet and numbers and to recognize colors and shapes. Health care is an important part of the program, and children in Head Start are surveyed to keep them up-to-date on their immunizations, and testing is also available for hearing and vision. Most centers are accredited by the National Association for Education of Young Children (NAEYC).

What Head Start programs offer

Head Start provides children with work that helps them grow mentally, socially, emotionally, and physically. The staff in these programs recognizes parents as the first and most important teachers of their children. They welcome parental involvement in the programs and will work as partners to help both the parent and child progress.

The staff create an environment that offers the child love, acceptance, understanding, and the opportunity to learn and to experience success. Head Start children socialize with others, solve problems, and have other experiences that help them become self-confident. The children also improve their listening and speaking skills.

The children spend time in stimulating settings where they form good habits and enjoy playing with **toys** and working on tasks with classmates. Children leave Head Start programs more prepared for kindergarten, excited about learning, and ready to succeed.

Head Start routine

When the children arrive at the Head Start center, they are greeted by their teachers or student aides. They put whatever they have brought from home in a place that is their own to use every day. Classroom time includes many different tasks. Some teachers begin the day by asking the children to sit in a circle. This encourages the children to talk about an idea or experience they want to share with others. In some centers, the children plan their work. They choose among art, playing and blocks or table toys, science, dancing to music, looking at books, or pretend housekeeping. Children can switch tasks when they are ready for a change.

Each day, the children have time to work in small groups with other children and to **play** outdoors on safe playground equipment when weather allows. In bad weather indoor play is planned. At lunchtime children

receive a nutritious meal and brush their teeth after eating. All children are taught to wash their hands before meals and are encouraged to develop good personal and health habits. If they come for an afternoon session, they receive a healthy snack.

Choice of programs

Several different choices are available to meet the varying needs of families:

- Five days per week with half-day preschool classrooms offer various developmental correct educational actions.
- Five days per week, extended-day classroom, are often the choice for working families.
- The combination program option (CPO) strongly focuses on involving the parent, guardian, or primary caregiver in the child's education. Children are invited to take part in a developmentally correct education classroom experience two or three days per week. A home visitor meets with every **family** in their home to provide continuing support and resources at least twice a month.
- Home-based programs provide families with one-hour home visits weekly, and children attend a classroom one day each week. In the home, parents and the visitor plan classwork together.

Family services

Head Start offers children other support services and a chance to be involved in programs designed to help the whole family. Some participating parents learn the English language; others learn to read. Head Start also offers support to parents interested in getting a high school General Equivalency Diploma (GED). If a family member has a special problem, such as drug or alcohol abuse, job loss, or other problem, the family can receive help through Head Start.

Head Start staffers refer families to medical, social welfare, or employment specialists they know in the community, and follow up to be sure the family receives help. Parents can become Head Start volunteers and learn more about child development. This experience may later qualify the volunteer for training that may lead to new employment in the childcare field. Parents also have a voice in the Head Start program by serving on various committees. Parents' experiences in Head Start have raised their own self-confidence and improved their ability to pursue a better life.

Support services for families take various forms. Family counseling advocates work with families to secure proper support to meet individual family needs. Referrals, crisis interventions, and short-term counseling provide families with the necessary tools to become self-sufficient. Health services employ at least one full-time nurse for children and other family members. Nurses screen children within 45 days of enrollment for vision and hearing problems, as well as check each child's height and weight. **Nutrition** and dental services provide students with breakfast and lunch daily. Children in an extended-day program also receive daily snacks. A registered dietitian provides individual nutrition counseling and nutrition workshops. Children learn about good eating habits through weekly nutrition education or cooking projects. A registered dental hygienist helps families find a dentist for their child if needed. Dental screenings are completed on each child within 45 days of enrollment, and hygienists work with children and families to achieve good **oral hygiene**. The program also includes a family liaison, a person who promotes parental participation in the children's education and in workshops on literacy, nutrition, budgeting, health, and other topics. Family orientations are scheduled regularly that give families an opportunity to share in an educational activity with their child. The program offers various educational programs for families such as English as a second language (ESL) and computer science. Disabilities staff serves children with special needs. Developmental screening and **assessment** are provided for the students. Some programs even offer limited bus transportation and interpreters as needed.

Common problems

School phobia and **separation anxiety** affects 3–5 percent of school-age children. The child with school phobia becomes anxious even at the thought of leaving home for school. Complaints of abdominal **pain**, **nausea**, **vomiting**, lack of appetite, and **headache** occur when it is time to go to school and resolve quickly once the child is allowed to remain home. Symptoms do not occur on weekends or holidays unless they are related to going other places, such as Sunday school. These children want to go to school and often earn good grades, but **fear** and **anxiety** prevent them from going.

School phobia in young children has been connected to separation anxiety. A child with separation anxiety is not afraid to go to school but is afraid to leave home. Sometimes children develop school phobia from bullying at school, an excessively critical teacher, and rejection by peers. Events such as marital conflict, moving to a new house, or the arrival of a new sibling can cause fear of going to school.

KEY TERMS

Peer influence—Peer approval or disapproval of the child's behavior or performance.

School phobia—Childhood anxiety about leaving home to attend school.

Separation anxiety—Childhood fear of leaving parents for any reason.

School phobia affects boys and girls equally. Almost all children with school **phobias** have average or above average **intelligence**. School phobia occurs most often at the start of school for children who are three to five years of age.

Most children who enroll in Head Start attend a half-day center-based program. This sometimes causes a problem for homebound or working parents who need to have another form of child care when the four-hour session ends. However, some communities may operate a full-day program or provide Head Start services at home. In a home-based program, a home visitor teaches parents how to provide learning experiences for their children.

Parents working or volunteering in Head Start programs must learn to work with children other than their own. Sometimes they have problems breaking the maternal attachment with their child if the child is attending the same class session. Teaching the child to be independent when their parents are present may not be difficult.

Staffing and funding Head Start programs is a common problem. Hiring qualified personnel in sufficient numbers may be a problem in schools with high enrollment. Staffing ratios and qualifications are established in federal guidelines and are checked by local boards or state department. The staffing needed may also vary with the age and mental health of the children.

Parental concerns

Parents are first concerned about finding a Head Start program in their service area. They can consult the Head Start directory (on the Head Start Bureau web site). Eligibility in Head Start is determined by the federally identified poverty line.

Parents need to communicate with teachers and stay informed about their child's progress. Visiting the classroom and attending parent-teacher conferences and school activities are important. Showing respect for the teacher and supporting the child's efforts helps the child learn.

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Headache

Definition

A headache involves **pain** in the head that can arise from many disorders or may be a disorder in and of itself.

Description

Headaches can be categorized as primary or secondary. Primary headaches occur independently and are not the result of another medical problem. Secondary headaches are caused by illness, infection, or injury and account for less than 10 percent of all headaches.

There are many classifications of headaches, including more than 150 diagnostic headache categories identified by the International Headache Society. In general, there are three types of primary headaches, including:

- Tension headaches—muscular contraction headaches that occur periodically or daily (chronic daily headache). The typical tension-type headache is described as a tightening around the head and neck, and an accompanying dull ache. The headache may last from 30 minutes to several days. Tension headaches usually are not associated with symptoms of **nausea** or **vomiting**.
- Migraine—moderate to severe throbbing pain occurring on one or both sides of the head. Migraines are often accompanied by other symptoms such as nausea, vomiting, blurred vision, and sensitivity to light, sound, strong odors, and movement. A migraine with aura has accompanying "warning signs" that indicate a pending attack. A hemiplegic migraine is associated with weakness on one side of the face, arm, or leg. A migraine may last from two to 48 hours and usually occurs two to four times per month.
- Cluster headaches—severe headaches characterized by pain centering around one eye, and eye tearing and nasal congestion occurring on the same side. The headache lasts from 15 minutes to four hours and may recur several times in a day. Cluster headaches have a characteristic grouping of attacks, which may last from two weeks to three months.

Some chronic tension headaches may start as migraines but become daily headaches. These are called transformed migraines. Drug rebound headaches are those that occur from over-using medications for headache pain; they result from exceeding labeling instructions or a physician's directions.

Headaches that occur along with other neurological symptoms, such as balance problems and vision changes, may be a sign of a disease process in the brain. These organic causes of headache may include **hydrocephalus** (abnormal build-up of fluid in the brain), infection of the brain, tumor, or other conditions.

Demographics

Headaches are very common in children and adolescents. One study reported that 56 percent of boys and 74 percent of girls between ages 12 and 17 have at least one headache within a 30-day period. Tension headaches are the most common type of headache, affecting 15–20 percent of adolescents. The American Council for Headache Education (ACHE) estimates 4–10 percent of children have migraine headaches. Many adults with headaches report that they first began in childhood, and 20 percent report headache onset before age 10. Before **puberty**, migraines occur equally in girls and boys. After puberty, girls are three times more likely to have migraines than boys because of associated hormonal changes and **menstruation**. Headaches are a major cause of missed school days.

Causes and symptoms

Causes

Most headaches in children and adolescents are benign and not the result of an underlying disease or disorder. Rather, most headaches in children are the result of stress and muscle tension, lack of **sleep**, or the **common cold**, flu, or sinus or ear infection.

Traditional theories about headaches link tension-type headaches to muscle contraction, and migraine and cluster headaches to blood vessel dilation (swelling). Pain-sensitive structures in the head include blood vessel walls, membranous coverings of the brain, and scalp and neck muscles. Brain tissue itself has no sensitivity to pain. Therefore, headaches may result from contraction of the muscles of the scalp, face or neck; dilation of the blood vessels in the head; or brain swelling that stretches the brain's coverings. Involvement of specific nerves of the face and head may also cause characteristic headaches. Sinus inflammation is a common cause of headache.

Tension-type headaches are often brought on by emotional or mental stress, overexertion, poor posture, loud noise, and other external factors.

In post-puberty girls, a hormonal connection is likely, since headaches occur at specific points in the menstrual cycle.

Secondary headaches are caused by a wide range of conditions, including some rare diseases and other more treatable conditions. Secondary headaches may be the result of infection, **meningitis**, tumors, or localized **head injury**.

Some headaches have a genetic link; sensitivities to certain environmental triggers and migraines also have been identified in one or both parents.

HEADACHE TRIGGERS Migraines are often triggered by food and environmental factors. Known food triggers include chocolate; aged cheeses; pizza; monosodium glutamate (MSG); bananas; nuts; peanut butter; ice cream; yogurt; fatty or fried foods; processed meats containing nitrates, such as hot dogs and pepperoni; certain food dyes; artificial sweeteners such as aspartame; and **caffeine**. Environmental triggers include weather changes; **smoking**; strong odors; and bright lights. Other triggers include sudden changes in sleep patterns and changes in hormone levels. By keeping a headache diary, the child and parents

can identify and then avoid the specific substances that seem to cause headache symptoms.

When to call the doctor

The parent or caregiver should call the child's pediatrician or neurologist when the child has these symptoms or conditions:

- headache pain that interrupts sleep
- early morning vomiting without an upset stomach
- worsening headache symptoms
- headaches that prevent the child from participating in usual activities
- frequent headaches, occurring three or more times per week
- headache characteristics that are completely different or new
- headache caused by strenuous activity, bending, coughing, or exertion
- headaches that become more severe and/or frequent over time
- **family** history of neurological disease
- headache pain requiring a pain reliever daily or almost every day
- headache pain requiring more than the recommended dose of over-the-counter pain relievers

The parent or caregiver should seek prompt medical attention when the child has these symptoms or conditions:

- Headache is described as the "Worst headache of my life." This may indicate an aneurysm or other neurological emergency.
- Headache accompanied by weakness, **numbness**, paralysis, visual loss, speech difficulty, loss of balance, falling, seizures, shortness of breath, mental confusion, or loss of consciousness. These symptoms could indicate a pending **stroke**.
- Sudden onset of headache, especially if accompanied by a **fever** and stiff neck. These symptoms could indicate meningitis.
- Visual changes, including blurry vision, "blind spots," or double vision.
- Headaches that persist after a head injury or accident.
- Personality changes or inappropriate or unusual behavior.
- Headaches accompanied by severe nausea or vomiting.
- A fever, rash, or stiff neck that occurs with a headache.

Diagnosis

All children who experience headaches on a relatively regular basis should be evaluated. Since headaches arise from many causes, a physical exam assesses general health and a neurological exam evaluates the possibility of neurological disease that is causing the headache. The doctor will look for signs of illness, including fever, high blood pressure, muscle weakness, difficulties with balance, or visual problems.

If the headache is the primary illness, the doctor elicits a thorough history of the headache to help classify the headache, including:

- age of onset
- duration and frequency
- types of headaches experienced
- when the headaches occur
- pain intensity and location
- accompanying symptoms or warning signs of headache onset
- possible triggers or causes of the headaches
- types of headache treatments used and their effectiveness
- presence of any prior symptoms
- impact on school and activities

The child's medical and family history help the physician determine if the child has any conditions or disorders that might contribute to or cause the headache. A family history of migraines or neurological disease might suggest a genetic predisposition to the condition.

The diagnostic evaluation for headache may include blood tests and urinalysis to rule out other medical conditions that may be causing the headaches. Neurological imaging tests such as **computed tomography** (CT) scan or **magnetic resonance imaging** (MRI) may be performed to rule out the presence of neurological diseases or disorders. Other tests may include a sinus x ray and ophthalmology examination. If a condition affecting the brain and spinal cord is suspected, a lumbar puncture or spinal tap may be performed.

A psychological **assessment** is not part of a routine headache evaluation but may be performed to identify stress triggers.

Treatment

The specific treatment prescribed will depend upon the type and frequency of the headache, its cause, and the child's age.

Headache diary

A headache diary can be used to record the characteristics of headaches, including possible triggers, such as foods, weather changes, odors, mood, stressful situations, emotions, or menstrual phases. It also can help the doctor identify the appropriate treatment.

Lifestyle changes

Making certain dietary and lifestyle changes can significantly improve the child's headache symptoms. **Exercise** is an important part of a healthy lifestyle. It aids in stress reduction and improves circulation, which may help reduce headache symptoms. Relaxation and stress management techniques may help the child cope with headache symptoms. Getting enough sleep is equally important; most children and adolescents need at least eight to 10 hours of sleep per night. Counseling can help the child identify stressful situations or events that cause the headaches. It also can teach the child various coping strategies.

Medications

Some children may find enough relief with over-the-counter pain relievers in the right dose. Other children need more aggressive treatment that includes preventive (prophylactic) medication.

Headache medications are classified as abortive, prophylactic, or symptom relief. Abortive medications treat a headache in progress, prophylactic medications prevent a headache, and symptom relief medications relieve associated headache symptoms.

Abortive medications are taken with the onset of the first sign of a migraine. Some prescribed abortive medications include the triptan drugs such as sumatriptan (Imitrex), zolmitriptan (Zomig), naratriptan (Amerge), and ergotamine tartrate and caffeine (Caffergot).

Prophylactic medications are prescribed to treat frequent tension headaches or migraines, or the combination of both headaches. These medications must be taken daily to reduce the frequency and severity of headaches, and they may take a few weeks to be fully effective. Some prophylactic treatments include **antidepressants**, **antihistamines**, nonsteroidal anti-inflammatories (NSAIDs), prednisone, beta-blockers, and calcium channel blockers.

Symptom relief medications are used to relieve symptoms associated with headaches, including headache pain or nausea. These drugs may include over-the-counter pain-relieving medications such as **acetaminophen**, ibuprofen, naproxen, or anti-nausea medications (called antiemitics). Prescribed symptom relief medications may include sedatives (to induce sleep) and muscle relaxants. If symptom relief medications are needed more than twice a week, the child should see his or her doctor, who can make adjustments to the treatment plan. When taken more than three times per week, symptom relief medications can actually cause a type of headache called a rebound headache. To treat rebound headaches, all pain-relieving medications are usually discontinued for a few weeks (as advised by the physician), then used no more than two to three times per week to relieve symptoms.

Alternative treatment

Alternative headache treatments include:

- relaxation techniques, such as meditation, deep breathing exercises, progressive muscle relaxation, guided imagery, and relaxation to music
- **yoga**
- acupuncture or acupressure
- biofeedback
- chiropractic
- homeopathic remedies chosen specifically for the individual and his or her type of headache
- hydrotherapy
- massage to reduce stress and tension and relieve tight muscles in the neck and shoulders
- essential oils such as lavender, ginger, peppermint, and wintergreen that can provide relief by simply smelling them or applying them to the temples or neck
- regular physical exercise

Biofeedback, which teaches patients how to direct mental thoughts to influence physical functions, may be helpful for some patients. For example, patients can use certain relaxation techniques to help them learn how their personal response to muscle tension is related to their headache symptoms. By practicing biofeedback, a patient may be able to stop a migraine attack before it occurs or prevent headache symptoms from becoming worse.

Follow-up care

It is important for the child to keep a regular follow-up appointment schedule so the doctor can monitor the effects of treatment and make any necessary medication adjustments.

KEY TERMS

Abortive—Referring to treatment that relieves symptoms of a disorder. Abortive headache medications are used to stop the headache process and prevent symptoms of migraines, including pain, nausea, sound and light sensitivity, and other symptoms.

Acupuncture—Based on the same traditional Chinese medical foundation as acupressure, acupuncture uses sterile needles inserted at specific points to treat certain conditions or relieve pain.

Acute—Refers to a disease or symptom that has a sudden onset and lasts a relatively short period of time.

Analgesics—A class of pain-relieving medicines, including aspirin and Tylenol.

Aneurysm—A weakened area in the wall of a blood vessel which causes an outpouching or bulge. Aneurysms may be fatal if these weak areas burst, resulting in uncontrollable bleeding.

Anti-inflammatory—A class of drugs, including nonsteroidal anti-inflammatory drugs (NSAIDs) and corticosteroids, used to relieve swelling, pain, and other symptoms of inflammation.

Antidepressant drug—A medication prescribed to relieve major depression. Classes of antidepressants include selective serotonin reuptake inhibitors (fluoxetine/Prozac, sertraline/Zoloft), tricyclics (amitriptyline/Elavil), MAOIs (phenelzine/Nardil), and heterocyclics (bupropion/Wellbutrin, trazodone/Desyrel).

Antiemetic drug—A medication that helps control nausea; also called an antinausea drug.

Antihistamine—A drug used to treat allergic conditions that blocks the effects of histamine, a substance in the body that causes itching, vascular changes, and mucus secretion when released by cells.

Aura—A subjective sensation or motor phenomenon that precedes and indicates the onset of a neurological episode, such as a migraine or an epileptic seizure. This term also is used to refer to the emanation of light from living things (plants and animals) that can be recorded by Kirlian photography.

Biofeedback—A training technique that enables an individual to gain some element of control over involuntary or automatic body functions.

Chiropractic—A method of treatment based on the interactions of the spine and the nervous system. Chiropractors adjust or manipulate segments of the patient's spinal column in order to relieve pain.

Chronic—Refers to a disease or condition that progresses slowly but persists or recurs over time.

Cyclic vomiting—Uncontrolled vomiting that occurs repeatedly over a certain period of time.

Decongestants—A group of medications, such as pseudoephedrine, phenylephrine, and phenylpropanolamine, that shrink blood vessels and consequently mucus membranes.

Episodic—Occurring once in a while, without a regular pattern.

Homeopathy—A holistic system of treatment developed in the eighteenth century. It is based on the idea that substances that produce symptoms of sickness in healthy people will have a curative effect when given in very dilute quantities to sick people who exhibit those same symptoms. Homeopathic remedies are believed to stimulate the body's own healing processes.

Hydrotherapy—The use of water (hot, cold, steam, or ice) to relieve discomfort and promote physical well-being. Also called water therapy.

Inflammation—Pain, redness, swelling, and heat that develop in response to tissue irritation or injury. It usually is caused by the immune system's response to the body's contact with a foreign substance, such as an allergen or pathogen.

Lumbar puncture—A procedure in which the doctor inserts a small needle into the spinal cavity in the lower back to withdraw spinal fluid for testing. Also known as a spinal tap.

Magnetic resonance imaging (MRI)—An imaging technique that uses a large circular magnet and radio waves to generate signals from atoms in the body. These signals are used to construct detailed images of internal body structures and organs, including the brain.

Meningitis—An infection or inflammation of the membranes that cover the brain and spinal cord. It is usually caused by bacteria or a virus.

Nervous system—The system that transmits information, in the form of electrochemical impulses, throughout the body for the purpose of activation, coordination, and control of bodily functions. It is comprised of the brain, spinal cord, and nerves.

KEY TERMS (contd.)

Neurologist—A doctor who specializes in disorders of the nervous system, including the brain, spinal cord, and nerves.

Neurology—The study of nerves.

Nitrate—A food additive, commonly found in processed meats, that may be a headache trigger for some people.

Prophylactic—Preventing the spread or occurrence of disease or infection.

Stroke—Interruption of blood flow to a part of the brain with consequent brain damage. A stroke may be caused by a blood clot or by hemorrhage due to a burst blood vessel. Also known as a cerebrovascular accident.

Trigger—Any situation (people, places, times, events, etc.) that causes one to experience a negative emotional reaction, which is often accompanied by a display of symptoms or problematic behavior.

Prognosis

Most headaches are benign (not the result of a severe disease). Headaches are typically resolved through the use of **analgesics** and other treatments. As a child grows, the headaches may disappear.

Prevention

Some headaches may be prevented if the child avoids triggering substances and situations, or practices alternative therapies, such as yoga or biofeedback. Regular exercise and good sleep habits also can help prevent headaches.

Nutritional concerns

Since **food allergies** are often linked with headaches, especially cluster headaches and migraines, identifying and eliminating the allergy-causing food(s) from the diet can be an important preventive measure. To help control migraines, the child should eat three balanced meals at regular intervals, take a multi-vitamin supplement to maintain adequate nutrient needs, and drink four to eight glasses of non-caffeinated fluids per day. Sports drinks during exercise and during a headache can help balance sugar and sodium levels. To prevent headache symptoms associated with certain foods, parents should work with a registered dietitian to facilitate specific dietary changes. They also should carefully read food labels to identify and avoid dietary triggers.

Parental concerns

It is important for parents to reassure their child that most headaches are not caused by a serious illness. Parents can help their child create and maintain a headache diary to record headache symptoms, triggers, as well as the duration and frequency of the headaches. Parents should make sure their child drinks enough fluids, eats three well-balanced meals each day, gets plenty of sleep, and balances activities to avoid an over-crowded schedule that may cause stress and lead to a headache. When headaches occur, parents should allow the child to take a nap; a dark, quiet room is usually preferred by the child. In addition, parents can help the child learn relaxation techniques to help relieve or prevent headache symptoms. If the headaches are linked to **anxiety** or depression, the parents should ask the child's doctor for a referral to a counselor who can provide additional assistance.

Resources

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ORGANIZATIONS

American Council for Headache Education (ACHE). 19 Mantua Road, Mt. Royal, NJ 08061. (856) 423-0258. Web site: <www.achenet.org>.

American Headache Society. 19 Mantua Rd., Mt Royal, NJ 08061.(856) 423-0043. Web site: <www.ahsnet.org>.

MAGNUM (Migraine Awareness Group: A National Understanding for Migraineurs). 113 South St. Asaph St., Suite 300, Alexandria, VA 22314. (703) 739-9384. Web site: <www.migraines.org>.

National Headache Foundation. 820 N. Orleans, Suite 217, Chicago, IL 60610. (888) NHF-5552. Web site: <www.headaches.org>.

National Institutes of Health (NIH). National Institute of Neurological Disorders and Stroke. NIH Neurological Institute. P.O. Box 5801, Bethesda, MD 20824. (800) 352-9424. Web site: <www.ninds.nih.gov>.

WEB SITES

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Headache Impact Test. A tool to measure the impact headaches are having on patients' lives, to track headaches over time, and to share this information with the physician. Available online at: <www.headachetest.com>.

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Hearing impairment

Definition

Hearing impairment is the temporary or permanent loss of some or all hearing in one or both ears.

Description

There are three types of hearing impairment that occur in young children:

- conductive hearing loss, a usually temporary interference with the reception of sound from the outer ear to the middle or inner ear
- sensorineural hearing impairment, a permanent abnormality of the cochlear hair cells of the inner ear, the auditory nerve, or the auditory center of the brain
- mixed hearing impairment, a combination of conductive and sensorineural impairments

Hearing impairments also are classified as prelingual (occurring before a child learns to speak) and postlingual (occurring after the child has acquired language).

Normal hearing in children is defined as the ability to hear sounds in the range of 0–25 decibels (dB). Hearing impairments are classified in the following degrees:

- Mild, in which a child hears sounds from 26–40 dB. Speech and conversation are usually unaffected but distant sounds may be difficult to hear.
- Moderate, in which a child hears sounds from 41–70 dB. The ability to form sounds and hear normal conversation is affected.
- Severe, in which a child hears sounds from 71–90 dB. The child requires a hearing aid to hear conversations.
- Profound, in which a child can only hear sounds above 90 dB. A hearing aid may help but the child will not be able to articulate words normally.

Demographics

Temporary and permanent hearing impairments are not uncommon among children.

Conductive hearing impairment is most often caused by **otitis media**, an infection of the middle ear. This is very common in children between the ages of six months and four years. About 20 percent of children have an episode of acute otitis media every year. It affects boys and girls equally. Otitis media is more common among children of Eskimo or Native American descent and among children whose parents smoke. The condition is less common in children over the age of eight. Chronic secretory otitis media, also called otitis media with effusion or suppurative otitis media, is the most common cause of temporary hearing impairment in children under eight. It is more common in boys and rare in children over age eight.

About 12,000 American infants annually are born with some degree of hearing impairment. Although congenital (present at birth) deafness is the rarest form of deafness, it is the most common congenital abnormality in newborns. Three out of every 1,000 children are born with significant hearing impairment. About 65 percent of these children are born deaf and an additional 12 percent become deaf before the age of three. In the United States 14.9 percent of children aged six to 19 have measurable hearing impairment in one or both ears.

Noise-induced hearing impairment is increasing in the United States. It is not uncommon for teenagers to become permanently hearing impaired in the high-frequency range above 4,000 hertz.

Causes and symptoms

Conductive hearing impairment

Children develop otitis media because the eustachian tubes that connect the middle ear with the back of the mouth and equalize air pressure and drain fluid are small and easily obstructed. Acute otitis media can result

from a respiratory infection such as a cold that causes an inflammation that blocks a eustachian tube. The fluid that builds up in the middle ear is susceptible to bacterial and viral infection. If the blockage persists it causes chronic secretory otitis media, the most common cause of conductive hearing impairment in children.

A painful earache and temporary hearing impairment in one ear are common symptoms of acute otitis media. The symptoms of secretory otitis media develop gradually and fluctuate. They are usually worse in the winter. Symptoms of partial hearing loss from secretory otitis media may go unnoticed for some time and may include the following symptoms:

- immature speech
- behavioral problems resulting from frustration at not being able to hear well
- sitting close to the television or turning up the volume
- poor school performance

Otitis media sometimes runs in families, indicating that there may be a hereditary component. Second-hand smoke also is a risk factor for otitis media. Conductive hearing impairment from middle ear infections may be associated with other medical conditions including the following problems:

- asthma or allergic rhinitis
- **cleft palate**, which impairs drainage of the middle ears through the eustachian tubes (Some 30% of children with cleft palate have conductive hearing loss.)
- other head or facial abnormalities
- **Down syndrome**, which is characterized by narrow ear canals resulting in susceptibility to middle ear infections (About 80% of children with Down syndrome have some hearing impairment.)

Another cause of conductive hearing impairment is an excessive build-up of earwax that prevents sound waves from reaching the eardrum. Although earwax, produced by glands in the outer ear canal, normally works its way out of the ear, sometimes excessive amounts build-up and harden in the outer ear canal, gradually impairing hearing.

Sensorineural hearing impairment

Sensorineural hearing impairments result from abnormal development or disorders of the cochlea, the spiral cavity of the inner ear, disorders of the auditory nerve that transmits electrical impulses from the inner ear to the brain, or abnormalities of the auditory center of the brain. Such conditions have a variety of causes. For example, more than 70 known inherited disorders account for about one-half of all severe sensorineural hearing impairments; however, 90 percent of children with congenital hearing impairment are born to parents with normal hearing. In addition, the following problems are associated with sensorineural hearing impairment:

- craniofacial anomalies
- Down syndrome, in many of which cases the child has some immune deficiency that leads to frequent ear infections resulting in hearing loss
- problems during or shortly after birth that may damage the inner ear or auditory nerve
- low birth weigh, below 3.5 lb (1.6 kg)
- incubator noise affecting premature infants
- neonatal exposure to aminoglycoside **antibiotics**
- bacterial infections such as **meningitis** during infancy
- cytomegalovirus (CMV) infection during childhood
- accidents involving head injuries

High-frequency hearing impairment in teenagers most often results from exposure to loud noise such as amplified music.

While about 50 percent of congenital hearing impairments have no known cause, prenatal risk factors for congenital hearing impairment include:

- **rubella** (German **measles**) (More than 50% of children born to mothers who contracted rubella during the first ten weeks of pregnancy suffer from congenital malformations.)
- CMV, the most common viral infection in fetuses, a leading cause of congenital deafness (CMV affects 1% or 40,000 newborns annually; about 8,000 of these newborns have birth defects.)
- other infections, including **toxoplasmosis**, herpes, syphilis, or flu
- drug or alcohol consumption
- drugs that are ototoxins

Symptoms of congenital deafness in newborns include:

- lack of response to loud noises
- lack of response to voices or noise when sleeping in a quiet room
- failure to calm down at the sound of the mother's voice
- failure to make normal baby sounds including cooing by six weeks of age
- failure to look for the source of a noise by three to six months of age

- failure to **play** with noisy **toys**, such as a rattle, by four to eight months
- failure to babble by about six months of age

Symptoms that a baby or young child may have a hearing impairment include:

- lack of reaction to loud noises
- failure to imitate sounds
- lack of response to the child's name during the first year of life
- failure to vocalize (to imitate simple words, enjoy games that involve speech, or talk in two-word sentences during the second year)
- failure to understand simple directions during the third year

When to call the doctor

A physician should be consulted immediately if a parent suspects that a child has a hearing impairment.

Diagnosis

Parents are usually the first to suspect a hearing impairment in their child. Early detection of and intervention for hearing impairments are crucial for preventing or minimizing developmental and educational delays. Hearing-impaired children who are identified and receive early intervention before six months of age develop significantly better language skills than children identified after six months of age. However, in the United States, the average age of diagnosis is at two years of age, and significant hearing impairments have gone undiagnosed in children as old as six.

Newborn hearing tests often are administered only if an infant is considered at risk for congenital deafness. However, routine screening of sleeping newborns is on the increase. If a problem is detected, additional tests are used to determine the type and severity of the impairment. Tests used are as follows:

- An evoked otoacoustic emissions (OAE) test that detects an echo emitted by the inner ear in response to sound; the echo is produced only if the inner ear is healthy and functioning normally.
- An automated auditory brainstem response (ABR) test, or brainstem auditory-evoked response (BAER) test, in which brainstem responses to sounds are monitored through small electrodes taped to the child's head.

Pediatricians may examine a child's ears with a viewing instrument called an otoscope. Age-appropriate hearing tests may be performed routinely throughout childhood. Test administrators who suspect a hearing impairment may cover their mouths to prevent the child from lip reading, also called speech reading. Types of hearing tests include:

- behavioral tests that measure the quietest sound that the child can hear and the ability to understand words
- speech discrimination tests for children with simple vocabularies
- the McCormick toy discrimination test for three-year-olds, in which the child is asked to identify words that sound similar, such as tree and key
- a simple form of **audiometry** that assesses frequency perception through earphones
- tympanometry, in which a probe inserted into the ear measures sound waves bouncing off the eardrum
- acoustical impedance tests to identify middle ear problems including otitis media

Treatment

Conductive hearing impairment

Acute otitis media may be treated with antibiotics. Secretory otitis media usually disappears without treatment. However, a procedure called **myringotomy** or tympanostomy may be used for recurrent acute otitis media or secretory otitis media that persists for several months. A small plastic tube is inserted through the eardrum to drain fluid and equalize the air pressure between the middle ear and the ear canal. The tube usually falls out within six to 12 months and the hole in the eardrum closes. Myringotomy is an outpatient procedure performed under general anesthesia.

Excessive earwax usually can be removed at home, following a doctor's instructions. Special drops are used to soften the wax, and the ears are flushed with water. If necessary a doctor may remove earwax using suction or a metal probe.

Sensorineural hearing impairment

Sensorineural hearing impairment and congenital deafness are incurable. However, any residual hearing can be maximized with a hearing aid. Many types of hearing aids are available for children as young as three months. A postauricular hearing aid fits behind the ear and is connected to a plastic mold that is custom-fitted for the child's ear. These must be replaced as the child grows.

An older child with sufficient residual hearing can use an in-the-ear or in-the-canal hearing aid, in which the entire apparatus fits inside the ear. Hearing aids may be programmed to match a child's particular type of hearing

loss. A transposer can change high-pitched sounds that are inaudible to many hearing-impaired children into lower-pitched sounds.

Cochlear implants may be used in children who are profoundly deaf and thus are not candidates for hearing aids. Electrodes are surgically implanted into the cochlea through a hole drilled in the mastoid bone. Cochlear implants rely on three external components: a microphone to pick up sound, a speech processor to select and arrange the sounds, and a transmitter and receiver/stimulator that converts the signals from the processor into electrical impulses. The electrodes in the cochlea collect the impulses from the stimulator and send them to the brain. Although they do not restore normal hearing, cochlear implants can provide substantial improvement in speech recognition and production, as well as the ability to hear and identify common sounds such as doorbells. Most children receive implants between the ages of two and six. As of 2002 about 10,000 American children had cochlear implants. Children with cochlear implants have been found to be at an increased risk for bacterial meningitis.

Various educational approaches are employed for children with hearing impairments:

- lip reading and sign language, particularly for children with severe hearing impairment
- a bilingual-bicultural (bi-bi) approach that considers the deaf community as a separate culture with its own language (American Sign Language [ASL])
- the auditory-oral approach, which relies on powerful hearing aids or cochlear implants, supplemented with lip reading, and uses spoken rather than sign language
- the auditory-verbal (A-V) approach, which relies on enhanced residual hearing and one-on-one teaching to develop auditory skills without lip reading or sign language
- cued speech, a simple visual phonetic-based system of eight handshapes, each representing several consonant sounds, and four positions around the mouth, each representing several vowel sounds
- the total communication approach, which uses multiple methods of communication, including hearing amplification, gestures, lip reading, finger spelling, and one of several English-based sign languages known collectively as Manually Coded English (MCE)

Prognosis

Symptoms of acute otitis media usually disappear within a few days, although a ruptured eardrum may take several weeks to heal. Sometimes hearing is affected for three months or more until all of the fluid has drained from the ear. Following a myringotomy hearing in the affected ear usually returns to normal, often within a few days. As a child grows the eustachian tubes widen and stiffen, allowing air to enter and fluid to drain from the middle ear more efficiently. However, recurrent or chronic otitis media can result in ongoing moderate hearing impairment, often at a stage in which hearing is essential for **language development**.

Children who receive early intervention for hearing impairments can develop at nearly the same rate as other children. However, even a minor hearing impairment can significantly affect a baby's ability to understand and communicate and to acquire speech and language. The effects of hearing impairment on learning depend on the following:

- the severity of the impairment
- the affected frequency range
- the age at which the impairment occurred
- how early the impairment was detected
- how early treatment was initiated

Prevention

Couples with **family** histories of congenital deafness may seek genetic counseling to assess the risks for their children. If they have not already had rubella, women should be vaccinated before becoming pregnant. During pregnancy women should take only drugs that are known to be safe for the fetus.

It is very important for the hearing-impaired to protect residual hearing from loud noise. Teenagers should be encouraged to avoid very loud music. Those at risk for hearing impairment from other loud noises should be encouraged to wear earplugs.

Parental concerns

Hearing is very important for the development of emotional relationships between a child and the family. Families of hearing-impaired children must find additional means of connecting emotionally. Support groups often are very helpful for hearing-impaired children and their families.

Because hearing impairments may delay speech and language acquisition, interfere with **cognitive development**, and disrupt progress in school, the educational decisions that parents make for their child are of special significance. About 50 percent of all children with congenital deafness attend regular schools; the other 50 percent receive some type of specialized schooling.

KEY TERMS

Audiometry—The measurement of hearing ability, usually with the an audiometer.

Auditory brainstem response (ABR)—Brainstem auditory evoked response (BAER), brainstem evoked response (BSER), auditory evoked response (AER); a hearing test that records electrical activity in the brain in response to sound via electrodes on the scalp; used for newborns, infants, and young children.

Cochlea—The hearing part of the inner ear. This snail-shaped structure contains fluid and thousands of microscopic hair cells tuned to various frequencies, in addition to the organ of Corti (the receptor for hearing).

Cochlear implantation—A surgical procedure in which a small electronic device is placed under the skin behind the ear and is attached to a wire that stimulates the inner ear, allowing people who have hearing loss to hear useful sounds.

Conductive hearing impairment—Hearing impairment associated with the outer or middle ear, often caused by infection.

Cytomegalovirus (CMV)—A common human virus causing mild or no symptoms in healthy people, but permanent damage or death to an infected fetus, a transplant patient, or a person with HIV.

Decibel—A unit of the intensity of sound or a measure of loudness. Normal speech is typically spoken in the range of about 20-50 decibels.

Eustachian tube—A thin tube between the middle ear and the pharnyx. Its purpose is to equalize pressure on either side of the ear drum.

Myringotomy—A surgical procedure in which an incision is made in the ear drum to allow fluid or pus to escape from the middle ear.

Otitis media—Inflammation or infection of the middle ear space behind the eardrum. It commonly occurs in early childhood and is characterized by ear pain, fever, and hearing problems.

Otoacoustic emission (OAE)—Sounds or echoes created by vibrations of hair cells in the cochlea in response to sound; used to screen for hearing impairment in newborns.

Otoscope—A hand-held instrument with a tiny light and a funnel-shaped attachment called an ear speculum, which is used to examine the ear canal and eardrum.

Sensorineural hearing loss—Hearing loss caused by damage to the nerves or parts of the inner ear governing the sense of hearing. Sound is conducted normally through the external and middle ear.

Tympanometry—A test where air pressure in the ear canal is varied to test the condition and movement of the ear drum. This test is useful in detecting disorders of the middle ear.

See also Cochlear implants.

Resources

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ORGANIZATIONS

Alexander Graham Bell Association for the Deaf and Hard of Hearing (AG Bell). 3417 Volta Place, NW, Washington, DC 20007. Web site: <www.agbell.org>.

American Speech-Language-Hearing Association. 10801 Rockville Pike, Rockville, MD 20852. Web site: <http://asha.org>.

Deafness Research Foundation. 1050 17th St., NW, Suite 701, Washington, DC 20036. Web site: <www.drf.org>.

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Margaret Alic, PhD

Hearing test with an audiometer *see* **Audiometry**

Heart disease, congenital *see* **Congenital heart disease**

Heart murmurs

Definition

A heart murmur is an abnormal swishing or whooshing sound made by blood moving through the heart, heart valves, or blood vessels near the heart during the heartbeat cycle. It is heard through a stethoscope by a physician.

Description

When the heart beats, normally it makes two sounds, "lubb" when the valves between the atria and ventricles close, and "dupp" or "dub" when the valves between the ventricles and the major arteries close. A heart murmur is a series of vibratory sounds made by turbulent blood flow through the heart. The sounds are longer than normal heart sounds and can be heard between the normal sounds of the heartbeat.

Heart murmurs can be present at birth or develop later in life. Murmurs are common in infants and children. Nearly two-thirds of heart murmurs in children are produced by a normal, healthy heart and are harmless. This condition is called an innocent heart murmur. It also may be called functional, physiologic, or benign. Innocent heart murmurs are usually very faint, intermittent, and occur in a small area of the chest. They can disappear and reappear from one examination to the next. Most innocent murmurs disappear by adulthood, but some adults may still have them.

Less commonly, heart murmurs can result from a valve defect, narrowed blood vessel, or other cardiovascular defect. These conditions may have been present since birth (congenital) or developed as the result of another medical illness. These conditions, called pathologic heart murmurs, may indicate the presence of a serious heart defect, especially when accompanied by other signs and symptoms of a heart problem such as shortness of breath, rapid heartbeats, or fainting. They are louder, continual, and may be accompanied by a click or gallop sound. **Failure to thrive** is an accompanying symptom.

Some heart murmurs are continually present; others happen only when the heart is working harder than usual, for example during **exercise** or with certain illnesses. Heart murmurs can be diastolic, systolic, or continuous. Diastolic murmurs occur during relaxation of the heart between beats, and systolic murmurs occur during contraction of the heart muscle. Continuous murmurs occur during both the relaxation and contraction of the heart. The characteristics of the murmur may suggest specific alterations in the heart or its valves.

Demographics

Heart murmurs are most commonly discovered in children from ages two to four, although they can be diagnosed at any age. Congenital cardiovascular defects that may cause pathologic heart murmurs affect 36,000 infants (about nine of every 1,000 infants or 1 percent of live births) annually in the United States.

Causes and symptoms

Causes

Many children have heart murmurs that are heard by their doctors at some time in their lives. Innocent heart murmurs are caused by blood flowing faster than normal through the chambers and valves of the heart or the blood vessels near the heart. An increased amount of blood flowing through the heart can also cause an innocent heart murmur. Innocent murmurs may be heard in children because their hearts are very close to their chest walls. Sometimes **anxiety**, stress, **fever**, anemia, and overactive thyroid cause innocent murmurs.

Pathologic heart murmurs are caused by structural abnormalities of the heart. These include defective heart valves, hypertrophic cardiomyopathy (enlarged heart muscle), holes or abnormal openings in the walls of the heart (septal defects), aortic aneurysm, or other **congenital heart disease**.

Heart valve disease is the most common cause of pathologic heart murmurs. Valves that are narrow, tight, or stiff (valvular stenosis) do not open completely and limit the forward flow of blood through the valve. Valves that do not close properly may cause blood to leak back through the valve (called valve regurgitation). Bacterial endocarditis (an infection of the heart) or **rheumatic fever** can damage heart valves or other structures of the heart and lead to heart murmurs.

A septal defect or aortic aneurysm can cause heart murmurs. The most common types of septal defects are **atrial septal defect**, an opening between the two upper heart chambers (atria), and ventricular septal defect, an opening between the two lower heart chambers (ventricles). Some septal defects close on their own; others require surgical treatment to prevent progressive damage to the heart. An aortic aneurysm is an abnormal bulging of part of the aorta that may cause blood to leak through the aortic valve and flow the wrong direction.

Symptoms

The symptoms of heart murmurs differ, depending on the cause of the heart murmur. Innocent heart murmurs and those that do not impair the function of the heart usually do not have symptoms. Murmurs caused by severe abnormalities of a heart valve or another congenital cardiovascular defect may cause feeding problems or failure to grow normally in infants, shortness of breath, **dizziness**, fainting, chest **pain**, palpitations or rapid heartbeats, fatigue with exertion or exercise, and lung congestion.

When to call the doctor

The parent or caregiver should call the child's pediatrician if the child has these symptoms or conditions, which could be the sign of an underlying heart problem:

- feeding problems in infants
- poor weight gain
- swelling in the ankles or feet
- swollen abdomen
- poor exercise tolerance
- recurrent chest colds and respiratory infections
- abnormal blood pressure
- signs of infection including **sore throat**, general body aches or fever

The parent or caregiver should seek emergency treatment by calling 911 in most areas when the child has these symptoms or conditions:

- bluish skin tone
- bluish coloration around the lips, fingernail beds, and tongue
- breathing difficulties or rapid breathing
- dizziness or fainting
- uncontrolled coughing or coughing with blood
- irregular heart beats or palpitations (abnormal heart beats that feel like fluttering in the chest)
- chest pain (although rare in children)

Diagnosis

Heart murmurs can be heard when a physician listens to the heart through a stethoscope during a regular physical exam or check-up. While listening to the heartbeat, the physician carefully evaluates several factors, including the loudness, frequency, pitch, duration, location, and timing of the murmur with the patient's heartbeat. A systolic heart murmur may be classified according to how loud it is, based on a scale from one to six, with a grade 6 being the loudest. However, this scale is not a precise measurement, since it is based on each physician's judgment. If a suspicious heart sound is detected, the physician will evaluate how breathing, exercise, or change of body position affect the sound.

Murmurs caused by congenital cardiovascular disease are often heard at birth or during infancy.

Very loud heart murmurs and those with clicks or extra heart sounds should be evaluated further. Infants who have heart murmurs and do not thrive, eat, or breathe properly, and older children who lose consciousness suddenly or are intolerant to exercise should be evaluated. Children with these symptoms may be referred to a pediatric heart specialist, called a pediatric cardiologist. The cardiologist will perform a physical examination, review the child's personal and **family** medical history, and order tests to evaluate the source of the heart murmur.

The physical exam will be performed to identify signs of illness or physical problems. The child's blood pressure, pulse, reflexes, and height and weight are measured and recorded. Internal organs are palpated, or felt from the outside, to determine if they are enlarged.

To determine if the child has any conditions or disorders that might increase the risk of a cardiovascular defect, the physician will review the child's family medical history.

Tests may include a chest x ray, echocardiogram, or electrocardiogram. A chest x ray is used to look at the size, shape, and location of the heart and lungs. The chest x ray can indicate if the heart is enlarged and can help the doctor identify some heart and lung problems.

An echocardiogram, or echo (cardiac ultrasound) may be used to distinguish an innocent murmur from a pathologic one. On the echo, the doctor may be able to identify a structural heart or vascular problem that is causing the heart murmur. An echo uses ultrasound, or high-frequency sound waves, to create an image of the heart's internal structures. The technician applies gel to a hand-held transducer then presses it against the patient's chest. The sound waves are converted into an image that

can be displayed on a monitor. Performed in a cardiology outpatient diagnostic laboratory, the test takes 30 minutes to an hour.

An electrocardiogram (ECG) shows the heart's electrical activity and may reveal muscle thickening, damage, or a lack of oxygen. Electrodes (small, sticky patches) covered with conducting jelly are placed on the patient's chest, arms, and legs. They send impulses of the heart's activity through a monitor (oscilloscope) to a recorder that traces them on a moving strip of paper. The test takes about 10 minutes and is commonly performed in a physician's office. An exercise ECG can reveal additional information.

Cardiac **magnetic resonance imaging** (MRI) is a scanning method that uses magnetic fields and radio waves to create three-dimensional images of the heart. The MRI reveals how blood flows through the heart and how the heart is working. Although not commonly used to diagnose heart murmurs, it may be used to help physicians evaluate certain congenital cardiovascular defects.

In rare cases when the chest x ray, echo, or ECG tests are not conclusive enough to confirm the presence of an underlying congenital cardiovascular defect, a more invasive diagnostic procedure such as angiography and cardiac catheterization may be performed to show the type and severity of heart disease. These procedures should be performed by a specially trained physician and diagnostic team in a well-equipped heart center.

During the catheterization, a long, slender tube called a catheter is inserted into a vein or artery and slowly directed to the heart, using x-ray guidance. To better view the heart and blood vessels, contrast material (dye) is injected through the catheter and viewed and recorded on an x-ray video as it moves through the heart. This imaging technique is called angiography.

Treatment

Innocent heart murmurs do not affect the patient's health and require no treatment. If a septal defect is causing the heart murmurs, corrective surgery may be required. If heart valve disease is causing the heart murmurs, treatment may include medications or surgery. Valve replacement or valve repair surgery are two treatment options for severely damaged or diseased valves. The pediatric cardiologist can recommend the appropriate type and timing of treatment, based on the child's age, condition, and overall health. Patients with heart disease need prophylactic antibiotic for any dental work or medical procedures they undergo.

Alternative treatment

Alternative and complementary therapies include approaches that are considered to be outside the mainstream of traditional health care. If a heart murmur requires surgical treatment, there are no alternative treatments, although there are alternative therapies that are helpful for pre- and post-surgical support of the patient, such as guided imagery for relaxation.

If the heart murmur is innocent, heart activity can be supported using the herb hawthorn (*Crataegus laevigata* or *C. oxyacantha*) or coenzyme Q10. These remedies improve heart contractility and the heart's ability to use oxygen. If the murmur is valvular in origin, herbs that act like **antibiotics** and build resistance to infection in the valve areas may be considered.

Before using any particular technique or remedy, it is important for the parent/caregiver and child to learn about the therapy, its safety and effectiveness, potential side effects, and the expertise and qualifications of the practitioner, if applicable. Although some practices are beneficial, others may be harmful to certain patients. Alternative treatments should not be used as a substitute for medical therapies prescribed by a doctor. Parents should discuss these alternative treatments with the child's doctor to determine the techniques and remedies that may be beneficial for the child.

Nutritional concerns

Children with an underlying congenital cardiovascular defect tend to gain weight slowly. An 8-ounce to 1-pound (225–450-gram) weight gain in a month may be acceptable. The physician will monitor the child's weight gain and advise the parents of the goal weight gain and any necessary dietary changes. The most common reason for poor growth among children with congenital cardiovascular defects is they are not taking in enough calories or nutrients. Some other factors that may interfere with growth are the following:

- rapid heart beat and increased breathing rate
- poor appetite
- decreased food intake due to rapid breathing and fatigue
- frequent respiratory infections
- poor absorption of nutrients from the digestive tract
- decreased oxygen in the blood

For infants with congenital cardiovascular defects, **nutrition** supplements may need to be added to regular formula or breast milk. Sometimes additional feedings are required with the aid of a nasogastric tube to provide

KEY TERMS

Anemia—A condition in which there is an abnormally low number of red blood cells in the bloodstream. It may be due to loss of blood, an increase in red blood cell destruction, or a decrease in red blood cell production. Major symptoms are paleness, shortness of breath, unusually fast or strong heart beats, and tiredness.

Artery—A blood vessel that carries blood away from the heart to the cells, tissues, and organs of the body.

Atrial—Referring to the upper chambers of the heart.

Atrial fibrillation—A type of heart arrhythmia in which the upper chamber of the heart quivers instead of pumping in an organized way. In this condition, the upper chambers (atria) of the heart do not completely empty when the heart beats, which can allow blood clots to form.

Bacterial endocarditis—An infection caused by bacteria that enter the bloodstream and settle in the heart lining, a heart valve, or a blood vessel. People with congenital cardiovascular defects have an increased risk of developing bacterial endocarditis, so preventive antibiotics are prescribed before surgery, invasive tests or procedures, and dental work to reduce this risk.

Congenital—Present at birth.

Echocardiogram—A record of the internal structures of the heart obtained from beams of ultrasonic waves directed through the wall of the chest.

Electrocardiagram (ECG, EKG)—A record of the electrical activity of the heart, with each wave being labeled as P, Q, R, S, and T waves. It is often used in the diagnosis of cases of abnormal cardiac rhythm and myocardial damage.

Hypertension—Abnormally high arterial blood pressure, which if left untreated can lead to heart disease and stroke.

Hypothyroidism—A disorder in which the thyroid gland produces too little thyroid hormone causing a decrease in the rate of metabolism with associated effects on the reproductive system. Symptoms include fatigue, difficulty swallowing, mood swings, hoarse voice, sensitivity to cold, forgetfulness, and dry/coarse skin and hair.

Pathologic—Characterized by disease or by the structural and functional changes due to disease.

Pericardium—The thin, sac-like membrane that surrounds the heart and the roots of the great vessels. It has two layers: the inner, serous (or visceral) pericardium and the outer, fibrous (or parietal) pericardium.

Phenylketonuria—A condition caused by a genetic error of the body's metabolism, characterized by the absence of phenylalanine hydroxylase (an enzyme that converts phenylalanine into tyrosine). Phenylalanine accumulates in blood and seriously impairs early neuronal development. The defect can be effectively controlled by diet.

Rheumatic fever—An illness that arises as a complication of an untreated or inadequately treated streptococcal infection of the throat. It ususally occurs among school-aged children and cause serious damage to the heart valves.

Septal—Relating to the septum, the thin muscle wall dividing the right and left sides of the heart. Holes in the septum are called septal defects.

Septum—A wall or partition. Often refers to the muscular wall dividing the left and right heart chambers or the partition in the nose that separates the two nostrils. Also refers to an abnormal fold of tissue down that center of the uterus that can cause infertility.

Stenosis—A condition in which an opening or passageway in the body is narrowed or constricted.

Ventricles—The lower pumping chambers of the heart. The ventricles push blood to the lungs and the rest of the body.

enough calories and promote weight gain. The nasogastric tube is placed in the baby's nose and passes to the stomach, and formula or breast milk is delivered through the tube. Breastfeeding may not be possible right after delivery, depending on the child's condition, so a breast pump may be used to maintain the mother's milk supply during times when the baby cannot nurse.

Babies with congenital cardiovascular defects tire quickly during feedings, so frequent feedings are necessary. Feedings should be on-demand and may need to be as frequent as every two hours in the first few months. Some babies have difficulty feeding from a regular bottle nipple, so different brands may need to be tried. If medications are prescribed, they should be given before a

feeding. Medications should not be mixed in the formula or breast milk unless the doctor advises to do so.

The pediatrician will advise when solid foods can be started, usually around six months of age. Fat should not be restricted in the diet, especially in the first two years. High-calorie foods and snacks can play an important role in providing good nutrition and helping the child grow at a healthy rate.

Follow-up care

Along with routine medical care and standard immunizations, periodic heart check-ups are necessary in children who have congenital cardiovascular defects. Usually, heart check-up appointments are scheduled more frequently just after the diagnosis or after treatment. Additional immunizations, such as the **influenza** vaccine, may be recommended.

Prognosis

Most children with innocent heart murmurs grow out of them by the time they reach adulthood. Children with complex heart disease may continue to need special medical attention throughout **adolescence** and into adulthood for survival and to maintain quality of life.

Prevention

Heart murmurs cannot be prevented. However, if a child has been diagnosed with valve disease or another congenital cardiovascular defect, the American Heart Association recommends regular dental check-ups to prevent infections of the mouth as well as the preventive use of antibiotics to reduce the risk of heart infections (endocarditis). Preventive antibiotics should be taken before surgery, invasive tests or procedures, and dental work.

Parental concerns

It is reassuring to know that two-thirds of heart murmurs are produced by a normal heart and do not require treatment.

If an underlying congenital cardiovascular defect is diagnosed, there are many treatment options that allow children to be fully active and grow up to be healthy adults. If treatment is needed, there is help available to cover medical expenses. Parents can discuss financial aid with the child's doctor or hospital, and some organizations, including the Heart of a Child Foundation and Little Hearts on the Mend Fund, provide financial assistance to children in need of heart surgery. Support groups are available to help parents and caregivers cope with the challenges of providing care for a child with a congenital cardiovascular defect. It is important for parents to take care of themselves, too, by eating properly, exercising regularly, taking care of personal hygiene, keeping in contact with friends and family members for support, and managing stress by practicing relaxation techniques.

So that the proper treatment can be provided in the event of an emergency, children with congenital cardiovascular defects should wear a medical identification bracelet or necklace to alert healthcare providers of their condition.

See also Congenital heart disease.

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American College of Cardiology. Heart House, 9111 Old Georgetown Rd., Bethesda, MD 20814–1699. Web site: <www.acc.org>.

American Heart Association. 7320 Greenville Ave., Dallas, TX 75231–4596. <www.americanheart.org/children>.

Children's Heart Services. PO Box 8275, Bartlett, IL 60108–8275. Web site: <www.childrensheartservices.org>.

The Cleveland Clinic Heart Center. The Cleveland Clinic Foundation, 9500 Euclid Ave., F25, Cleveland, Ohio, 44195. Web site: <www.clevelandclinic.org/heartcenter>.

Congenital Heart Disease Information and Resources. 1561 Clark Dr., Yardley, PA 19067. Web site: <www.tchin.org>.

Heart Support of America. 4873 N. Broadway, Knoxville, TN 37918. Web site: <www.heartsupport.com>.

International Children's Heart Foundation. 1750 Madison, Suite 100, Memphis, TN 38104. Web site: <www.babyhearts.com>.

National Heart, Lung, and Blood Institute. PO Box 30105, Bethesda, MD 20824–0105. Web site: <www.nhlbi.nih.gov>.

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Heat disorders

Definition

Heat disorders are a group of physically related illnesses caused by prolonged exposure to hot temperatures, restricted fluid intake, or failure of temperature regulating mechanisms of the body. Disorders of heat exposure include heat cramps, heat exhaustion, and heat stroke (also called sunstroke).

Description

Hyperthermia is the general name given to heat-related illnesses. The two most common forms of hyperthermia are heat exhaustion and heat stroke, the latter of which is especially dangerous and requires immediate medical attention.

The thermal regulation centers of the brain help to maintain the body's internal temperature. Regardless of extreme weather conditions, the healthy human body keeps a steady temperature of approximately 98.6°F (37°C). In hot weather or during vigorous activity, the body perspires. As perspiration evaporates from the skin, the body is cooled. The thermal regulating centers in the brain help the body adapt to high temperatures by adjusting the amount of salts (electrolytes) in the perspiration. Electrolytes help the cells in body tissues maintain water balance. In hot weather, a healthy body will lose enough water to cool the body while creating the lowest level of electrolyte imbalance. If the body loses too much salt and fluid, symptoms of **dehydration** will occur.

Heat cramps

Heat cramps are the least severe of the heat-related illnesses. This heat disorder is often the first signal that the body is having difficulty with increased temperature. Individuals exposed to excessive heat should view heat cramps as a warning sign of a potential heat-related emergency.

Heat exhaustion

Heat exhaustion is a more serious and complex condition than heat cramps. Heat exhaustion can result from prolonged exposure to hot temperatures, restricted fluid intake, or failure of temperature regulation mechanisms of the body. Heat exhaustion requires immediate attention, as it can rapidly progress to heat stroke.

Heat stroke

Heat stroke is life threatening, and because a high percentage of individuals who experience heat stroke

die, immediate medical attention is critical when symptoms first appear. Heat stroke, like heat exhaustion, is also a result of prolonged exposure to hot temperatures, restricted fluid intake, or failure of temperature regulation mechanisms of the body. However, the severity of impact on the body is much greater with heat stroke.

Demographics

Heat disorders are harmful to people of all ages, but their severity is greatest in young children and the elderly. Young children are at risk because, in relation to their weight, they have a large surface area of skin through which to lose water. In addition, until about age two, children's kidneys are not able to concentrate urine and preserve body fluids as efficiently as adult kidneys. The elderly also often have reduced kidney function or underlying diseases, or take medications that make them more vulnerable to dehydration. In healthy adults, heat stroke and heat exhaustion often affect athletes, firefighters, construction workers, factory workers, and anyone who exercises heavily and/or wears heavy clothing in hot, humid weather. Obese individuals and those with poor circulation or who take medications to reduce excess body fluids (diuretics) can be at risk when conditions are hot and humid.

Causes and symptoms

Heat cramps

Heat cramps are painful **muscle spasms** caused by the excessive loss of electrolytes due to heavy perspiration. The correct balance of electrolytes is crucial to many body functions, including muscle contraction and nerve impulse transmission. Heavy exertion in extreme heat and/or restricted fluid intake may lead to heat cramps.

With heat cramps, muscle tissue becomes less flexible, causing **pain**, difficult movement, and involuntary tightness. Cramps occurs more often in the legs and abdomen than in other areas of the body.

Heat exhaustion

Heat exhaustion is caused by exposure to high heat and humidity for many hours, resulting in excessive loss of fluids and salts through heavy perspiration. The skin may appear cool, moist, and pale. The child may complain of **headache** and **nausea**, with a feeling of overall weakness and exhaustion. **Dizziness**, faintness, and mental confusion are often present, as is a rapid, weak pulse. Breathing becomes fast and shallow. Fluid loss reduces blood volume and lowers blood pressure. Intense thirst and a highly concentrated, reduced volume of deep yellow or orange urine are signs of inadequate fluid intake.

Heat stroke

Before heat stroke occurs, an individual experiences heat exhaustion and the associated symptoms. When the body can no longer maintain a normal temperature, heat exhaustion escalates and becomes heat stroke. Heat stroke is a life-threatening medical emergency that requires immediate life-saving measures.

Heat stroke is caused by overexposure to extreme heat, resulting in a breakdown of the body's heat regulating mechanisms. Body temperature reaches a dangerous level. An individual with heat stroke has a body temperature higher than 104°F (40°C), and possibly as high as 106°F (41.1°C).

Other symptoms of heat stroke include mental confusion with possible combativeness and bizarre behavior, staggering, and faintness. The pulse becomes strong and rapid (160–180 beats per minute). The skin takes on a dry and flushed appearance. There is often very little perspiration. The individual can quickly lose consciousness or have convulsions.

When to call the doctor

The doctor should be called when the child shows any symptoms of heat exhaustion of if he or she has been exposed to heat and dehydrating conditions and has a body temperature of over 102°F (38.9°C). Emergency medical services should be called immediately if the individual has any symptoms of heat stroke or is having difficulty breathing.

Diagnosis

Diagnosis of heat cramps usually involves observation of symptoms such as muscle cramping and thirst. Diagnosis of heat exhaustion or heat stroke, however, may require a healthcare worker to review the child's medical history, document symptoms, and obtain blood pressure and temperature readings. A physician may take blood and urine samples for further laboratory testing. A test to measure the body's electrolytes can also give valuable information about chemical imbalances caused by the heat-related illness.

Treatment

Heat cramps

The care of heat cramps includes placing the child at rest in a cool environment, while giving cool water with a teaspoon of salt per quart, or a commercial sports drink (e.g. Gatorade). Usually, rest and liquids are all that is needed for the child to recover. Mild stretching and massaging of the muscles may be helpful once the condition improves. The child should not take salt tablets, because such a high concentration of salt may actually worsen the condition. When the cramps stop, the person usually can begin light activity again if there are no other signs of illness. The child needs to continue drinking fluids and should be watched carefully for further signs of heat-related illnesses.

Heat exhaustion

The child suffering from heat exhaustion should stop all physical activity and move immediately to a cool place out of the sun, preferably a cool, air-conditioned location. He or she should lay down with feet slightly elevated, remove or loosen clothing, and drink cold (but not iced), slightly salty water or a commercial sports drink. Rest and replacement of fluids and salt is usually all the treatment that is needed, and **hospitalization** is rarely required. Following rehydration, the child usually recovers rapidly.

Heat stroke

Simply moving the individual experiencing heat stroke to a cooler place is not enough to reverse internal overheating. Emergency medical assistance should be called immediately. While waiting for help to arrive, quick action to lower body temperature must take place.

Immediate treatment involves getting the child to a cool place, loosening clothing or undressing the person, and allowing air to circulate around the body. The next step is to wrap the child in wet towels or clothing, and place ice packs in areas with the greatest blood supply. These areas include the neck, under the arm and knees, and in the groin. Once the patient is under medical care, cooling treatments continue as appropriate. The child's body temperature is monitored constantly to guard against overcooling. Breathing and heart rate are monitored, and fluids and electrolytes are replaced intravenously. Anti-convulsant drugs may be given. After severe heat stroke, hospitalization may be necessary and bed rest is recommended for several days.

Prognosis

Prompt treatment for heat cramps is usually very effective, allowing the individual to return rapidly to activity. Treatment of heat exhaustion usually brings full recovery in one to two days. Heat stroke is a very serious condition and its outcome depends upon the general health and age of the individual. Due to the high body temperature resulting from heat stroke, permanent damage to the brain, kidneys, heart, and other internal organs is possible. Heat stroke can be fatal, especially for infants and toddlers.

Prevention

Because heat cramps, heat exhaustion, and heat stroke have a cascade effect, the prevention of the onset of all heat disorders is similar.

- Avoid strenuous **exercise** when it is very hot.
- Drink plenty of fluids, especially water, and avoid drinking alcoholic beverages; drink frequently, even if not thirsty.
- Wear light and loose-fitting clothing that allows the air to circulate around the body in hot weather.
- Eat lightly salted foods which can help replace salts lost through perspiration.
- Provide proper ventilation of hot areas (fan, open window, air conditioning).
- Use sunblocks and **sunscreens** with a protection factor of SPF 15 or greater when exposed to direct sunlight.
- Never leave a child locked in a hot environment such as a car, even for a minute.
- Monitor children's activity and fluid intake frequently.
- Offer infants supplemental bottles of water in hot weather.
- Soak bandanas or other clothing in water to wear while working or playing in the heat.
- Wear a hat that allows air circulation (mesh, straw) in the sun.

Parental concerns

Parents need to be especially alert to dehydration and the development of heat disorders in infants who cannot ask for something to drink. Parents need to take the initiative in encouraging children to drink frequently in hot weather. Water or sports drinks are a better choice of liquids than soft drinks.

KEY TERMS

Convulsions—Also termed seizures; a sudden violent contraction of a group of muscles.

Electrolytes—Salts and minerals that produce electrically charged particles (ions) in body fluids. Common human electrolytes are sodium chloride, potassium, calcium, and sodium bicarbonate. Electrolytes control the fluid balance of the body and are important in muscle contraction, energy generation, and almost all major biochemical reactions in the body.

Rehydration—The restoration of water or fluid to a body that has become dehydrated.

Before the 1970s, some coaches felt it was good training to limit the amount of fluids athletes drank at practices. As a result, there were 39 documented heat-related deaths in athletes between 1964 and 1973. As the water-electrolyte balance of the body became better understood, most coaches have recognized that water should be freely available during athletic practices and events. As a result, documented heat-related deaths declined substantially in the 1980s. When children participate in athletics, parents need to be aware of the potential for heat disorders and assure that appropriate measures for prevention are taken by coaches.

See also Sunburn.

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Heavy metal poisoning

Definition

Heavy metal **poisoning** is the toxic accumulation of heavy metals in the soft tissues of the body.

Description

Heavy metals are chemical elements that have a specific gravity (a measure of density) at least five times that of water. The heavy metals most often implicated in human poisoning are lead, mercury, arsenic, and cadmium. Some heavy metals, such as zinc, copper, chromium, iron, and manganese, are required by the body in small amounts, but these same elements can be toxic in larger quantities.

Heavy metals may enter the body in food, water, or air, or by absorption through the skin. Once in the body, they compete with and displace essential **minerals** such as zinc, copper, magnesium, and calcium, and interfere with organ system function. People may come in contact with heavy metals in industrial work, pharmaceutical manufacturing, and agriculture. Children may be poisoned as a result of playing in contaminated soil.

Demographics

Heavy metal poisoning is relatively uncommon. In children, lead ingestion is the major culprit of heavy metal poisoning. In 2000, an estimated one in 22 American children had high levels of lead in their blood. Children in urban areas with old lead water pipes and lead-painted homes are especially at risk. Mercury poisoning is possible from eating contaminated fish.

Causes and symptoms

Symptoms will vary, depending on the nature and the quantity of the heavy metal ingested. Affected people may complain of **nausea**, **vomiting**, **diarrhea**, stomach **pain**, **headache**, sweating, and a metallic taste in the mouth. Depending on the metal, there may be blue-black lines in the gum tissues. In severe cases, people exhibit obvious impairment of cognitive, motor, and language skills. The expression "mad as a hatter" comes from the mercury poisoning prevalent in seventeenth-century France among hat makers who soaked animal hides in a solution of mercuric nitrate to soften the hair.

When to call the doctor

A healthcare professional should be contacted whenever exposure to any heavy metal is suspected. The Centers for Disease Control and Prevention (CDC) recommends testing all children for lead exposure at 12 months of age and, if possible, again at 24 months. Testing should start at six months for children at higher risk for **lead poisoning**.

Diagnosis

Heavy metal poisoning may be detected using blood and urine tests, hair and tissue analysis, or x ray.

In childhood, blood lead levels above 80 µg/dL generally indicate lead poisoning; however, significantly lower levels (>30 µg/dL) can cause **mental retardation** and other cognitive and behavioral problems in affected children. The Centers for Disease Control and Prevention considers a blood lead level of 10 µg/dL or higher in children a cause for concern. In adults, symptoms of lead poisoning are usually seen when blood lead levels exceed 80 µg/dL for a number of weeks.

Blood levels of mercury should not exceed 3.6 µg/dL, while urine levels should not exceed 15 µg/dL. Symptoms of mercury poisoning may be seen when mercury levels exceed 20 µg/dL in blood and 60 µg/dL in urine. Mercury levels in hair may be used to gauge the severity of chronic mercury exposure.

Since arsenic is rapidly cleared from the blood, blood arsenic levels may not be very useful in diagnosis. Arsenic in the urine (measured in a 24-hour collection following 48 hours without eating seafood) may exceed 50 µg/dL in people with arsenic poisoning. If acute arsenic poisoning is suspected, an x ray may reveal ingested arsenic in the abdomen (since arsenic is opaque to **x rays**). Arsenic may also be detected in the hair and nails for months following exposure.

Cadmium toxicity is generally indicated when urine levels exceed 10 µg/dL of creatinine and blood levels exceed 5 µg/dL.

Treatment

The treatment for most heavy metal poisoning is chelation therapy. A chelating agent specific to the metal involved is given orally, intramuscularly, or intravenously. The three most common chelating agents are calcium disodium edetate, dimercaprol (BAL), and penicillamine. The chelating agent encircles and binds to the metal in the body's tissues, forming a complex; that complex is then released from the tissue to travel in the bloodstream. The complex is filtered out of the blood by the kidneys and excreted in the urine. This process may be lengthy and painful and typically requires **hospitalization**. Chelation therapy is effective in treating lead, mercury, and arsenic poisoning, but it is not useful in treating cadmium poisoning. As of 2004, no treatment had been proven effective for cadmium poisoning.

In cases of acute mercury or arsenic ingestion, vomiting may be induced. Washing out the stomach (gastric lavage) may also be useful. The affected person may also require treatment such as intravenous fluids for complications of poisoning such as shock, anemia, and kidney failure.

Prognosis

The chelation process can only halt further effects of the poisoning; it cannot reverse neurological damage already sustained.

Prevention

Because exposure to heavy metals is often an occupational hazard, protective clothing and respirators should be provided and worn on the job. Protective clothing should then be left at the work site and not worn home, where it could carry toxic dust to **family** members. Industries are urged to reduce or replace the heavy metals in their processes wherever possible. For the sake of children's health along with everyone else's, exposure to environmental sources of lead, including lead-based paints, plumbing fixtures, vehicle exhaust, and contaminated soil, should be reduced or eliminated.

Nutritional concerns

Parents should avoid preparing or serving food in containers that have lead in their glazing.

Parental concerns

Parents living in homes built prior to 1978 should be vigilant in removing flaking or peeling paint because it might contain lead. Simply repainting such surfaces will not solve the problem. Parents must monitor the environments in which their children **play** and the objects that go into their children's mouths. Cleanliness is a must if old paint is in a child's environment. Removal (stripping paint to bare metal or bare wood) of lead is the best way to prevent lead exposure in children. Areas where removal is taking place should be sealed off from the rest of the house. In addition, children should be kept away from occupational sources of other heavy metals. Parents

KEY TERMS

Chelation—The process by which a molecule encircles and binds to a metal and removes it from tissue.

Heavy metal—One of 23 chemical elements that has a specific gravity (a measure of density) at least five times that of water.

who are concerned about their child's exposure to lead should have the child tested.

See also Lead poisoning.

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American Academy of Family Physicians. 11400 Tomahawk Creek Parkway, Leawood, KS 66211–2672. Web site: <www.aafp.org/>.

American Academy of Pediatrics. 141 Northwest Point Boulevard, Elk Grove Village, IL 60007–1098. Web site: <www.aap.org/default.htm>.

American Association of Poison Control Centers. 3201 New Mexico Avenue NW, Washington, DC 20016. Web site: <www.aapcc.org/>.

American College of Occupational and Environmental Medicine. 55 West Seegers Road, Arlington Heights, IL 60005. Web site: <www.acoem.org/>.

Multiple Sclerosis Foundation. 6350 North Andrews Ave., Fort Lauderdale, Fl 33309–2130. Web site: <www.msfacts.org/>.

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Heimlich maneuver

Definition

The Heimlich maneuver is an emergency technique for removing a foreign object lodged in the airway that is preventing a child or an adult from breathing.

Purpose

The Heimlich maneuver is used when a person is **choking** on a foreign object to the extent that he/she cannot breathe. Oxygen deprivation from a foreign body airway obstruction can result in permanent brain damage or death in four minutes or less. Using the Heimlich maneuver can save a choking victim's life. The Heimlich

maneuver is not performed on infants under one year of age (see below for technique for infants). Indications that a choking victim's airway is blocked include the following:

- inability to **cough**, cry, or speak
- blue or purple face color from lack of oxygen
- grabbing at throat
- weak cough and labored breathing that produces a high-pitched noise
- all of the above, followed by loss of consciousness

Each year, more than 17,000 infants and children are treated in hospital emergency departments for choking-related incidents, and more than 80 percent of these occur in children aged four years and younger. Airway obstruction death and injury are especially prevalent in children under age four due to their anatomy (small airway), natural curiosity and tendency to put objects in their mouths, and incomplete chewing. In infants, choking usually results from inhalation of small objects (e.g., coins, small **toys**, deflated balloons, buttons) that they place in their mouth.

Description

In 1974, Henry Heimlich first described an emergency technique for expelling foreign material blocking the trachea. This technique, now called the Heimlich maneuver, is simple enough that it can be performed immediately by anyone trained in the maneuver. The Heimlich maneuver is a standard part of all first-aid and **cardiopulmonary resuscitation** (CPR) courses.

The theory behind the Heimlich maneuver is that by compressing the abdomen below the level of the diaphragm with quick abdominal thrusts, an ''artificial cough'' is created. Air is forced out of the lungs to dislodge the obstruction in the trachea and bring the foreign object back up into the mouth.

The Heimlich maneuver can be performed on all people; however, modifications are necessary infants, children, obese individuals, and pregnant women.

Performing the Heimlich maneuver on children

To perform the Heimlich maneuver on a conscious child, the rescuer stands or kneels behind the child, who may be seated or standing. The rescuer makes a fist with one hand, and places it, thumb toward the child, below the rib cage and above the waist. The rescuer encircles the child's waist, placing his other hand on top of the fist then gives a series of five quick and distinct inward and upward thrusts. If the foreign object is not dislodged, the cycle of five thrusts is repeated until the object is expelled or the child becomes unresponsive. As the child is deprived of oxygen, the muscles of the trachea relax slightly, and it is possible that the foreign object may be expelled on a second or third attempt.

If the victim is unconscious or becomes unconscious, the rescuer should lay him or her on the floor, bend the chin forward, make sure the tongue is not blocking the airway, and feel in the mouth for the foreign object, being careful not to push any farther into the airway. The rescuer kneels astride the child's thighs and places his fists between the bottom of the victim's breastbone and the navel. The rescuer then executes a series of five quick compressions by pushing inward and upward.

After the abdominal thrusts, the rescuer repeats the process of lifting the chin, moving the tongue, feeling for and possibly removing the foreign material. If the airway is not clear, the rescuer repeats the abdominal thrusts as often as necessary. If the foreign object has been removed, but the victim is not breathing, the rescuer starts CPR.

The technique in children over one year of age is the same as in adults, except that the amount of force used is less than that used with adults in order to avoid damaging the child's ribs, breastbone, and internal organs.

OBESE CHILDREN AND ADOLESCENTS The main difference in performing the Heimlich maneuver on an obese victim is in the placement of the fists. Instead of using abdominal thrusts, chest thrusts are used. The fists are placed against the middle of the breastbone, and the motion of the chest thrust is in and downward, rather than upward. If the victim is unconscious, the chest thrusts are similar to those used in CPR.

Foreign body obstruction in infants under age one year

The Heimlich maneuver as described above is not performed on infants under one year of age. Instead, a series of back blows and chest thrusts are used. The rescuer sits down and lays the infant along his or her forearm with the infant's face pointed toward the floor and tilted downward lower than the body. The rescuer's hand supports the infant's head, and his or her forearm rests on his or her own thigh for additional support. Using the heel of the other hand, the rescuer administers five rapid blows to the infant's back between the shoulder blades.

After administering the back blows, the rescuer sandwiches the infant between his or her arms and turns the infant over so that the infant is lying face up supported by the opposite arm. Using the free hand, the rescuer places the index and middle finger on the center of

the breastbone just below the nipple line and makes gives five quick chest thrusts. This series of back blows and chest thrusts is alternated until the foreign object is expelled. If the infant becomes unconscious, CPR should be initiated.

Precautions

Any lay person can be trained to perform the Heimlich maneuver and to know how may save someone's life. Before doing the maneuver, it is important to determine if the airway is completely blocked. If the victim choking can talk or cry, the Heimlich maneuver should not be administered. If the airway is not completely blocked, the choking victim should be allowed to try to cough up the foreign object on his or her own.

Aftercare

Vomiting may occur after being treated with the Heimlich maneuver. All infants and children who experience a choking episode severe enough to require the Heimlich maneuver should be taken to the hospital emergency room to be examined for airway injuries.

Risks

Incorrectly applied, the Heimlich maneuver can break bones or damage internal organs. In infants, the rescuer should never attempt to sweep the baby's mouth without looking to remove foreign material. This is likely to push the material farther down the trachea. Following the Heimlich maneuver, dysphagia (swallowing difficulty) and obstructive pulmonary edema (fluid accumulation in the lungs) may occur.

Normal results

In many cases the foreign material is dislodged from the throat, and the choking victim suffers no permanent effects of the episode. If the foreign material is not removed, the choking victim may suffer permanent brain damage from lack of oxygen or may die.

Parental concerns

Because most choking incidents occur in the home, all parents and infant/child caregivers should be trained in the Heimlich maneuver. Training is available through the American Red Cross and American Heart Association at local schools, YMCAs, and community centers.

The likelihood of choking incidents can be reduced by closely supervising infants and children while eating and playing. Most choking incidents are associated with food items, especially hot dogs, candies, grapes, nuts, popcorn, and carrots. Common non-food items that are choking hazards include deflated balloons, buttons, coins, small balls, small toys, and toy parts. All toys should be examined to make sure they are age-appropriate and do not have loose parts.

KEY TERMS

Diaphragm—The thin layer of muscle that separates the chest cavity containing the lungs and heart from the abdominal cavity containing the intestines and digestive organs. This term is also used for a dome-shaped device used to cover the back of a woman's vagina during intercourse in order to prevent pregnancy.

Trachea—The windpipe. A tube composed of cartilage and membrane that extends from below the voice box into the chest where it splits into two branches, the bronchi, that lead to each lung.

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Hemangiomas *see* **Birthmarks**
Hemoglobin F test *see* **Fetal hemoglobin test**

Hemophilia

Definition

Hemophilia is a coagulation disorder arising from a genetic defect of the X chromosome; the defect can either be inherited or result from spontaneous gene mutation. In each type of hemophilia (hemophilias A, B, and C), a critical coagulation protein is missing, causing individuals to bleed for long periods of time before clotting occurs. Depending on the degree of the disorder in the affected individual, uncontrolled bleeding may occur spontaneously with no known initiating event, or occur after specific events such as surgery, dental procedures, immunizations, or injury.

Description

The body's normal mechanism for blood clotting is a complex series of events (coagulation cascade) involving interaction between the injured blood vessel, blood cells called platelets, 13 specific coagulation factors (designated by Roman numerals I through XIII), and other substances that circulate in the blood.

When blood vessels are injured in a way that causes bleeding, platelets collect over the injured area, forming a temporary plug to prevent further bleeding. This temporary plug, however, is too disorganized to serve as a long-term solution, so a series of chemical events occurs that results in the formation of a more reliable plug. The final plug or clot involves tightly woven fibers of a material called fibrin. The production of fibrin requires the interaction of a series of proteins, clotting factors I through XIII, in a process called amplification to rapidly produce the proper-sized fibrin clot from the small number of molecules initially activated by the injury. In the complex coagulation process, the absence or inactivity of just one clotting factor can greatly increase bleeding time. In hemophilia, certain clotting factors are either decreased in quantity, absent altogether, or improperly formed, preventing the formation of a clot and resulting in uncontrolled bleeding.

Hemophilia A is the most common type of coagulation disorder and involves decreased activity of factor VIII. There are three levels of factor VIII deficiency: severe, moderate, and mild. This classification is based on the percentage of normal factor VIII activity present:

- Individuals with less than 1 percent of normal factor VIII activity level have severe hemophilia. Half of all people with hemophilia A fall into this category. Such individuals frequently experience spontaneous musculoskeletal bleeding into their joints, skin, and muscles. Surgery or trauma can result in life-threatening hemorrhage and must be carefully managed.
- Individuals with 1–5 percent of normal factor VIII activity level have moderate hemophilia and are at risk for heavy bleeding after seemingly minor traumatic injuries.
- Individuals with 5–40 percent of normal factor VIII activity level have mild hemophilia and must prepare carefully for any surgery or dental procedures.

In hemophilia B, or Christmas disease, the deficient clotting factor is factor IX, but the symptoms are very similar to those of hemophilia A. Factor IX is produced in the liver and is dependent on interaction with vitamin K in order to function properly. A deficiency in vitamin K can affect the clotting factor's performance as well as a deficiency in the factor itself.

Hemophilia C is rare and much milder than hemophilia A or B. It involves reduced activity of factor XI and is characterized by mild bleeding such as nosebleeds (epistaxis) or prolonged menstrual bleeding, or mild bleeding after tonsillectomies or dental extractions.

Demographics

Hemophilia A affects between one in 5,000 to one in 10,000 males in most populations. Hemophilia B occurs in one in 40,000 to 50,000. The prevalence of hemophilia is estimated to be 13.4 cases per 100,000 U.S. males (10.5 hemophilia A and 2.9 hemophilia B). By race/ethnicity, the prevalence is 13.2 cases in 100,000 among white males, 11.0 among African-American males, and 11.5 among Hispanic males. Hemophilia C occurs primarily among individuals of Jewish descent.

Causes and symptoms

Hemophilia A and B are both caused by a genetic defect present on the X chromosome. (Hemophilia C is inherited in a different fashion.) About 70 percent of all people with hemophilia A or B inherited the disease. The

other 30 percent develop from a spontaneous genetic mutation.

Both factors VIII and IX are produced by a genetic defect of the X chromosome, so hemophilia A and B are both sex-linked diseases passed on from a female to male offspring. (All humans have two chromosomes determining their gender: females have XX, males have XY. Because the trait is carried only on the X chromosome, it is called sex-linked.) Because a female child always receives two X chromosomes, she will nearly always receive at least one normal X chromosome. Therefore, even if she receives one flawed X chromosome, she will still be capable of producing a sufficient quantity of factors VIII and IX to avoid the symptoms of hemophilia. Such a person who has one flawed chromosome but does not actually suffer from the disease is called a carrier. She carries the flaw that causes hemophilia and can pass it on to her offspring. If, however, she has a son who receives her flawed X chromosome, he will be unable to produce the right quantity of factors VIII or IX, and he will suffer some degree of hemophilia. (Males inherit one X and one Y chromosome and, therefore, have only one X chromosome.)

In rare cases, a hemophiliac father and a carrier mother can pass on the right combination of parental chromosomes to result in a hemophiliac female child. However, the vast majority of people with either hemophilia A or B are male.

About 30 percent of all people with hemophilia A or B are the first member of their **family** to ever have the disease. These individuals have had the unfortunate occurrence of a spontaneous mutation, meaning that in their early development, some random genetic accident affected their X chromosome, resulting in the defect that causes hemophilia A or B. Once such a spontaneous genetic mutation takes place, offspring of the affected person can inherit the newly created, flawed chromosome.

In the case of severe hemophilia, the first bleeding event usually occurs prior to 18 months of age. In some babies, hemophilia is suspected immediately when a routine **circumcision** (removal of the foreskin of the penis) results in unusually heavy bleeding. Toddlers are at particular risk because they fall frequently and may bleed into the soft tissue of their arms and legs. These small bleeds result in bruising and noticeable lumps but do not usually require treatment. As a child becomes more active, bleeding may occur into the muscles, a much more painful and debilitating situation. These muscle bleeds result in **pain** and pressure on the nerves in the area of the bleed. Damage to nerves can cause **numbness** and decreased ability to use the injured limb.

Christmas disease varies from mild to severe, but mild cases are more common. The severity depends on the degree of deficiency of factor IX. Hemophilia B symptoms are similar to those of hemophilia A, including numerous large and deep **bruises** and prolonged bleeding.

Some of the most problematic and frequent bleeds occur into the joints, particularly the knees and elbows. Repeated bleeding into joints can result in scarring within the joints and permanent deformities. Individuals may develop arthritis in joints that have suffered continued irritation from the presence of blood. Mouth injuries can result in compression of the airway, which interrupts breathing and can be life-threatening. A blow to the head, which might be totally insignificant in a normal child, can result in bleeding into the skull and brain. Because the skull has no room for expansion, the hemophiliac is at risk for brain damage due to blood taking up space and exerting pressure on the delicate brain tissue.

People with hemophilia are at very high risk of severe, heavy, uncontrollable bleeding (hemorrhage) from injuries such as motor vehicle accidents and also from surgery.

Some other rare clotting disorders such as von Willebrand's disease present similar symptoms but are not usually called hemophilia.

When to call the doctor

Hemophilia is usually discovered when an injury initiates bleeding and the bleeding will not stop. In very young children, spontaneous musculoskeletal bleeding may occur around the time the child begins to walk; these episodes may be the first sign of hemophilia. In some children, a simple surgical procedure, such as a tooth extraction or injection, may present with uncontrolled bleeding. Any signs of deep bruises or the presence of prolonged bleeding after a bump or an injury that breaks the skin should be reported to a physician or emergency service immediately. Bleeding under the skin (hematoma), which looks like a severe bruise, should also be reported and medical care sought immediately.

Diagnosis

Various diagnostic tests are available to measure, under carefully controlled conditions, the length of time it takes to produce certain components of the final fibrin clot. The activated partial thromboplastin time (APTT) is performed and will typically be prolonged while a prothrombin time (PT) will likely be normal. Factor assays, measurement methods performed by the clinical laboratory, can determine the percentage of factors VIII and IX present compared to normal percentages. This

information helps to confirm a diagnosis of hemophilia and identifies the type and severity of hemophilia present.

Families with a history of hemophilia can also have tests done during a pregnancy to determine whether the fetus will have hemophilia. Chorionic villous sampling is a test that examines proteins for deficiencies or defects that are characteristic of hemophilia. The test can be performed at 10 to 14 weeks; test performance is associated with a 1 percent risk of miscarriage. **Amniocentesis** is a method of withdrawing amniotic fluid from the placenta to allow examination of fetal cell DNA shed into the amniotic fluid, helping to identify genetic mutations. Amniocentesis can be performed at 15 to 18 weeks gestation and is associated with a one in 200 risk of miscarriage.

Treatment

The treatment of hemophilia involves replacing or supplementing the deficient coagulation factors. Various preparations of factors VIII and IX are available to replace missing factors as needed. Cryoprecipitate, for example, is a single- or multiple-donor human plasma preparation rich in coagulation factors; it is made available as a frozen concentrate. Fresh frozen plasma is a single-donor preparation of factor-rich plasma; it is used primarily for replacing factor XI in individuals with hemophilia C. Concentrated factor preparations may be obtained from a single donor, by pooling the donations of as many as thousands of donors, or by laboratory creation through highly advanced genetic techniques. These preparations are administered directly into the individual's veins (intravenous administration).

The frequency of treatment with coagulation factors depends on the severity of the individual's disease. Relatively mild disease will only require treatment in the event of injury, or to prepare for scheduled surgical or dental procedures. More severe disease will require regular treatment to avoid spontaneous bleeding.

Appropriate treatment of hemophilia can decrease suffering and be lifesaving in the presence of hemorrhage. Complications associated with treatment, however, can also be quite serious. About 20 percent of all individuals with hemophilia A begin to produce antibodies in their blood against the specific factor protein; the presence of antibodies may then rapidly destroy infused factor VIII. The presence of such antibodies may greatly hamper efforts to prevent or stop a major hemorrhage.

Individuals who receive coagulation factors prepared from pooled donor blood were once at risk for serious infections that could be passed through the infusion of human blood products, such as the hepatitis virus and HIV. Concern has also been raised about the possibility

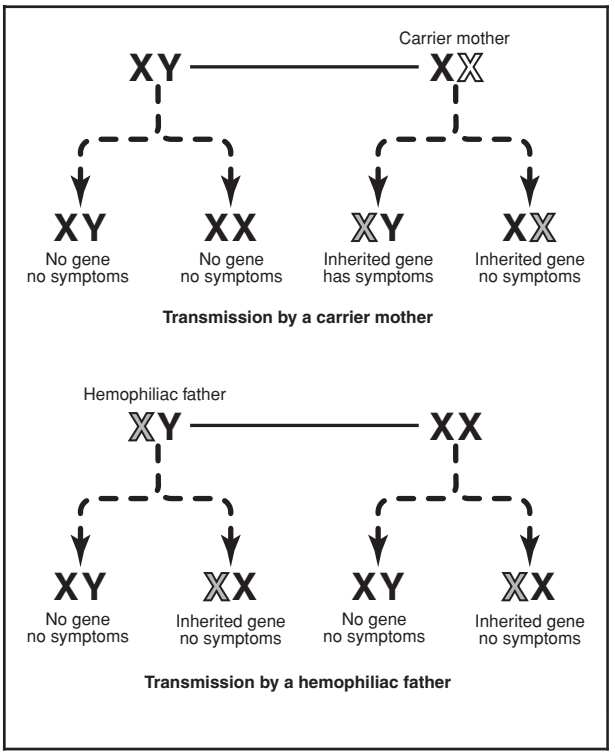

Chart showing how hemophilia is transmitted on the X chromosome. However, some 30 percent of hemophilia cases are caused by random genetic mutation. *(Illustration by Electronic Illustrators Group.)*

of hemophiliacs contracting a fatal slow virus infection of the brain (Creutzfeldt-Jakob disease) from blood products. However, more sensitive testing techniques have been developed and as of 2004 were employed by the companies producing pooled precipitates from human plasma. These improved methods of donor testing, as well as methods of inactivating viruses present in donated blood, have greatly lowered the risk of infection.

Molecular biological techniques have introduced gene therapies as new treatment possibilities for hemophilia. Gene therapy involves sophisticated methods of transferring new genes to hemophiliacs, correcting deficiencies or defects in the clotting mechanism. These methods are being researched in the early 2000s.

Prognosis

Variations in the type and severity of hemophilia makes it difficult to generalize a prognosis, however, for individuals with mild hemophilia, the prognosis is quite good. Those with more severe hemophilia can also live relatively normal lives with careful management and avoidance of injury. Many individuals achieve normal life expectancy. Without treatment of bleeding episodes,

KEY TERMS

Amplification—A process by which something is made larger. In clotting, only a very few chemicals are released by the initial injury; they trigger a cascade of chemical reactions which produces increasingly larger quantities of different chemicals, resulting in an appropriately-sized, strong fibrin clot.

Coagulation factors—Specific coagulation proteins in the blood required for clotting. Coagulation proteins are designated with roman numerals I through XIII.

Fibrin—The last step in the blood coagulation process. Fibrin forms strands that add bulk to a forming blood clot to hold it in place and help "plug" an injured blood vessel wall.

Hemorrhage—Severe, massive bleeding that is difficult to control. The bleeding may be internal or external.

Mutation—A permanent change in the genetic material that may alter a trait or characteristic of an individual, or manifest as disease. This change can be transmitted to offspring.

Platelet—A cell-like particle in the blood that plays an important role in blood clotting. Platelets are activated when an injury causes a blood vessel to break. They change shape from round to spiny, "sticking" to the broken vessel wall and to each other to begin the clotting process. In addition to physically plugging breaks in blood vessel walls, platelets also release chemicals that promote clotting.

Trauma—Serious physical injury. Also refers to a disastrous or life-threatening event that can cause severe emotional distress, including dissociative symptoms and disorders.

severe muscle and joint pain and eventually permanent damage can occur. Much depends upon the physical activity level of the individual and the possibility of accidental injuries or surgeries required for other conditions, which cannot be predicted.

Prevention

Because of its genetic origins, hemophilia cannot be prevented in those born with the inherited defects or factor deficiencies. However, individuals who have a family history of hemophilia may benefit from genetic testing and counseling before deciding to have a baby.

The most important way for individuals with hemophilia to prevent complications of the disease is to avoid activities that may lead to injury. Those individuals who require dental work or any type of surgery may need to be pre-treated with an infusion of factor VIII to avoid hemorrhage. Hemophiliacs should also avoid medications or drugs that promote bleeding; aspirin is one such medication and many prescription drugs have anticoagulant properties.

Parental concerns

When a child has an inherited coagulation disorder such as hemophilia, parents will be concerned about the possibility of trauma or injury that may lead to potentially dangerous bleeding episodes. The watchfulness of parents along with effective management of hemophilia by physicians can help the child to lead a relatively normal life. Careful avoidance of injury is essential. Counseling is available to help children handle the psychosocial aspects of living with hemophilia. Education is available from public health organizations to help parents be informed about their child's condition.

See also Coagulation disorders.

Resources

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Hemophilus infections

Definition

Hemophilus infections, most of which are due to *Haemophilus influenzae* infections, are a group of contagious diseases that are caused by a bacterium and affect only humans. Some hemophilus infections are potentially fatal.

Description

H. influenzae is a common organism worldwide; it has been found in the nasal secretions of as many as 90 percent of healthy individuals in the general population. Hemophilus infections are characterized by acute inflammation with a discharge (exudate). They may affect almost any organ system but are most common in the respiratory tract. The organism can be transmitted by person-to-person contact or by contact with nasal discharges and other body fluids. Hemophilus infections in the United States are most likely to spread in the late winter or early spring.

Demographics

The primary factor influencing the rate of infection is age; children between the ages of six months and four years are most vulnerable to *H. influenzae*. In the twentieth century, about 50 percent of children would acquire a hemophilus infection before reaching one year of age; almost all children would develop one before age three. In the United States, these figures have declined, however, as a result of the increasing use of hemophilus vaccines for children. Worldwide, however, *Haemophilus influenzae* remains a significant childhood pathogen. It is the primary cause of childhood **meningitis** and the second most common cause of childhood **pneumonia**. In developing countries, *Haemophilus influenzae* is responsible for 500,000 annual deaths in children under the age of five.

Causes and symptoms

Hemophilus infections are primarily caused by *Haemophilus influenzae*, a bacterium that is capable of spreading from the nasal tissues and upper airway, where it is usually found, to the chest, throat, or middle ear. The organism sometimes invades localized areas of tissue, producing meningitis, infectious arthritis, **conjunctivitis**, cellulitis, **epiglottitis**, or inflammation of the membrane surrounding the heart. The most serious infections are caused by a strain called *H. influenzae* b (Hib). Before routine **vaccination**, Hib was the most common cause of bacterial meningitis and responsible for most of the cases of acquired **mental retardation** in the United States.

Bacterial sepsis in the newborn

Bacterial sepsis (the presence of illness-causing microorganisms, or their poisons, in the blood) is a potentially fatal illness in newborn infants. The child may acquire the disease organism as it passes through the mother's birth canal or from the hospital environment. *H. influenzae* can also produce inflammations of the eye (conjunctivitis) in newborn children. The signs of sepsis may include **fever**, fussiness, feeding problems, breathing difficulties, pale or mottled skin, or drowsiness. Premature birth is the most significant risk factor for hemophilus infections in newborns.

Epiglottitis

Epiglottitis is a potentially fatal hemophilus infection. Although children are more likely to develop epiglottitis, it can occur in adults as well. When the epiglottis (the flap that covers the trachea during swallowing so that food odes not enter the lungs) is infected, it can swell to the point where it blocks the windpipe. The symptoms of epiglottitis include a sudden high fever, drooling, the feeling of an object stuck in the throat, and **stridor** (a high-pitched, noisy respiratory sound). The epiglottis will look swollen and bright red if the doctor examines the patient's throat with a laryngoscope (a viewing device).

Meningitis

Meningitis caused by Hib is most common in children between nine months and four years of age. The child usually develops upper respiratory symptoms followed by fever, loss of appetite, **vomiting**, **headache**, and a stiff or sore neck or back. In severe cases, the child may have convulsions or go into shock or coma.

Other infections

Hib is the second most common cause of middle ear infection and **sinusitis** in children. The symptoms of sinusitis include fever, **pain**, bad breath, and coughing. Children may also develop infectious arthritis from Hib. The joints most frequently affected are the large weight-bearing ones.

Diagnosis

The diagnosis is usually based on a combination of the patient's symptoms and the results of blood counts, cultures, or antigen detection tests.

Laboratory tests

Laboratory tests can be used to confirm the diagnosis of hemophilus infections. The bacterium can be grown on chocolate agar or identified by blood cultures or Gram stain of body fluids. Antigen detection tests can be used to identify hemophilus infections in children. These tests include latex agglutination and electrophoresis.

Other laboratory findings that are associated with hemophilus infections include anemia (low red blood cell count) and a drop in the number of white blood cells in children with severe infections. Adults often show an abnormally high level of white blood cells; cell counts of 15,000 to 30,000/mm^3 are not unusual.

Treatment

Because some hemophilus infections are potentially fatal, treatment is started without waiting for the results of laboratory tests.

Medications

Hemophilus infections are treated with **antibiotics**. Patients who are severely ill are given ampicillin or a third-generation cephalosporin, such as cefotaxime or ceftriaxone, intravenously. Patients with milder infections are given oral antibiotics, including amoxicillin, cefaclor, erythromycin, or trimethoprim-sulfamethoxazole. Patients who are allergic to penicillin are usually given cefaclor or trimethoprim-sulfamethoxazole.

Patients with Hib strains that are resistant to ampicillin may be given chloramphenicol. Chloramphenicol is not a first-choice drug because of its side effects, including interference with bone marrow production of blood cells.

The duration of antibiotic treatment depends on the location and severity of the hemophilus infection. Adults with respiratory tract infections, or Hib pneumonia, are usually given a 10 to 14 day course of antibiotics. Meningitis is usually treated for 10 to 14 days, but a seven-day course of treatment with ceftriaxone appears to be sufficient for infants and children. Ear infections are treated for seven to 10 days.

Supportive care

Patients with serious hemophilus infections require bed rest and a humidified environment (such as a **croup** tent) if the respiratory tract is affected. Patients with epiglottitis frequently require intubation (insertion of a breathing tube) or a tracheotomy to keep the airway open. Patients with inflammation of the heart membrane, pneumonia, or arthritis may need surgical treatment to drain infected fluid from the chest cavity or inflamed joints.

Supportive care also includes monitoring of blood cell counts for patients using chloramphenicol, ampicillin, or other drugs that may affect production of blood cells by the bone marrow.

Prognosis

The most important factors in the prognosis are the severity of the infection and promptness of treatment. Untreated hemophilus infections—particularly meningitis, sepsis, and epiglottitis—have a high mortality rate. Bacterial sepsis of the newborn has a mortality rate of 13–50 percent. The prognosis is usually good for patients with mild infections who are treated without delay. Children who develop Hib arthritis sometimes have lasting problems with joint function.

Prevention

Hemophilus vaccines

There are three different vaccines for hemophilus infections used to immunize children in the United States: PRP-D, HBOC, and PRP-OMP. PRP-D is used only in children older than 15 months. HBOC is administered to infants at two, four, and six months after birth, with a booster dose at 15 to 18 months. PRP-OMP is administered to infants at two and four months, with the third dose at the child's first birthday. All three vaccines are given by intramuscular injection. About 5 percent of children may develop fever or soreness in the area of the injection.

Other measures

Other preventive measures include isolating patients with respiratory hemophilus infections; treating appropriate contacts of infected patients with rifampin; maintaining careful standards of cleanliness in hospitals,

KEY TERMS

Bacteria—Singular, bacterium; tiny, one-celled forms of life that cause many diseases and infections.

Epiglottitis—Inflammation of the epiglottis, most often caused by a bacterial infection. The epiglottis is a piece of cartilage behind the tongue that closes the opening to the windpipe when a person swallows. An inflamed epiglottis can swell and close off the windpipe, thus causing the patient to suffocate. Also called supraglottitis.

Exudate—Cells, protein, fluid, or other materials that pass through cell or blood vessel walls. Exudates may accumulate in the surrounding tissue or may be discharged outside the body.

Intubation—A procedure in which a tube is inserted through the mouth and into the trachea to keep the airway open and to help a patient breathe.

Nosocomial infection—An infection acquired in a hospital setting.

Sepsis—A severe systemic infection in which bacteria have entered the bloodstream or body tissues.

Stridor—A term used to describe noisy breathing in general and to refer specifically to a high-pitched crowing sound associated with croup, respiratory infection, and airway obstruction.

Tracheotomy—An surgical procedure in which the surgeon cuts directly through the patient's neck into the windpipe below a blockage in order to keep the airway open.

including proper disposal of soiled tissues; and washing hands properly.

See also Hib vaccine.

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Henoch-Schönlein purpura *see* **Allergic purpura**

Hepatitis A

Definition

Hepatitis A is a liver disease caused by the hepatitis A virus (HAV).

Description

Hepatitis A is a form of viral hepatitis also known as infectious hepatitis, due to its ability to be spread through personal contact. Hepatitis A is a milder liver disease than **hepatitis B**, and asymptomatic infections are very common, especially in children. Hepatitis A does not cause a carrier state or chronic liver disease. Once the infection ends, there is no lasting phase of illness. However, it is not uncommon to have a second episode of symptoms about a month after the first; this is called a relapse.

Transmission

HAV is found in the stool (feces) of persons infected with hepatitis A. HAV is usually spread from person to person by putting something in the mouth that has been contaminated with the stool of a person infected with hepatitis A. This is called fecal-oral transmission. Thus, the virus spreads more easily in areas where there are poor sanitary conditions or where good personal hygiene is not observed. Most infections result from contact with a household member who has hepatitis A. Blood-borne infection has been documented but is rare in the United States. The common modes of transmission of hepatitis A are as follows:

- consuming food made by someone who touched infected feces
- drinking water that is contaminated by infected feces (a problem in communities with poor sewage treatment facilities)
- touching an infected person's feces, which may occur with poor hand washing
- having direct contact in large daycare centers, especially where there are children in diapers
- being a resident of states in which hepatitis A is more common
- sexual contact with an infected person

Demographics

Hepatitis A has a worldwide distribution and is endemic in most countries. However, the incidence of the disease is declining in developed countries. There is a very high incidence in developing countries and rural areas. For example, in rural areas of South Africa, the rate of infection is 100 percent.

According to the Centers for Disease Control, HAV infects up to 200,000 Americans each year with the highest rate of hepatitis A being among children five to 14 years of age. Almost 30 percent of reported cases of hepatitis A occur among children under 15 years of age, chiefly because they are frequently in close contact with other children in school and at daycare. Approximately 15 percent of reported cases of hepatitis A occur among children or employees in daycare centers. The states with the highest incidence of hepatitis A account for 50 percent of the reported cases. According to the American Academy of Pediatrics, 11 states have a rate of HAV infection that is at least twice the national average, or 20 cases per every 100,000 people. The states are: Arizona, Alaska, California, Idaho, Nevada, New Mexico, Oklahoma, Oregon, South Dakota, Utah, and Washington.

Causes and symptoms

Hepatitis A is caused by HAV, also called *Enterovirus 72*, which was identified in 1973. The virus has an incubation period of three to five weeks. It enters the body via the gut and replicates in the digestive tract and spreads to infect the liver, where it multiplies. HAV is excreted in the stools for two weeks preceding the onset of symptoms.

Persons with hepatitis A may not have signs or symptoms of the disease and older persons are more likely to have symptoms than children. If present, symptoms are non-specific and usually include **fever**, tiredness, loss of appetite, **nausea**, abdominal discomfort, dark urine, and **jaundice** (yellowing of the skin and eyes). Symptoms usually last less than two months, but some persons can be ill for as long as six months.

When to call the doctor

Parents should call the doctor immediately if any of the following occurs:

- A child has changes in symptoms, is confused, is difficult to wake up, is lethargic (sluggish), or irritable.
- A child is unable to drink fluids.
- A child's skikn is yellow in color.
- A child has signs of **dehydration** such as no urine in over eight hours or a dry mouth.
- A child starts to look very sick.

Diagnosis

Hepatitis A symptoms often go unrecognized because they are not specific to hepatitis A, thus a blood test (IgM anti-HAV) is required to diagnose HAV infection. This test detects a specific antibody, called hepatitis A IgM, that develops when HAV is present in the body.

Treatment

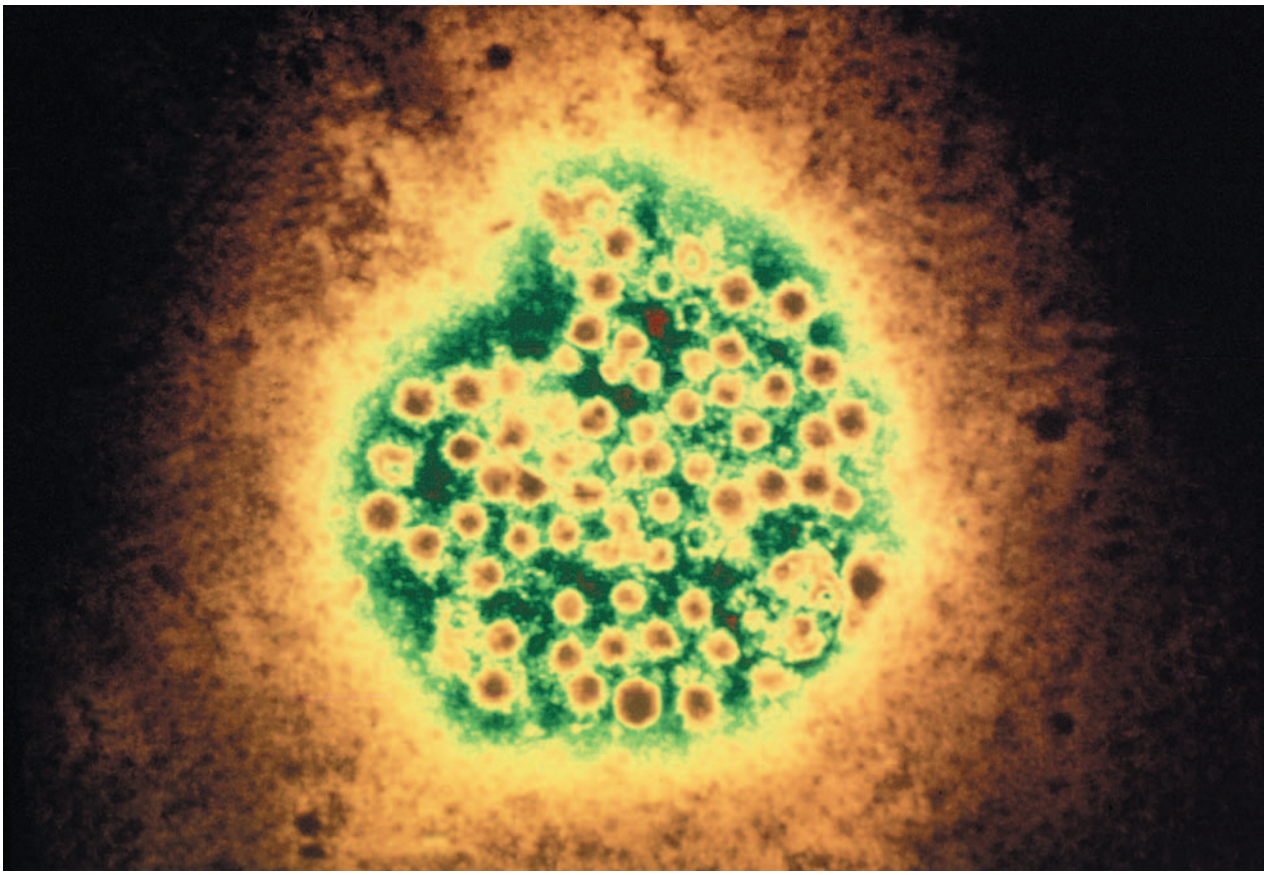
No specific treatment is available for hepatitis A. However, the following guidelines are often recommended:

- Fluids and diet. The best treatment is to make sure that the child drinks a lot of fluids and eats well.
- Rest. The child should rest while he or she has fever or jaundice. When fever and jaundice are gone, activity may be gradually increased as with the healthcare provider's approval.
- Medications. The body's immune system fights the HAV infection. Once the child recovers from hepatitis A, the virus leaves the body. Medications, prescription or nonprescription, should not be given without consulting the doctor.

Nutritional concerns

Parents should ensure that their infected child has a well-balanced diet. Children with advanced liver disease need to follow specific diets issued by the treating physician. However, most children are not in this category, and no special diet is currently recommended for them, except that they should avoid eating fatty foods because the body has difficulty digesting fat when the liver is not working well.

However, adequate protein intake is important to regenerate liver cells. Children without liver cirrhosis require about 2–3 grams of protein per kilogram of body weight. Children with cirrhosis need an individual **nutrition** plan from their pediatric specialist or nutritionist.



Hepatitis A virus magnified 225,000 times. *(© 1990 Custom Medical Stock Photo, Inc.)*

Prognosis

Viral hepatitis symptoms usually last three weeks to two months but may last up to six months. Children may return to daycare one week after symptoms first appear, with the doctor's permission. Most children with hepatitis get better naturally without liver problems later in life. However, some children do have subsequent liver problems. For this reason, it is important to keep in close touch with the treating physician and to keep all follow-up appointments. Chronic, or relapsing, infection does not occur with hepatitis A. In the United States, serious complications are infrequent, and deaths are very rare.

Prevention

According to the Centers for Disease Control and Prevention (CDC), routine **vaccination** of children is the most effective way to lower the incidence of hepatitis A nationwide. The CDC encourages implementation of routine hepatitis A vaccination programs for children in the 17 states which have the highest rates of hepatitis A. Hepatitis A vaccine has been licensed in the United States for use in persons two years of age and older. The vaccine is recommended (before exposure to hepatitis A virus) for persons who are more likely to get hepatitis A virus infection or are more likely to get seriously ill if they do get hepatitis A. The vaccines licensed in the United States as of 2004 were HAVRIX(r) (manufactured by Glaxo SmithKline) and VAQTA(r) (manufactured by Merck & Co., Inc).

Parents should teach their children always to wash their hands with soap and water after using the bathroom and before preparing and eating food. Travelers should avoid water and ice if unsure of their purity, or they can boil water for one minute before drinking it.

Short-term protection against hepatitis A is available from immune globulin, a preparation of antibodies that can be given before exposure for short-term protection against hepatitis A and for persons who have already been exposed to HAV. It can be given before and within two weeks after suspected contact with the virus.

KEY TERMS

Antibody—A special protein made by the body's immune system as a defense against foreign material (bacteria, viruses, etc.) that enters the body. It is uniquely designed to attack and neutralize the specific antigen that triggered the immune response.

Antigen—A substance (usually a protein) identified as foreign by the body's immune system, triggering the release of antibodies as part of the body's immune response.

Asymptomatic—Persons who carry a disease and are usually capable of transmitting the disease but who do not exhibit symptoms of the disease are said to be asymptomatic.

Endemic—Natural to or characteristic of a particular place, population, or climate.

Hepatitis A—Commonly called infectious hepatitis, caused by the hepatitis A virus (HAV). Most often spread by food and water contamination.

Immune globulin—Preparation of antibodies that can be given before exposure for short-term protection against hepatitis A and for persons who have already been exposed to hepatitis A virus. Immune globulin must be given within two weeks after exposure to hepatitis A virus for maximum protection.

Jaundice—A condition in which the skin and whites of the eyes take on a yellowish color due to an increase of bilirubin (a compound produced by the liver) in the blood. Also called icterus.

Vaccine—A substance prepared from a weakened or killed microorganism which, when injected, helps the body to form antibodies that will prevent infection by the natural microorganism.

Parental concerns

The best way to prevent exposure to HAV is good habits in washing hands. Children should wash their hands every time they go to the bathroom. Good hand washing should be enforced at home and at daycare facilities. It is also very important to keep a clean environment, such as clean toilets, bathrooms, and clothing. If a child is diagnosed with HAV, other **family** members should be treated to prevent spread of the disease. The healthcare provider can help parents to plan treatment for the entire family.

See also Hepatitis B; Vaccination.

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Monique Laberge, Ph.D.

Hepatitis B

Definition

Hepatitis B is a liver disease caused by the hepatitis B virus (HBV).

Description

Hepatitis B is a form of viral hepatitis that is also known as serum hepatitis, due to its ability to be spread through body fluids and blood. HBV can cause lifelong infection, cirrhosis (scarring) of the liver, liver **cancer**, liver failure, and death. Hepatitis B is a more severe liver disease than **hepatitis A**, and asymptomatic infections occur frequently. Chronic hepatitis B infection may take one of two forms: chronic persistent hepatitis, a condition characterized by persistence of the virus but in which liver damage is minimal; and chronic active hepatitis, in which there is aggressive destruction of liver tissue and rapid progression to cirrhosis or liver failure.

Transmission

Transmission of HBV occurs through blood and body fluid exposure such as blood, semen, vaginal secretions, or saliva. Hepatitis B is not spread through food or water or by casual contact. Infants may also develop the disease if they are born to a mother who has the virus. Infected children often spread the virus to other children if there is frequent contact or a child has many scrapes or cuts. The common modes of transmission of hepatitis B are as follows:

- children born to mothers who have hepatitis B (the illness may present up to five years after the child is born)
- children who are born to mothers who have emigrated from a country where hepatitis B is widespread such as southeast Asia and China
- individuals who live in households where another member is infected with the virus
- infection through intravenous (IV) drug use and/or unprotected heterosexual or homosexual sexual contact
- infection through blood transfusions from infected donors

Demographics

Worldwide there are 450 million carriers of hepatitis B, 50 million of which are in Africa. Carriage rates vary markedly in different areas. In South Africa, infection is much more common in rural communities than in the cities.

According to the Centers for Disease Control (CDC), an estimated 78,000 persons in the United States were infected with HBV in 2001. People of all ages get hepatitis B, and about 5,000 die per year of sickness caused by HBV. An estimated 1.25 million Americans are chronically infected, of whom 20 to 30 percent acquired their infection in childhood. It is estimated that hepatitis B accounts for 20 to 25 percent of all acute viral hepatitis in children. Infected newborns rarely suffer but have 90 percent chance of becoming carriers. Twenty-five percent of all HAV positive newborns develop chronic liver disease by the third to fourth decade of life.

Causes and symptoms

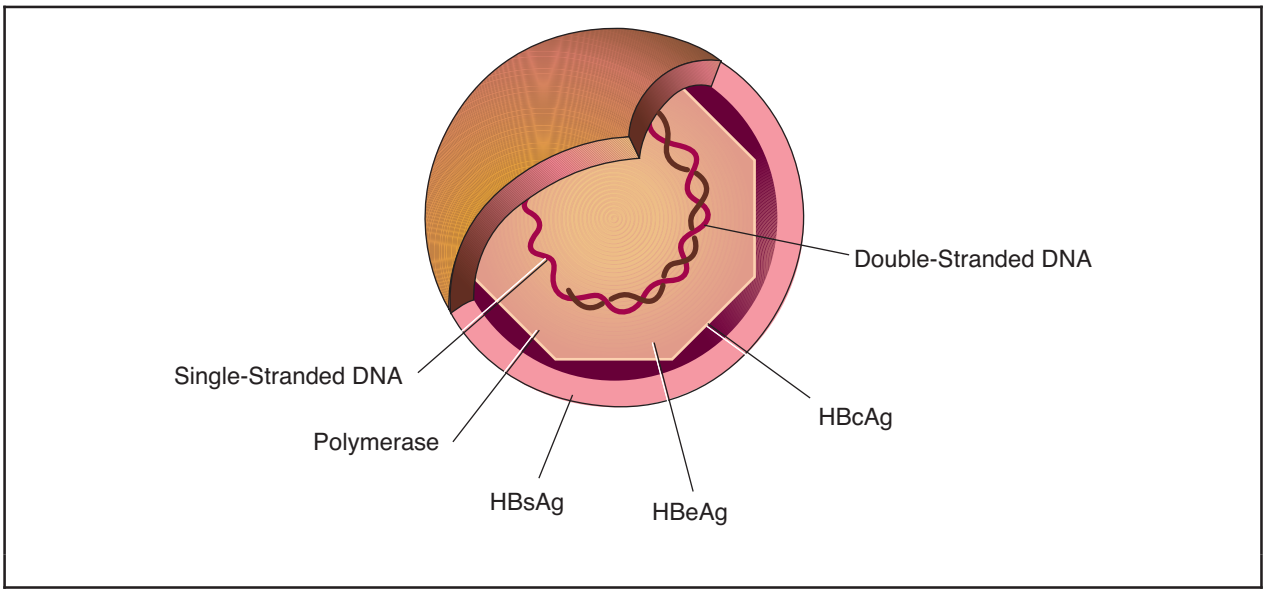
Hepatitis B is caused by HBV, also called Hepadna virus. The virus has an incubation period of two to five months. It replicates in the liver, and virus particles are shed in large amounts into the blood. The blood of infected individuals is thus highly infectious.

Hepatitis B has a wide range of symptoms. It can also be mild, without symptoms. When present, the symptoms are non-specific and usually include **fever**, tiredness, loss of appetite, **nausea**, abdominal discomfort, dark urine, clay-colored bowel movements, and **jaundice** (yellowing of the skin and eyes).

When to call the doctor

Parents should call the doctor immediately if any of the following occurs:

- A child has changes in symptoms, is confused, is difficult to wake up, is lethargic (sluggish), or irritable.
- A child is unable to drink fluids.
- A child's skin becomes much more yellow in color.
- A child has signs of **dehydration** such as no urine in over eight hours or a dry mouth.
- A child starts to look very sick.



Hepatitis B virus (HBV) is composed of an inner protein core and an outer protein capsule. The outer capsule contains the hepatitis B surface antigen (HBsAg). The inner core contains HBV core antigen (HBcAg) and hepatitis B e-antigen (HBeAg). This cell also contains polymerase, which catalyzes the formation of the cell's DNA. HBV is the only hepatitis-causing virus that has DNA, instead of RNA. *(Illustration by Electronic Illustrators Group.)*

Diagnosis

A blood test is required to diagnose hepatitis B. The test detects one of the viral antigens called hepatitis B surface antigen (HBsAg) in the blood. Later on, HBsAg may no longer be present, in which case a test for antibodies to a different antigen, called hepatitis B core antigen, is used. If HBsAg can be detected in the blood for longer than six months, chronic hepatitis B is diagnosed.

Treatment

There is no cure for hepatitis B and no specific treatment is available. However, the following guidelines are often recommended:

- Fluids and diet. The best treatment is to ensure that the child drinks a lot of fluids and eats well.
- Rest. The child should rest while he or she has fever or jaundice. When fever and jaundice are gone, activity may be gradually increased as with the healthcare provider's approval.
- Medications. There is no medicine that gets rid of HBV or heals the liver. There are medications available to treat chronic HBV-infection. These work for some people, but experience with children is limited. Three drugs are licensed, as of 2004, for the treatment of chronic hepatitis B: Adefovir dipivoxil, alpha interferon, and lamivudine.

Nutritional concerns

Parents should ensure that their infected child has a well-balanced diet. Children with advanced liver disease need to follow specific diets issued by the treating physician. However, most children are not in this category, and no special diet is recommended for them, except that they should avoid eating fatty foods because the body has difficulty digesting fat when the liver is not working well.

However, adequate protein intake is important to regenerate liver cells. Children without liver cirrhosis require about 1–2 grams of protein per pound (2–3 grams per kilogram) of body weight. Children with cirrhosis need an individual **nutrition** plan from their pediatric specialist or nutritionist.

There is some evidence that iron can lower the response to interferon treatment in adults. Although no results have been reported for children, the issue of restricting iron intake should be discussed with the treating physician.

Prognosis

Viral hepatitis symptoms usually last three weeks to two months but may last up to six months. Children may return to daycare one week after symptoms first appear, with the doctor's permission. Most children with hepatitis get better naturally without liver problems later on in life. However, some children do have subsequent liver

KEY TERMS

Cirrhosis—A chronic degenerative disease of the liver, in which normal cells are replaced by fibrous tissue and normal liver function is disrupted. The most common symptoms are mild jaundice, fluid collection in the tissues, mental confusion, and vomiting of blood. Cirrhosis is associated with portal hypertension and is a major risk factor for the later development of liver cancer. If left untreated, cirrhosis leads to liver failure.

Hepatitis B virus (HBV)—Also called Hepadna virus, the pathogen responsible for hepatitis B infection.

Jaundice—A condition in which the skin and whites of the eyes take on a yellowish color due to an increase of bilirubin (a compound produced by the liver) in the blood. Also called icterus.

Vaccine—A substance prepared from a weakened or killed microorganism which, when injected, helps the body to form antibodies that will prevent infection by the natural microorganism.

problems. Thus, it is important to keep in close touch with the treating physician and to keep all follow-up appointments. Chronic, or relapsing, infection occurs with hepatitis B in about 5–10 percent of cases.

Prevention

A vaccine for hepatitis B is as of 2004 widely used in the United States for routine childhood immunization. Children usually receive the first vaccine between birth and two months of age, the second vaccine at one to four months, and the third vaccine at six to 18 months. The vaccine is generally required for all children born on or after January 1, 1992, before they enter school. The vaccine is available for older children who may have not been immunized before 1992 and is recommended before age 11 or 12.

Parental concerns

If mothers have HBV in their blood, they can give hepatitis B to their baby during **childbirth**. Babies who get HBV at birth may have the virus for the rest of their lives, can spread the disease, and can get cirrhosis of the liver or liver cancer. The CDC recommends that all pregnant women be tested for HBV early in their pregnancy. If the blood test is positive, the baby should receive vaccine along with hepatitis B immune globulin (HBIG) at birth. The second dose of vaccine should be given at one to two months of age and the third dose at six months of age.

See also Hepatitis A; Hepatitis B vaccine; Vaccination.

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Monique Laberge, Ph.D.

Hepatitis B vaccine

Definition

The **hepatitis B** vaccine (HBV or HepB) is an injection that protects children from contracting hepatitis B, a serious disease caused by the hepatitis B virus.

Description

The hepatitis B vaccine consists of a small protein from the surface of the hepatitis B virus called the hepatitis B surface antigen (HBsAg). After **vaccination** with HBV, the child's immune system recognizes HBsAg as foreign and produces antibodies that attach to the protein (anti-HBs). These specific antibodies remain in the blood. Later, if the child becomes infected with the hepatitis B virus, the antibodies recognize the protein and stimulate the immune system to produce large quantities of specific antibodies that attach to and destroy the virus and prevent the disease.

HBV is usually the first vaccine a child receives, most often before leaving the hospital after birth. The second and third HBV immunizations are administered by the age of 18 months, in conjunction with other routine childhood vaccinations.

Vaccine formulations

The HBsAg in HBVs is referred to as recombinant because it is genetically engineered. The gene encoding the DNA for HBsAg is introduced into common baker's yeast. The yeast is grown in vats in which large amounts of HBsAg are produced. The yeast cells are broken, and the HBsAg is isolated and purified. It is adsorbed into aluminum hydroxide.

Packaged hepatitis B vaccine contains the following:

- up to 95 percent HBsAg, with 10 to 40 micrograms of HBsAg per milliliter of vaccine
- no more than 5 percent yeast protein
- a small amount of aluminum hydroxide (0.5 mg/ml)
- very small amounts of other additives to stabilize and preserve the vaccine

Two HBVs are approved for use in the United States. Recombivax HB, manufactured by Merck & Company, is as of 2004 available as a pediatric/adolescent formulation (orange cap) and as an adult formulation (green cap). Engerix-B, made by SmithKline Beecham Biologicals, is as of 2004 available as a pediatric formulation (blue cap) and as an adult formulation (orange cap). In general these HBVs are interchangeable and either or both can be used in an individual immunization series. An HBV derived from the blood serum of people with hepatitis B was as of 2004 no longer produced in the United States.

Dosages

The immune response to HBV varies among individual children. Therefore, the HBV dose should be determined by a medical professional. In general, the recommended doses are as follows:

- Newborns: 2.5 to 20 micrograms injected into the anterolateral thigh muscle within seven days of birth or at the first visit to the physician's office, and one month and six months after the first dose, for a total of three doses.
- Newborns: 10 to 20 micrograms injected into the thigh muscle within seven days of birth and one month, two months, and 12 months after the first dose, for a total of four doses.
- Older child or adolescent: 2.5 to 20 micrograms injected into the deltoid arm muscle, with additional doses one month and six months after the first injection, for a total of three doses.

Safety

Although the vast majority of parents believe that vaccinations are important for their children, the majority of parents are also concerned about the safety of vaccines including HBV. Although controversy over the

safety of HBV resulted in congressional hearings in 1999, the National Academy of Science's Institute of Medicine, as well as other authorities, considers HBV to be safe. Repeated studies have found no association between HBV and **sudden infant death syndrome** (SIDS) or other medical conditions, including neurological or immune system disorders.

Effectiveness

HBV usually is effective in protecting against hepatitis B. (HBV also protects against the related hepatitis D virus, which occurs as a co-infection with hepatitis B and usually results in more severe disease symptoms.) However, the immune response to HBV varies among children, apparently due to genetic variations in individual immune systems. In addition, the following medical conditions may cause children to benefit less from HBV:

- stomach **pain**
- cirrhosis (scarring) of the liver
- immune system impairment
- medical conditions requiring kidney dialysis

The duration of hepatitis B immunity following infant vaccination is not known. A 2004 study found that most low-risk children vaccinated at birth did not have antibodies against hepatitis B in their blood by the time they reached the age of five. Although the majority of these children responded positively to a booster HBV immunization, one-third of them did not respond. Likewise, a 2003 Israeli study found a steady decline in anti-hepatitis-B antibodies over time in children vaccinated as infants. The steepest decline in the antibodies occurred between five and eight years after vaccination.

General use

Hepatitis B in children

The U.S. Centers for Disease Control and Prevention (CDC) estimates that, prior to the launch of the infant HBV immunization program, about 33,000 American children of non-infected mothers acquired hepatitis B by the age of ten. Hepatitis B is a potentially serious disease caused by the hepatitis B virus. It may result in inflammation and damage to the liver. Hepatitis B infection may be without symptoms or with acute or short-lived symptoms that can include:

- jaundice (a yellowing of the skin and whites of the eyes)
- joint pain
- stomach pain
- itchy red **hives** on the skin

The hepatitis B virus is eventually cleared from the bodies of most infected adolescents and adults. Only about 2–6 percent of infected older children and adults develop chronic hepatitis B and can continue to transmit the virus to other people. By contrast 90 percent of infants and 30 percent of young children infected with hepatitis B develop chronic disease: the younger the child, the more likely that a hepatitis B infection will become chronic. The consequences of chronic hepatitis B infection may include:

- chronic liver disease
- cirrhosis
- liver **cancer**
- liver failure

There is no cure for hepatitis B and approximately one-fourth of chronic hepatitis B victims die of cirrhosis or liver cancer, including children who do not survive to young adulthood. Of the approximately 1.25 million Americans with chronic hepatitis B, 20–30 percent were infected as infants or children.

Risk of childhood infection

Those with the highest risk for infection are older adolescents and adults engaging in high-risk behaviors such as drug use and unprotected sex with multiple partners.

Far less common sources of childhood hepatitis B infection include:

- breast milk from an infected mother
- contact with blood, saliva, tears, or urine from an infected household member
- cuts
- blood transfusions

However, the following children are at particular risk for hepatitis B infection:

- children of immigrants and refugees or children adopted from regions where hepatitis B is endemic, including Asia, Sub-Saharan Africa, the Amazon Basin, Eastern Europe, and the Middle East
- Alaskan natives and Pacific Islanders
- children living in households with a chronically hepatitis-B-infected person
- children living in institutions
- children receiving hemodialysis
- children receiving certain blood products

Children born to infected mothers

Children of hepatitis B-infected mothers are at a 10–85 percent risk of becoming infected during birth. The CDC estimates that, prior to the infant HBV immunization program, about 12,000 American infants per year were infected by their mothers at birth. In addition, children of hepatitis B-infected mothers are at high risk of becoming infected before the age of five.

Children under the age of five who become infected with hepatitis B are at high risk for chronic infection and severe liver damage and disease later in life, even though initially they may have no symptoms. These infected children have a 90 percent risk of chronic hepatitis B infection and as many as 25 percent of them will die of chronic liver disease as adults. Mothers who have emigrated from countries with high rates of endemic hepatitis B are more likely to be infected.

Mothers with acute or chronic infectious hepatitis B can be identified by a blood test for HBsAg. Children born to mothers who have hepatitis B or whose hepatitis B status is unknown should receive their first HBV dose within 12 hours of birth. The second and third doses are given at two and six months of age. In many parts of the world, vaccine intervention before birth is required to prevent hepatitis B infection and its consequences in newborns.

It is recommended that newborns whose mothers are HBsAg-positive receive hepatitis B immune globulin (HBIG)—a preparation of serum containing high levels of antibodies to hepatitis B—as well as HBV within 12 hours of birth. About 70 percent of these newborns will be protected from chronic hepatitis B. A child's immune response to either hepatitis B infection or to HBV can be measured by a blood test for antibodies to HBsAg (anti-HBs). If a vaccinated child is exposed to hepatitis B, a measure of the anti-HBs in the blood will indicate whether another dose of HBV is required. Infants born to mothers who are HBsAg-positive should be tested for anti-HBs three to nine months following their last dose of vaccine. Their anti-HB levels should be at least 10 milli-international units per milliliter (mIU/ml), indicating that they are immune due to vaccination.

Mass immunization

HBV first became available in the United States in 1982. Between 1979 and 1989, the incidence of acute hepatitis B increased in the United States by 37 percent. There were 200,000–300,000 new infections annually between 1980 and 1991. In 1991 the CDC developed a strategy for eliminating the transmission of hepatitis B via universal childhood vaccination. The World Health Organization also declared the goal of immunizing all infants worldwide.

Nearly all states enacted laws requiring hepatitis B vaccination for enrollment in daycare, schools, and colleges. All these laws include exemptions for medical reasons and most include exemptions for religious reasons; however, only a few states allow exemptions from vaccination on philosophical grounds. Most states do not have laws mandating the screening of pregnant women for HBsAg.

By 2002, 90 percent of American children had been vaccinated against hepatitis B. The number of children carrying the virus was subsequently reduced substantially. Infant death from hepatitis B and the incidence of liver disease in children also decreased significantly. The CDC estimates that in 1998 the vaccine prevented 6,800 infections during birth and 18,700 infections in infants and children up to the age of nine. About 12,900 of these children would have developed chronic hepatitis and 3,000 of them eventually would have died of cirrhosis or liver cancer. The CDC expects the overall incidence of hepatitis B in the American population to fall throughout the early 2000s as a result of mass childhood vaccination. However, as of 2004, infants receiving HBV since 1991 had not yet reached the age when high-risk behaviors increase the likelihood of hepatitis B infection. In Pacific Island nations—where rates of hepatitis B infection are among the highest in the world—a regionally coordinated immunization program has significantly reduced the incidence of chronic infection.

Costs

HBV usually is covered by health insurance. In the United States the Vaccines for Children program covers the cost of hepatitis B vaccination for those without health insurance and for other specific groups of children, including Native Americans. The CDC estimates that infant hepatitis B vaccination saves fifty cents in direct medical costs for every dollar spent on HBV.

Precautions

Because most children are not at high risk for hepatitis B infection, and because the duration of immunity provided by HBV is not known, some parents and medical professionals question the need for and the effectiveness of childhood vaccination against hepatitis B. Some also continue to question the safety of the vaccine.

Children should not receive HBV if they are allergic to baker's yeast or thimerosal, are allergic to any other components in a combination vaccine, or have had a previous allergic reaction to HBV. A 2003 study found that

HBV was safe and effective in children with **asthma**, even those on inhaled steroid therapy.

Side effects

Although most children experience no side effects from HBV, the most common side effects are as follows:

- fatigue or irritability in up to 20 percent of children
- soreness at the point of the injection, lasting one to two days, in about one out of eleven children and adolescents
- a mild to moderate **fever** in one out of 14 children and adolescents

Other less common side effects of HBV include:

- a purple spot, hard lump, redness, swelling, pain, or **itching** at the point of injection
- unusual tiredness or weakness
- dizziness
- fever of 100°F (37.7 °C) or higher
- headache

Other rare reactions to HBV include:

- general feeling of discomfort or illness
- aches or pain in joints or muscles
- skin rash or welts that may occur days or weeks after receiving the vaccine
- blurred vision or other vision changes
- muscle weakness or **numbness** or **tingling** in the arms and legs
- back pain or stiffness or pain in the neck or shoulder
- chills
- diarrhea or stomach cramps
- nausea or vomiting
- increased sweating
- sore throat or runny nose
- itching
- decreased or lost appetite
- sudden redness of the skin
- swelling of glands in the armpit or neck
- difficulty sleeping

Although allergic reactions to HBV are rare, if they occur emergency medical help should be sought immediately. Symptoms of an allergic reaction include:

- reddening of the skin, especially around the ears
- swelling of the eyes, face, or inside of the nose
- hives
- itching, especially of the feet or hands
- sudden and severe tiredness or weakness
- difficulty breathing or swallowing

Parental concerns

Preparing a child for an injection

Most children are afraid of injections; however, there are simple methods for easing a child's **fear**. Prior to the vaccination parents should take the following steps:

- Tell children that they will be getting a shot and that it will feel like a prick; however, it will only sting for a few seconds.
- Explain to children that the shot will prevent them from becoming sick.
- Have older siblings comfort and reassure a younger child.
- Bring along the child's favorite toy or blanket.
- Never threaten children by telling them they will get a shot.
- Read the vaccination information statement and ask questions of the medical practitioner.

During the vaccination parents should take the following steps:

- Hold the child.
- Make eye contact with the child and smile.
- Talk softly and comfort the child.
- Distract the child by pointing out pictures or objects or using a hand puppet.
- Sing or tell the child a story.
- Have the child tell a story.
- Teach the child to focus on something other than the shot.
- Help the child take deep breaths.
- Allow the child to cry.
- Stay calm.

Comforting restraint

Parents may choose to use a comforting restraint method while their child is receiving an injection. These methods enable the parent to control and steady the child's arm while not holding the child down. With

infants and toddlers, the following holds may be effective:

- The child is held on the parent's lap.
- The child's arm is behind the parent's back, held under the parent's arm.
- The parent's arm and hand control the child's other arm.
- The child's feet are held between the parent's thighs and steadied with the parent's other arm.

With older children, the following positions may be effective:

- The child is held on the parent's lap or stands in front of the seated parent.
- The parent's arms embrace the child.
- The child's legs are between the parent's legs.

After the injection

Following an injection parents should help in the following ways:

- Hold and caress a child or breastfeed an infant.
- Talk soothingly and reassuringly.
- Hug and praise the child for doing well.
- Review the information for possible side effects.
- Use a cool, wet cloth to reduce soreness or swelling at the injection site.
- Check the child for **rashes** over the following few days.

In addition, parents should remember the following:

- The child may eat less during the first 24 hours following a vaccination.
- The child should drink plenty of fluids.
- The medical practitioner may suggest a non-aspirin pain reliever for the child.

KEY TERMS

Antibody—A special protein made by the body's immune system as a defense against foreign material (bacteria, viruses, etc.) that enters the body. It is uniquely designed to attack and neutralize the specific antigen that triggered the immune response.

Antigen—A substance (usually a protein) identified as foreign by the body's immune system, triggering the release of antibodies as part of the body's immune response.

Booster immunization—An additional dose of a vaccine to maintain immunity to the disease.

Cirrhosis—A chronic degenerative disease of the liver, in which normal cells are replaced by fibrous tissue and normal liver function is disrupted. The most common symptoms are mild jaundice, fluid collection in the tissues, mental confusion, and vomiting of blood. Cirrhosis is associated with portal hypertension and is a major risk factor for the later development of liver cancer. If left untreated, cirrhosis leads to liver failure.

Comvax—Hib-HepB, a combination vaccine that protects against the *Haemophilus influenzae* type B bacterium and the hepatitis B virus.

***Haemophilus influenzae* type B**—An anaerobic bacteria associated with human respiratory infections, conjunctivitis, and meningitis.

Hepatitis B immune globulin—HBIG, a blood serum preparation containing anti-hepatitis-B antibodies (anti-HBs) that is administered along with HBV to children born to hepatitis-B-infected mothers.

Immunity—Ability to resist the effects of agents, such as bacteria and viruses, that cause disease.

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National Immunization Program. NIP Public Inquiries, Mailstop E-05, 1600 Clifton Rd. NE, Atlanta, GA 30333. Web site: <www.cdc.gov/nip>.

National Vaccine Information Center. 421-E Church St., Vienna, VA 22180. Web site: <www.909shot.com>

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Hereditary fructose intolerance

Definition

Hereditary fructose intolerance is a metabolic disorder in which the small intestine cannot process fructose (fruit sugar) into a source of energy because of an enzyme deficiency that prevents fructose absorption.

Description

Fructose is a simple sugar found naturally in fruits, vegetables, and honey. Synthetic fructose (in the form of corn syrup) is used as a sweetener in many foods, including baby food, and sweetened beverages. Other simple sugars include glucose (the form in which sugar circulates in the blood) and galactose (produced by the digestion of milk). Simple sugars can be absorbed by the small intestine.

Digestion of food begins in the mouth, moves to the stomach, and then into the small intestine. Along the way, specific enzymes are needed to process different types of sugars. An enzyme is a substance that acts as a catalyst to produce chemical changes without being changed itself. People with fructose intolerance do not have the enzyme 1-phosphofructaldolase (also called aldolase B enzyme and fructose 1-phosphate aldolase). This enzyme is necessary for the absorption of fructose.

When people with fructose intolerance ingest fructose or sucrose (cane or beet sugar, table sugar), complicated chemical changes occur in the body due to the absence of the enzyme needed to process these sugars. The undigested fructose accumulates in the liver, kidneys, and small intestine, progressively causing damage that can lead to liver and kidney failure. The accumulated fructose interferes with the conversion of glycogen, the body's energy storage material, into glucose. As a result, the blood sugar falls to abnormal levels (**hypoglycemia**).

An interesting feature of fructose intolerance is that children affected by the disorder develop a powerful protective aversion (feeling of intense dislike) to sweet-tasting foods and beverages. In addition, they have an exceptionally good record of dental hygiene, which is thought to be the result of diminished sugar and carbohydrate intake.

Demographics

Hereditary fructose intolerance is estimated to affect one in about 20,000 people. It is reported more frequently in the United States and northern European countries than in other parts of the world. It occurs with equal frequency in males and females.

Causes and symptoms

Causes

Fructose intolerance is an inherited disorder. Both the mother and father have the gene that causes the condition but may not have symptoms of fructose intolerance themselves. (This is called an autosomal recessive pattern of inheritance.)

Symptoms

The disorder is not apparent until the infant is fed formula, juice, fruits, or baby foods that contain fructose. Many soy-based formulas contain sucrose as a carbohydrate source. Initial symptoms include severe abdominal **pain**, **vomiting** that can lead to **dehydration**, and unexplained **fever**. Other symptoms include extreme thirst and excessive urination and sweating. There is also a loss of appetite and a failure to grow. Tremors and seizures caused by low blood sugar can occur. The liver becomes swollen, and the patient becomes jaundiced with yellowing of the eyes and skin. Left untreated, this condition can lead to coma and death.

When to call the doctor

If a child develops the following symptoms after he or she begins eating formula or solid food, the parent should contact the child's pediatrician:

- persistent or severe vomiting
- severe abdominal pain
- unexplained fever
- intolerance for fruits or avoidance of fruits/sucrose-containing foods
- extreme thirst
- excessive urination
- jaundice (yellowing of the eyes and skin)
- loss of appetite
- failure to grow
- unexplained weight loss

Early symptoms of hypoglycemia include:

- confusion
- dizziness
- feeling shaky or trembling
- hunger
- headache
- irritability
- fast heartbeat
- pale skin
- sweating
- sudden drowsiness, weakness, or fatigue

These symptoms can be treated by giving the child an oral glucose tablet, available from most pharmacies.

Symptoms of late hypoglycemia should be treated immediately. Parents should give the child a glucagon injection (a medication used in an emergency to increase blood glucose) and seek emergent treatment when the child has the following symptoms:

- inability to swallow
- numbness in mouth or tongue
- poor coordination
- poor concentration, confusion
- unconsciousness

When hypoglycemia is severe, it can lead to convulsions and coma. The child's doctor can advise the parents on how to manage the child's blood glucose levels to avoid hypoglycemic reactions as much as possible.

Diagnosis

The diagnosis includes a physical exam and evaluation of the child's **family** medical history. A family history of fructose intolerance may suggest a genetic predisposition to the disease. Several gene mutations causing hereditary fructose intolerance have been identified. Genetic testing with DNA analysis may be available to identify one of the common gene mutations that lead to this disorder. Positive results of the DNA test and the presence of clinical symptoms can serve as strong indicators of the condition. However, negative results are not a guarantee that the person does not have hereditary fructose intolerance.

Urine tests can be used to detect fructose sugar in the urine. Blood tests can also be used to detect hyperbilirubinemia and high levels of liver enzymes and uric acid in the blood. A liver biopsy may be performed to test for levels of enzymes present (aldolase assay) and to evaluate the extent of damage to the liver. A fructose tolerance test may also be used to confirm fructose intolerance. In this test, a dose of fructose is given to the patient in a well-controlled hospital or clinical setting. This test should only be performed on an asymptomatic patient. Both the biopsy and the fructose tolerance test are very risky, particularly in infants who are already sick.

Treatment

With early diagnosis, fructose intolerance can be successfully treated by eliminating fructose, sucrose, and sorbitol from the diet (less than 40 mg/kg per day). Sorbitol is an artificial sweetener found in many sugar-free products, such as sugarless gum or diet foods. Patients usually respond favorably within a few weeks and can make a complete recovery if fructose-containing foods are avoided. Early recognition and treatment of the disorder is important to avoid damage to the liver, kidneys, and small intestine.

Early symptoms of hypoglycemia can be treated with oral glucose tablets or gel, available at most pharmacies. The doctor can provide more information about how to manage a hypoglycemic reaction, as well as how to monitor the child's blood glucose levels using a blood glucose meter to prevent a hypoglycemic reaction. Severe hypoglycemia should be treated with a glucagon injection to increase the blood glucose level. In some cases, the child may need an intravenous glucose solution, given in the hospital.

Children with this condition should be managed by a medical specialist in biochemical genetics or metabolism, as well as a registered dietitian who can provide **nutrition** support and information.

Nutritional concerns

It is important for the child to avoid fructose, sucrose, and sorbitol sources and yet maintain proper nutrition. A registered dietitian can work with the parents and child to identify and avoid fructose and sucrose foods and beverages. This is very important, since many unsuspected food sources, such as potatoes when prepared a certain way, are significant sources of fructose. The dietitian can provide instructions for reading food and medication labels to detect these problem-causing substances. Regular follow-up appointments with a registered dietitian should be part of the child's overall treatment program.

Prognosis

There is no cure for hereditary fructose intolerance, since the enzyme needed to process fructose is missing at birth. The prognosis depends on how soon the diagnosis is made and how soon fructose and sucrose are eliminated from the child's diet. If the condition is not recognized and the diet is not well controlled, death can occur in infants or young children. With a well-controlled diet, the child will thrive and develop normally. In the absence of liver damage, the child's life expectancy is normal. Most of the damaging effects of the disorder can be prevented by strictly following the fructose-free diet.

Prevention

Carriers of the gene for hereditary fructose intolerance can be identified through DNA analysis. Anyone who is known to carry the disorder or who has the disorder in his or her family may benefit from genetic counseling. At-risk individuals can be assisted with family planning and reproductive decisions.

Nutritional concerns

To prevent complications from this disorder, parents should take the following steps:

- work with a registered dietitian to facilitate specific dietary changes
- carefully read food labels to identify and avoid dietary sources of fructose and sucrose
- maintain a regular follow-up schedule with the child's metabolic specialist

Parental concerns

Fructose intolerance can be a life-threatening condition if strict dietary guidelines are not followed. If the diet is relaxed, the child may fail to grow normally or may develop liver or kidney complications.

Preliminary evidence suggests that parents of a child with this disorder, and other carriers of the mutant gene,

KEY TERMS

Aldolase B—Also called fructose 1-phosphate aldolase, this chemical is produced in the liver, kidneys, and brain. It is needed for the breakdown of fructose, a sugar found in fruits, vegetables, honey, and other sweeteners.

Digestion—The mechanical, chemical, and enzymatic process in which food is converted into the substances suitable for use by the body.

DNA—Deoxyribonucleic acid; the genetic material in cells that holds the inherited instructions for growth, development, and cellular functioning.

Enzyme—A protein that catalyzes a biochemical reaction without changing its own structure or function.

Hyperbilirubinemia—A condition characterized by a high level of bilirubin in the blood. Bilirubin is a natural byproduct of the breakdown of red blood cells, however, a high level of bilirubin may indicate a problem with the liver.

Lactose—A sugar found in milk and milk products.

Liver biopsy—A surgical procedure where a small piece of the liver is removed for examination. A needle or narrow tube may be inserted either directly through the skin and muscle or through a small incision and passed into the liver for collection of a sample of liver tissue.

Metabolism—The sum of all chemical reactions that occur in the body resulting in growth, transformation of foodstuffs into energy, waste elimination, and other bodily functions. These include processes that break down substances to yield energy and processes that build up other substances necessary for life.

Nutrient—Substances in food that supply the body with the elements needed for metabolism. Examples of nutrients are vitamins, minerals, carbohydrates, fats, and proteins.

Sugars—Those carbohydrates having the general composition of one part carbon, two parts hydrogen, and one part oxygen.

may have an increased risk for gout. Gout is a form of arthritis caused by excess uric acid in the body and uric acid crystals in the joints.

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National Institute of Diabetes and Digestive and Kidney Diseases (NIDDK). 2 Information Way, Bethesda, MD 20892–3570. Web site: <www.niddk.nih.gov>.

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Hereditary hemorrhagic telangiectasia

Definition

Hereditary hemorrhagic telangiectasia is a condition characterized by abnormal blood vessels which are delicate and prone to bleeding. Hereditary hemorrhagic telangiectasia is also known as Osler-Weber-Rendu disease.

Description

The term telangiectasia refers to a spot formed, usually on the skin, by a dilated capillary or terminal artery. Telangiectasia is an arterial-venous malformation (AVM) composed of small blood vessels. In hereditary hemorrhagic telangiectasia these spots occur because the blood vessel is fragile and bleeds easily. The bleeding may appear as small, red or reddish-violet spots on the face, lips, inside the mouth and nose, or the tips of the fingers and toes. Besides the skin and mouth, telangiectasias may occur in the gastrointestinal tract (GI tract), the brain, and the lungs. Unlike **hemophilia**, where bleeding is caused by an ineffective clotting mechanism in the blood, bleeding in hereditary hemorrhagic telangiectasia is caused by fragile blood vessels. However, like hemophilia, bleeding may be extensive and can occur without warning.

Causes and symptoms

Hereditary hemorrhagic telangiectasia, an autosomal dominant disorder, occurs in one in 50,000 people. Recurrent nosebleeds are a nearly universal symptom in this condition. Usually the nosebleeds begin in childhood and become worse with age. A patient may begin to **cough** up blood or pass blood in stools. The skin changes begin at **puberty**, and the condition becomes progressively worse until about 40 years of age, when it stabilizes.

When to call the doctor

Parents should notify a doctor if a child's bleeding does not stop, or bleeding is severe, or the child has severe headaches or becomes unresponsive. Frequent nosebleeds should be followed up with the pediatrician.

Diagnosis

The physician looks for red spots on all areas of the skin, but especially on the upper half of the body and in the mouth and nose and under the tongue. Bleeding in

KEY TERMS

Autosomal dominant—A pattern of inheritance in which only one of the two copies of an autosomal gene must be abnormal for a genetic condition or disease to occur. An autosomal gene is a gene that is located on one of the autosomes or non-sex chromosomes. A person with an autosomal dominant disorder has a 50% chance of passing it to each of their offspring.

Chromosome—A microscopic thread-like structure found within each cell of the human body and consisting of a complex of proteins and DNA. Humans have 46 chromosomes arranged into 23 pairs. Chromosomes contain the genetic information necessary to direct the development and functioning of all cells and systems in the body. They pass on hereditary traits from parents to child (like eye color) and determine whether the child will be male or female.

the GI tract can cause the stool (feces) to be darker than normal.

Treatment

There is no specific treatment for hereditary hemorrhagic telangiectasia. The bleeding resulting from the condition can be stopped by applying compresses or direct pressure to the area. If necessary, a laser can be used to destroy the vessel. In severe cases, the leaking artery can be plugged or covered with a graft from normal tissue. In some cases, estrogen therapy is used to reduce bleeding episodes.

Prognosis

In most people, recurrent bleeding results in an iron deficiency. It is usually necessary to take iron supplements. Patients have a normal lifespan, and many people are not aware they are affected by the disease until a **family** is diagnosed.

Prevention

Because it is an inherited disorder, hereditary hemorrhagic telangiectasia cannot be prevented.

Parental concerns

Parents should be aware that frequent nosebleeds are a common sign of hereditary hemorrhagic telangiectasia. Genetic counseling is recommended for those who know the disease runs in their families. There are also support groups for those who are affected by the disease.

Resources

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HHT Foundation International Inc. PO Box 329, Monkton, MD 21111. Web site: <www.hht.org/web/>.

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Hermaphroditism *see* **Intersex states**

Hernia

Definition

A hernia is the protrusion of an organ through the structure or muscle that usually contains it.

Description

There are many different types of hernias in children. The most common are direct inguinal hernias, indirect inguinal hernias, and umbilical hernias. A direct inguinal hernia occurs when a small section of bowel herniates, or protrudes, through the groin muscle. Indirect inguinal hernia occurs when part of the bowel protrudes through the muscles of the groin into a sac left over from fetal development. An umbilical hernia occurs when a portion of the bowel protrudes through a small defect in the abdominal wall muscle near where the umbilical cord attaches to the baby's abdomen. More serious defects involving herniation of abdominal contents outside the infant's body are omphalocele and gastroschisis. These are not a result of an organ protruding through weakened muscle tissue but rather are a result of a much larger defect of the muscles of the abdomen that causes the internal organs to develop outside the body.

Omphalocele and gastroschisis are considered **abdominal wall defects** and are not called hernias.

While an umbilical hernia usually resolves spontaneously as the abdominal muscles grow and requires no further treatment, in children with direct and indirect inguinal hernia, surgery is almost always required to prevent the herniated bowel from becoming incarcerated or strangulated. When an inguinal hernia is incarcerated, the bowel becomes swollen and trapped outside the body. If the hernia remains incarcerated for too long, strangulation can occur. In strangulation, the blood supply to the section of bowel that has herniated is cut off, and the tissue begins to die. When this happens, the intestines cannot function properly and are said to be obstructed. If the bowel perforates, or develops a hole in it, emergency surgery is required to repair the intestine and prevent infection.

A more severe, but less common, hernia is a diaphragmatic hernia. This occurs inside the body when the diaphragm, the large muscle that separates the abdominal cavity from the chest cavity, fails to develop fully. In children with diaphragmatic hernia, the contents of the abdomen protrude into the chest cavity. These children may have difficulty breathing. During fetal development the presence of abdominal organs in the fetal chest cavity prevents the lungs from growing normally. A diaphragmatic hernia can occur as an isolated defect or as part of a more complex syndrome. Children with diaphragmatic hernias are usually very ill and require immediate treatment after birth. Some of these children have other defects such as cardiac anomalies, chromosomal abnormalities, kidney and genital anomalies, and neural tube defects, such as **spina bifida**.

Demographics

Estimates of the true incidence of inguinal hernias vary, but they may affect 1–5 percent of all births in the United States. International rates appear to be similar. Males are more than seven times more likely to have an inguinal hernia than females, and premature infants are more likely than full term infants to have inguinal hernias and to have incarcerated hernias. While inguinal hernias seem to affect all racial groups at the same rate, umbilical hernias occur more frequently in African Americans.

Diaphragmatic hernias occur in approximately one in every 3,000 births. These hernias do not seem to affect any race or nationality more than another.

Causes and symptoms

A direct inguinal hernia is caused when the muscles of the floor of the groin area are weak and allow the bowel to press through. An indirect inguinal hernia is caused when remnants of early fetal genital development stay within the body after this development is complete. In early fetal development male and female genitalia are identical. At around the seventh week of gestation, the gonads (sex organs) begin to change, or differentiate, into the characteristic genitalia of males and females. Males develop testes, and females develop ovaries. During this process, in some fetuses, a small sac may form near the genitalia. Most often the opening to this sac, called the processus vaginalis, closes. However, in children with inguinal hernia, this sac remains patent, or open, becoming a container into which bowels may be herniated.

The main symptom of inguinal hernias (both direct and indirect) in infants is an obvious bulge in the groin in the inguinoscrotal region (near the scrotum) in boys and in the inguinolabial (near the labia) in girls. The bulge may or may not be painful. It will usually appear after straining or crying and then disappear after a period of time. If the hernia has incarcerated, the infant will be in obvious **pain**, appearing fussy, crying, and refusing to eat. The skin over the hernia may be discolored and swollen.

Umbilical hernia is caused by a small defect in the muscles of the abdominal wall. These hernias are usually small and have no symptoms other than a small protrusion near the base of the umbilical cord.

Like inguinal hernias, diaphragmatic hernias are caused early in fetal development. The structures that form the diaphragm do not properly form, allowing the contents of the lower abdomen to migrate up near the heart and lungs. The increased pressure these organs place on the lungs causes the lungs to remain small and underdeveloped. When the infant is born and must breathe air, the lungs are not able to work properly.

Children with diaphragmatic hernia have the following symptoms immediately after birth: breathing difficulty, a bluish skin color (cyanosis), rapid breathing, rapid heat rate, and asymmetrical chests—one side is not the same size as the other. These infants are often critically ill and are be placed on a ventilator—a machine to help them breath. Because the lungs have not had enough room to grow and are small, doctors must stabilize the baby's breathing before the hernia can be repaired.

When to call the doctor

If a small child, especially an infant, has a bulge in the abdominal or groin area, the child's pediatrician should be consulted. If the child is in severe pain, and the skin is discolored or swollen, medical help should be sought immediately.

Diagnosis

Umbilical and inguinal hernias are diagnosed by physical examination. For some children with inguinal hernia, a laparoscopic examination may be performed. A laparoscopy is an exploratory surgical procedure in which the doctor makes an incision and inserts a small tube connected to a camera to view the herniated area. This procedure is used most often in patients who have already had one hernia repair to see if the hernia has returned in a new location.

Diaphragmatic hernia may be diagnosed while the fetus is still in the womb using prenatal ultrasonography. After birth, physical symptoms of respiratory distress, cyanosis, and chest asymmetry can indicate the presence of a diaphragmatic hernia. In children with less severe diaphragmatic hernias, the diagnosis may be made later in childhood if the child develops **intestinal obstructions**. An x ray showing bowel loops within the chest cavity confirms the diagnosis.

Treatment

Umbilical hernia is generally a benign condition that will resolve spontaneously as the muscles of the abdomen grow. No treatment is usually required. For children in whom the umbilical hernia does not resolve, surgery is not usually performed until after the age of five. The only treatment necessary is observation of the hernia during routine physical examinations.

The standard treatment for inguinal hernias is a surgical repair called herniorrhaphy. Unlike umbilical hernias, inguinal hernias do not resolve spontaneously. Because of the risk of incarceration and strangulation, most doctors prefer to repair these hernias as soon after the initial diagnosis as possible. Herniorrhaphies are performed as an outpatient procedure in otherwise healthy full-term infants and children.

Prior to repair surgery, parents may be taught how to apply pressure to the hernia, thereby reducing it temporarily and preventing incarceration. If the hernia has already become incarcerated, the doctor will attempt to force the hernia out of the sac and back into the body manually. This process is called manual reduction. With the child on his back, the doctor will use his fingers to press the hernia back into the body. If successful, manual reduction relieves the child's pain and prevents strangulation until surgery can be scheduled. Repair surgery is usually performed within 72 hours. If an incarcerated hernia is not reducible, surgery must be performed much sooner to prevent strangulation. If strangulation occurs, emergency surgery is the only treatment.

Treatment for diaphragmatic hernia involves treatment of the other accompanying health issues. First and foremost, the infant's respiratory distress must be addressed. Most newborns with diaphragmatic hernias require intubation and ventilation. A tube is inserted through the mouth into the throat, and breathing is assisted by a ventilation machine. A feeding tube may be inserted through the nose and into the stomach to insure the infant receives sufficient **nutrition**. After the infant is stabilized, surgery to repair the hernia is performed. In diaphragmatic hernia repair surgery, the herniated abdominal organs are forced back into their proper position within the abdomen. If the bowels are injured or malrotated, this will be repaired, and the hole in the diaphragm is sewn closed and patched, if necessary, with surgical mesh.

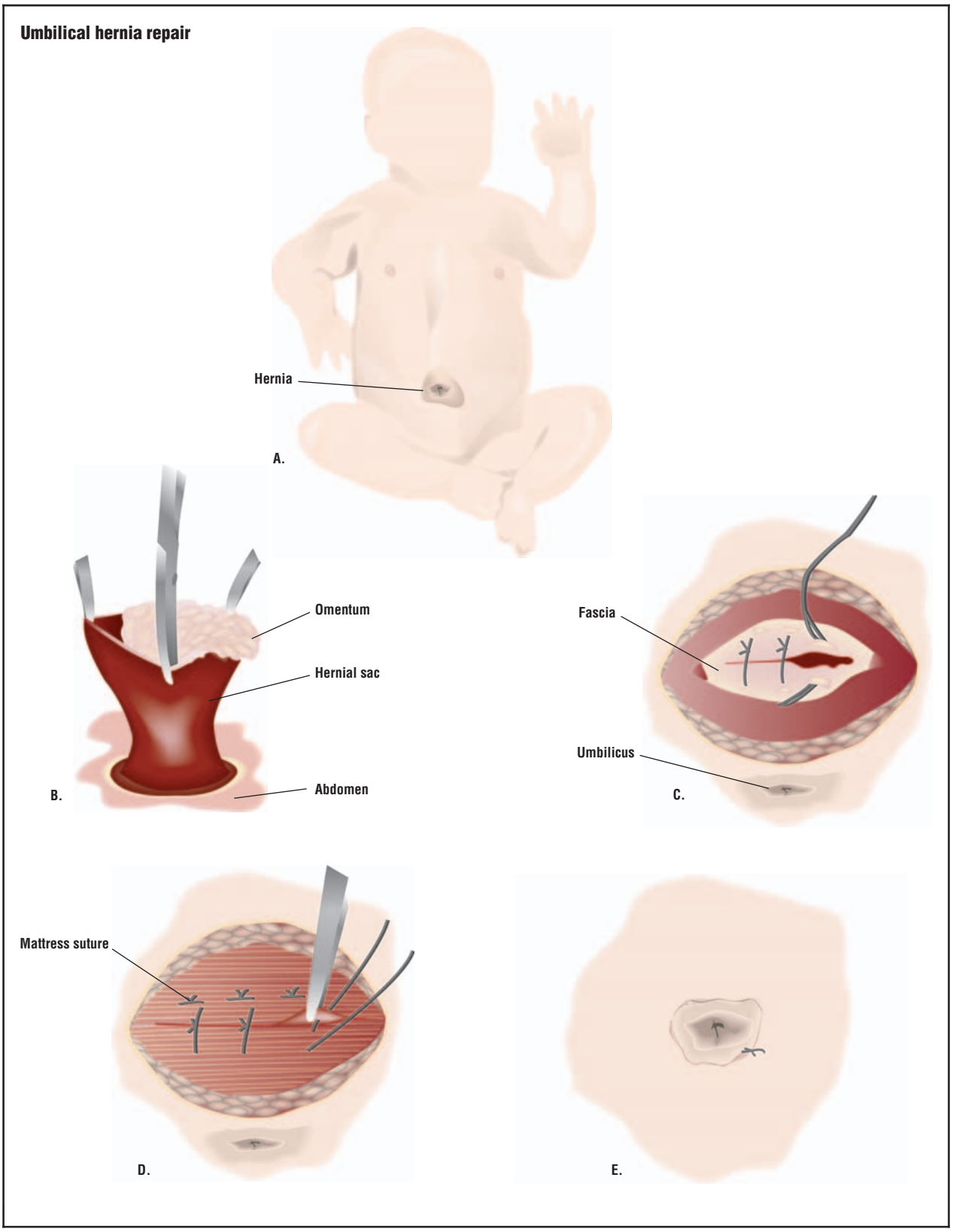
Prognosis

If diagnosed early in childhood, the prognosis for children who have had a surgically repaired inguinal hernia is excellent. Occasionally there are complications associated with inguinal hernias including death, but these are rare, occurring most often in children who were diagnosed later in childhood or whose hernias were strangulated.

The prognosis for children with diaphragmatic hernia depends on the extent of the defects of the lungs and the impact of the treatments necessary to save their lives. Children with diaphragmatic hernias have an increased incidence of chromic lung disease. These children also have an increased risk for slow growth and development. The survival rate of these children is also related to the other anomalies these children may have. If the diaphragmatic hernia is part of a syndrome, the other birth defects may be life threatening. The survival rate after surgical repair of a diaphragmatic hernia is 60–80 percent.

Prevention

The exact cause of umbilical hernias, inguinal hernias, and diaphragmatic hernias is as of 2004 unknown. Until a cause is discovered, no prevention is available.



Baby with an umbilical hernia (A). To repair, the hernia is cut open (B), and the contents replaced in the abdomen. Connecting tissues, or fascia, are sutured closed (C), and the skin is repaired (D). *(Illustration by GGS Information Services.)*

KEY TERMS

Herniorrhaphy—Surgical repair of a hernia.

Incarcerated hernia—A hernia of the bowel that can not return to its normal place without manipulation or surgery.

Laparoscopy—A surgical procedure in which a small incision is made, usually in the navel, through which a viewing tube (laparoscope) is inserted. This allows the doctor to examine abdominal and pelvic organs. Other small incisions can be mad to insert instruments to perform procedures. Laparoscopy is done to diagnose conditions or to perform certain types of surgeries.

Reducible hernia—A hernia that can be gently pushed back into place or that disappears when the person lies down.

Strangulated hernia—A hernia that is so tightly incarcerated outside the abdominal wall that the intestine is blocked and the blood supply to that part of the intestine is cut off.

Ultrasonography—A medical test in which sound waves are directed against internal structures in the body. As sound waves bounce off the internal structure, they create an image on a video screen. Ultrasonography is often used to diagnose fetal abnormalities, gallstones, heart defects, and tumors. Also called ultrasound imaging.

Parental concerns

Prior to surgery, parents of a child with an inguinal hernia can be taught to apply pressure to the hernia, preventing incarceration. Parents should be aware of the circumstances under which to seek immediate medical attention for their child.

See also Abdominal wall defects.

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Deborah L. Nurmi, MS

Herpes simplex

Definition

Herpes is an infection caused by a herpes simplex virus 1 or 2, and it primarily affects the mouth or genital area.

Description

There are two strains of herpes simplex viruses. Herpes simplex virus type 1 (HSV-1) is usually associated with infections of the lips, mouth, and face. It is the most common herpes simplex virus among the general population and is usually acquired in childhood. Herpes simplex virus 2 (HSV-2) is sexually transmitted and is usually associated with genital ulcers or sores. Individuals may harbor HSV-1 and or HSV-2 and not have developed any symptoms.

Transmission

HSV-1 causes lesions inside the mouth that are often referred to as cold sores or fever blisters, and it is transmitted by contact with infected saliva. By adulthood, up to 90 percent of the population has antibodies to HSV-1. HSV-2 is sexually transmitted and not everyone develops symptoms when they have it. Up to 30 percent of adults in the United States have antibodies against HSV-2. Cross infection of type 1 and 2 viruses may occur from oral-genital contact. Herpes viruses can be transmitted to a newborn during vaginal delivery in

mothers infected with herpes viruses, especially if the infection is primary (first occurrence) and is active at the time of delivery. The virus can lead to complications such as meningoencephalitis, which is an infection of the lining of the brain and the brain itself. It can also cause eye infections, in particular, of the conjunctiva and cornea.

Demographics

The prevalence of herpes simplex in the United States is as follows:

- Seventy to ninety percent of adults test seropositive (present in blood serum) for HSV-1.
- Up to 30 percent of adults test seropositive for HSV-2.
- The highest incidence of HSV-1 is in children six months to three years of age.
- The highest incidence of HSV-2 is in young adults between the age of 18 and 25 years.
- HSV-2 antibodies are present in approximately 20 percent of Caucasians and about 65 percent of African-American adults.

Causes and symptoms

A primary infection of HSV-1 typically occurs between six months and five years of age and is systemic (affecting the whole body). Transmission is generally via respiratory droplets (HSV-1) or direct contact (HSV-1 and HSV-2). The virus enters the body through mucosal surfaces, replicates in the cell nucleus, and then kills the host cell. The initial infection is self-limiting, but the immune system does not destroy the virus. The virus migrates along nerves to an area of regional ganglia (nerve centers) and then typically enters into a latent (sleeping) phase. Reactivation of the virus occurs in 50 percent of patients within five years, and it can be triggered by various factors:

- fatigue
- stress
- trauma
- immunocompromise (lack of normal immune response)
- illness, such as a cold
- fever
- sunburn
- menstruation
- sexual intercourse

The symptoms of a herpes infection can vary tremendously. Many infected individuals have few, if any, noticeable symptoms. Those who do have symptoms usually notice them from two to 20 days after being exposed to someone with HSV infection. Symptoms can last for several weeks, but the first episode of herpes is usually worse than subsequent outbreaks. The predominant symptom of herpes is the outbreak of painful, **itching** blisters filled with fluid on and around the external sexual organs or, for oral herpes, on or very near the lip. Females may have a vaginal discharge and experience flu-like symptoms with HSV2 outbreaks, including fever, **headache**, muscle aches, and fatigue. There may be painful urination, and swollen and tender lymph glands in the groin. More often than not the blisters disappear without treatment in two to 10 days, but the virus remains in the body, lying dormant among clusters of nerve cells until another outbreak is triggered.

Many people are able to anticipate an outbreak when they notice a warning sign (a **tingling** sensation, called a prodrome) of the approaching illness. It is when they feel signs that an outbreak is about to start that they are particularly contagious, even though the skin still appears normal. Most people with genital herpes have five to eight outbreaks per year, but not everyone has recurrent symptoms. In time, the number of outbreaks usually decreases. Oral herpes can recur as often as monthly or only one or two times each year. Sores typically come back near the site of the first infection, but there are fewer sores with recurrences that heal faster and are less painful.

When to call the doctor

Anyone who has a history of herpes infection and current lesions should notify the physician if the lesions do not resolve after seven to ten days or if a condition exists that weakens the immune system. Children with a herpes infection most commonly have sores in the mouth usually caused by HSV-1. This infection causes fever, irritability, **pain**, decreased appetite, and ulcers in the mouth. The most common complication is **dehydration** secondary to a refusal to drink fluids because of mouth pain and difficulty swallowing. Treatment is usually not required, and symptoms generally improve in three to five days. If, however, the child does not improve, develops a fever, and becomes lethargic, the pediatrician should be called immediately.

Herpes infections that spread throughout the body in a newborn are usually more serious, but fortunately less common than the other types of neonatal infections. They typically occur in the first week of life, with symptoms including fever, difficulty breathing, seizures,

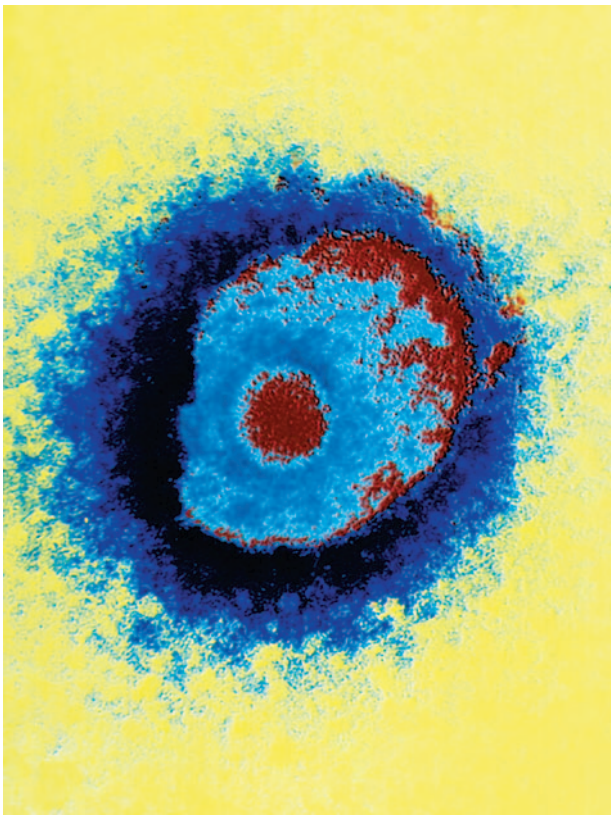
lethargy, and irritability. Since many infants in the first month of life can have a herpes infection and not have skin lesions, it takes a great deal of time and effort to diagnose and treat these infections early. Herpes should be considered in any acutely ill newborn, especially if bacterial cultures are negative and the baby is not improving after two to three days. Parents should be informed to watch the baby closely if either one of them has a history of herpes infections.

Diagnosis

Testing for neonatal herpes infections may include special smears and/or viral cultures, blood antibody levels, and polymerase chain reaction (PCR) testing of spinal fluid. Cultures are usually obtained from skin vesicles, eyes, mouth, rectum, urine, stool, and blood. For older children and adults, if there is a question as to the cause of a sore, a tissue sample or culture can be taken to determine what type of virus or other microorganism is responsible. For herpes, it is preferable to have this test done within the first 48 hours after symptoms first show up for a more accurate result.

Treatment

There are three drugs proven to treat genital herpes symptoms: acyclovir, sold under the brand name Zovirax, Famvir, and Valtrex. These are all taken in pill form. Formulas applied to the surface of the skin provide little benefit, and they are not recommended. Drug therapy is not a cure, but it can make living with the condition easier. For an initial outbreak with symptoms such as sores, a doctor should begin a brief course of antiviral therapy to relieve the symptoms or prevent them from getting worse. Seven to ten days of treatment is recommended but if the lesions do not heal, a longer period of time may be required. Following the initial outbreak there are two options to consider for further outbreaks. One is intermittent treatment, which involves the physician prescribing an antiviral drug to keep on hand in case an individual has a flare-up. The pills can be taken for three to five days as soon as sores are noticed or when an outbreak tingling sensation occurs. Sores heal and disappear on their own, but taking the drugs helps to alleviate the symptoms. For individuals who have frequent outbreaks, a suppressive treatment may work better. This treatment involves taking an antiviral drug every day. For example, someone who typically has more than six outbreaks a year, suppressive therapy reduces the number of outbreaks by 70 to 80 percent. Moreover, many who take the **antiviral drugs** daily have no outbreaks at all.



A false-color transmission electron microscopy (TEM) image of a herpes simplex virus. *(Custom Medical Stock Photo, Inc.)*

In the early 2000s herpes vaccines are being investigated, and an effective vaccine may be available in before 2010. Vaccines will only function to prevent the infection in new patients. Those who already have the simplex virus disease will probably not benefit.

Nutritional concerns

Diet is a very important factor in keeping herpes in remission. It has been found that foods high in arginine may cause herpes outbreaks. Supplementation with free-form lysine has shown to be beneficial in controlling herpes along with a diet high in lysine and low in arginine. The amount of lysine required to control herpes varies from case to case, but a typical adult dose to maintain remission is 500 mg daily, and active herpes requires 1–6 g between meals to induce healing.

Prognosis

There is no cure for herpes simplex. Once it is contracted, it is always in a person's system. However, with treatment therapies, the problems previously encountered are lessened considerably.

KEY TERMS

Conjunctiva—Plural, conjunctivae. The mucous membrane that covers the white part of the eyes (sclera) and lines the eyelids.

Cornea—The clear, dome-shaped outer covering of the eye that lies in front of the iris and pupil. The cornea lets light into the eye.

Ganglion—Plural, ganglia. A mass of nerve tissue or a group of neurons.

Latent virus—A nonactive virus that is in a dormant state within a cell. The herpes virus is latent in the nervous system.

Lesion—A disruption of the normal structure and function of a tissue by an injury or disease process. Wounds, sores, rashes, and boils are all lesions.

Meningoencephalitis—Inflammation of the brain and its membranes; also called cerebromeningitis or encephalomeningitis.

Mucosal—Refers to the mucous membrane.

Seropositive—Showing a positive reaction to a test on blood serum for a disease; exhibiting seroconversion.

Virus—A small infectious agent consisting of a core of genetic material (DNA or RNA) surrounded by a shell of protein. A virus needs a living cell to reproduce.

Prevention

Whereas it is almost impossible to keep a baby or child from being exposed to herpes simplex due to its universal presence, there are conditions that can be used to prevent its transmission. Hand washing is one of the biggest factors in the transmission of all diseases, and it is especially true of herpes simplex since it is spread by respiratory droplets through mucosal membranes. In terms of genital herpes, education regarding the use of condoms is the best tool. Young adults should also be reminded that herpes simplex can be transferred from oral-genital contact. Since many teenagers do not consider oral or anal sex as sexual intercourse per se, it is imperative to spell out exactly what, when, and how these viruses can be spread.

Parental concerns

It is important that the pediatrician discusses the possibility of herpes infections with new parents, particularly if they have a history of genital herpes. Signs and symptoms need to be gone over, i.e., lethargy, fever, as well as the fact that there may or may not be lesions present. A newborn's own immune system begins to function around the third month, and if a mother is breastfeeding, she is passing antibodies to her baby. The primary concern is that a herpes infection does not become systemic. Thus, if the child seems to be getting sicker instead of better, parents should call the doctor immediately.

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Hib vaccine

Definition

The Hib vaccine is an injection that helps protect children from contracting infections due to *Haemophilus influenzae* type B (Hib), a bacterium that is capable of causing serious illness and potential death in children under age five.

Description

H. influenzae type B (Hib) is a common organism worldwide; it is found in most healthy individuals in the general population. Small children can pick up the bacteria from people who are not aware that they are carriers. When the bacteria spread to the lungs and bloodstream, serious illness, including **pneumonia** and **meningitis**, can result. Another serious disease stem-

ming from this pathogen is **epiglottitis**, an infection of the epiglottis that cause swelling of the airways and potential death.

Pediatricians and the Centers for Disease Control (CDC) recommend that all infants are vaccinated against Hib disease. In general, the vaccine is considered highly effective, with few side effects. Before the institution of routine infant vaccinations in the United States in the 1990s, Hib was the leading of bacterial meningitis among children younger than five years of age. Nearly 1,000 people in the United States died every year from Hib disease. As of the early 2000s, the disease has largely disappeared due to these vaccinations. In developing countries, however, Hib disease is a major cause of serious disease and death in young children.

General use

The Hib vaccine is given in three or four doses during infancy, depending on the brand used. The first dose starts at about two months of age, and the last is usually scheduled at a 12- or 15-month check-up. Children older than five years do not need **vaccination**, unless the child or adolescent has a serious health problem that lowers immunity, such as **HIV infection**, sickle cell disease, or is being treated for **cancer**.

Precautions

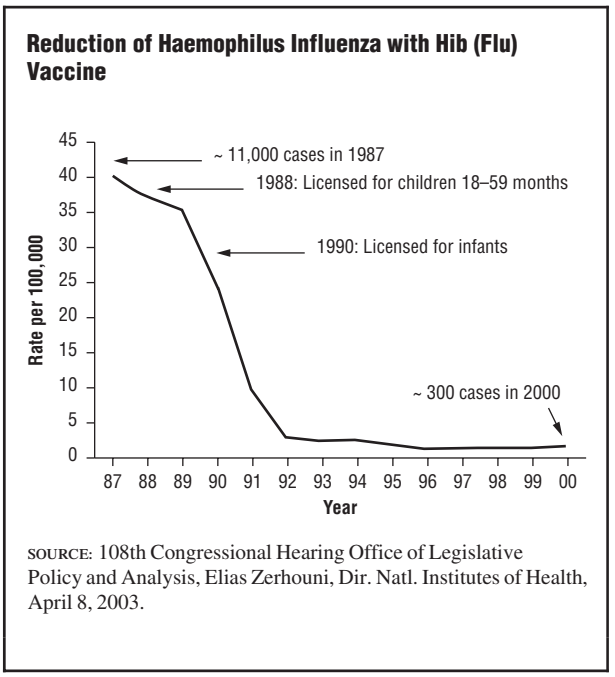
Infants under six weeks of age should not receive the Hib vaccine. Those babies with moderate to severe illness should wait for vaccination until they are well. If a baby has had a severe reaction to Hib vaccine, another dose should not be administered.

Side effects

Like any vaccine or medication, Hib vaccine is capable of causing an allergic reaction. However, this is extremely rare. More common side effects include inflammation at the injection site and slight **fever**, which can usually occur within 24 hours of the injection and can last two or three days.

Interactions

Interactions with medicines or food have not been reported. The Hib vaccine is routinely given at the same time as other childhood vaccines.



SOURCE: 108th Congressional Hearing Office of Legislative Policy and Analysis, Elias Zerhouni, Dir. Natl. Institutes of Health, April 8, 2003.

This graph shows the dramatic reduction of *Haemophilus influenzae* infection with the introduction of the Hib vaccine. *(Illustration by GGS Information Services.)*

Parental concerns

Parents should be aware of the benefits this vaccine provides, as well as its overall safety and effectiveness. Any low-grade fever or soreness can be treated with ibuprofen or **acetaminophen** as recommended by the pediatrician. Serious reactions, including high fever, difficulty breathing, and fast heartbeat should receive immediate medical treatment.

See also Hemophilus infections.

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Kristine Krapp

High blood pressure *see* **Hypertension**

High cholesterol *see* **Cholesterol, high**

High-risk pregnancy

Definition

Although as of 2004 there was no formal or universally accepted definition of a "high-risk" pregnancy, it is generally thought of as one in which the mother or the developing fetus has a condition that places one or both of them at a higher-than-normal-risk for complications, either during the pregnancy (antepartum), during delivery (intrapartum), or following the birth (postpartum).

Description

Certain conditions, called risk factors, make a pregnancy high risk. Maternal conditions can be identified with preconception counseling and from the maternal history. Maternal physical and social characteristics that can contribute to a high-risk pregnancy include:

- age younger than 15 years and older than 35 years
- pre-pregnancy weight under 100 lbs (45 kg) or obesity
- height under 5 ft (1.5 m)
- incompetent cervix
- uterine malformations
- small pelvis
- being a single woman
- being a smoker
- using illicit drugs
- having no access to early prenatal care
- using alcohol
- having low socioeconomic status

For women who do not have health insurance, obtaining early prenatal care is extremely difficult, and these same women are often from a socioeconomic level that prevents adequate or appropriate nutritional intake. There is a scoring system that can be used by healthcare professionals to determine the degree of risk for a pregnant woman, but it is difficult to rate risk by degrees. Nevertheless, identification of a high-risk pregnancy helps to ensure that those women who need the most care receive it.

One of the initial factors to consider when evaluating risk is the obstetrical history. If this is not the woman's first pregnancy, outcomes of her previous pregnancies are of importance in relation to the outcome of this one. An obstetrical history with any of the following conditions would be considered high risk:

- previous stillbirth
- previous neonatal death
- previous premature infant
- previous post-term (over 42 weeks) pregnancy
- fetal blood transfusion for hemolytic disease
- repeated miscarriages
- previous infant over 10 lbs (4.5 kg)
- six or more completed pregnancies
- history of preeclampsia
- history of eclampsia
- previous **cesarean section**
- history of a fetus with anomalies

Next to be considered is the medical history factor. A pregnant woman with any of the following medical conditions would be considered at risk:

- abnormal PAP test
- chronic **hypertension**
- heart disease (class II-IV, symptomatic)
- insulin-dependent diabetes
- moderate to severe kidney disease
- endocrine gland removal or ablation by autoimmune disease
- sickle cell disease
- epilepsy
- history of tuberculosis
- positive serology for syphilis
- pulmonary disease
- thyroid disease
- family history of diabetes
- HIV
- other chronic diseases
- autoimmune diseases, such as lupus

Current pregnancy risk factors would be considered as follows:

- abnormal fetal position
- mild to severe preeclampsia
- multiple pregnancy
- placenta abruption
- placenta previa
- polyhydramnios or oligiohydramnios
- gestational diabetes
- kidney infection
- Rh sensitization only
- mild (>9g/dl hemoglobin) or severe (<9g/dl hemoglobin) anemia
- vaginal spotting
- bladder infection
- emotional problems
- moderate alcohol use
- smoking more than one pack per day
- infection with parvovirus B19 (fifth disease), cytomegalovirus (CMV), **toxoplasmosis**, listeria, rubella
- exposure to damaging medications, esp., phenytoin, **folic acid** antagonists, lithium, streptomycin, tetracycline, warfarin

If prenatal testing indicates the baby has a serious congenital anomaly as a heart defect or spinal cord defect, the mother may need additional testing to determine the extent of the problem. Certain maternal or fetal problems may require the physician to deliver a baby early or to choose a surgical delivery (cesarean section) rather than a vaginal delivery.

Most women will see one healthcare provider during pregnancy, either an obstetrician, a midwife, or a nurse practitioner. Women who have a medical problem may need to see a medical specialist as well. Women diagnosed with a high-risk pregnancy should seek the care of an expert in the field of high-risk obstetrics, called a perinatologist. Perinatologists have additional training beyond the education required for an obstetrician. They care for women who have pre-existing medical problems, women who develop complications during pregnancy, and women whose fetus has problems.

Diagnosis

Labeling a woman with the diagnosis of high-risk pregnancy requires that one of the previous conditions be met. Thus, the diagnosis may be determined during history taking or if it is the fetus, during the morphological ultrasound at 16–19 weeks gestation. A woman with a high-risk pregnancy will need closer monitoring than pregnant women who are not high risk. Such monitoring may include frequent visits with the primary caregiver, tests to monitor the medical problem, blood tests to check the levels of medication, **amniocentesis**, serial ultrasound examination, and fetal monitoring. These tests are designed to follow the original condition, survey for complications, verify that the fetus is growing adequately, and make decisions regarding whether labor may need to be induced for early delivery of the fetus.

Treatment

Treatment varies widely with the type of disease, the effect that pregnancy has on the disease, and the effect that the disease has on pregnancy. If it is the fetus that has a problem, serial ultrasounds may be performed. Fetal heart rate monitoring may be necessary, or amniocentesis may be required. In addition, it may be essential to give the mother medications to act on the baby.

Prognosis

The prognosis is usually dependent on the specific medical condition. Some medical conditions make it difficult for women to get pregnant and lead to a higher risk of problems in the baby. In thyroid disease, the thyroid gland (located in the neck) may produce too much or too little thyroid hormone. Abnormal levels of this hormone can affect fertility and/or cause problems with the pregnancy and possibly affect the health of the baby. Fortunately, thyroid disease can be treated with medication. As long as the level of thyroid hormone is controlled throughout pregnancy, there should be no problems for mother or baby.

There are other medical conditions that do not interfere with pregnancy but are themselves affected by pregnancy. This group includes **asthma**, epilepsy, and ulcerative colitis. Some women with ulcerative colitis experience a worsening of their symptoms during pregnancy, while others will have no change or may get better during pregnancy. The same is true of asthma: some women notice that their asthma symptoms are better during pregnancy, some find their asthma worse, and some women notice no change in symptoms. It is not immediately apparent why this discrepancy occurs, but due to the unpredictability of diseases, all women with chronic illnesses should be monitored throughout the course of a pregnancy.

Some autoimmune diseases constitute a group of medical conditions that have a major impact on

KEY TERMS

Ablation—To remove or destroy tissue or a body part, such as by burning or cutting.

Amniocentesis—A procedure performed at 16–18 weeks of pregnancy in which a needle is inserted through a woman's abdomen into her uterus to draw out a small sample of the amniotic fluid from around the baby for analysis. Either the fluid itself or cells from the fluid can be used for a variety of tests to obtain information about genetic disorders and other medical conditions in the fetus.

Amniotic sac—The membranous sac that contains the fetus and the amniotic fluid during pregnancy.

Antepartum—The time period of the woman's pregnancy from conception and onset of labor.

Cytomegalovirus (CMV)—A common human virus causing mild or no symptoms in healthy people, but permanent damage or death to an infected fetus, a transplant patient, or a person with HIV.

Eclampsia—Coma and convulsions during or immediately after pregnancy, characterized by edema, hypertension, and proteinuria.

Endocrine—Refers to glands that secrete hormones circulated in the bloodstream or lymphatic system.

Gestational diabetes—Diabetes of pregnancy leading to increased levels of blood sugar. Unlike diabetes mellitus, gestational diabetes is caused by pregnancy and goes away when pregnancy ends. Like diabetes mellitus, gestational diabetes is treated with a special diet and insulin, if necessary.

Intrapartum—Refers to labor and delivery.

Listeria—An uncommon food-borne, life-threatening pathogen that can cause perinatal infection, which is associated with a high rate of fetal loss (including full-term stillbirths) and serious neonatal disease.

Oligohydramnios—A reduced amount of amniotic fluid. Causes include non-functioning kidneys and premature rupture of membranes. Without amniotic fluid to breathe, a baby will have underdeveloped and immature lungs.

Parvovirus B19—A virus that commonly infects humans; about 50 percent of all adults have been infected sometime during childhood or adolescence. Parvovirus B19 infects only humans. An infection in pregnancy can cause the unborn baby to have severe anemia and the woman may have a miscarriage.

Perinatal—Referring to the period of time surrounding an infant's birth, from the last two months of pregnancy through the first 28 days of life.

Phenytoin—An anti-convulsant medication used to treat seizure disorders. Sold under the brand name Dilantin.

Polyhydramnios—A condition in which there is too much fluid around the fetus in the amniotic sac.

Postpartum—After childbirth.

Preeclampsia—A condition that develops after the twentieth week of pregnancy and results in high blood pressure, fluid retention that doesn't go away, and large amounts of protein in the urine. Without treatment, it can progress to a dangerous condition called eclampsia, in which a woman goes into convulsions.

Premature labor—Labor beginning before 36 weeks of pregnancy.

Rubella—A mild, highly contagious childhood illness caused by a virus; it is also called German measles. Rubella causes severe birth defects (including heart defects, cataracts, deafness, and mental retardation) if a pregnant woman contracts it during the first three months of pregnancy.

Streptomycin—An antibiotic used to treat tuberculosis.

Tetracycline—A broad-spectrum antibiotic.

Toxoplasmosis—A parasitic infection caused by the intracellular protozoan *Toxoplasmosis gondii*. Humans are most commonly infected by swallowing the oocyte form of the parasite in soil (or kitty litter) contaminated by feces from an infected cat; or by swallowing the cyst form of the parasite in raw or undercooked meat.

Warfarin—An anticoagulant drug given to treat existing blood clots or to control the formation of new blood clots. Sold in the U.S. under the brand name Coumadin.

pregnancy. Women with lupus (a disease caused by alterations in the immune system that result in inflammation of connective tissue and organs) or kidney disease face serious risks during pregnancy. Pregnancy can cause their symptoms to worsen significantly and lead to severe complications for the mother and the baby. With systemic autoimmune diseases or vasculitis, the mother's blood circulation can be impaired and thus the ability to

supply oxygen and nutrients to the baby through the placenta is affected. As a result, fetal intrauterine growth becomes restricted (IUGR). Since chronic hypertension or pregnancy-induced hypertension (preeclampsia, eclampsia) similarly affect blood circulation to the placenta, women with these problems are also at risk for IUGR. If the condition is not determined early enough to provide constant monitoring, there is increased risk of stillbirth. Other autoimmune diseases, (antiphospholipid antibody, APA; anticardiolipin antibody, ACLA) are associated with miscarriages.

Diabetes is a medical condition that is affected by pregnancy and, likewise, affects pregnancy. Diabetes can lead to miscarriages, birth defects, and stillbirths. Women with diabetes should have preconception counseling with a perinatologist. Birth defects can result from the variation in a woman's blood sugar level during the first eight to 12 weeks, which is the time period when the embryo is developing. Cardiac defects are not unusual in the babies of women with abnormal blood sugars during that time. Insulin requirements vary tremendously during pregnancy due to placenta hormones that may inhibit the action of insulin. A perinatologist who specializes in diabetes is well aware of what the pregnant woman needs in each trimester and usually recommends the use of an insulin pump for better control. Women with symptomatic cardiac disease face one of the biggest challenges in pregnancy.

Before the advent of perinatology training, women with medical problems such as chronic hypertension, diabetes, and epilepsy were advised to not get pregnant because they could die. With the advancement of technology, it is in the early 2000s possible for these women to have a baby with just a modicum of risk.

Prevention

Women who have health problems and start specific care before conception have the best chance of a healthy pregnancy. A pre-pregnancy visit with a healthcare provider is, therefore, of the utmost importance for a woman with a medical problem. Together, the perinatologist and the woman can start therapies that will improve the woman's health prior to conception. There may be medications that are safer to take during pregnancy, and the physician can discuss how other women with a specific condition fare during pregnancy. For some diseases, pregnancy can mean increased risk of health problems for mother and baby. In fact, with lupus, preconception counseling is essential to determine the optimum time period for getting pregnant, which is when the disease is in remission. The bottom line is that a woman must always weigh the risks to herself and the baby when deciding whether or not to become pregnant and she can only do this by becoming informed.

See also Amniocentesis; Cesarean section; Electronic fetal monitoring.

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American College of Obstetricians and Gynecologists. 409 12th Street, SW, PO Box 96920, Washington, DC 20090. Web site: <www.acog.org>.

Association of Women's Health, Obstetric and Neonatal Nursing. 2000 L Street, NW Suite 740, Washington, DC 20036. Web site: <www.awhonn.org.

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Hip dysplasia, congenital *see* **Congenital hip dysplasia**

Hirschsprung's disease

Definition

Hirschsprung's disease, also known as congenital megacolon or aganglionic megacolon, is an abnormality in which certain nerve fibers are absent in segments of the bowel, resulting in severe bowel obstruction.

Description

Hirschsprung's disease is caused when certain nerve cells (called parasympathetic ganglion cells) in the wall of the large intestine (colon) do not develop before birth. Without these nerves, the affected segment of the colon lacks the ability to relax and move bowel contents along. This causes a constriction and as a result, the bowel above the constricted area dilates due to stool becoming trapped, producing megacolon (enlargement of the colon). The disease can affect varying lengths of bowel segment, most often involving the region around the rectum. In up to 10 percent of children, however, the entire colon and part of the small intestine are involved.

Demographics

Hirschsprung's disease occurs once in every 5,000 live births, and it is about four times more common in males than females. Between 4 percent and 50 percent of siblings are also afflicted. The wide range for recurrence is due to the fact that the recurrence risk depends on the gender of the affected individual in the **family** (i.e., if a female is affected, the recurrence risk is higher) and the length of the aganglionic segment of the colon (i.e., the longer the segment that is affected, the higher the recurrence risk).

Causes and symptoms

Hirschsprung's disease occurs early in fetal development when, for unknown reasons, there is either failure of nerve cell development, failure of nerve cell migration, or arrest in nerve cell development in a segment of bowel. The absence of these nerve fibers, which help control the movement of bowel contents, is what results in intestinal obstruction accompanied by other symptoms.

There is a genetic basis to Hirschsprung's disease, and it is believed that it may be caused by different genetic factors in different subsets of families. Proof that genetic factors contribute to Hirschsprung's disease is that it is known to run in families, and it has been seen in association with some chromosome abnormalities. For example, about 10 percent of children with the disease have **Down syndrome** (the most common chromosome abnormality). Molecular diagnostic techniques have identified many genes that cause susceptibility to Hirschsprung's disease. As of the early 2000s, there are a total of six genes: the RET gene, the glial cell line-derived neurotrophic factor gene, the endothelin-B receptor gene, endothelin converting enzyme, the endothelin-3 gene, and the Sry-related transcription factor SOX10. Mutations that inactivate the RET gene are the most frequent, occurring in 50 percent of familial cases (cases which run in families) and 15 to 20 percent of sporadic (non-familial) cases. Mutations in these genes do not cause the disease, but they make the chance of developing it more likely. Mutations in other genes or environmental factors are required to develop the disease, and these other factors are not understood. Also, among children with Hirschsprung's disease, some 2–5 percent have cardiac defects.

For persons with a ganglion growth beyond the sigmoid segment of the colon, the inheritance pattern is autosomal dominant with reduced penetrance (risk closer to 50 percent). For persons with smaller segments involved, the inheritance pattern is multifactorial (caused by an interaction of more than one gene and environmental factors, risk lower than 50 percent) or autosomal recessive (one disease gene inherited from each parent, risk closer to 25 percent) with low penetrance.

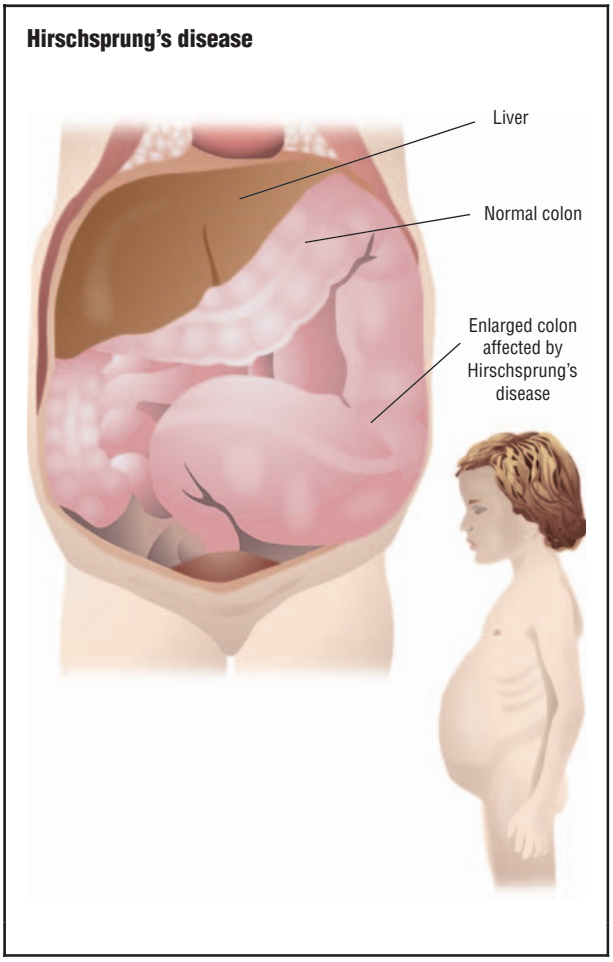
The initial symptom is usually severe, continuous **constipation**. A newborn may fail to pass meconium (the first stool) within 24 hours of birth, may repeatedly vomit yellow or green colored bile, and may have a distended (swollen, uncomfortable) abdomen. Occasionally, infants may have only mild or intermittent constipation, often with **diarrhea**.

While two-thirds of cases are diagnosed in the first three months of life, Hirschsprung's disease may also be diagnosed later in infancy or childhood. Occasionally, even adults are diagnosed with a variation of the disease. In older infants, symptoms and signs may include anorexia (lack of appetite or inability to eat), lack of the urge to move the bowels or empty the rectum on physical examination, distended abdomen, and a mass in the colon that can be felt by the physician during examination. It should be suspected in older children with abnormal bowel habits, especially a history of constipation dating back to infancy and ribbon-like stools.

Occasionally, the presenting symptom may be a severe intestinal infection called enterocolitis, which is life threatening. The symptoms are usually explosive, watery stools and **fever** in a very ill-appearing infant. It is important to diagnose the condition before the intestinal obstruction causes an overgrowth of bacteria that evolves into a medical emergency. Enterocolitis can lead to severe diarrhea and massive fluid loss, which can cause death from **dehydration** unless surgery is done immediately to relieve the obstruction.

Diagnosis

Hirschsprung's disease in the newborn must be distinguished from other causes of intestinal obstruction. The



In Hirschsprung's disease, the flow of contents through the large intestine is halted, causing some areas to enlarge greatly. This causes many symptoms in the patient, including a distended abdomen. *(Illustration by GGS Information Services.)*

KEY TERMS

Anus—The opening at the end of the intestine through which solid waste (stool) passes as it leaves the body.

Barium enema—An x-ray procedure that involves the administration of barium into the intestines by a tube inserted into the rectum. Barium is a chalky substance that enhances the visualization of the gastrointestinal tract on x ray.

Colostomy—A surgical procedure in which an opening is made in the wall of the abdomen to allow a part of the large intestine (the colon) to empty outside the body. Colostomies are usually required because portions of the intestine have been removed or an intestinal obstruction exists.

Enterocolitis—Severe inflammation of the intestines that affects the intestinal lining, muscle, nerves and blood vessels.

Manometry—A technique for measuring changes in pressure.

Meconium—A greenish fecal material that forms the first bowel movement of an infant.

Megacolon—Abnormal dilation of the colon.

Parasympathetic ganglion cell—Type of nerve cell normally found in the wall of the colon.

diagnosis is suspected by the child's medical history and physical examination, especially the rectal exam. The diagnosis is confirmed by a barium enema x ray, which shows a picture of the bowel. The x ray will indicate if a segment of bowel is constricted, causing dilation and obstruction. A biopsy of rectal tissue will reveal the absence of the nerve fibers. Adults may also undergo manometry, a balloon study (device used to enlarge the anus for the procedure) of internal anal sphincter pressure and relaxation.

Treatment

Hirschsprung's disease is treated surgically. The goal is to remove the diseased, nonfunctioning segment of the bowel and restore bowel function. This is often done in two stages. The first stage relieves the intestinal obstruction by performing a colostomy. This procedure creates an opening in the abdomen (stoma) through which bowel contents can be discharged into a waste bag. When the child's weight, age, or condition is deemed appropriate, surgeons close the stoma, remove the diseased portion of bowel, and perform a pull-through procedure, which repairs the colon by connecting functional bowel to the anus. This usually establishes fairly normal bowel function.

Prognosis

Overall, prognosis is very good. Most infants with Hirschsprung's disease achieve good bowel control after surgery, but a small percentage of children may have lingering problems with soilage or constipation. These infants are also at higher risk for an overgrowth of bacteria in the intestines, including subsequent episodes of enterocolitis, and should be closely followed by a physician. Mortality from enterocolitis or surgical complications in infancy is 20 percent.

Prevention

Hirschsprung's disease is a congenital abnormality that has no known means of prevention. It is important to diagnose the condition early in order to prevent the development of enterocolitis. Genetic counseling can be offered to a couple with a previous child with the disease or to an affected individual considering pregnancy to discuss recurrence risks and treatment options. Prenatal diagnosis was not available as of 2004.

Parental concerns

Parents should understand that toilet teaching may be delayed in children who have had surgery for Hirschsprung's disease. Children who have had surgical correction of Hirschsprung's disease are also at a higher risk for constipation and dehydration. Increased fluid and fiber intake are usually sufficient to improve these problems.

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Histiocytosis X

Definition

Histiocytosis X is a generic term that refers to an increase in the number of histiocytes, a type of white blood cell that acts as a scavenger to remove foreign material from the blood and tissues. Research that demonstrated Langerhans cell involvement as well as histiocytes led to a proposal that the term Langerhans cell histiocytosis (LCH) be used in place of histiocytosis X. Either term refers to three separate illnesses (listed in order of increasing severity): eosinophilic granuloma, Hand-Schuller-Christian disease, and Letterer-Siwe disease.

Description

Epidermal (skin) Langerhans cells (a form of dendritic cell) accumulate with other immune cells in various parts of the body and cause damage by the release of chemicals. Normally, Langerhans cells recognize foreign material, including bacteria, and stimulate the immune system to react to them. Langerhans cells are usually found in skin, lymph nodes, lungs, and the gastrointestinal tract. Under abnormal conditions these cells affect skin, bone, and the pituitary gland as well as the lungs, intestines, liver, spleen, bone marrow, and brain. Therefore, the disease is not confined to areas where Langerhans cells are normally found. The disease is more common in children than adults and tends to be most severe in very young children.

Histiocytosis X or LCH is a family of related conditions, which are characterized by a distinct inflammatory and proliferative process but which differ from each other regarding what parts of the body are involved. The least severe of the histiocytosis X/LCH family is eosinophilic granuloma. Approximately 60 to 80 percent of all diagnosed cases are in this classification, which usually occurs in children aged five to ten years. The bones are involved 50 to 75 percent of the time, which includes the skull, or mandible, and the long bones. If the bone marrow is involved, anemia can result. With skull involvement, growths can occur behind the eyes, bulging them forward. The lungs are involved less than 10 percent of the time, and this involvement signals the worst prognosis.

Next in severity is Hand-Schuller-Christian disease, a chronic, scattered form of histiocytosis. It occurs most commonly from the age of one to three years and is a slowly progressive disease that affects the softened areas of the skull, other flat bones, the eyes, and skin. Letterer-Siwe disease is the acute form of this series of diseases. It is generally found from the time of birth to one year of age. It causes an enlarged liver, bruising and skin lesions, anemia, enlarged lymph glands, other organ involvement, and extensive skull lesions.

KEY TERMS

Anemia—A condition in which there is an abnormally low number of red blood cells in the bloodstream. It may be due to loss of blood, an increase in red blood cell destruction, or a decrease in red blood cell production. Major symptoms are paleness, shortness of breath, unusually fast or strong heart beats, and tiredness.

Biopsy—The surgical removal and microscopic examination of living tissue for diagnostic purposes or to follow the course of a disease. Most commonly the term refers to the collection and analysis of tissue from a suspected tumor to establish malignancy.

Computed tomography (CT)—An imaging technique in which cross-sectional x rays of the body are compiled to create a three-dimensional image of the body's internal structures; also called computed axial tomography.

Cytokines—Chemicals made by the cells that act on other cells to stimulate or inhibit their function. They are important controllers of immune functions.

Dendritic—Branched like a tree.

Eosinophil—A type of white blood cell containing granules that can be stained by eosin (a chemical that produces a red stain). Eosinophils increase in response to parasitic infections and allergic reactions.

Epidermal—The outermost layer of the skin.

Inflammatory—Pertaining to inflammation.

Magnetic resonance imaging (MRI)—An imaging technique that uses a large circular magnet and radio waves to generate signals from atoms in the body. These signals are used to construct detailed images of internal body structures and organs, including the brain.

Pituitary gland—The most important of the endocrine glands (glands that release hormones directly into the bloodstream), the pituitary is located at the base of the brain. Sometimes referred to as the "master gland," it regulates and controls the activities of other endocrine glands and many body processes including growth and reproductive function. Also called the hypophysis.

Prostaglandins—A group of hormone-like molecules that exert local effects on a variety of processes including fluid balance, blood flow, and gastrointestinal function. They may be responsible for the production of some types of pain and inflammation.

Serous—Pertaining to or resembling serum.

Demographics

Histiocytosis X is a rare disorder affecting only approximately one in 200,000 children or adults each year.

Causes and symptoms

Because histiocytosis X is so rare, little research has been done to determine the cause. Over time, it may lessen in its assault on the body, but there are still problems from damage to the tissues. There are no apparent inheritance patterns in these diseases with the exception of a form involving the lymphatic system.

The symptoms of histiocytosis are caused by substances called cytokines and prostaglandins, which are normally produced by histiocytes and act as messengers between cells. When these chemicals are produced in excess amounts and in the wrong places, they cause tissue swelling and abnormal growth. Thus, symptoms may include painful lumps in the skull and limbs as well as **rashes** on the skin. General symptoms may include: poor appetite, failure to gain weight, recurrent **fever**, and irritability. Symptoms from other possible sites of involvement include:

- gums: swelling, usually without significant discomfort
- ears: chronic discharge
- liver or spleen: abdominal discomfort or swelling
- pituitary gland: affected at some stage in approximately 20 percent to 30 percent of children, causing a disturbance in water balance and producing thirst and frequent urination
- eyes: behind-the-eye bulging may occur (exophthalmos)
- lungs: breathing problems

Diagnosis

The diagnosis can only be made by performing a biopsy (taking a tissue sample under anesthesia from a site in the patient thought to be involved and having it tests). Blood and urine tests, chest and other **x rays**, **magnetic resonance imaging** (MRI) and **computed tomography** scans (CT scans) (to check the extent of involve-

ment), and possibly bone marrow or breathing tests may be required to confirm the diagnosis.

Treatment

Although this disease is not **cancer**, most patients are treated in cancer clinics. There are two reasons for this. Historically, cancer specialists treated it before the cause was known. Moreover, the treatment requires the use of drugs typically required to treat cancer.

Any cancer drugs utilized are usually given in smaller doses, which diminishes the severity of their side effects. Radiation therapy is rarely used, and special drugs may be prescribed for skin symptoms. If there is only one organ affected, steroids may be injected locally, or a drug called indomethacin may be used. Indomethacin is an anti-inflammatory medication that may achieve a similar response with less severe side effects.

Prognosis

The disease fluctuates markedly. If only one system is involved, the disease often resolves by itself. Multisystem disease usually needs treatment, although it may disappear spontaneously. The disease is not normally fatal unless organs vital to life are damaged. In general, the younger the child at diagnosis and the more organs involved, the poorer the outlook. If the condition resolves, there can still be long-term complications because of the damage done while the disease was active.

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Hitting *see* **Aggressive behavior**

HIV infection and AIDS

Definition

Human **immunodeficiency** virus (HIV) is a retrovirus that causes acquired immune deficiency syndrome (AIDS) by infecting helper T cells of the immune system. The most common serotype, HIV-1, is distributed worldwide, while HIV-2 is primarily confined to West Africa. AIDS is a severe immunological disorder caused by the retrovirus HIV, resulting in a defect in cell-mediated immune response that is manifested by increased susceptibility to opportunistic infections and to certain rare cancers, especially Kaposi's sarcoma. It is transmitted primarily by exposure to contaminated body fluids, especially blood and semen. Everybody who has AIDS also has HIV disease, but not everybody with HIV disease is classified by the United States (U.S.) government as having AIDS. The U.S. government uses CD4 cell counts (part of the immune system) to make this distinction.

Description

The earliest known case of HIV-1 came from a human blood sample collected in 1959 from a man in Kinshasa, Democratic Republic of Congo. The method by which he became infected is not known; however, genetic analysis of his blood sample suggested that HIV-1 might have stemmed from a single virus in the late 1940s or early 1950s. HIV has existed in the United States since the mid to late 1970s. During 1979 to 1981, rare types of **pneumonia**, **cancer**, and other illnesses were reported by physicians in Los Angeles and New York among a number of male patients who had sex with other men. Since it is rare to find these diseases in people with a healthy immune system, public health representatives became concerned that a new virus was emerging.

In 1982, the term AIDS was introduced to describe the occurrences of opportunistic infections, Kaposi sarcoma, and *Pneumocystis carinii* pneumonia in previously healthy persons and formal tracking of these cases in the United States began that year. The virus that causes AIDS was discovered in 1983 and named human T-cell lymphotropic virus-type III/lymphadenopathy-associated virus (HTLV-III/LAV) by an international scientific committee who later changed it to HIV. Many theories as to the origins of HIV and how it appeared in the human population have been suggested. The majority of scientists believed that HIV originated in other primates and was somehow transmitted to man. In 1999, an international group reported the discovery of the origins of HIV-1, the predominant strain of HIV in the devel-

oped world. A subspecies of chimpanzees native to west equatorial Africa were identified as the original source of the virus. The researchers believe that HIV-1 was introduced into the human population when hunters became exposed to infected blood.

Most scientists believe that HIV causes AIDS by directly inducing the death of CD4+ T cells (helper T cells in the immune system) or interfering with their normal function and by triggering other events that weaken a person's immune function. For example, the network of signaling molecules that normally regulates a person's immune response is disrupted during HIV disease, impairing a person's ability to fight other infections. The HIV-mediated destruction of the lymph nodes and related immunologic organs also plays a major role in causing the immunosuppression seen in persons with AIDS.

In the absence of antiretroviral therapy, the median time from HIV infection to the development of AIDS-related symptoms has been approximately 10 to 12 years. A wide variation in disease progression, however, has been noted. Approximately 10 percent of HIV-infected persons have progressed to AIDS within the first two to three years after infection, whereas up to 5 percent of persons have stable CD4+ T cell counts and no symptoms even after 12 or more years. Factors such as age or genetic differences among persons with HIV, the level of virulence of an individual strain of virus, and co-infection with other microbes may influence the rate and severity of disease progression. Drugs that fight the infections associated with AIDS have improved and prolonged the lives of HIV-infected persons by preventing or treating conditions such as *Journal of Infectious Diseases*. This approach is known formally as short-cycle structured intermittent antiretroviral therapy (SIT) or colloquially as the "7-7" approach. Dr. Mark Dybul, of the National Institute of Allergy and Infectious Diseases (NIAID) and the study author, noted that this approach together with high patient adherence could be a powerful and cost-effective tool in HIV treatment. This regimen uses half as much antiretroviral medication so not only are drug costs reduced but drug-related toxicities may be less in the long run. He believes that this is particularly important to countries with few resources around the world.

Nutritional concerns

Nutrition is definitely a concern for the individual who is HIV infected and even more so for the individual who has progressed to AIDS. The antiretroviral drugs have numerous side effects that make eating an adequate diet difficult and the disease itself affects nutritional intake. It is important for HIV individuals to supplement their diet with **vitamins** and **minerals** as well as protein drinks to maintain their energy. There are many supplements on the market and in health food stores that can be of benefit. The patient needs to go in search of what best suits their tastes.

Prognosis

The prognosis for individuals with AIDS in recent years has improved significantly because of new drugs and treatments, and educational and preventive efforts. Women whose HIV infections are detected early and receive appropriate treatment survive as long as infected men. There are several studies that have shown HIV-infected women to have shorter survival times than men. Women may be less likely than men to be diagnosed early, which may account for shorter survival times. In an analysis of several studies involving more than 4,500 people with HIV infection, women were one-third more likely than men to die within the study period. The investigators could not definitively identify the reasons for excess mortality among women in this study, but they speculated that poorer access to or use of health care resources among HIV-infected women as compared to men, domestic violence, homelessness, and lack of social supports for women may have been important factors.

Researchers have observed two general patterns of illness in HIV-infected children. About 20 percent of children develop serious disease in the first year of life; most of these children die by age four years. The remaining 80 percent of infected children have a slower rate of disease progression, many not developing the most serious symptoms of AIDS until school entry or even **adolescence**. A recent report from a large European registry of HIV-infected children indicated that half of the children with perinatally acquired HIV disease were alive at age nine. Another study, of 42 perinatally HIV-infected children who survived beyond nine years of age, found about one-quarter of the children to be asymptomatic with relatively intact immune systems.

Prevention

Because no vaccine for HIV is available, the only way to prevent infection by the virus is to avoid behaviors that put a person at risk of infection, such as sharing needles and having unprotected sex. Many people infected with HIV have no symptoms; therefore, there is no way of knowing with certainty whether a sexual partner is infected unless he or she has repeatedly tested negative for the virus and has not engaged in any risky behavior. Individuals should either abstain from having

KEY TERMS

B-cell lymphomas—Non-Hodgkin's lymphomas that arise from B cells.

CD4+ cells—Called helper T-cells, these cells work in cell-mediated immunity by causing a form of inflammation to wall off and destroy foreign material as with a bacterial infection.

Co-infection—Concurrent infection of a cell or organism with two microorganisms (pneumonia caused by coinfection with a cytomegalovirus and streptococcus).

Immunosuppression—Techniques used to prevent transplant graft rejection by the recipient's immune system.

Kaposi's sarcoma—A cancer characterized by bluish-red nodules on the skin, usually on the lower extremities, that often occurs in people with AIDS.

Lymphadenopathy—A disorder characterized by local or generalized enlargement of the lymph nodes or lymphatic vessels.

Perinatal—Referring to the period of time surrounding an infant's birth, from the last two months of pregnancy through the first 28 days of life.

Pneumocystis carinii—A parasite transitional between a fungus and protozoan, frequently occurring as aggregate forms existing within rounded cystlike structures. It is the causative agent of pneumocystosis.

Retrovirus—A family of RNA viruses containing a reverse transcriptase enzyme that allows the viruses' genetic information to become part of the genetic information of the host cell upon replication. Human immunodeficiency virus (HIV) is a retrovirus.

T cell—A type of white blood cell that is produced in the bone marrow and matured in the thymus gland. It helps to regulate the immune system's response to infections or malignancy.

sex or use male latex condoms or female polyurethane condoms, which may offer partial protection, during oral, anal, or vaginal sex. Only water-based lubricants should be used with male latex condoms. Although some laboratory evidence shows that spermicides can kill HIV, researchers have not found that these products can prevent a person from getting HIV.

The risk of HIV transmission from a pregnant woman to her baby can be significantly reduced with the use of antiretroviral drugs taken during pregnancy, labor, and delivery and administered to the baby for the first six weeks of life. In addition, the International Perinatal HIV Group reported in 1999 that elective **cesarean section** delivery could help reduce vertical transmission of HIV, although it is not without risk to certain women.

Parental concerns

Parental concerns are reflected in the status of a reproductive couple to prevent the transmission of the virus before pregnancy and if this is not possible, to obtain adequate prenatal care to prevent the transmission to the baby.

See also High-risk pregnancy.

Resources

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Hives

Definition

Hives is an allergic skin reaction causing localized redness, swelling, and **itching**.

Description

Hives is a reaction of the body's immune system that causes areas of the skin to swell, itch, and become reddened (wheals). When the reaction is limited to small areas of the skin, it is called urticaria. Involvement of larger areas, such as whole sections of a limb, is called angioedema.

Demographics

Many children and adults experience hives at various times during their lives. As hives is not a reportable event, no accurate prevalence statistics are available.

Causes and symptoms

Hives is an allergic reaction. The body's immune system is normally responsible for protection from foreign invaders. When it becomes sensitized to normally harmless substances, the resulting reaction is called an allergy. An attack of hives is set off when such a substance, called an allergen, is ingested, inhaled, or otherwise contacted. It interacts with immune cells called mast cells, which reside in the skin, airways, and digestive system. When mast cells encounter an allergen, they release histamine and other chemicals, both locally and into the bloodstream. These chemicals cause blood vessels to become more porous, allowing fluid to accumulate in tissue and leading to the swollen and reddish appearance of hives. Some of the chemicals released sensitize **pain** nerve endings, causing the affected area to become itchy and sensitive.

A wide variety of substances may cause hives in sensitive people, including foods, drugs, and insect **bites** or **stings**. Common culprits include:

- nuts, especially peanuts, walnuts, and Brazil nuts
- fish, mollusks, and shellfish
- eggs
- wheat
- milk
- strawberries
- food additives and preservatives
- penicillin or other antibiotics
- flu vaccines
- tetanus toxoid vaccine
- gamma globulin
- bee, wasp, and hornet stings
- bites of mosquitoes, fleas, and scabies

Urticaria is characterized by redness, swelling, and itching of small areas of the skin. These patches usually grow and recede in less than a day but may be replaced by others in other locations. Angioedema is characterized by more diffuse swelling. Swelling of the airways may cause wheezing and respiratory distress. In severe cases, airway obstruction may occur.

When to call the doctor

A doctor or other healthcare professional should be called when hives do not spontaneously clear within a day of their appearance or when they include swelling of the throat. If the reactions are severe, as in anaphylactic reaction or shock, immediate medical care is needed.

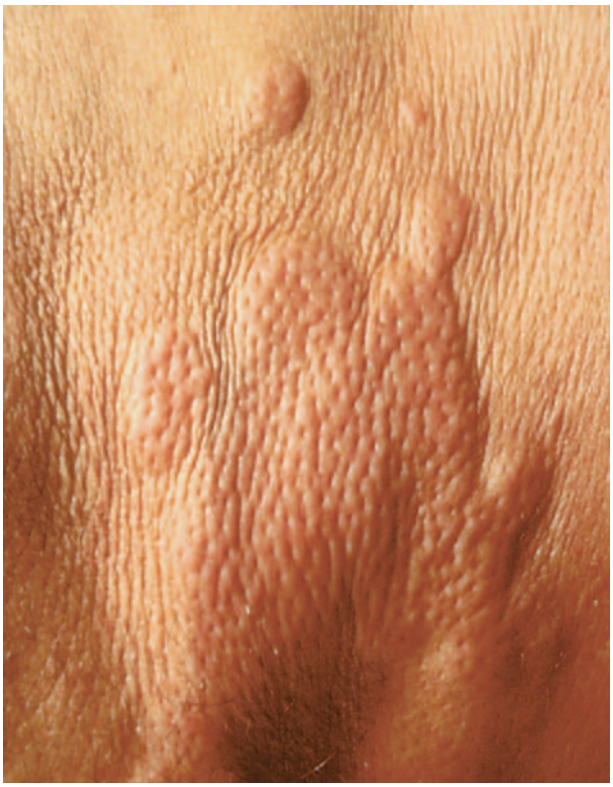
Diagnosis

Hives are easily diagnosed by visual inspection. The cause of hives is usually apparent but may require a careful medical history in some cases.

Treatment

Mild cases of hives are treated with **antihistamines**, such as diphenhydramine (Benadryl). More severe cases may require oral corticosteroids, such as prednisone. Topical corticosteroids are not effective. Airway swelling may require emergency injection of epinephrine (adrenaline).

An alternative practitioner will try to determine what allergic substance is causing the reaction and help the person eliminate or minimize its effects. To deal with the symptoms of hives, an oatmeal bath may help to relieve itching. Chickweed (*Stellaria media*), applied as a poultice (crushed or chopped herbs applied directly to the skin) or added to bath water, may also help relieve itching. Several homeopathic remedies, including *Urtica urens* and *Apis* (*Apis mellifica*), may help relieve the itch, redness, or swelling associated with hives.



Hives on the back of a young woman's legs. The accompanying inflammation develops as an allergic reaction which ranges in size from small spots to patches measuring several inches across. *(© 1994 Caliendo/Custom Medical Stock Photo, Inc.)*

Prognosis

Most cases of hives clear up within one to seven days without treatment, providing the cause (allergen) is found and avoided.

Prevention

Preventing hives depends on avoiding the allergen causing them. Analysis of new items in the diet or new drugs taken may reveal the likely source of the reaction. Chronic hives may be aggravated by stress, **caffeine**, alcohol, or tobacco; avoiding these may reduce the frequency of reactions.

Nutritional concerns

Hives may be triggered or worsened by caffeine or alcohol (in adults), or specific allergenic foods, which depend entirely on the patient. Avoiding these substances may reduce the occurrence of hives.

KEY TERMS

Allergen—A foreign substance that provokes an immune reaction or allergic response in some sensitive people but not in most others.

Wheal—A smooth, slightly elevated area on the body surface that is redder or paler than the surrounding skin.

Parental concerns

Parents should monitor their children to ensure that any attack of hives does not involve the throat area. Young children should be encouraged not to stratch their skin too vigorously. If hives are a recurrent problem, parents should keep track of the foods the child eats in an attempt to discover the allergen.

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American Academy of Pediatrics. 141 Northwest Point Blvd., Elk Grove Village, IL 60007–1098. Web site: <www.aap.org/>.

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Home schooling

Definition

Home schooling is the process of educating school-aged children at home rather than at a school. As of the early 2000s, it is perhaps one of the fastest growing trends in education in the United States. Since 1993, the practice has been legal in all 50 states. About 1.1 million students were being home-schooled in the spring of 2003, according to the National Household Education Surveys Program (NHES), which was conducted by the United States Department of Education. In addition, the percentage of the school-age population that was being home-schooled increased from 1.7 percent in 1999 to 2.2 percent in 2003. Parents choose to home-school their children for a variety of reasons, though certain factors appear to be more prevalent than others. Nearly two-thirds of the parents of home-schooled students reported that their primary reason for home schooling was either concern regarding the environment of schools or a wish to provide moral or religious instruction.

Description

Societies have practiced home schooling for centuries. In North America, home schooling was widespread until the 1870s, when compulsory school attendance laws and the development of professional educators came together to institutionalize education in the form recognized in the early 2000s as the school. Some pre-eminent historical figures who were home-schooled include several presidents, such as George Washington, John Quincy Adams, Abraham Lincoln, and Franklin Delano Roosevelt. Other home-schooling successes in American history include Thomas Edison, General Robert E. Lee, Booker T. Washington, and Mark Twain.

Although home schooling was practiced in a limited way after the 1870s, it was not until the 1960s that this practice claimed attention from a large number of parents and educators. The writings of Raymond Moore, a former U.S. Department of Education official, and John Holt, author of several books on education, gave credence and national presence to a growing home school movement. Moore began researching the institutionalization of children's education and concluded that a child's first foray into formal education should not begin until sometime between eight and 12 years of age. Holt advocated the decentralization of schools and a greater degree of parental involvement. He believed that the most civilized way to educate a child was through home schooling.

Prior to 1993, when home schooling became legal in all states, many parents who taught their children at home often faced arrest and jail time, amidst accusations of neglect and abuse. Most of that changed over the following decade. Even so, attitudes about home schooling vary widely from state to state, and there is a patchwork of regulation across the country. Some states may require a state-approved curriculum, conduct home visits periodically, and require that home-schooling parents be certified teachers. Others may not require a parent to have any contact with the state and have no minimum educational standards for the home-schooling parent.

Despite greater acceptance, home schooling has its critics, such as the National Education Association (NEA). This organization sees the **safety** of children and the economics of public schools as potential home school problems. They cite a few well-publicized incidences of abuse and state a **fear** that in states where there is no accountability of the home-schooling parents to the government, some children may be placed at higher risk for

A mother home schools her children. *(© Ed Kashi/Corbis.)*

abuse, neglect, and other problems. The NEA is also concerned that home schooling will eventually lead to a diversion of funding from the public schools.

Characteristics of home schoolers

In the 1960s and 1970s, most home-schooling parents were members of the counter-cultural left. By the 1980s, however, most home-schooling parents were part of what is often called the Christian Right. In the early 2000s, approximately 75 percent of American home schoolers are practicing Christians. However, not all home-schooling parents are Christians. The rise in home schooling is reaching a much broader range of families. For example, the fastest growing number of practicing home schoolers is among Muslim Americans. Some surveys show that the average home-schooling **family** has an above average income. Others indicate that the household income of home schoolers is very similar to that of non-home-schooling families. Most home-schooling families have above-average levels of education. One important factor is that home-schooling families are 97-percent two-parent families, and most home-schooling mothers do not work outside the home. The average size of a home-schooling family is three children or more.

Reasons parents choose homeschooling

The decision to home-school is not based solely on conservative religious or political views. Although parents homeschool for a variety of reasons, the primary reason is dissatisfaction with public education. Other reasons stated by home-schooling parents include the following:

- the opportunity to impart a certain set of beliefs and morals
- higher academic performance through one-on-one instruction
- the ability to develop stronger parent-child relationships
- the lack of **discipline** in public schools
- the opportunity to escape negative **peer pressure** through more controlled interactions with a student's peers
- an inability to pay private school tuition

• a physically safer environment in which to learn

Home schooling involves a tremendous commitment from the parents. At least one parent must be willing to work closely with the child, develop lesson plans, keep current with government requirements, and sometimes negotiate issues with the local school district. The most common home-schooling arrangement is for the mother to teach while the father works outside the home. There are numerous educational materials available that are geared for home-schooled children. These include correspondence courses, full curricula, and single topic books in areas such as math or phonics. There are both religious and non-religious publishers of these materials. Some parents do not use these materials and develop individualized lessons based on their children's unique learning needs.

Performance of home-schooled students

One of the questions many people have is how home-schooled children perform academically. According to the U.S. Department of Education, virtually all of the data available illustrate that home-schooled students perform at an above average level on a variety of tests, including the Scholastic Aptitude Test (SAT). Interestingly, one study found that students whose parents are certified teachers performed no better than other students and that neither parental income nor parents' educational background had a significant impact on student performance. In the late 1990s and early 2000s, home-schooled students have gained admission and scholarships to such prestigious universities as Harvard, Yale, Stanford, and MIT. In 2000, Patrick Henry College opened, a university established especially for home-schooled children.

Common problems

One disadvantage to home schooling is the loss of an income in a family, since many families make the decision to live on a single income so that one parent can devote time to educating the children. Some home-schooling families find the practice of home-schooling confining. It takes a great deal of dedication and preparation for instruction and schoolwork. One of the most often voiced concerns is that children who are home schooled are not properly socialized. However, there are numerous opportunities for home-schooled students to interact with others, including libraries, scouting, 4-H, **sports** teams, and a variety of church activities. In addition, many local communities have formed home-schooling associations in which children have many outlets for interacting with their peers.

Parental concerns

Parents interested in teaching their children at home should thoroughly research what is involved before making the decision to do so. They need to be informed regarding the laws in their state and local school district, which may affect their decision.

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Homosexuality and bisexuality

Definition

Homosexuality is the consistent sexual and emotional attraction, including fantasy, interest, and arousal to a person of the same sex. Bisexuality is the sexual and emotional attraction to members of both sexes.

Description

References to homosexuality and bisexuality can be found in recorded history and literature dating back thousands of years. They are part of a trio of classifications referred to collectively as sexual orientation. The third is heterosexuality, the sexual and emotional attraction to members of the opposite sex. Both male and female homosexuals are commonly referred to as gay while homosexual females are called lesbians.

The earliest documentation of homosexuality in Western civilization occurs in ancient Greece, where same-sex relationships were considered normal by society. Although there were some homosexual relationships between adult males, most were between men and boys. Although there is some disagreement among historians, a number of historical figures were believed to be gay, including Alexander the Great (356 B.C.–323 B.C.), Plato (20 B.C.–45 A.D.) Virgil (Vergil) (70 B.C.–19 B.C.), Leonardo da Vinci (1452–1519), and Michelangelo (1475–1564). Homosexuality in Asian, especially Japanese and Chinese, cultures has been documented since at least 600 B.C.

Social attitudes towards homosexuality and bisexuality have varied over the centuries, from complete rejection, or homophobia, through covert acceptance, to complete normalization, with many degrees in between. The religious response to homosexuality varies, though in the three major Western religions (Judaism, Christianity, and Islam) homosexuality and bisexuality are considered sins.

In some cultures, especially those influenced by homophobic religions, homosexuality is considered a perversion and has been outlawed; in some jurisdictions homosexual behavior is a crime punishable by death. Persecution of homosexuals in such cultures is common. In Nazi Germany, homosexuals and bisexuals were sent to concentration camps and were murdered in gas chambers along with other minority and religious groups.

The modern gay rights movement began in the late 1960s and included the development of the often activist academic treatment of sexuality in colleges and universities. This focus led to changes in social acceptance and in the media portrayal of homosexuality and bisexuality. In 1973, the American Psychiatric Association (APA) removed homosexuality from its list of mental disorders. The legalization of same-sex marriage and non-gender-specific civil unions is one of the major goals of gay rights activism. Toward the end of the 1990s and into the early 2000s, a number of jurisdictions relaxed or eliminated laws curbing homosexual behavior, including sodomy laws and laws preventing homosexuals from serving in armed forces. This trend culminated on June 26, 2003, with the landmark U.S. Supreme Court decision *Lawrence v. Texas* which overturned all sodomy laws in the United States.

In 2003, Canada legalized same-sex marriages, according the same rights to gay married couples as to heterosexual married couples. Gay marriage is also legal in The Netherlands and Belgium. In 2004, due to several local and state actions, gay marriages were legalized in San Francisco; Massachusetts; Portland, Oregon; and several other areas. They were as of 2004 all under legal challenge, and the California Supreme Court nullified the San Francisco gay marriages in mid-2004. The Defense of Marriage Act, signed by President Bill Clinton in 1996, prevents federal recognition of same-sex marriage and allows states to ignore same-sex licenses from outside their borders.

The correct term or terms to use when referring to homosexuals varies widely by location and culture. In the United States and Europe, even the use of the word homosexual can be seen as insulting. In Washington state, The Safe Schools Coalition of Washington's *Glossary for School Employees* advises that *gay* is the preferred synonym for homosexual and goes on to advise avoiding the term homosexual, because it is clinical, distancing, and archaic.

The causes of homosexuality and bisexuality are unknown, although there are many controversial theories. These include genetic, biological, psychological, and social factors, as well as conscious choice. A majority of researchers believe sexual orientation is most likely determined by a combination of factors. Since about the 1970s, researchers have tended to rule out conscious choice. The reason can be answered in a question: Why would anyone choose a lifestyle which may well bring them discrimination, hatred, and even violence?

Much research suggests sexual orientation is set in early childhood. In surveys of gay men and lesbians, most say they believe they were born that way. This awareness usually occurs during **puberty** but sometimes earlier. Many experts believe sexual orientation, whether homosexual, bisexual, or heterosexual, is determined by a complex interaction between anatomical and hormonal influences during fetal development.

There is also no definitive research on the percentage of the population that is homosexual or bisexual. Studies in the 1940s and 1950s by biologist and sex researcher Alfred C. Kinsey (1894–1956) found that 2 percent of women and 10 percent of men were exclusively homosexual and that 37 percent of men reported having at least one same-sex experience after **adoles-**

cence. The validity of this research, while often cited in scientific literature, is questionable, since most of the study subjects were over 30 years old, white, and not randomly selected.

School age

Several studies suggest that first sexual attraction, whether for homosexuals, bisexuals, or heterosexuals, begins in middle childhood at about age 10. At this time, the adrenal glands begin to produce sex steroids, which motivate sexual attraction as well as social and emotional behavior.

Development of sexual identity in middle childhood and early adolescence is a natural process but is more stressful for homosexual adolescents, according to the American School Health Association (ASHA). To avoid rejection and hostility, homosexual adolescents feel obliged to hide their sexual identities. Professionals generally agree that homosexual identity development usually occurs in stages, according to a March 2003 article in ASHA *Journal of School Health*.

The first stage is identified as ''sensitization'' or early awareness, where, around age 10, a child experiences same-sex attraction and feelings of being different than other children. The second stage is ''identity confusion,'' in which simple awareness is no longer ignored. Gay male and lesbian children usually try to hide their sexual identities because society encourages heterosexuality. This stage is usually resolved by denying or hiding homosexual feelings, repressing same-sex attraction, or taking on a homosexual identity.

The next stage is ''identity assumption,'' in which the person accepts their homosexuality but usually limits disclosure to others. The final stage is ''identity consolidation,'' also known as ''coming out,'' in which disclosure may be expanded and the homosexual identity may be incorporated into social activities.

After identifying themselves as homosexual or bisexual, adolescents face the often-difficult problem of deciding whom to tell that they are gay or bisexual. According to a 1999 report by Cornell University, the average coming-out age for a gay and lesbian young person in the United States is 14–15 years, significantly younger than the average age of 19–23 during the late 1970s and early 1980s, according to the advocacy group Tolerance.com. Confidence and openness about their sexual orientation at a younger age, however, almost invariably exposes young people to homophobia and abuse at an early age, the group states on its Web site (<www.tolerance.org>).

Common problems

In hiding their sexual identities, homosexual and bisexual adolescents deprive themselves and each other of positive role models. However, disclosure to **family** members may lead to pressure to change through psychological or religious ''conversion'' therapies, which the ASHA regards as ineffective. The ASHA and most other professional organizations say family support when an adolescent discloses that they are gay or bisexual is crucial to the child's mental and emotional health. Children and teens who reveal that they are gay or bisexual to non-supportive families are much more likely to become runaways and resort to prostitution for financial support.

Research shows inconsistencies regarding disclosure. To test how a family will react, a gay or bisexual child or teen will often first tell a sibling whom they feel they can trust and whom they believe is most likely to be supportive, most often a sister. Mothers are more often disclosed to than fathers because fathers tend to have a more negative reaction. Studies also show that parents react more negatively when a son tells them he is gay or bisexual than when a daughter reveals she is lesbian or bisexual.

Numerous studies show that gay and bisexual youth are at a higher risk of dropping out of school, of being kicked out of their homes, and becoming prostitutes, than their heterosexual peers. They also have a higher incidence of drug, alcohol, and tobacco use. Studies have also shown that gay and bisexual adolescents are two to seven times more likely to commit or attempt **suicide** compared to heterosexual children and teens. Other studies have found that 45 percent of gay males and 20 percent of lesbians were victims of verbal or physical abuse in middle and high school and were two to four times more likely to be threatened with a weapon compared to heterosexual students.

Parental concerns

Studies have shown that parents usually go through a series of stages when they learn a child is gay or bisexual. In the first stage, nearly all parents go through a grieving period after learning their child is gay or bisexual. The parents mourn the loss of what they assumes was their child's heterosexuality and ''traditional'' lifestyle, the lack of grandchildren and their role as potential grandparents, and the improbability of changing their child's sexual orientation.

Soon after disclosure, parents often experience **fear** and guilt and may deny their child is gay or bisexual. They may urge their child to change their sexual orienta-

tion or urge them to keep their sexuality secret. Also, parents often become angry and seek to blame someone for their child's sexual orientation, such as a gay teacher, a sexual abuser, or as often is the case with gay males, to blame the father for a lack of engaging the child in perceived masculine activities such as **sports**. During this anger stage, parents often threaten or abuse the child or try to force them to change. Any of these actions tends to drive a wedge between the parents and child and is the primary reason many gay and bisexual youth run away from home. Sometimes they are thrown out of the house by their parents and are forced to live on the streets, often turning to prostitution to survive.

The anger stage is usually followed by the bargaining stage, where parents try to get their child to change their sexual orientation, sometimes through God or religion, or through psychological intervention. In this stage, parents sometimes experience one or a combination of emotions, including shame, guilt, and depression.

The final stage is resolution, where the parents either accept or deny that their child is gay or bisexual, though studies show few fully accept it. Some families remain in denial indefinitely. Others ostracize the child through eviction from the home or family.

When to call the doctor

Gay and bisexual adolescents may need psychological help in dealing with their sexual orientation. The vast majority of experts say this counseling should be supportive and not seek to change the child's sexual orientation. Counseling that offers emotional support may be helpful for teens who are uncomfortable with their sexual orientation. Therapy may also help the adolescent adjust to personal, family, or school-related problems.

Therapy directed specifically at changing homosexual or bisexual orientation is not recommended and may be harmful for an unwilling teen, according to a behavioral health advisory issued in 2002 by the journal *Clinical Reference Systems*. It may create more confusion and **anxiety** by reinforcing negative thoughts and emotions with which the child is already struggling, the advisory states.

Signs that a child or teem may be gay or bisexual and is having problems dealing with it include social isolation, avoiding school, threats of **running away**, poorly developed dating skills, low **self-esteem**, self hatred, alcohol and/or drug abuse, harassment at school or home, feelings of inferiority, depression, threats of suicide, and eating disorders.

Advice for healthcare professionals

The American Academy of Pediatrics (AAP) has issued guidelines for pediatricians in dealing with gay and bisexual adolescents. An article in the June 2004 issue of the AAP journal *Pediatrics* states: "Pediatricians should be aware that some youths in their care may have concerns about their sexual orientation or that of siblings, friends, parents, relatives, or others. Health care professionals should provide factual, current, nonjudgmental information in a confidential manner."

The article states that pediatricians and other healthcare professionals should be attentive of various psychological difficulties, offer counseling or refer for counseling when necessary, and ensure that all sexually active youths receive a physical examination, immunizations, appropriate laboratory tests, and counseling about **sexually transmitted diseases**, and appropriate treatment if necessary.

The *Pediatrics* article also states: "Not all pediatricians may feel able to provide the type of care [necessary]. Any pediatrician who is unable to care for and counsel nonheterosexual youth should refer these patients to an appropriate colleague."

Most gay and bisexual youth seen by pediatricians and other healthcare providers will not raise the issue of sexual orientation on their own. Therefore, healthcare professionals should raise issues of sexual orientation and sexual behavior with all adolescent patients or refer them to a colleague who can these issues, according to the AAP. Pediatricians should also consider displaying posters and offering brochures that demonstrate support for gay and bisexual teens. The AAP publishes a brochure dealing with sexual orientation, "Gay, Lesbian, and Bisexual Teens: Facts for Teens and their Parents."

Advice for teachers, counselors, and other school employees

Because students who discover they are gay or bisexual often experience rejection, discrimination, isolation, and violence, it is important for teachers and administrators to be supportive and highly sensitive to the stress gay and bisexual youth feel, according to the American School Health Association. Schools are legally obligated to protect students from discrimination and harassment from other students, from teachers, and from all other school employees. In 1996, a federal appeals court ruled that school officials can be held liable under the Equal Protection Clause of the U.S. Constitution for not protecting gay and bisexual students from harassment and discrimination.

The non-profit group Parents and Friends of Lesbians and Gays (PFLAG) makes the following recommendations for all schools:

- have a harassment policy or student bill of rights that explicitly includes sexual orientation
- provide annual, mandatory training for all school employees about sexual orientation and on intervention against bullying of gay and bisexual students
- have a support group for gay, bisexual, and straight students
- have information on display and readily available in the library on gay and bisexual issues
- include gay and bisexual issues in the curriculum, including history, social studies, literature, political science, health, and arts

Resources

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ORGANIZATIONS

The Gay, Lesbian, Straight Education Network. 121 W. 27th St., Suite 804, New York, NY 10001. Web site: <www.glsen.org>.

Gay-Straight Alliance Network. 160 14th St., San Francisco, CA 94103. Web site: <www.gsanetwork.org>.

The Healthy Lesbian, Gay, and Bisexual Students Project. American Psychological Association Education Directorate, 750 First St. NE, Washington, DC 20002. Web site: <www.apa.org/ed/hlgb/>.

Parents, Families, and Friends of Lesbians and Gays. 1726 M St. NW, Suite 400, Washington, DC 20036. Web site: <www.pflag.org>.

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KEY TERMS

Adrenal glands—A pair of endocrine glands (glands that secrete hormones directly into the bloodstream) that are located on top of the kidneys. The outer tissue of the glands (cortex) produces several steroid hormones, while the inner tissue (medulla) produces the hormones epinephrine (adrenaline) and norepinephrine.

Coming out—The process by which gays and bisexuals become public or tell others about their sexual orientation.

Gay bashing—Physical or verbal violence directed against homosexuals.

Homophobia—An irrational hatred, disapproval, or fear of homosexuality and homosexuals.

Sexual orientation—The direction of somebody's sexual desire, toward people of the opposite sex (heterosexual or straight) or of the same sex (homosexual or gay), or of both sexes (bisexual).

Sodomy—Anal intercourse.

Transgender—Any person who feels their assigned gender does not completely or adequately reflect their internal gender, such as a biological male who perceives himself to be female.

Hospitalization

Definition

Hospitalization is admittance to the hospital as a patient.

Purpose

Patients are admitted to the hospital for a variety of reasons, including scheduled tests, procedures, or surgery; emergency medical treatment; administration of medication; or to stabilize or monitor an existing condition.

Description

Preparation

Because no one can predict when a child may face an emergency hospital stay, it is a good idea for all parents to spend some time talking to their children about hospitals. Even though the information presented here is geared toward a planned hospitalization, the communication tips will also prove helpful to parents when their child's hospitalization is emergent.

Parents should describe and explain, as honestly and thoroughly as possible, what will happen to the child in the hospital. Parents should tell their child as much of the truth as he or she can understand. A toy doctor kit can help prepare a child for the experience. There are children's books about hospitalization, written for all age levels, that parents can read to their child before the hospital stay. Parents need to reassure young children, with their limited concept of time, that the hospital stay will be temporary. They can plan a party afterward or read a storybook part way through and mark the place where it will be resumed once the child comes home.

For **preschool** children, explanations should be simple and concrete. It will not ease the child's anxiety to try to explain that he or she will undergo a series of tests or will spend three weeks in the hospital. Instead, the parent might indicate the part or parts of the body that are to be "fixed," using a doll or stuffed animal.

By the time children reach five or six years of age, they can understand hospitalization on a more sophisticated level. They will be familiar with some medical instruments and concepts, and better able to grasp the time frame involved. Children of this age may feel they are going to the hospital because they have done something wrong, and parents need to reassure their child that hospitalization is not a punishment.

Teens should be given an honest explanation of what to expect during their hospital stay. They should be included in the discussions about their care. They also should be encouraged to ask their health care providers questions.

Overall, the best reassurance parents can give children of any age is the promise that they will be there to help them through the experience, even if they cannot be physically present during the entire ordeal. Parents should encourage their child to ask questions and talk about their feelings.

LEARNING ABOUT THE HOSPITAL Many hospitals allow parents and children to tour the pediatric facilities before the hospital stay, further reassuring the child. Children may be shown rooms similar to that in which they will stay. The tour may include a visit to the unit's playroom, a chance to meet the nursing staff, and the opportunity to become familiar with some of the hospital equipment. It is best for parents to be present during these tours, so the child can see that they approve of the facilities and trust the care providers.

Most hospitals provide information to the parents in advance of a planned hospital stay. This information may include directions to the hospital, parking information, and other services available. Parents should take advantage of the services offered during their child's hospital stay, especially support groups or educational classes that provide more information about the child's condition.

For a planned hospital stay, the parents need to contact their insurance company, if insured, to determine if the hospital is covered by their insurance plan. Once a hospitalization date is confirmed, the parents are required to notify the insurance company. If the hospital admission was emergent, the parents should notify the insurance company as soon as possible. Parents also need to review the credentials of the health care providers and hospital, gather information about the hospital, including services offered and specific policies (especially the visitation, boarding, and rooming-in policies), schedule the hospital stay, take the child to complete pre-admission testing, and receive and follow all of the appropriate pre-admission instructions. If certain medications need to be discontinued before the hospital stay, the hospital staff will notify the parents or send a complete list of medications to avoid.

MAKING SURE THE CHILD IS HEALTHY It is important for the child to be as healthy as possible before a planned hospitalization. The child should eat healthy foods, and rest and **exercise** as normal, unless given other instructions. The child needs to get extra **sleep** before the hospitalization, since his or her normal sleep patterns will likely be disrupted during the hospital stay. If the child has a **fever**, **cough**, or cold, the parents should call the child's doctor to determine if the hospitalization should be delayed.

PACKING FOR THE HOSPITAL STAY The child should help the parent pack items for the hospital stay. It

is helpful to pack familiar pajamas, **toys**, games, a special **family** photo, and other belongings that will provide comfort. Personal items should be labeled with the child's name. Valuables should be left at home.

Children should not bring latex (rubber) balloons to the hospital, as they can be a serious **safety** hazard, as well as a health hazard for children with a latex allergy. Shiny, metallic balloons (Mylar) are usually permitted. Parents should check the hospital's policies before packing any electronic items, such as **video games** or hair dryers. Some items may cause interference with the hospital equipment. Also, parents should check the specific unit's policy for bringing fresh flowers or plants. In most cases, bringing food from home is not permitted since certain foods may be restricted and the child's specific dietary intake may need to be recorded.

The parents should bring a complete list of the child's medications, medical conditions, and any known **allergies**. The child's medications should remain at home; all necessary medications will be provided in the hospital.

SELECTING A FAMILY SPOKESPERSON Because of privacy regulations established by the Health Insurance Portability and Accountability Act (HIPAA), some hospitals require families to select one spokesperson to communicate with health care providers. The spokesperson helps maintain the patient's privacy and also improves communication with the health care providers. The family spokesperson should be responsible for communicating information about the child's health to outside family members. Families and friends who call the child's nursing unit will not be able to obtain information about the patient, due to privacy regulations.

Aftercare

Before the child leaves the hospital, the health care providers will review discharge instructions with the child and parents. These instructions include incision care, signs of infection or complications to watch for, information on when to call the doctor, medication guidelines, activity guidelines, dietary restrictions, information about when the child may return to work or school, a follow-up appointment schedule, and other specific instructions as applicable to the patient's condition. Follow-up appointments may be scheduled, and the necessary prescriptions will be given to the parents.

If the child weighs less than 40 lbs (18 kg) or is under four years of age, he or she is required by most state laws to ride home in a safety seat. Parents should remember to bring the car seat with them on the day of the child's hospital discharge.

If health care services will be needed at home (home care), they can be arranged by a social worker or the nursing staff.

Risks

The risks of hospitalization are related to the type of treatment or procedure the child will be having. Every procedure has risks, which the parents should discuss with the child's doctor and health care team. Parents should make sure they understand the potential risks of any procedure prior to signing the informed consent form.

Normal results

Hospital admission

For planned hospitalizations, the parents register their child at the hospital registration or admitting area. For emergency hospital admissions, the parents register their child in the emergency department. The parents are required to complete paperwork and show an insurance card, if insured. Often, a pre-registration process performed before a planned hospitalization makes the registration process on the day of admission run smoothly.

A health care provider will review an informed consent form with the parents, and they will be asked to sign it. Informed consent is an educational process between health care providers and patients or their guardians. Parents are encouraged to ask questions. Before signing the form, the parents should understand the nature and purpose of the child's hospital stay and medical treatment, the risks and benefits, and alternatives, including the option of not proceeding with the medical treatment. Signing the informed consent form indicates that the parents permit a treatment to be administered to their child.

Upon admission, an identification bracelet that includes the patient's name and doctor's name will be placed on the child's wrist. The child may wear his or her own pajamas, or a hospital gown, depending on the hospital and nursing unit's policy. A nurse usually consults with the parents to learn about the child's dietary restrictions or preferences. Daily menu choices are available.

Hospital room

Usually, the child will share a room with one or more other children, unless a private room was requested or is required for health reasons. A typical feature of children's hospitals or children's hospital units is a playroom where children can interact with others who are undergoing similar experiences.

Most non-intensive care wards allow parents to stay overnight, at least initially, either in a special nearby unit or on a cot or chair in the child's room.

Health care team

Children's units in many hospitals are staffed by at least some nurses who specialize in caring for infants and children and understand their special needs. A children's activities specialist, also called a child life specialist, is usually on staff. This specialist has a background in child development and therapeutic **play**.

A variety of physicians, specialists, nurses, and teachers may make up the child's health care team. Parents should make sure they know who is providing care for their child during the hospital stay.

Hospital routine

The child's daily hospital experience will likely vary each day. The first day may be consumed by tests to determine the proper course of treatment. The health care team will discuss with the parents the anticipated length of stay in the hospital, based on the child's diagnosis.

Some of the typical hospital routines may be as follows: the nurse checks the child's vital signs (blood pressure, temperature, and heart rate) several times throughout the day and night; the attending physician and medical team usually visit in the morning (medical rounds); medications are administered; tests are performed; and meals are provided three times a day. During the entire hospital stay, the child will be made as comfortable as possible. The health care team will provide the parents with as much information as possible about the child's condition, care, and treatment throughout the hospital stay. Parents are encouraged to ask questions.

Most hospitals have designated visiting hours. The hospital staff is usually accommodating to close family members who want to visit at other times. Each hospital has its own visitation policy that should be investigated before the child's hospitalization. Parents should inquire about sibling visitation guidelines. Usually, only the child's parent or guardian may stay in the room with the child after a certain time in the evening. In some cases, the health care team may ask visitors to wait outside the patient's room during a procedure or treatment.

Depending on the reason for the hospital stay, certain medications may be prescribed or restricted. The health care team will provide specific guidelines and provide all necessary medications during the child's hospital stay.

Many children's hospitals will assign a teacher to any child able to do some assignments. Social workers can help arrange home-schooling as needed after the child is discharged from the hospital.

Resources for families

Educational classes may be available for family members to learn more about the child's condition and what to expect during the child's recovery at home.

Most hospitals have on-site pharmacies where family members can fill the patient's prescriptions; gift shops; and a cafeteria. Usually a list of on-site and off-site dining options can be obtained from the hospital's information desk or social work department.

Parental concerns

Before they can reassure their children, parents need to deal with their own fears about the impending experience. It may be easy for parents to unintentionally communicate these fears to the child. The parents should learn all they can about their child's condition and about the hospital, the child's health care team, and available services.

Even with preparation and support, it is normal for children to experience certain fears when they are hospitalized. The most common **fear** is **separation anxiety**. The hospital is a frightening place full of unfamiliar sights, sounds, and people, and the child's primary source of security and reassurance is a parent. Many young children have never spent even a single night away from their parents, and if the parents leave for the night, especially at the beginning of a hospital stay, a child can easily fear that they will never return. Even a parent's short absence during the day can prove upsetting to a child. Once the child has become familiar with the hospital environment and personnel, the parents can encourage attachments to particular staff members or playmates to prevent the child from feeling abandoned during periods when the parents must be away.

In addition to separation anxiety, it is common for hospitalized children to fear injury or even death. Children may or may not verbalize these fears. Besides comforting their children simply by their presence, parents can also help them cope by trying as much as possible to help the child feel he or she has some control over things, such as what toys to play with, or what to eat or wear. Parents should answer their child's questions as honestly and thoroughly as possible.

Children may cope with their emotions by being withdrawn or aggressive, or they may have an unnatural

KEY TERMS

Anesthesia—Treatment with medicine that causes a loss of feeling, especially pain. Local anesthesia numbs only part of the body; general anesthesia causes loss of consciousness.

Anesthesiologist—A medical specialist who has special training and expertise in the delivery of anesthetics.

Attending physician—The doctor who is in charge of the patient's overall care and treatment in the hospital. This doctor may or may not be the child's primary physician.

Case manager—A professional who designs and monitors implementation of comprehensive care plans (i.e., services addressing medical, financial, housing, psychiatric, vocational, social needs) for individuals seeking mental health or social services.

Clinical nurse specialist—A nurse with advanced training as well as a master's degree.

Discharge planner—A health care professional who helps parents arrange for health and home care needs after their child goes home from the hospital.

Electrocardiagram (ECG, EKG)—A record of the electrical activity of the heart, with each wave being labeled as P, Q, R, S, and T waves. It is often used in the diagnosis of cases of abnormal cardiac rhythm and myocardial damage.

Financial counselor—Professional who can provide assistance with financial matters associated with the patient's hospital stay. The financial counselor can help families evaluate their insurance plan's hospitalization coverage, determine a payment plan for medical expenses that are not covered, and discuss possible sources of financial aid.

General anesthesia—Deep sleep induced by a combination of medicines that allows surgery to be performed.

Home care—Health care services provided in the patient's home. If home health services will be needed after the patient is discharged, they can be arranged by the social worker or nursing staff.

Inpatient surgery—Surgery that requires an overnight stay of one or more days in the hospital. The number of days spent in the hospital after surgery depends on the type of procedure performed.

Local anesthesia—Pain-relieving medication used to numb an area while the patient remains awake. Also see general anesthesia.

Nursing unit—The floor or section of the hospital where patient rooms are located.

Nutrition therapy—Nutrition assessment, counseling, and education, usually provided by registered dietitians.

Outpatient surgery—Also called same-day or ambulatory surgery. The patient arrives for surgery and returns home on the same day. Outpatient surgery can take place in a hospital, surgical center, or outpatient clinic.

Patient education—Instruction and information that helps patients prepare for a procedure, learn about a disease, or manage their health. Patient education may include one-on-one instruction from a health care provider, educational sessions in a group setting, or self-guided learning videos or modules. Informative and instructional handouts are usually provided to explain specific medications, tests, or procedures.

Patient rights and responsibilities—Every hospital has an established list of patient rights and responsibilities, established by the American Hospital Association. They are usually posted throughout the hospital.

Pediatric intensivist—A physician who completed a three-year residency in pediatrics after medical school and an additional subspecialty fellowship training in intensive care.

Regional anesthesia—Blocking of specific nerve pathways through the injection of an anesthetic agent into a specific area of the body.

Registered nurses—Specially trained nurses who provide care during the patient's hospital stay. Registered nurses provide health care, administer medications, monitor the patient's condition, and educate the patient.

Social worker—Health care professional available to help patients and families manage the changes that may occur as a result of the patient's hospitalization. Socials workers provide referrals to community resources and can help the family make arrangements for care in the home as necessary after the patient is discharged from the hospital.

degree of obedience stemming from the fear that if they are "bad," worse things will happen. All of these emotions are normal and generally transient reactions that do not cause any long-lasting emotional harm.

It is common for children to experience some developmental regression in response to being hospitalized, either during the experience or after they come home. Often, they temporarily lose a recent advance, such as staying dry at night or overcoming certain fears. It can be helpful for children to cope with their feelings about hospitalization once they are home by playing with dolls and other toys. A doctor kit, selected to help prepare for the hospitalization, is sometimes very helpful in replaying children's reactions once they return home.

Whether planned or on an emergency basis, hospitalization causes disruption in the life of any child. However, with the special accommodations that hospitals usually make for children and their parents, a stay in the hospital need not be a traumatic event. If children receive proper support from family members and the hospital staff, hospitalization can even make them feel proud for having successfully negotiated a challenge to their maturity, self-discipline, and courage.

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American Hospital Association One North Franklin, Chicago, IL 60606. (312) 422-3000. Web site: <www.hospitalconnect.com>.

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Human bite infections

Definition

Human bite infections are potentially serious injuries that develop when a person's teeth break the skin of the hand or other body part and introduce saliva containing disease organisms below the skin surface.

Description

There are three common types of injuries caused by human **bites**:

- Closed-fist injuries: These are injuries to the hand sustained in a fight when the skin over the knuckles is broken and penetrated by the teeth in the opponent's mouth.
- Occlusional or chomping injuries: This type of injury results when a person bites down hard on another person's ear, nose, or finger.
- Avulsion injuries: Avulsion is the medical term for a ripping or tearing of the skin or body part. Human bites on the head and neck may cause avulsion injuries to the ears, nose, cheeks, or scalp.

Demographics

Exact statistics on human bite **wounds** are difficult to establish, although one figure for closed-fist bites in

the United States is 11.8 per 100,000 persons per year. Bite injuries account for about 1 percent of emergency room admissions in the United States and Canada, with human bites ranked third after dog and cat bites respectively. About 70 percent of all human bite wounds reported in North America involve adolescent males or adult males below the age of 40.

Closed-fist and chomping injuries account for most human bite infections. In one study done in a California hospital, closed-fist injuries accounted for 56 percent of the human bite infections treated, with the remaining 44 percent caused by occlusional bites.

Causes and symptoms

Causes

In children, bite infections result either from accidents during **play** or from fighting. Toddlers often bite one another when they are roughhousing; however, they usually do not bite hard enough to cause serious injury. Deep bite wounds on a young child may indicate abuse by an adolescent or adult.

Most infected human bites in adolescents and adults result from fighting, and some are inflicted on police officers or institutional staff. Alcohol or drug intoxication is an additional factor in closed-fist injuries.

The structure of the human hand contributes to the frequency with which closed-fist bites are likely to become infected. When a person closes the hand to make a fist, a tendon known as the extensor tendon is stretched. When the person hits the teeth in another person's mouth hard enough to break the skin, bacteria from the saliva in the mouth get into the tendon and its overlying sheath. After the hand is opened, the extensor tendon relaxes and returns to its normal position underneath the skin, but it is now carrying bacteria with it. The bacteria can then invade tissues that are very difficult to cleanse when the person finally seeks medical help. A similar chain of events is involved in infections of chomping injuries. Like the back of the hand, the fingers also have tendons lying just below the skin. A chomping bite that is hard enough to break the skin can also introduce bacteria into the finger tendons or their sheaths.

The infection itself can be caused by a number of bacteria that live in the human mouth. These include streptococci, staphylococci, anaerobic organisms, *Prevotella melaninogenica*, *Fusobacterium nucleatum*, *Candida spp.* and *Eikenella corrodens*. Infections that begin less than 24 hours after the injury are usually produced by a mixture of organisms and can produce a necrotizing infection (causing the death of a specific area of tissue), in which tissue is rapidly destroyed.

Symptoms

The most common sign of infection from a human bite is inflammation, which usually develops within eight to 24 hours following the bite. The skin around the wound is red and feels warm, and the wound may ooze pus or a whitish discharge. Nearby lymph glands may be swollen, and there may be red streaks running up the arm or leg from the wound toward the center of the body. Complications can arise if the infection is not treated and spreads into deeper structures or into the bloodstream.

Live disease-causing bacteria within the bloodstream and tissues may cause complications far from the wound site, including transmission of **HIV infection**. Deep bites or bites near joints can damage joints and bones, causing inflammation of the bone and bone marrow, necrotizing fasciitis, or septic arthritis.

When to call the doctor

Parents should call the doctor or take the child to the emergency room for examination and treatment of any human bite severe enough to break the skin, no matter what part of the body is affected. Even wounds that appear to be minor abrasion-type injuries may prove to be deeper puncture wounds when the doctor examines them.

Diagnosis

In most cases the diagnosis is made by an emergency room doctor on the basis of the patient's history.

The medical examination involves taking the history of the injury and assessing the type of wound and damage. The child's record of **tetanus** immunization and general health status are checked. An x ray may be ordered to assess bone damage and to check for **foreign objects** in the wound. In the case of a closed-fist injury, there may be fragments of teeth present in the wound. Wound cultures are done for infected bites if the victim is at high risk for complications or if the infection does not respond to treatment. If the child was bitten severely on the head, the emergency room doctor will call in a neurologist for consultation, particularly if the eyes, ears, or neck were injured or the skull was penetrated. Young children are particularly at risk for infection of puncture wounds from bites on the head because the skin on the scalp and forehead is relatively thin and soft. The doctor may also consult a plastic surgeon if the bites are extensive, if large pieces of tissue have been lost or if the

functioning or appearance of the injured part of the body is likely to be affected.

Because the human mouth contains a variety of bacteria, the doctor may order a laboratory culture in order to choose the most effective antibiotic. Cultures are most commonly done when the wound has begun to show signs of infection.

Treatment

Treatment depends on the wound type, its site, and such other risk factors for infection as the condition of the patient's immune system. All wounds from human bites are cleaned and disinfected as thoroughly as possible. The doctor will begin by injecting a local anesthetic in order to examine the wound thoroughly without causing additional **pain** to the child. The next step is to remove dead tissue, foreign matter, and blood clots, all of which can become sources of infection. This removal is called debridement. After debriding the wound, the doctor will cut away the edges of the tissue, as clean edges heal faster and are less likely to form scar tissue. The doctor will then irrigate, or flush, the wound with saline solution forced through a syringe.

Doctors do not usually suture a bite wound on the hand because the connective tissues and other structures in the hand form many small closed spaces that make it easy for infection to spread. Emergency room doctors often consult surgical specialists if a patient has a deep closed-fist injury or one that appears already infected.

Bites on the ear are also difficult to treat because the cartilage in the ear does not have a good blood supply. If the cartilage has been exposed by the bite, the doctor will administer intravenous **antibiotics** for 48 hours and delay closing the wound for 24 hours or longer following the injury.

The doctor will make sure that the patient is immunized against tetanus, which is routine procedure for any open wound. Because of risk of infection, all patients with human bite wounds should be given antibiotics. The usual choice for human bite wounds on the hand is a first-generation cephalosporin and either penicillin or amoxicillin-clavulanate (Augmentin). If the child has a weakened immune system, the doctor will prescribe either amoxicillin or erythromycin. Patients with severe closed-fist injuries may need inpatient treatment in addition to an intravenous antibiotic.

Prognosis

The prognosis depends on the location of the bite and whether it was caused by a child or an adult. Bites caused by children rarely become infected because they are usually shallow. Between 15 and 30 percent of bites caused by adults become infected, with a higher rate for closed-fist injuries.

The prognosis for restoring the function or appearance of a hand, ear, or other body part following a severe bite depends on the location of the bite, the promptness of treatment, and the availability of specialized surgical repair. Infections of the hand have a high rate of permanent scarring, tissue damage, and loss of function when treatment is delayed.

Prevention

Prevention of human bite infections depends upon prompt treatment of any bite caused by a human being, particularly a closed-fist injury. Long-term prevention, particularly in young males, requires teaching children how to resolve arguments or quarrels without resorting to violence.

Parental concerns

Biting is fairly normal behavior in toddlers and rarely reflects intentional malice. Children in this age group sometimes bite one another because they are overstimulated or tired or because they have not yet learned to use words to express their feelings. Suggestions for preventing biting in young children include scheduling naps after highly stimulating activities, giving the child a washcloth or similar object to bite, and observing the child to see whether a particular person or situation is triggering the biting behavior. As has been mentioned earlier, toddlers' bites are rarely severe enough to cause a serious infection unless the bitten child has a weakened immune system.

Parental concerns for older children and adolescents include the possibility that the child is abusing drugs or alcohol, has a **conduct disorder**, or is being bullied, as well as fears of disfigurement or lasting injury from an infected bite on the face or hand.

See also Animal bite infections.

Resources

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KEY TERMS

Avulsion—The forcible separation of a piece from the entire structure.

Closed-fist injury—A hand wound caused when the skin of the fist is torn open by contact with teeth.

Debridement—The surgical removal of dead tissue and/or foreign bodies from a wound or cut.

Fasciitis—Inflammation of the fascia (plural, fasciae), which refers to bands or sheaths of connective tissue that cover, support, or connect the muscles and internal organs. Human bites can lead to infection of the fasciae in the hand.

Fight bite—Another name for closed-fist injury.

Irrigation—Cleansing a wound with large amounts of water and/or an antiseptic solution. Also refers to the technique of removing wax (cerumen) from the ear canal by flushing it with water.

Necrotizing—Causing the death of a specific area of tissue. Human bites frequently cause necrotizing infections.

Occlusional—Referring to a type of injury caused by the closing of the teeth on a finger or other body part. Occlusional injuries are also called chomping injuries.

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Rebecca Frey, PhD

Hunter's syndrome *see* **Mucopolysaccharidoses**

Hurler's syndrome *see* **Mucopolysaccharidoses**

Hyaline membrane disease *see* **Respiratory distress syndrome**

Hydrocephalus

Definition

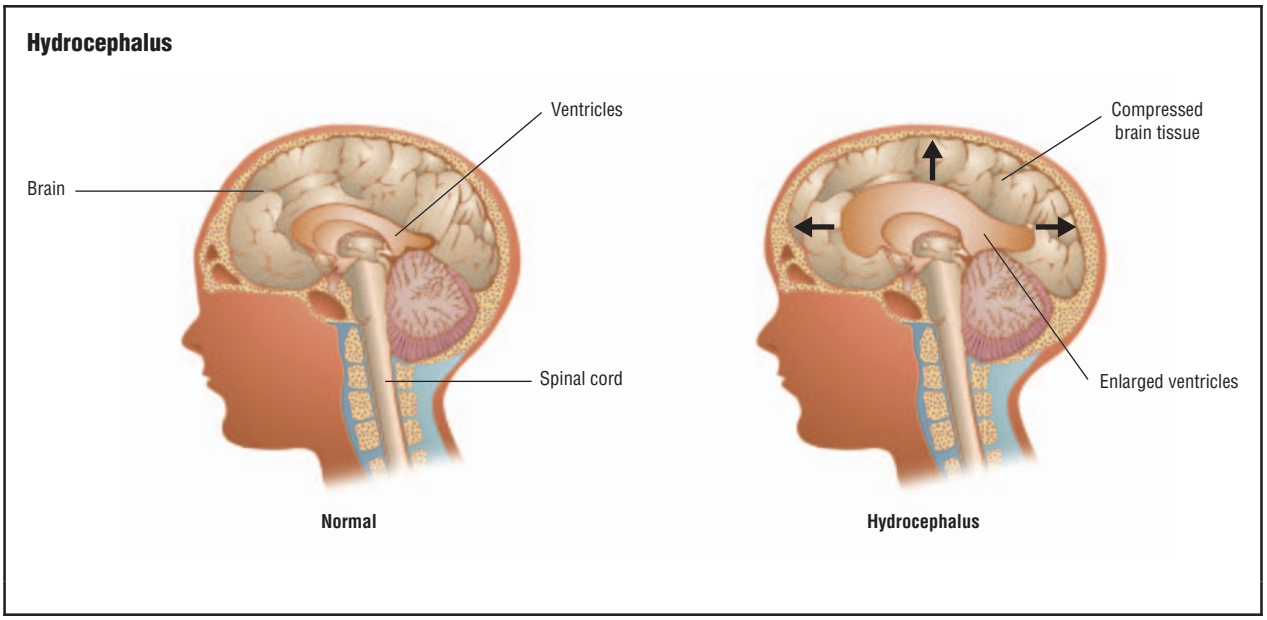
Hydrocephalus is an abnormal expansion of cavities, called ventricles, within the brain, which is caused by an abnormally large accumulation of cerebrospinal fluid (**CSF**).

Description

Hydrocephalus is the result of an imbalance between the formation and drainage of CSF. There are four ventricles in the human brain. CSF is formed by structures within these ventricles. Once formed, CSF circulates among all the ventricles before it is absorbed and returned to the circulatory system. When the ventricles are obstructed, the CSF cannot circulate and be absorbed. An elevated level of CSF in the brain leads to pressure within the ventricles. This pressure pushes against the soft tissues of the brain, resulting in damage to these tissues.

There are three different types of hydrocephalus: communicating hydrocephalus, noncommunicating hydrocephalus, and normal pressure hydrocephalus. Communicating hydrocephalus is the most common type and exists when one or more passages connecting the ventricles become blocked. This blockage prevents the movement of CSF to its drainage sites in the subarachnoid space just inside the skull. In noncommunicating hydrocephalus, the tissue within the brain responsible for absorption of CSF is damaged. Normal pressure hydrocephalus is marked by ventricle enlargement without an apparent increase in CSF pressure. This type affects mainly the elderly and will not be discussed in this entry.

Hydrocephalus may be either congenital (present at birth) or acquired. An obstruction within the brain is the most frequent cause of congenital hydrocephalus. Acquired hydrocephalus may result from other birth defects such as **spina bifida**, conditions related to **prematurity** such as intraventricular hemorrhage (bleeding



A normal brain (left) and one showing the enlarged ventricles of hydrocephalus. The additional fluid in the ventricles causes increased pressure on the brain. *(Illustration by GGS Information Services.)*

within the brain), infections such as **meningitis**, or other causes such as head trauma, tumors, and cysts.

Demographics

Hydrocephalus is believed to occur in approximately one to two of every 1,000 live births. It is not more prevalent in males or females, nor in any individual racial group.

Causes and symptoms

Hydrocephalus has a variety of causes including the following:

- congenital brain defects
- hemorrhage, either into the ventricles or the subarachnoid space
- infection of the central nervous system (syphilis, herpes, meningitis, **encephalitis**, or mumps)
- tumor

Signs and symptoms of elevated-pressure hydrocephalus include the following:

- headache
- nausea and **vomiting**, especially in the morning
- lethargy
- disturbances in walking (gait)
- double vision
- subtle difficulties in learning and memory
- delay in achieving childhood developmental milestones

Irritability is the most common sign of hydrocephalus in infants. If this is not treated, it may lead to lethargy. Bulging of the fontanelles, or the soft spots between the skull bones, may also be an early sign. When hydrocephalus occurs in infants, fusion of the skull bones is prevented, which leads to abnormal expansion of the skull.

Diagnosis

Imaging studies such as x ray, **computed tomography** scan (CT scan), ultrasound, and especially **magnetic resonance imaging** (MRI) are used to assess the presence and location of obstructions, as well as changes in brain tissue that have occurred as a result of the hydrocephalus. Lumbar puncture (spinal tap) may be performed to aid in determining the cause when infection is suspected.

Treatment

The primary method of treatment for hydrocephalus is surgical installation of a shunt. A shunt is a tube connecting the ventricles of the brain to an alternative drainage site, usually the abdominal cavity. A shunt contains a one-way valve to prevent reverse flow of fluid. In some cases of non-communicating hydrocephalus, a direct connection can be made between one of the ventricles and the subarachnoid space, allowing drainage without a shunt.

Installation of a shunt requires lifelong monitoring by the recipient or **family** members for signs of recurring hydrocephalus due to obstruction or failure of the shunt. Other than monitoring, no other management activity is usually required.

Some drugs may postpone the need for surgery by inhibiting the production of CSF. These include acetazolamide and furosemide. Other drugs that are used to delay surgery are glycerol, digoxin, and isosorbide.

Prognosis

The prognosis for elevated-pressure hydrocephalus depends on a wide variety of factors, including the cause, age of onset, and the timing of surgery. Studies indicate that about half of all children who receive appropriate treatment and follow-up will develop IQs greater than 85. Those with hydrocephalus at birth do better than those with later onset due to meningitis. For individuals with normal pressure hydrocephalus, approximately half will benefit by the installation of a shunt.

Prevention

There is no known prevention of congenital hydrocephalus. Some cases of elevated pressure hydrocephalus may be avoided by preventing or treating the infectious diseases that precede them. Prenatal diagnosis of congenital brain malformation is often possible, offering the option of family planning.

Parental concerns

Parents may be concerned about the intellectual development of a child with hydrocephalus. While nearly 50 percent of all children with hydrocephalus have average **intelligence**, many do not. Early intervention programs are important to the development of children with special needs and are available in most communities. In addition to developmental issues, many children with hydrocephalus require medical care. It is important for parents to prepare children for medical treatment and surgery. A healthcare team including a pediatrician, surgeon, and social worker is a valuable asset for parents and most children's hospitals can assist parents in finding the support and resources they need.

When to call the doctor

The most common treatment for hydrocephalus is the surgical installation of a shunt. If a child with a shunt has any of the following symptoms, parents should contact the child's doctor because the shunt may not be functioning properly. According to the Spina Bifida Association of America, nearly 40 percent of shunts malfunction and may need to be replaced within one year, 60 percent will require revision within five years, and 80 to 90 percent within ten years.

For this reason, parents need to be aware of the symptoms of shunt malfunction and contact their physician if they notice any of the following symptoms:

- headaches
- nausea
- vomiting
- seizures
- change in intellect or personality
- swallowing problems
- impaired muscle function, balance, or coordination

KEY TERMS

Cerebrospinal fluid—The clear, normally colorless fluid that fills the brain cavities (ventricles), the subarachnoid space around the brain, and the spinal cord and acts as a shock absorber.

Choroid plexus—Specialized cells located in the ventricles of the brain that produce cerebrospinal fluid.

Fontanelle—One of several "soft spots" on the skull where the developing bones of the skull have yet to fuse.

Shunt—A passageway (or an artificially created passageway) that diverts blood flow from one main route to another. Also refers to a small tube placed in a ventricle of the brain to direct cerebrospinal fluid away from a blockage into another part of the body.

Ventricles—Four cavities within the brain that produce and maintain the cerebrospinal fluid that cushions and protects the brain and spinal cord.

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Hyperbilirubinemia *see* **Jaundice**

Hyperglycemia

Definition

Hyperglycemia is a complex metabolic condition characterized by abnormally high levels of blood sugar (blood glucose) in circulating blood, usually as a result of **diabetes mellitus** (types 1 and 2), although it can sometimes occur in **cystic fibrosis** and **near-drowning** (submersion injury).

Description

Hyperglycemia, also known as diabetic ketoacidosis, is a condition that develops over a period of a few days as the blood glucose levels of a type 1 or type 2 diabetic gradually rise. Ketoacidosis occurs when increasing glucose levels are met by a lack of sufficient or effective insulin production, starting a sequence of physiologic events as follows:

- The combination of excess glucose production and low glucose utilization in the body raises levels of blood glucose, which leads to increased urinary output (diuresis) followed quickly by a loss of fluid and essential mineral salts (electrolytes) and, ultimately, **dehydration**. The loss of fluid may finally result in dehydration. If the entire process is severe enough over several hours (serum glucose levels over 800mg/dL), swelling can occur in the brain (cerebral edema), and coma can eventually result.
- In a metabolic shift to a catabolic (breaking down) process, cells throughout the body empty their electrolytes (sodium, potassium, and phosphate) into the bloodstream. Electrolytes control the fluid balance of the body and are important in muscle contraction, energy generation, and almost all major biochemical reactions in the body. As a result of electrolyte imbalance, many functions can become impaired.
- Free fatty acids from lipid stores are increased, encouraging the production of ketoacids in the liver, leading to an over-acidic condition (metabolic acidosis) that causes even more disruption in body processes.

Without effective treatment of the hyperglycemic episode, the child can lapse into a diabetic coma, which sometimes leads to death.

In diabetes type 2, which is characterized by insulin resistance, enhanced glucose production in the liver and decreased insulin secretion can be aggravated by low physical activity and/or a high-calorie, high-fat diet. Over time as glucose production accelerates, the child develops hyperglycemia or glucotoxicity and lipotoxicity (hyperlipidemia or high fat levels in the blood) as well. It is primarily thought to be a disease affecting sedentary, obese adults over age 40, but it is found in young people as well, most of them obese at the time of diagnosis. Pediatric type 2 diabetes is increasing in the early 2000s among adolescents and has become the fastest growing form of diabetes. Therefore, hyperglycemic episodes are also noted to be increasing in frequency among young people admitted to hospitals for treatment of diabetes.

Demographics

The incidence of hyperglycemia approximately parallels the incidence of diabetes type 1 cases, which represents about 70 percent of all diabetes cases (17 million Americans diagnosed) in the United States. It occurs more in whites than blacks or Asians. About 30 percent of all new cases of diabetes are children with diabetes type 2. Diabetes type 2 occurs more often in African American youth but also in Native Americans, white Americans, and Hispanic youth between ages 10 and 19. Those with type 2 have fewer symptoms and are not treated as frequently for hyperglycemia.

Causes and symptoms

Diabetes is a chronic metabolic disorder with hyperglycemia, gradually rising levels of glucose, as its primary characteristic. As diabetes develops and symptoms increase, hyperglycemia becomes progressive but will occur only occasionally in the carefully managed

diabetic patient. Hyperglycemia can be triggered by irregular self-administration of insulin, by insulin resistance or defective insulin response in the body, by stress or infection, and by the activation of certain autoimmune processes characteristic of type 1 diabetes. It occurs in 20 to 40 percent of children newly diagnosed with diabetes and in children who are not yet successfully managed. Many young type 2 diabetics do not have symptoms because their hyperglycemia is moderate compared to type 1 diabetics, and they are not taking insulin.

The first signs of hyperglycemia or ketoacidosis are frequent urination and increased thirst. The child may then show any of the following symptoms:

- flushed face
- dry skin
- dry mouth
- **headache**
- abdominal **pain**
- **nausea and vomiting**
- drowsiness and lethargy
- blurred vision
- fruity-smelling breath
- rapid heartbeat
- deep and labored breathing

When to call the doctor

The pediatrician or **family** doctor should be consulted about any sudden change in the child's urinary output, frequency of urination, or increased thirst, especially if accompanied by dry skin or mouth, flushed face, headache, abdominal pain, nausea or vomiting, unusual drowsiness and lack of movement, rapid heartbeat, or difficulty breathing. Parents should be aware of the last insulin injection if the child is on insulin therapy.

Diagnosis

Hyperglycemia can be diagnosed fairly quickly in known diabetic children. The non-fasting serum glucose will exceed 200mg/dL with classic symptoms such as increased urination, extreme thirst, dry skin or mouth, flushed face, headache, abdominal pain, nausea or vomiting, unusual drowsiness and lack of movement, rapid heartbeat, or difficulty breathing. If elevated glucose levels are present, the doctor will want to determine if ketoacidosis is also present by measuring levels of ketones in the blood serum and urine. Electrolyte levels will be measured along with carbon dioxide and pH and serum osmolality, which may indicate hypertonic dehydration. Routine screening of blood glucose levels and glucose tolerance tests is not recommended in children; symptoms are believed to help confirm hyperglycemia more readily. It is also not considered necessary to test non-obese children for autoimmune antibodies, which are more apt to be found in adult type 2 diabetics.

Treatment

Treatment for hyperglycemia must be delivered carefully and with close monitoring to avoid the risk of hypokalemia (higher than normal serum levels of potassium) and subsequent cerebral edema. Treatment will take place over a period of several days, including administration of insulin, usually in combination with administration of intravenous fluids and salts to restore fluid and electrolyte balance. Fluid intake and output is carefully monitored, and serum electrolytes are tested hourly or even more frequently to make sure balance is restored to support normal metabolic activity in the body. Children must be rehydrated very gradually; this can be done orally in mild hyperglycemia and over an extended period (30 to 36 hours) of intravenous administration with severe hyperglycemia. Administration of insulin helps move glucose back into cells, reduces glucose production by the liver, and stops the release of fatty acids. Insulin injections, while helping to normalize glucose production, also increase the risk of hypokalemia and abnormally low levels of glucose in the blood (**hypoglycemia**), the opposite of the hyperglycemic condition being corrected. Glucose is sometimes infused with the insulin to help avoid hypoglycemia. The insulin infusion will be slowed once hyperglycemia has been corrected (blood glucose levels less than 250mg/dL); in children with moderate hyperglycemia, this can often be accomplished within 24 hours. It may take several days, however, to restore normal cellular levels of potassium, sodium, and phosphate. The acidosis will be reversed, reflected in a gradual increase in pH.

In severe cases of hyperglycemia in which cerebral edema occurs, mannitol is administered at the first sign of edema, such as unconsciousness, difficulty breathing, severe headache, irregular heartbeat, or seizures. Changes can occur very rapidly. Children with moderate to severe hyperglycemia may be treated in an intensive care unit for continuous monitoring and rapid response capabilities.

In rare instances, a child may have a hyperglycemic episode that is triggered by a stressful situation or a physically challenging situation such as another illness. Transient hyperglycemia can be triggered by any type of stress that overtaxes the child's mental and physical resources. Stress hyperglycemia may be reversed com-

pletely when the stressors are removed or relieved. Temporary hyperglycemia of this type will still require careful monitoring for symptoms and testing and treatment as above if any symptoms occur.

Alternative treatment

Although alternative treatment for diabetes includes taking chromium picolinate to improve the efficiency of insulin in glucose metabolism and coenzyme Q10 to improve blood circulation and stabilize blood glucose levels, hyperglycemia and diabetic ketoacidosis require immediate measures such as insulin injections and rehydration and cannot be treated by nutritional means.

Nutritional concerns

Nutritional therapy along with insulin therapy can both help avoid hyperglycemia and relieve associated symptoms. Immediate medical attention is needed, however, and parents should not undertake correction of hyperglycemia or dehydration on their own. Basic nutritional requirements for children with diabetes can be provided by the pediatrician, based on the child's age, sex, weight, activity levels, food preferences, and ethnic or cultural factors. The recommended diet for those with diabetes calls for complex carbohydrates such as whole (unrefined) whole grains, plenty of fresh vegetables and fruits, with an overall intake of foods that are low in fat and high in fiber. This diet reduces the need for insulin and lowers fat levels in the blood, all helping to stabilize glucose levels.

Prognosis

The prognosis for children with mild to moderate hyperglycemia is good; the condition can usually be corrected within 24 hours. Severe hyperglycemia (serum glucose levels in the range of 800mg/dL) may lead to cerebral edema, coma, and death if not treated immediately. Hyperglycemia in children during severe illness is a risk factor for poor outcomes in the underlying illness and has been reported as a cause of increased mortality in pediatric intensive care units. Morbidity (the incidence of other diseases) and mortality are higher in adults than in children because of long-term complications that include vascular conditions, circulatory problems, nervous system disorders, liver problems, and heart disease.

Prevention

Occurrences of hyperglycemia can be prevented by careful monitoring of blood glucose levels and insulin injections while balancing **exercise** and diet. Diabetic adolescents are especially susceptible to hyperglycemia, since hormone levels are in flux and many adolescents exhibit erratic eating and sleeping patterns. Athletic activities can be beneficial, since exercise makes use of surplus blood glucose. Obese children must be encouraged to eat properly and to avoid the fats and sugary sweets that can lead to increased weight, decreased mobility, and hyperglycemia.

Nutritional concerns

Nutrition, of course, is important in the treatment of diabetes and accordingly can play a role in preventing hyperglycemic episodes. Diabetic children must avoid sweetened and high-carbohydrate foods such as white flour and white sugar products and generally eat a high-fiber, low-fat diet consisting of whole grains, high quality protein (lean meat, eggs, low-fat dairy), fresh vegetables, and fresh fruits.

Parental concerns

Parents of a diabetic child may live with the uncertainty of possible hyperglycemic episodes but can be reassured by knowing that continuous glucose monitoring, a proper diet as advised by the pediatrician, insulin therapy if prescribed, and appropriate exercise can control the disease and help avoid extremes that lead to hyperglycemia. It is important to maintain close contact with the child's diabetes team of professionals and to learn as much as possible about the disease and the symptoms to watch for in the child that may signal hyperglycemia. The parents of school-age children should make sure that teachers also understand the warning signs of hyperglycemia so that immediate medical attention can be given when needed.

See also Diabetes mellitus.

Resources

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National Institute of Diabetes & Digestive & Kidney Diseases (NIDDK). NIH Building 31, Room 9A, 4 Center Drive,

KEY TERMS

Antibody—A special protein made by the body's immune system as a defense against foreign material (bacteria, viruses, etc.) that enters the body. It is uniquely designed to attack and neutralize the specific antigen that triggered the immune response.

Autoimmune—Pertaining to an immune response by the body against its own tissues or types of cells.

Cerebral—Pertaining to the brain.

Coma—A condition of deep unconsciousness from which the person cannot be aroused

Diabetes—A disease characterized by an inability to process sugars in the diet, due to a decrease in or total absence of insulin production.

Edema—The presence of abnormally large amounts of fluid in the intercellular tissue spaces of the body.

Electrolytes—Salts and minerals that produce electrically charged particles (ions) in body fluids. Common human electrolytes are sodium chloride, potassium, calcium, and sodium bicarbonate. Electrolytes control the fluid balance of the body and are important in muscle contraction, energy generation, and almost all major biochemical reactions in the body.

Hypertonic saline solution—Fluid that contains salt in a concentration higher than that of healthy blood.

Ketones—Poisonous acidic chemicals produced by the body when fat instead of glucose is burned for energy. Breakdown of fat occurs when not enough insulin is present to channel glucose into body cells.

Metabolic—Refers to the chemical reactions in living organisms.

Osmolality—The concentration of osmolar particles in the blood (or other solutions) that can help determine if the body is dehydrated.

Physiologic—Refers to physiology, particularly normal, healthy, physical functioning.

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L. Lee Culvert

Hyperhidrosis

Definition

Hyperhidrosis is a medical condition characterized by excessive sweating in the armpits, palms, soles of the feet, face, scalp, and/or torso.

Description

Hyperhidrosis involves sweating in excess of the amount required normally for the body's level of activity and temperature. There are two types of hyperhidrosis—primary and secondary. In primary hyperhidrosis, the cause is unknown and excessive sweating is localized in the armpits, hands, face, and/or feet. Primary hyperhidrosis begins during childhood or early **adolescence**, gets worse during **puberty**, and lasts a lifetime. In secondary hyperhidrosis, which is less common than primary hyperhidrosis, excessive sweating is caused by another medical condition and usually occurs over the entire body. Medical conditions that can cause secondary hyperhidrosis include **hyperthyroidism**, menopause, **obesity**, psychiatric disorders, and diabetes. Secondary hyperhidrosis may also be caused by use of certain medications.

In about 60 percent of cases, the hands and feet are affected, and in about 30–40 percent of cases, the armpits are affected.

Demographics

Axillary (underarm) hyperhidrosis occurs more frequently in females and in individuals of Asian or Jewish ancestry. Hyperhidrosis of the hands and feet occurs 20 times more frequently in the Japanese. Previously, it was thought that hyperhidrosis was rare, occurring in only 0.6–1 percent of adolescents and young adults; however, a national survey conducted in 2004 found that up to 2.8 percent of Americans (approximately 7.8 million individuals) may have hyperhidrosis.

Causes and symptoms

The exact cause of hyperhidrosis is as of 2004 unknown. Excessive sweating in the affected area is caused by overactivity of the nerves linked to the sweat glands. Specifically, acetylcholine, a chemical in the body that transmits nerve signals, is released from nerve endings and stimulates secretion of sweat. Genetics may also be a factor, since 25–40 percent of individuals with hyperhidrosis also have a **family** member with the condition.

In hyperhidrosis, sweating may be continuous or start suddenly. Usually, excessive sweating does not occur in response to **exercise** and does not occur during **sleep**. Emotional stress, high room/environmental temperature, and digestion of certain foods can aggravate hyperhidrosis. Symptoms of hyperhidrosis vary depending on the body area affected:

- In palmar hyperhidrosis, the palms of the hands are excessively wet or moist and also cold to the touch.
- In axillary hyperhidrosis, excessive sweating in the underarm area occurs, leaving large wet marks and staining clothes.
- In scalp/facial hyperhidrosis, excessive sweating of the face and scalp occurs, as well as moderate to severe facial blushing.
- In plantar hyperhidrosis, the soles of the feet sweat excessively. This condition is often associated with hyperhidrosis in other body areas.
- In truncal hyperhidrosis, the torso area sweats excessively. This condition is rare alone and usually occurs with hyperhidrosis in other areas.

When to call the doctor

Parents should call the doctor if their child or adolescent experiences excessive sweating unrelated to an obvious medical condition (e.g., high **fever**) or physical exertion. Usually, consultation and treatment will be given by a dermatologist.

Diagnosis

Hyperhidrosis is diagnosed by physical examination. For suspected secondary hyperhidrosis, laboratory and imaging tests may be performed to determine the underlying medical condition causing the hyperhidrosis.

Treatment

Topical agents applied to the skin in the affected area are the first course of treatment for hyperhidrosis. Topical applications include anticholinergic drugs, boric acid, tannic acid solutions, and glutaraldehyde. Drysol, an aluminum chloride solution, is the most commonly used and most effective topical application; it is applied nightly on dry skin. Systemic medications may be taken orally and include anticholinergic drugs, sedatives or tranquilizers, and calcium channel blockers. These oral drugs do have side effects, such as dry mouth and eyes, blurry vision, and **constipation**, and may not be appropriate for pediatric patients.

Iontophoresis, which involves the application of an electrical current across the skin, can be used to treat plantar and palmar hyperhidrosis but requires daily treatment for about 30 minutes, often multiple times daily.

As a last resort, surgery is used to treat palmar, plantar, and axillary hyperhidrosis. Surgical procedures involve removing portions of the nerves responsible for excessive sweating and removing sweat glands during an open or minimally invasive surgical procedure. Liposuction may be used to remove sweat glands in the underarm area.

In 2004, the U.S. Food and Drug Administration approved the use of botulinum toxin (Botox) for treatment of axillary (underarm) hyperhidrosis that resists treatment with topical drugs. Botox is commonly used for cosmetic treatment of wrinkles but is also used to treat neuromuscular problems, including migraine and cervical dystonia. In the early 2000s researchers are also investigating the use of Botox to treat hyperhidrosis of the hands, feet, and face. Although most studies of Botox for hyperhidrosis included adult patients, some physicians use Botox to treat hyperhidrosis in children with some success. Even though Botox has only been approved to treat axillary hyperhidrosis, physicians can legally use Botox "off-label" to treat other affected areas of the body. Botox is injected into the affected area, and one series of injections may last for several months. Botox is a likely treatment when topical applications fail.

In 2004, guidelines were proposed by expert physicians for treating primary hyperhidrosis. Topical treatments followed by Botox if the topical agent fails is recommended for treating axillary and facial hyperhidrosis. For palmar and plantar hyperhidrosis, topical treatment and iontophoresis, followed by Botox are recommended. Surgery is mentioned as an option only for palmar and axillary hyperhidrosis and only as a last resort.

Alternative treatment

Although no evidence has documented an effective alternative treatment for hyperhidrosis, acupuncture, homeopathy, and/or herbal preparations are used by some individuals with hyperhidrosis. A common home remedy involves soaking the affected body parts in home-brewed tea, which contains tannic acid, a natural antiperspirant. Because stress can trigger sweating, relaxation techniques such as **yoga**, massage, and meditation can help with stress reduction.

Prognosis

Hyperhidrosis is not a life-threatening condition. However, it can severely affect quality of life and comfort in social situations. Children and adolescents who receive early treatment have a better quality of life. If left untreated, hyperhidrosis can result in physical, social, and occupational impairments.

Prevention

Hyperhidrosis treatments help to prevent excessive sweating but may not entirely eliminate the condition. Hyperhidrosis can be managed by using simple daily personal hygiene methods, such as the following:

- bathing daily to reduce bacteria
- washing and changing clothes frequently
- changing socks or pantyhose at least twice daily
- airing out shoes and rotating shoes worn each day
- wearing absorbent socks, clothing shields, and natural fabrics
- using antiperspirants in the evening and gently massaging them into the skin
- using foot powders and going barefoot frequently to air out feet

Nutritional concerns

Although no foods cause hyperhidrosis, certain foods and food ingredients can stimulate sweating and should be avoided. These include **caffeine**, alcohol, and spicy foods. Hot beverages, like coffee and hot chocolate, may also increase sweating. Consuming foods with strong odors, such as those containing garlic and onions, should be avoided because it can cause a person's sweat to smell stronger.

Parental concerns

Children and adolescents with hyperhidrosis suffer extreme social embarrassment related to their condition, and hyperhidrosis can result in low **self-esteem**, difficulties in school, and difficulties in and avoidance of social situations. For example, children with palmar hyperhidrosis may have difficulties holding a pen to write, and adolescents may be reluctant to shake or hold hands with others. Children with axillary hyperhidrosis may be made fun of for excessive body odor and sweat stains. Early treatment is essential to improve children's quality of life. Joining a support group or participating in online hyperhidrosis chat groups may help individuals better manage their condition through peer support.

KEY TERMS

Anticholinergic drug—Drugs that block the action of the neurotransmitter acetylcholine. They are used to lessen muscle spasms in the intestines, lungs, bladder, and eye muscles.

Axillary—Located in or near the armpit.

Liposuction—A surgical technique for removing fat from under the skin by vacuum suctioning.

Resources

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Hyper-IgM syndrome

Definition

Hyper-IgM syndrome is a primary **immunodeficiency** disorder in which the child's body fails to produce certain specific types of antibodies. The term primary means that the disorder is present from birth, in contrast to secondary immunodeficiencies (such as **AIDS**), which are acquired later in life by previously healthy persons. Hyper-IgM syndrome is caused by mutations in a gene or genes in the body's T cells, which are a type of white blood cell or lymphocyte. T cells regulate the production of antibodies, which are protein molecules produced as the first line of the immune system's defense against disease-causing organisms. Hyper-IgM syndrome is also known as hypogammaglobulinemia with hyper IgM.

There are two forms of hyper-IgM syndrome, defined by their patterns of inheritance. The more common of the two, known as X-linked hyper-IgM syndrome (XHIM), is caused by an abnormal gene on the X chromosome and affects only boys. The less common form, autosomal recessive hyper-IgM syndrome (ARHIM), occurs in children who have inherited an abnormal gene from both parents. ARHIM affects girls as well as boys.

Description

Hyper-IgM syndrome appears during the first year of life when the child develops recurrent infections of the respiratory tract that do not respond to standard antibiotic treatment, along with chronic **diarrhea**. Other early symptoms may include enlarged tonsils; swelling of the liver and spleen; enlarged lymph nodes; or opportunistic infections. Children with XHIM are more likely to develop enlarged lymph nodes than children with other primary immunodeficiency disorders. Opportunistic infections are caused by organisms that do not usually cause disease in people with normally functioning immune systems. The most common opportunistic infection in children with XHIM is a lung disease known as *Pneumocystis carinii* **pneumonia** (PCP). Children with either XHIM or ARHIM who are not diagnosed early may show delays in growth and normal weight gain.

Hyper-IgM syndrome is a disorder with a high degree of morbidity, which means that patients diagnosed with it often suffer from other diseases or disorders. The most common morbid conditions associated with XHIM include the following:

- Recurrent and chronic infections of the lungs and sinuses leading to chronic dilation of the bronchi (the larger air passageways) in the lungs. This condition, called bronchiectasis, is marked by frequent attacks of coughing that bring up pus-streaked mucus.
- Chronic diarrhea leading to weight loss and **malnutrition**. The diarrhea is usually caused by opportunistic infections of the digestive tract; the most common disease agents are *Cryptosporidium parvum*, *Giardia lamblia*, *Campylobacter*, or rotaviruses.
- Frequent mouth ulcers, skin infections, and inflammation of the area around the rectum (proctitis). These complications are associated with neutropenia, a condition in which the blood has an abnormally low number of neutrophils. Neutrophils are a special type of white blood cell that ingests bacteria and other foreign substances. The connection between hyper-IgM syndrome and neutropenia was not as of 2004 yet fully understood.
- Infections of the bones and joints leading to arthritis or osteomyelitis.
- Disorders of the nervous system caused by meningoencephalitis, or inflammation of the brain and its overlying layers of protective tissue. Patients with these disorders may have problems with thinking clearly, have difficulty walking normally, or develop paralysis on one side of the body (hemiplegia).
- Liver disease. About 70 percent of patients with XHIM develop liver disease by age 30, usually as a result of recurrent *Cryptosporidium* infections.
- Malignant tumors, most commonly non-Hodgkin's lymphoma or cancers of the gall bladder and liver.

Demographics

Both XHIM and ARHIM are rare disorders. One group of researchers at Johns Hopkins University estimates the incidence of XHIM in the general North American population as one in 1,030,000 males. In the early 2000s, however, it is thought that the disorder may be underdiagnosed. The incidence of ARHIM has not been established as of 2004, but it is known to be much less common than XHIM. As of the early 2000s, researchers do not know whether these disorders are more common in some racial or ethnic groups than others or whether they are equally common in all parts of the world. The only registries of patients as of 2004 diagnosed with hyper-IgM syndrome are located in Europe and the United States. The registry that was established in the United States in 1997 contains the records of 79 patients from 60 unrelated families, while the European database contains the records of XHIM patients from 130 unrelated families.

Causes and symptoms

Causes

Hyper-IgM syndrome is caused by a mutation in a gene on the X chromosome that affects the patient's T cells. The gene has been identified at locus Xq27. Normal T cells produce a ligand (a small molecule that links to larger molecules) known as CD40. CD40 is a protein found on the surface of T cells that signals B cells to stop producing IgM, which is the antibody that is first produced in response to invading organisms and switch to producing IgG and IgA, which are more specialized antibodies. As a result, boys with XHIM have abnormally low levels of IgG and IgA in their blood, with normal or higher than normal levels of IgM. Because they lack these "second line of defense" antibodies, they are more vulnerable to infections.

About 70 percent of patients diagnosed with XHIM have inherited the disorder through their mother; about 30 percent of cases, however, are caused by new mutations. Females who carry the defective gene have a 50 percent chance of passing it on to their sons but are not affected themselves by the disorder. The daughters of carriers have a 50 percent risk of carrying the defective gene to the next generation.

Symptoms

The symptoms of hyper-IgM syndrome usually become noticeable after the baby is six months to a year old. At this point the antibodies received from the mother during pregnancy are no longer present in the baby's blood. The child develops a series of severe ear, throat, or chest infections that do not clear up with standard antibiotic treatment. Another early warning sign is recurrent or chronic diarrhea. In addition, the child may have more than one infection at the same time. The most common telltale symptom, however, is PCP; in fact, the frequency of *Pneumocystis carinii* pneumonia in children with hyper-IgM syndrome was a useful clue to geneticists searching for the mutation that causes the disorder.

When to call the doctor

The Jeffrey Modell Foundation (JMF) and the American Red Cross have drawn up a list of 10 warning signs of hyper-IgM syndrome and other primary immunodeficiency disorders:

- The child has eight or more ear infections within one year.
- The child has two or more serious sinus infections within one year.
- The child has been treated with **antibiotics** for two months or longer with little effect.
- The child has been diagnosed with pneumonia more than twice within the past year.
- If an infant, the child is not growing or gaining weight normally.
- The child has repeatedly developed deep skin abscesses.
- If older than 12 months, the child has persistent thrush.
- The child needs intravenous antibiotics to clear infections.
- The child has two or more deep-seated infections (**meningitis**, osteomyelitis, sepsis, or cellulitis).
- Other **family** members have been diagnosed with a primary immunodeficiency disorder.

Diagnosis

Most children with hyper-IgM syndrome are diagnosed before they are a year old; about 40 percent have PCP at the time of diagnosis. If the doctor has seen the child on a regular basis since birth, there will be a record of the number of infections the child has had, the length of time the child has had each infection, and the child's response to treatment. If the doctor suspects a primary immunodeficiency disorder, he or she will ask the parents about a family history of such disorders.

The next step in diagnosis is a thorough physical examination. Children with primary immunodefiencies are often underweight or small for their age and may look pale or generally unwell. The doctor will listen for unusual sounds in the lungs when the child breathes in and out and will check the child's skin and the inside of the mouth for **rashes**, ulcers, or sores. As the doctor palpates or feels the child's abdomen, he or she will pay particular attention to the size of the spleen and liver. The doctor will also examine the child's joints and the lymph nodes in the neck for signs of swelling.

The doctor will order a blood test to screen the child for an immunodeficiency disorder. The most common tests performed to screen for hyper-IgM syndrome are a complete blood count (CBC) and a quantitative immunoglobulin test. The CBC will help to determine whether the child has neutropenia. The quantitative test measures the levels of the different types of immunoglobulins in the blood as well as the total level of all immunoglobulins. A child with hyper-IgM syndrome will be found to have abnormally low levels of IgA and IgG antibodies and a normal or elevated level of IgM.

The doctor may also order x-ray studies of the child's chest or sinuses in order to determine whether lung damage has already occurred or to make a baseline evaluation of the child's lungs.

The diagnosis of hyper-IgM syndrome can be confirmed by molecular genetics testing for the defective CD40 gene. The test involves DNA sequencing and has been available since the early 2000s.

Treatment

Intravenous immunoglobulin (IVIG) therapy

Intravenous immunoglubulin (IVIG) has been the mainstay of treatment for a number of primary immunodeficiencies since it was first approved by the Food and Drug Administration (FDA) in the early 1980s. IVIG involves the infusion of immunoglobulins derived from donated blood plasma directly into the patient's bloodstream as a protection against infection. In the case of children with XHIM, IVIG is given to replace the missing IgG antibodies and to reduce or normalize the IgM level. IVIG infusions are usually given every three to four weeks for the remainder of the patient's life. They can be given in an outpatient clinic or in the patient's home. Patients with neutropenia may be treated with G-CSF (Neupogen), a protein given by injection that stimulates the body to produce more neutrophils.

IVIG therapy is the only effective treatment for ARHIM as of the early 2000s.

Antibiotics

Boys diagnosed with XHIM are given antibiotics as a prophylactic (preventive) treatment to protect them against *Pneumocystis carinii* pneumonia. They are usually started on a regimen of trimethoprim-sulfamethoxazole (Bactrim or Septra) as soon as they are diagnosed.

Bone marrow transplantation

A subsequent treatment for XHIM is bone marrow transplantation (BMT), which is also referred to as hematopoietic stem cell transplantation (HSCT). It is considered to be a cure for primary immunodeficiency disorders. Although BMT has been performed on children with severe immunodeficiency disorders since the 1980s, it was usually restricted to those with limited life expectancy because of complications associated with transplantation. Several advances since the late 1990s, however, have made this form of treatment more feasible for boys with XHIM. These advances include better matching of potential donors and recipients through more accurate tissue typing and improved surgical techniques. As of 2004, however, doctors recommended that boys with XHIM be given BMT before significant infections or organ damage occur. This form of treatment is not recommended for patients who already have signs of liver damage.

The best source of bone marrow for transplantation is the affected child's siblings. They will be tissue-typed to determine whether their bone marrow has the same human leukocyte antigens (HLA) as the affected child. Human leukocyte antigens are genetically determined proteins that allow the body to distinguish between its own cells and those from an outside source. The closer the HLA match between a bone marrow donor and recipient, the lower the chances that the recipient's body will reject the transplanted tissue. In addition to siblings, another choice is bone marrow from one of the parents, who shares half the affected child's HLA antigens. With the expansion of bone marrow registries since the early 2000s, it is also possible to use bone marrow from an unrelated donor whose tissues closely match those of the affected child. These are called matched unrelated donor (MUD) transplants. The most successful bone marrow transplants in hyper-IgM children, however, have used marrow donated by HLA-identical siblings.

Cord blood stem cell transplantation

Another approach to transplantation as a cure for hyper-IgM syndrome is the use of stem cells from cord blood. This technique was first used for immunodeficiency disorders in 1988. Stem cells are undifferentiated precursor cells whose daughter cells can differentiate into more specialized cells. The stem cells used for transplantation are taken from blood collected from a baby's umbilical cord or the placenta (afterbirth) immediately following delivery. Cord blood from healthy siblings can be used for transplantation to treat XHIM patients. Stem cell transplants from cord blood have two advantages over bone marrow transplants: they have a lower rate of rejection in recipients, and they can be stored ahead of time. Families with a history of primary immunodeficiency disorders can save cord blood in private storage facilities for later use if needed.

Experimental and investigational treatments

Researchers have found that giving artificial CD40 ligand to specially bred immunodeficient mice improves their ability to make IgA and IgG antibodies. The National Institutes of Health (NIH) is in the early 2000s conducting studies to evaluate the effectiveness of this treatment in humans.

As of the early 2000s, researchers at the National Institutes of Health and the University of Pennsylvania are investigating the possibility of treating hyper-IgM syndrome with gene therapy. Gene therapy involves the insertion of a normal gene into a targeted cell to replace an abnormal gene by means of a vector or carrier molecule. The most common vectors are genetically altered viruses. Reports on this research published in 2004, however, indicate that gene therapy for hyper-IgM syndrome will be more complicated and take longer to develop than was originally expected.

Nutritional concerns

Nutritional concerns for children with XHIM are related to infections of the digestive tract resulting in chronic diarrhea. The primary risks with chronic diarrhea are **dehydration** and malnutrition. To prevent dehydration, the doctor may recommend a clear liquid diet for infants and toddlers during episodes of diarrhea. If the child is able to keep the clear liquids down, milk or diluted formula can be given. The child's rectal area should be coated with petroleum jelly to reduce irritation.

If the diarrhea is caused by *Cryptosporidium* or another infectious organism, the child's diapers and bed linens should be washed separately from the rest of the family's laundry, and the bathroom should be cleaned regularly with a disinfectant.

Prognosis

The prognosis for children diagnosed with XHIM is poor as of the early 2000s; morbidity and mortality for this disorder are significantly higher than for other primary immunodeficiency disorders. A study done in 2000 indicated that only 20 percent of patients with hyper-IgM syndrome survived to the age of 25. However, researchers expect the outlook to improve for children in treatment in 2004, particularly those patients who are good candidates for bone marrow transplantation. In one Japanese study, five out of seven patients who received BMT survived, with four of the five producing T cells with normal CD40 ligand without supplementary IVIG therapy. In general, children who are treated with IVIG and/or BMT as infants have a better prognosis than those who are diagnosed after the age of two years.

Prevention

As both forms of hyper-IgM syndrome are caused by genetic mutations, there is no way to prevent the disorders after the child is born. Parents who already have a child with hyper-IgM syndrome or who come from families with a history of primary immunodeficiency disorders may wish to consider genetic counseling and prenatal genetic testing with future pregnancies.

There are also some preventive measures that families can take to lower the risk of opportunistic infections and other complications in affected children. These precautions include the following:

- Practicing good hygiene, including careful washing of the hands before and after meals and after using the toilet. Antibacterial hand wipes can be packed with the child's school lunches.
- Careful cleansing of even small cuts or scrapes with an antiseptic liquid or cream.
- Proper dental care. Children with primary immunodeficiency syndromes are at increased risk of **tooth decay** and gum disorders as well as thrush and mouth ulcers.
- Avoiding the use of vaccines made from live viruses (**measles**, poliovirus, **mumps**, **rubella**). Vaccines made with killed viruses should be given regularly.
- Having the home water supply tested for possible contamination by *Cryptosporidium parvum*.
- Avoiding crowded stores, theaters, or athletic events during flu season.
- Giving the affected child his own room if possible.
- Having other family members take primary responsibility for the care of household pets. Children with hyper-IgM syndrome are highly susceptible to infection from animal bites.

Nutritional concerns

Proper **nutrition** is a lifelong concern for children with hyper-IgM syndrome because of the possibility of malnutrition caused by chronic diarrhea. A normal well-balanced diet is recommended for older children, with multivitamin supplements as prescribed by the child's primary doctor. Older children can be given a nutritional supplement (Ensure) during episodes of severe diarrhea. In extremely severe cases the child may require parenteral nutrition, which is a liquid food given intravenously. Parenteral nutrition is also known as hyperalimentation.

Parental concerns

Hyper-IgM syndrome has a major impact on the family of a child diagnosed with the disease. The following are some of the important concerns for parents:

- High financial costs. Children diagnosed with hyper-IgM syndrome require careful monitoring for liver

KEY TERMS

Antibody—A special protein made by the body's immune system as a defense against foreign material (bacteria, viruses, etc.) that enters the body. It is uniquely designed to attack and neutralize the specific antigen that triggered the immune response.

B cell—A type of white blood cell derived from bone marrow. B cells are sometimes called B lymphocytes. They secrete antibodies and have a number of other complex functions within the human immune system.

Bronchiectasis—A disorder of the bronchial tubes marked by abnormal stretching, enlargement, or destruction of the walls. Bronchiectasis is usually caused by recurrent inflammation of the airway.

Cord blood—The blood that remains in the umbilical cord and placenta after birth. Stem cells from cord blood can be used in place of bone marrow for treating primary immunodeficiency disorders.

Gene therapy—An experimental treatment for certain genetic disorders in which a abnormal gene is replaced with the normal copy. Also called somatic-cell gene therapy.

Human leuckocyte antigen (HLA)—A group of protein molecules located on bone marrow cells that can provoke an immune response. A donor's and a recipient's HLA types should match as closely as possible to prevent the recipient's immune system from attacking the donor's marrow as a foreign material that does not belong in the body.

Immunoglobulin G (IgG)—Immunoglobulin type gamma, the most common type found in the blood and tissue fluids.

Ligand—Any type of small molecule that binds to a larger molecule. Hyper-IgM syndrome is caused by a lack of a ligand known as CD40 on the surfaces of the T cells in the child's blood.

Lymphocyte—A type of white blood cell that participates in the immune response. The two main groups are the B cells that have antibody molecules on their surface and T cells that destroy antigens.

Morbidity—A disease or abnormality. In statistics it also refers to the rate at which a disease or abnormality occurs.

Neutropenia—A condition in which the number of neutrophils, a type of white blood cell (leukocyte) is abnormally low.

Opportunistic infection—An infection that is normally mild in a healthy individual, but which takes advantage of an ill person's weakened immune system to move into the body, grow, spread, and cause serious illness.

Osteomyelitis—An infection of the bone and bone marrow, usually caused by bacteria.

Primary immunodeficiency disease—A group of approximately 70 conditions that affect the normal functioning of the immune system.

Prophylactic—Preventing the spread or occurrence of disease or infection.

Stem cell—An undifferentiated cell that retains the ability to develop into any one of a variety of cell types.

T cell—A type of white blood cell that is produced in the bone marrow and matured in the thymus gland. It helps to regulate the immune system's response to infections or malignancy.

Thrush—An infection of the mouth, caused by the yeast *Candida albicans* and characterized by a whitish growth and ulcers.

function, lung function, nutritional status, **oral hygiene**, and normal growth patterns as well as blood antibody levels. In most cases the child will be seen every three to six months by a clinical immunologist as well as his primary care doctor. Such procedures as bone marrow or cord blood transplantation are also costly.

- Emotional wear and tear on the family. In addition to the extra time and attention required for the affected child, parents are likely to confront emotional problems in the family, ranging from resentment on the part of siblings to **anxiety** about the affected child's survival and decisions about future pregnancies. Support groups and/or **family therapy** may be helpful.
- Genetic testing. Genetic counselors recommend having the affected child's siblings tested to see whether they are carriers of the defective gene.
- Education and future employment. Children who are receiving IVIG treatment can attend a regular school and participate in most **sports** provided that minor injuries are treated promptly. Adults with hyper-IgM syndrome can attend college or graduate school and work in most fields of employment.

- Peer pressure in **adolescence**. It is important for parents to warn children with hyper-IgM syndrome that **smoking**, alcohol consumption, and the use of recreational drugs are far more dangerous for them than for adolescents with normal immune systems.

See also Immunodeficiency; Immunoglobulin deficiency syndromes.

Resources

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Immune Deficiency Foundation (IDF). 40 West Chesapeake Avenue, Suite 308, Towson, MD 21204. Web site: <www.primaryimmune.org>.

Jeffrey Modell Foundation (JMF). 747 Third Avenue, New York, NY 10017. Web site: <www.jmfworld.com>.

National Institute of Allergy and Infectious Diseases (NIAID). Building 31, Room 7A50, 31 Center Drive, MSC 2520, Bethesda, MD 20892–2520. Web site: <www.niaid.nih.gov>.

National Institute of Child Health and Human Development (NICHD). 31 Center Drive, Room 2A32, Bethesda, MD 20892–2425. Web site: <www.nichd.nih.gov>.

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Rebecca Frey, PhD

Hypertension

Definition

Hypertension is high blood pressure. Blood pressure is the force of blood pushing against the walls of arteries. Arteries are the blood vessels that carry oxygenated blood from the heart to the body's tissues.

Description

As blood flows through arteries, it pushes against the inside of artery walls. The more pressure the blood exerts on the artery walls, the higher the blood pressure is. The size of arteries also affects the blood pressure. When the muscular walls of arteries are relaxed, or dilated, the pressure of the blood flowing through them is lower than when the artery walls narrow, or constricted.

Blood pressure is highest when the heart beats to push blood out into the arteries. When the heart relaxes to fill with blood again, the pressure is at its lowest point. Blood pressure when the heart beats is called systolic pressure. Blood pressure when the heart is at rest is called diastolic pressure. When blood pressure is measured, the systolic pressure is stated first and the diastolic pressure second. Blood pressure is measured in millimeters of mercury (mm Hg). For example, if a person's systolic pressure is 120 and diastolic pressure is 80, it is written as 120/80 mm Hg.

Blood pressure measurements

The National Heart, Lung, and Blood Institute in Bethesda, Maryland released clinical guidelines for blood pressure in 2003, lowering the standard normal readings for adults to less than 120 over less than 80.

Although there are set blood pressure ranges for adults, normal blood pressure ranges for children vary according to age, gender, and height so that different levels of growth are considered when evaluating blood pressure. In children, blood pressure normally rises during growth and maturation and varies greatly during **adolescence**.

Specific systolic and diastolic blood pressure percentiles have been established for each age, gender, and height group. In children ages six to 12, up to 125/80 mm Hg is considered normal. In youth ages 12–15, 126/78 mm Hg is normal, and for ages 16–18, 132/82 mm Hg is normal.

Children whose blood pressure is above the 95th percentile for their age/gender/height group are diagnosed with hypertension. Children whose blood pressure is between the 90th and 95th percentile are diagnosed with pre-hypertension. Adolescents whose blood pressure is greater than 120/80 also may be diagnosed with pre-hypertension.

Complications

Childhood hypertension is serious because it increases the risk of heart disease, **stroke**, and other medical problems in adulthood. Serious complications can be avoided by ensuring the child gets regular blood pressure checks and by treating hypertension as soon as it is diagnosed.

If left untreated, hypertension can lead to the following long-term complications:

- atherosclerosis, also called arteriosclerosis
- peripheral vascular disease
- heart attack
- stroke
- enlarged heart and heart failure
- kidney damage or kidney failure
- retinopathy or blindness

Atherosclerosis is hardening of the arteries. The walls of arteries have a layer of muscle and elastic tissue that makes them flexible and able to dilate and constrict as blood flows through them. High blood pressure can make the artery walls thicken and harden. When artery walls thicken, the inside of the blood vessel narrows. Cholesterol and fats are more likely to build up on the walls of damaged arteries, making them even narrower. Blood clots also can get trapped in narrowed arteries, blocking the flow of blood. When atherosclerosis occurs in the blood vessels leading to the legs and feet, it is called peripheral vascular disease. Blood flow is decreased to the legs and feet with peripheral vascular diseases and can cause poor circulation in the legs, claudication, or aneurysm.

Arteries narrowed by atherosclerosis may not deliver enough blood to organs and other tissues. Reduced or blocked blood flow to the heart can cause a heart attack. If an artery to the brain is blocked, a stroke can result.

Hypertension makes the heart work harder to pump blood through the body. The extra workload can make the heart muscle thicken and stretch. When the heart becomes enlarged it cannot pump enough blood. If the hypertension is not treated, the heart may fail.

The kidneys remove the body's wastes from the blood. If hypertension thickens the arteries to the kidneys, less waste can be filtered from the blood. As the condition worsens, the kidneys fail and wastes build up in the blood. Dialysis or a kidney transplant is needed when the kidneys fail.

Hypertension can cause damage to blood vessels in the eyes, leading to retinopathy, or damage to the retina. Retinal damage becomes severe when blood pressure levels are high and remain elevated for a prolonged period of time.

Demographics

In the United States, an estimated 5–10 percent of children have hypertension, and one in four adults (about 50 million) have hypertension. About 30 percent of those with hypertension do not know they have it. Hypertension is more common in men than women and in people over age 65 than in younger persons. It also is more frequent and severe in African-American and Mexican-American adults and children than in white Americans. The prevalence of high blood pressure among African-Americans and whites in the southeastern United States

is greater, and death rates from stroke are higher than among those in other regions.

In the early 2000s, high blood pressure in children and adolescents is on the rise. A 2003 report indicated this increase is most likely due to a greater number of overweight and obese children and adolescents. The U.S. Centers for Disease Control and Prevention studied the health and **nutrition** of Americans in the National Health and Nutrition Examination Surveys for more than 40 years, and the last data were collected in 2000. Researchers found a trend of high blood pressure in children ages eight to 17 years who were overweight or obese.

Causes and symptoms

Causes

Many different actions or situations can normally raise blood pressure. Physical activity and changes in position can temporarily raise blood pressure. Stressful situations can make blood pressure go up. When the stress goes away, blood pressure usually returns to normal. Certain medications also may change blood pressure, but usually blood pressure returns to normal when the drug is discontinued. These temporary increases in blood pressure are not considered hypertension. A diagnosis of hypertension is made only when a person has at least three separate high blood pressure readings performed one to several weeks apart.

Hypertension without a known cause is called primary or essential hypertension. Although the cause of hypertension is unknown in 90–95 percent of adults, primary hypertension is uncommon in children, occurring in less than 1–2 percent of hypertensive children.

When a child has hypertension caused by another medical condition, it is called secondary hypertension. Secondary hypertension can be caused by a number of different illnesses. Kidney disease causes hypertension in 80–85 percent of childhood cases. The kidneys regulate the balance of salt and water in the body. If the kidneys cannot rid the body of excess salt and water, blood pressure goes up. Kidney infections, a narrowing of the arteries that carry blood to the kidneys, called renal artery stenosis, and other kidney disorders can disturb the salt and water balance.

As body weight increases, blood pressure rises. Being overweight or obese is the strongest predictor of hypertension in young adults. **Obesity** has steadily increased in children and adolescents over the years. An estimated 16 percent of school-age children are overweight. High blood pressure develops about 10 years after a young person becomes overweight. Obesity may cause other cardiovascular diseases if it is not managed or treated properly.

Risk factors

Risk factors are conditions that increase the chance of developing hypertension. Some of these risk factors can be changed to reduce the risk of developing hypertension or to lower blood pressure:

- being overweight or obese
- lack of physical activity
- a diet high in fat, salt, and sugar
- heredity
- low birth weight and subsequent rapid weight gain
- male sex
- race
- congenital conditions, such as coarctation of the aorta
- diabetes
- kidney disease
- in adolescents, heavy alcohol consumption and use of oral contraceptives
- in adults, being over the age of 60

Although **smoking** is not directly related to high blood pressure in children and adolescents, those who smoke should stop to reduce their risk of developing other health problems such as coronary artery disease.

Some risk factors for hypertension can be changed, while others cannot. Some children inherit a tendency to develop hypertension, and the risk increases if both parents are hypertensive. Children who have the risk factors above can work with their doctor and **family** to manage the controllable risk factors.

Symptoms

Hypertension generally does not cause symptoms. When symptoms occur, they are usually mild and nonspecific. In young children (age three and younger), symptoms may include:

- irritability
- excessive crying
- failure to gain weight
- poor feeding
- low-grade fever

In older children, symptoms may include:

- dizziness

- headaches
- vomiting
- heart palpitations

In severe and acute (sudden-onset) cases, hypertension can cause seizures, swelling throughout the body, blindness, or renal (kidney) failure. All of these symptoms require immediate medical attention and **hospitalization**.

When to call the doctor

If a child has any of the following symptoms, the parent or caregiver should call the child's doctor:

- unexplained headache
- sudden or gradual changes in vision
- dizziness or light-headedness that does not resolve with rest
- nausea associated with headache
- unexplained or uncontrollable vomiting
- heart palpitations

If a child has any of these symptoms, the parent or caregiver should immediately seek emergency medical attention:

- severe headache
- fainting
- seizures or convulsions
- swelling throughout the body
- unexplained blurred vision or vision loss
- severe chest **pain** or shortness of breath
- unexplained sudden weakness

Diagnosis

Blood pressure in children should be checked regularly: at least at every doctor's visit after age three. Early detection and treatment of hypertension improve the child's overall health and decrease the risk of future health problems associated with hypertension.

Blood pressure is measured with an instrument called a sphygmomanometer. A cloth-covered rubber cuff is wrapped around the upper arm and inflated. When the cuff is inflated, an artery in the arm is squeezed to momentarily stop the flow of blood. Then, the air is let out of the cuff while a stethoscope placed over the artery is used to detect the sound of the blood spurting back through the artery. This first sound is the systolic pressure, the pressure when the heart beats. The last sound heard as the rest of the air is released is the diastolic pressure, the pressure between heartbeats. Both sounds are recorded on the mercury gauge on the sphygmomanometer.

The arm cuff used to measure blood pressure in children must be appropriate to the child's size, or the reading may be inaccurate.

A typical physical examination to evaluate hypertension includes:

- medical and family history
- physical examination
- ophthalmoscopy: examination of the blood vessels in the eye
- blood and urine tests

The physical exam may include several blood pressure readings at different times and in different positions. For at least five minutes before the blood pressure reading is taken, the child should be seated in a chair, with feet on the floor and arms supported at heart level. For best results, the child should not eat or drink caffeinated products within the 30 minutes prior to the exam. The physician uses a stethoscope to listen to sounds made by the heart and blood flowing through the arteries.

During the physical exam, the child's pulse, reflexes, and height and weight are checked and recorded. Internal organs are palpated to determine if they are enlarged.

Because hypertension can cause damage to the blood vessels in the eyes, the eyes may be checked with a instrument called an ophthalmoscope. The physician will look for thickening, narrowing, or hemorrhages in the blood vessels.

Urine and blood tests may be done to evaluate health and to detect the presence of certain substances that may indicate an underlying condition that is causing the hypertension.

Usually blood tests and urine tests, along with the physical examination and medical history, are enough to make the diagnosis of hypertension. If necessary, to rule out other medical conditions or to assess any damage from hypertension and/or its treatment, the following tests may be performed:

- Chest x ray: To detect an enlarged heart, other vascular abnormalities, or lung disease.
- Electrocardiogram (ECG): To measure the electrical activity of the heart. It can detect if the heart muscle is enlarged and if there is damage to the heart muscle from blocked arteries.

- Echocardiogram (echo): To produce a graphic outline of the heart's movement, valves, and chambers, used to evaluate the function of the heart and valves. Echo is often combined with Doppler ultrasound and color Doppler. During the echo, an ultrasound transducer (hand-held wand placed on the skin of the chest) emits high-frequency sound waves to produce pictures of the heart's valves and chambers. An echo is used in pediatric patients diagnosed with hypertension to determine the extent of left ventricular hypertrophy, a condition in which the heart's main pumping vessel is enlarged.

Treatment

There is no cure for primary hypertension, but blood pressure can almost always be lowered with the correct treatment. The goal of treatment is to lower blood pressure to levels that will prevent heart disease and other complications of hypertension that could manifest in adulthood. In secondary hypertension, the disease that is responsible for the hypertension is treated in addition to the hypertension itself. Successful treatment of the underlying disorder may cure the secondary hypertension.

Clinicians should work with the child and the parents or caregivers to develop an individual treatment plan. Specific treatment goals vary. Treatment should be provided by a pediatric cardiologist or pediatrician with special knowledge and experience in the treatment of high blood pressure.

Lifestyle changes

Depending on the results of diagnostic tests, childhood hypertension is generally treated with lifestyle changes, including diet and **exercise**, before antihypertensive medication is prescribed. Lifestyle changes that may reduce blood pressure include:

- losing weight
- exercising regularly
- reducing fat, salt, and sugar in the diet
- managing stress and anxiety
- quitting smoking and reducing alcohol consumption, as applicable in older children

Reaching and maintaining a healthy body weight is important. Overweight children with hypertension are recommended to lose weight until they are within 15 percent of their healthy body weight. Even a small amount of weight loss can make a major difference. Physical activities should be encouraged, and sedentary activities such as watching television or playing **video games** should be limited. The recommended exercise goal is aerobic activity, such as brisk walking, at least 30 minutes per day, most days of the week.

A pediatrician can calculate a healthy range of body weight for the child, recommend dietary guidelines, and provide activity guidelines to help the child safely and effectively lose weight. A consultation with a registered dietitian also may assist the parent or caregiver in implementing dietary changes.

Nutritional concerns

Dietary guidelines are individualized, based on the child's blood pressure levels and specific needs. In children older than two years of age, the following low-fat dietary guidelines are recommended:

- Total fat intake should comprise 30 percent or less of total calories consumed per day.
- Calories consumed as saturated fat should equal no more than 8 to 10 percent of total calories consumed per day.
- Total cholesterol intake should be less than 300 mg/dl per day.

Elevated blood pressure can be reduced by an eating plan that emphasizes fruits, vegetables, and low-fat dairy foods, and which is low in saturated fat, total fat, and cholesterol. The DASH diet is recommended for patients with hypertension and includes whole grains, poultry, fish, and nuts. Fats, red meats, sodium, sweets, and sugar-sweetened beverages are limited. Sodium should also be reduced to no more than 1,500 milligrams per day.

A gradual transition to a heart-healthy diet can help decrease a child's risk of coronary artery disease and other health conditions in adulthood. Parents can replace foods high in fat with grains, vegetables, fruits, lean meat, and other foods low in fat and high in complex carbohydrates and protein. They can resist adding salt to foods while cooking and avoid highly processed foods that are usually high in sodium, such as fast foods, canned foods, boxed mixes, and frozen meals.

Alternative treatment

Alternative and complementary therapies include approaches that are considered to be outside the mainstream of traditional health care.

Techniques that induce relaxation and reduce stress, such as **yoga**, tai chi, meditation, guided imagery, and relaxation training, may be helpful in controlling blood pressure. Acupuncture and biofeedback training also may help induce relaxation. Before learning or practicing

any particular technique, it is important for the parent/caregiver and child to learn about the therapy, its safety and effectiveness, potential side effects, and the expertise and qualifications of the practitioner. Although some practices are beneficial, others may be harmful to certain patients.

Dietary supplements, including garlic, fish oil (omega-3 fatty acids), L-arginine, soy, coenzyme Q10, phytosterols, and chelation therapy may be beneficial, but the exact nature of their effects on blood pressure is unknown. There is little scientific evidence that these therapies lower blood pressure or prevent the complications of high blood pressure, and most of these supplements have not been studied extensively in children and adolescents.

Vitamin E and beta carotene supplements were once thought to help prevent the development of heart disease, but subsequent studies disprove that assumption.

Medications

Medications usually are not prescribed for children as a first-line treatment for hypertension. Medications are prescribed, however, to treat hypertension when the child has significant high blood pressure or organ damage, or when diet and exercise are not adequately controlling the child's blood pressure.

Follow-up care

Follow-up care for hypertension includes home blood pressure monitoring. The parent or caregiver checks the child's blood pressure at different times of the day and records the readings. The doctor reviews this blood pressure record during the child's check-ups to evaluate the effectiveness of the child's treatment and to make any necessary adjustments.

Depending on the child's blood pressure levels and presence of other medical conditions such as diabetes, the doctor may recommend annual eye exams to detect the presence of vision changes and the development of retinopathy.

Prognosis

There is no cure for hypertension. However, it can be well controlled with the proper treatment. Therapy with a combination of lifestyle changes and sometimes antihypertensive medicines usually can manage blood pressure. For most children, early primary hypertension causes no immediate risk of serious health problems, but it does increase the risk for future organ damage. The key to avoiding serious complications of hypertension is to detect and treat it at the earliest possible age so that preventive treatment can be initiated.

Prevention

Avoiding or eliminating known risk factors helps reduce the risk of developing hypertension. Making the same changes recommended for treating hypertension can reduce a child's risk of developing hypertension:

- losing weight if overweight or obese
- exercising regularly
- reducing salt, fat, and sugar in the diet
- reducing fat intake
- managing stress and anxiety
- quitting smoking and limiting alcohol, as applicable in older children

Parental concerns

Parents should reinforce with the child that hypertension is a serious condition that can cause more health problems later in life. Parents should work with their child to make dietary changes and increase their activity level to manage hypertension and prevent it from getting worse. Everyone can benefit when a heart-healthy lifestyle is followed, so the dietary and activity changes made for the hypertensive child will benefit the entire family.

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KEY TERMS

Aneurysm—A weakened area in the wall of a blood vessel which causes an outpouching or bulge. Aneurysms may be fatal if these weak areas burst, resulting in uncontrollable bleeding.

Aorta—The main artery located above the heart that pumps oxygenated blood out into the body. The aorta is the largest artery in the body.

Arteriosclerosis—A chronic condition characterized by thickening, loss of leasticity, and hardening of the arteries and the build-up of plaque on the arterial walls. Arteriosclerosis can slow or impair blood circulation. It includes atherosclerosis, but the two terms are often used synonymously.

Artery—A blood vessel that carries blood away from the heart to the cells, tissues, and organs of the body.

Atrial—Referring to the upper chambers of the heart.

Claudication—Cramping or pain in a leg caused by poor blood circulation. This condition is frequently caused by hardening of the arteries (atherosclerosis). Intermittent claudication occurs only at certain times, usually after exercise, and is relieved by rest.

Coarctation of the aorta—A congenital defect in which severe narrowing or constriction of the aorta obstructs the flow of blood.

Dialysis—A process of filtering and removing waste products from the bloodstream, it is used as a treatment for patients whose kidneys do not function properly. Two main types are hemodialysis and peritoneal dialysis. In hemodialysis, the blood flows out of the body into a machine that filters out the waste products and routes the cleansed blood back into the body. In peritoneal dialysis, the cleansing occurs inside the body. Dialysis fluid is injected into the peritoneal cavity and wastes are filtered through the peritoneum, the thin membrane that surrounds the abdominal organs.

Diastolic blood pressure—Diastole is the period in which the left ventricle relaxes so it can refill with blood; diastolic pressure is therefore measured during diastole.

Heart attack—Damage that occurs to the heart when one of the coronary arteries becomes narrowed or blocked.

Obesity—An abnormal accumulation of body fat, usually 20% or more over an individual's ideal body weight.

Overweight—Being 25 to 29 percent over the recommended healthy body weight for a specific age and height, as established by calculating body mass index.

Retinopathy—Any disorder of the retina.

Sphygmomanometer—An instrument used to measure blood pressure.

Stroke—Interruption of blood flow to a part of the brain with consequent brain damage. A stroke may be caused by a blood clot or by hemorrhage due to a burst blood vessel. Also known as a cerebrovascular accident.

Systolic blood pressure—Blood pressure when the heart contracts (beats).

Ventricles—The lower pumping chambers of the heart. The ventricles push blood to the lungs and the rest of the body.

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American College of Cardiology. Heart House, 9111 Old Georgetown Rd., Bethesda, MD 20814–1699. Web site: <www.acc.org>.

American Heart Association. 7320 Greenville Ave., Dallas, TX 75231. Web site: <www.americanheart.org>.

American Society of Hypertension. 148 Madison Ave., 5th Floor, New York, NY 10016. Web site: <www.ash-us.org>.

The Cleveland Clinic Heart Center. The Cleveland Clinic Foundation, 9500 Euclid Ave., F25, Cleveland, OH 44195. Web site: <www.clevelandclinic.org/heartcenter>.

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Hyperthyroidism

Definition

Hyperthyroidism is the overproduction of thyroid hormones by an overactive thyroid.

Description

The term hyperthyroidism covers any disease which results in overabundance of thyroid hormone. Other names for hyperthyroidism, or specific diseases within the category, include Graves' disease, diffuse toxic goiter, Basedow's disease, Parry's disease, and thyrotoxicosis.

Located in the front of the neck, the thyroid gland produces the hormones thyroxin (T4) and triiodothyronine (T3), which regulate the body's metabolic rate by helping to form protein ribonucleic acid (RNA) and increasing oxygen absorption in every cell. In turn, the production of these hormones is controlled by thyroid-stimulating hormone (TSH) that is produced by the pituitary gland. When production of the thyroid hormones increases despite the level of TSH being produced, hyperthyroidism occurs. The excessive amount of thyroid hormones in the blood increases the body's metabolism, creating both mental and physical symptoms.

Demographics

Only about 5 percent of all individuals with hyperthyroidism are younger than 15 years of age. About five times as many girls as boys develop hyperthyroidism. Almost all cases of pediatric hyperthyroidism are the form called Graves' disease. There is a form of hyperthyroidism called neonatal Graves' disease, which occurs in infants born of mothers with Graves' disease. Children with other conditions, such as trisomy 21, Addison's disease, diabetes, systemic lupus erythematosus, rheumatoid arthritis, myasthenia gravis, vitiligo, pernicious anemia, and immune thrombocytopenic purpura are more likely to develop Graves' disease.

Causes and symptoms

Hyperthyroidism is often associated with the body's production of autoantibodies in the blood which causes the thyroid to grow and secrete excess thyroid hormone. This condition, as well as other forms of hyperthyroidism, may be inherited. Regardless of the cause, hyperthyroidism produces the same symptoms, including weight loss with increased appetite, shortness of breath and fatigue, intolerance to heat, heart palpitations, increased frequency of bowel movements, weak muscles, tremors, **anxiety**, and difficulty sleeping. Adolescent girls may also notice decreased menstrual flow and irregular menstrual cycles.

Patients with Graves' disease often have a goiter (visible enlargement of the thyroid gland), although as many as 10 percent do not. These patients may also have bulging eyes. Thyroid storm, a serious form of hyperthyroidism, may show up as sudden and acute symptoms, some of which mimic typical hyperthyroidism, as well as the addition of **fever**, substantial weakness, extreme restlessness, confusion, emotional swings or psychosis, or coma. Fortunately, such a fulminant course of Graves' disease is rare in children and adolescents.

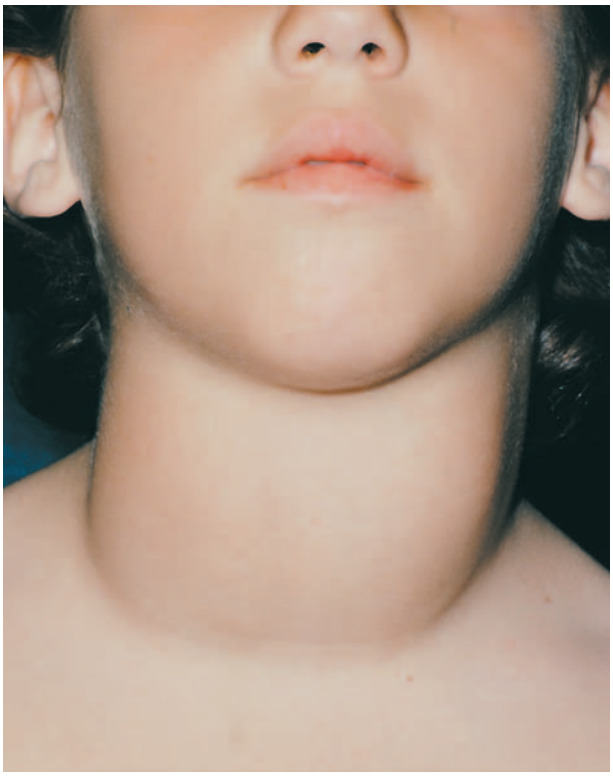
Babies with neonatal Graves' disease may suffer from **prematurity**, airway obstruction, and heart failure. Death occurs in as many as 16 percent of these babies, and other complications from which survivors may suffer include **craniosynostosis** (early closure of the sutures of the skull, which can result in compression of the growing brain), and **developmental delay**.

When to call the doctor

Parents should contact a child's pediatrician if the child shows the following symptoms: rapid weight loss, shortness of breath, intolerance to heat, heart palpitations, increased frequency of bowel movements, weak muscles, tremors, anxiety, and difficulty sleeping. An enlarged thyroid gland, seen as a bulge in the neck, should be examined by a doctor.

Diagnosis

Physicians will look for physical signs and symptoms indicated by patient history. On inspection, the physician may note symptoms such as a goiter or eye bulging. Other symptoms or **family** history may be clues to a diagnosis of hyperthyroidism. An elevated body temperature (basal body temperature) above 98.6°F (37°C) may be an indication of a heightened metabolic rate (basal metabolic rate) and hyperthyroidism. A simple blood test can be performed to determine the amount of thyroid hormone in the patient's blood. The diagnosis is usually straightforward with this combination of clinical history, physical examination, and routine blood hormone tests. Radioimmunoassay (a test to show concentrations of thyroid hormones with the use of a radioisotope mixed with fluid samples) helps confirm the diagnosis. A thyroid scan is a nuclear medicine procedure involving injection of a radioisotope dye, which tags the thyroid and helps produce a clear image of



A symptom of hyperthyroidism is the enlargement of the thyroid gland, as seen in this youngster's neck. *(Photograph by Lester V. Bergman. Corbis)*

inflammation or involvement of the entire thyroid. Other tests can determine thyroid function and thyroid-stimulating hormone levels. Ultrasonography, **computed tomography** scans (CT scan), and **magnetic resonance imaging** (MRI) may provide visual confirmation of a diagnosis or help to determine the extent of involvement.

Treatment

Treatment depends on the specific disease and individual circumstances such as age, severity of disease, and other conditions affecting a patient's health.

Antithyroid drugs

Antithyroid drugs are often administered to help the patient's body cease overproduction of thyroid hormones. This medication may work for young adults, pregnant women, and others. Women who are pregnant should be treated with the lowest dose required to maintain thyroid function in order to minimize the risk of **hypothyroidism** in the infant.

Radioactive iodine

Radioactive iodine is often prescribed to damage cells that make thyroid hormone. The cells need iodine to make the hormone, so they absorb any iodine found in the body. The patient may take an iodine capsule daily for several weeks, resulting in the eventual shrinkage of the thyroid, reduced hormone production, and a return to normal blood levels. Some patients may receive a single larger oral dose of radioactive iodine to treat the disease more quickly. This should only be done for patients who are not of reproductive age or are not planning to have children, since a large amount can concentrate in the reproductive organs (gonads).

Surgery

Some patients may undergo surgery to treat hyperthyroidism. Most commonly, patients treated with thyroidectomy, in the form of partial or total removal of the thyroid, suffer from large goiter and have suffered relapses, even after repeated attempts to address the disease through drug therapy. Some patients may be candidates for surgery because they were not good candidates for iodine therapy or refused iodine administration. Patients receiving thyroidectomy or iodine therapy must be carefully monitored for years to watch for signs of hypothyroidism (insufficient production of thyroid hormones), which can occur as a complication of thyroid production suppression.

Prognosis

Hyperthyroidism is generally treatable and carries a good prognosis. Most patients lead normal lives with proper treatment. Thyroid storm, however, can be life-threatening and can lead to heart, liver, or kidney failure. Luckily, this form of fulminant hyperthyroidism is rare in children and adolescents.

KEY TERMS

Goiter—Chronic enlargement of the thyroid gland.

Gonads—Organs that produce gametes (eggs or sperm), i.e. the ovaries and testes.

Palpitations—Rapid and forceful heartbeat.

Radioisotope—One of two or more atoms with the same number of protons but a different number of neutrons with a nuclear composition. In nuclear scanning, radioactive isotopes are used as a diagnostic agent.

Thyroidectomy—Surgical removal of all or part of the thyroid gland.

Prevention

As of 2004 there are no known prevention methods for hyperthyroidism; its causes are either inherited or not completely understood. The best prevention tactic is knowledge of family history and close attention to symptoms and signs of the disease. Careful attention to prescribed therapy can prevent complications of the disease.

Parental concerns

Parents should be aware that hyperthyroidism is very rare in young children. However, once diagnosed, the condition needs short- and long-term treatment, with regular follow-up visits to the doctor.

Resources

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PERIODICALS

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ORGANIZATIONS

Thyroid Foundation of America. 350 Ruth Sleeper Hall, RSL 350, Parkman St., Boston, MA 02114. Web site: <www.clark.net/pub/tfa>.

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Hypertonia *see* **Spasticity**

Hypoglycemia

Definition

Hypoglycemia is a condition characterized by low blood sugar, or abnormally low levels of glucose in the blood.

Description

Hypoglycemia (also known as a hypo, insulin shock, and a low) is brought on by abnormally low levels of glucose in the blood (i.e., 70 mg/dl or less). The condition is common among children with type 1 diabetes, but may also occur less frequently in children or teens with type 2 diabetes who are taking a sulfonylurea drug. An inadequate diet, improperly calculated insulin dose, minor illnesses, or excessive activity without adequate sustenance can contribute to the condition. If unchecked, hypoglycemia can lead to unconsciousness. In very rare cases, the victim may suffer a seizure.

A hypoglycemic child will appear irritable, sweaty, shaky, and confused and may complain of being very hungry. In most cases, a snack of quick-acting carbohydrates (e.g., juice or hard candy) will remedy the situation. Glucose tablets or gel can also be taken. A child who has lost consciousness due to hypoglycemia may require a glucagon shot to return blood sugar levels to normal.

Newborns of women with gestational, type 1, or type 2 diabetes during pregnancy may also experience hypoglycemia at birth, particularly if the mother's blood glucose levels were not well controlled in late pregnancy. High levels of maternal glucose cause the fetus to generate equally high levels of insulin to handle the overload, and when the maternal glucose source is disconnected at birth with the cutting of the umbilical cord, all of that insulin causes the newborn's blood sugar levels to plummet. Intravenous administration of a glucose solution to the newborn can help re-establish normal blood sugar levels.

A rare type of hypoglycemia, known as reactive hypoglycemia, may occur in children and teens without diabetes. In reactive hypoglycemia, blood glucose levels drop to 70 mg/dl approximately four hours after a meal is eaten, causing the same symptoms of low blood sugars that can occur in people with diabetes.

Also rare is fasting hypoglycemia, a condition in which blood sugars are 50 mg/dl or lower after an overnight fast or between meals. Certain medications and medical conditions can cause this problem in children who do not have diabetes.

Demographics

Among children with diabetes, hypoglycemia is much more common in those with type 1 diabetes (also known as insulin-dependent diabetes or juvenile diabetes) than in those with type 2 diabetes (formerly known as adult-onset diabetes).

Causes and symptoms

Hypoglycemia in children and teens with diabetes can be triggered by too much insulin, excessive **exercise** without proper food intake, certain oral medications, skipping meals, and drinking alcoholic beverages.

Symptoms of hypoglycemia include:

- shakiness
- nervousness
- irritability
- dizziness
- sweating
- confusion
- fatigue
- hunger
- feelings of anxiety

Reactive hypoglycemia can be triggered by enzyme disorders and by gastric bypass surgery. Causes of fasting hypoglycemia in children without diabetes may include insulin-producing tumors, certain hormonal deficiencies, medications (including sulfa drugs and large doses of aspirin), and critical illnesses. Fasting hypoglycemia is more likely to occur in children under the age of 10.

When to call the doctor

Children who are experiencing frequent episodes of hypoglycemia should see their diabetes care doctor as soon as possible as they may require an insulin adjustment, medication change, or another change in their treatment regimen.

If a child or teen with diabetes starts experiencing low blood sugars without any symptoms, he or she may be developing hypoglycemic unawareness and the child's physician should be notified immediately. In hypoglycemic unawareness, the body stops sending its normal warning signs of hypoglycemia, and a child may not realize that blood glucose levels are dangerously low until he or she loses consciousness.

Diagnosis

Episodes of hypoglycemia in children and adolescents with diabetes can be confirmed with a blood test on a home blood glucose monitor. A small needle or lancet is used to prick the finger or an alternate site and a small drop of blood is collected on a test strip that is inserted into the monitor. The monitor then calculates and displays the blood glucose reading on a screen. Although individual blood glucose targets should be determined by a medical professional in light of a child's medical history, the general goal is to keep them as close to normal (i.e., 90 to 130 mg/dl or 5 to 7.2 mmol/L before meals) as possible. Glucose levels that are below 70 mg/dl (3.9 mmol/L) are typically considered hypoglycemic.

In order to diagnose reactive hypoglycemia in those without diabetes, a blood sample must be drawn while a child is experiencing symptoms. If the blood glucose levels are 70 mg/dl or lower and the symptoms subside after food or drink is provided, reactive hypoglycemia is diagnosed.

Treatment

Children with diabetes who exhibit symptoms of hypoglycemia should check their blood glucose levels on a home glucose meter immediately. If levels are 70 mg/dl (3.9 mmol/L) or lower, they should take 15 grams of a fast-acting carbohydrate (e.g., glucose tablets, Life Savers, regular cola), wait 15 minutes, and test their blood sugars again. If levels are still too low, repeating the procedure is necessary until blood glucose is within a safe range.

Giving an unconscious child or teen food or drink by mouth can be potentially dangerous due to the possibility of **choking**. A glucagon injection should be used on a child that has lost consciousness due to hypoglycemia. Glucagon is a hormone manufactured by the pancreas that triggers the release of blood glucose by the liver. The synthetic version of the hormone is used to rapidly raise blood glucose levels in people with diabetes experiencing a severe low. A glucagon injection kit contains a syringe of sterile water and a vial of powdered glucagon. The water is injected into the glucagon vial and then mixed, and the resulting solution is drawn back into the syringe for injection into any muscular area (e.g., arm, buttock, thigh). Glucagon can cause **vomiting**, so a child that is given a glucagons injection should be monitored carefully to prevent aspiration.

Episodes of reactive and fasting hypoglycemia in children without diabetes can also be treated with a fast-acting carbohydrate.

Nutritional concerns

For children with diabetes, eating or drinking large quantities of carbohydrates in an attempt to push blood glucose levels back to normal can result in **hyperglycemia**, or blood sugars that are too high. The 15 grams/15 minutes rule is important to follow to avoid dramatic blood sugar swings.

Eating small, frequent meals and spreading carbohydrate intake throughout the day may help keep blood glucose levels from bouncing too high or too low.

Prognosis

With early detection and immediate and appropriate treatment, children will recover quickly from hypoglycemia.

Prevention

The best way to prevent hypoglycemia is to check blood glucose levels frequently and treat falling blood sugars before they become dangerously low. However, even the most dedicated child or parent may be faced with situations that trigger lows, such as a delay in restaurant service after an insulin injection has been taken or a broken hotel elevator that requires one to climb 20 flights of stairs after a vigorous workout in the pool. Because hypoglycemia can be predictable, children with diabetes and their parents should always have a source of fast-acting carbohydrate on hand for treatment.

A child diagnosed with reactive hypoglycemia can alleviate the problem by consuming small, frequent meals (about every three hours) that are heavy in high-fiber, low-sugar foods. Some physicians may also recommend a high-protein, low-carbohydrate diet.

Parental concerns

Parents of children with diabetes must work with their child's teachers and school administrators to ensure that their child is able to test his or her blood sugars regularly, take insulin as needed, and have access to food or drink to treat hypoglycemia when necessary. Someone at school should also be trained in how to administer a glucagon injection, an emergency treatment for a hypoglycemic episode when a child loses consciousness. Caregivers of children with type 1 diabetes should have access to an emergency glucagon kit and be trained in its use. This should include a responsible adult at the child's school and at any **extracurricular activities** where parents are not present.

Section 504 of the Rehabilitation Act of 1973 enables parents to develop both a Section 504 plan (which describes a child's medical needs) and an individualized education plan, or IEP (which describes what special accommodations a child requires to address those needs). An IEP should cover issues surrounding hypoglycemia detection and treatment, and should outline how these episodes should be handled.

Because children who are self-conscious about their differences may not comply with their treatment routines as well as they should when their peers are around, parents should work with schools and caregivers to ensure that their child has a clean and private place to test blood glucose levels and take injections.

KEY TERMS

Fast-acting carbohydrate—A carbohydrate that causes blood sugar levels to rise quickly rather than slowly and steadily. Also called simple sugars. Examples include glucose tablets, honey, fructose, hard candy, and cake frosting.

Glucagon—A hormone produced in the pancreas that changes glycogen, a carbohydrate stored in muscles and the liver, into glucose. It can be used to relax muscles for a procedure such as duodenography. An injectable form of glucagon is sometimes used to treat insulin shock.

Hypoglycemic unawareness—A condition in which normal warning signals of a blood sugar low, such as shakiness, sweating, or rapid heartbeat, are no longer felt.

Reactive hypoglycemia—A rare condition in which blood sugars drop below normal levels approximately four hours after eating.

Sulfonylurea drug—A medication for type 2 diabetes that causes the pancreas to produce more insulin, and may trigger hypoglycemia in some people.

Teens who drive and have type 1 diabetes should always test their blood glucose levels before getting behind the wheel, and should have a snack before driving if their levels have fallen below the low range of normal (i.e., 90 mg/dl or 5 mmol/L or lower). Keeping the glove compartment stocked with a roll of glucose tablets can help in the case of an unexpected low on the road.

Because alcohol can also trigger hypoglycemia, adolescents should be informed of the risks of drinking. Parents should let their children know that alcohol is both illegal for minors and potentially dangerous to their health, but they should also ensure that teens know what to do to avoid a dangerous low if they do choose to drink. Food should always accompany alcohol, and anyone who drinks in the evening should consider setting an alarm to test blood sugar levels during the night. Many of the symptoms of hypoglycemia can mimic intoxication, so even those teens who do not drink but do attend parties where alcohol is available should always make sure they are with someone whom they can trust who knows what to do in case of hypoglycemia.

See also Diabetes mellitus.

Resources

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ORGANIZATIONS

American Diabetes Association. 1701 North Beauregard Street, Alexandria, VA 22311. (800) 342–2383. Web site: <www.diabetes.org>.

American Dietetic Association. 216 W. Jackson Blvd., Chicago, IL 60606–6995. (312) 899–0040. Web site: <www.eatright.org>.

Children With Diabetes. Diabetes 123, Inc. 5689 Chancery Place, Hamilton, OH 45011. info@diabetes123.com. Web site: <www.childrenwithdiabetes.org>.

Juvenile Diabetes Research Foundation. 120 Wall St., 19th Floor, New York, NY 10005. (800) 533–2873. Web site: <www.jdrf.org>.

National Diabetes Information Clearinghouse. 1 Information Way, Bethesda, MD 20892–3560. (800) 860–8747. Ndic@info.niddk.nih.gov. Web site: <www.niddk.nih.gov/health/diabetes/ndic.htm>.

Paula Ford-Martin

Hypogonadism

Definition

Hypogonadism is the condition in which the production of sex hormones and germ cells (sperm and eggs) is inadequate.

Description

Gonads are the organs of sexual differentiation: in the female, they are ovaries; in the male, the testes. Along with producing eggs and sperm, they produce sex hormones that generate all the differences between men and women. If they produce too little sex hormone, then either the growth of the sexual organs or their function is impaired.

The gonads are not independent in their function, however. They are closely controlled by the pituitary gland. The pituitary hormones are the same for males and females, but the gonadal hormones are different. Men produce mostly androgens, and women produce mostly estrogens and progesterone. Androgens regulate the development of the embryo, determining whether it is a male or a female (male in the presence of androgens and female in the absence of androgens). They also direct the adolescent maturation of sex organs into their adult form. Further, they sustain other sexual organs and their function throughout the reproductive years. Estrogen and testosterone help to maintain bone mass and strength and may protect the cardiovascular system.

Hormones can be inadequate during or after each stage of development—embryonic and adolescent. During each stage, inadequate hormone stimulation will prevent normal development. After each stage, a decrease in hormone stimulation will result in failed function and perhaps some shrinkage. The organs affected principally by sex hormones are the male and female genitals, both internal and external, and the female breasts. Body hair, fat deposition, bone and muscle growth, and some brain functions are also influenced.

Demographics

Hypogonadism may occur at any age; however, consequences differ according to the age at onset. If hypogonadism occurs prenatally (even if incomplete), sexual ambiguity may result. If hypogonadism occurs before **puberty**, puberty does not progress. If hypogonadism occurs after puberty, infertility and sexual dysfunction result. The demographics of hypogonadism vary depending on the cause. XYY syndrome has an incidence of one in 1,000 newborn males. However, since many males with XYY syndrome look like other males without XYY syndrome, they may never be identified.

Kallman's syndrome (KS) is the most frequent cause of hypogonadotropic hypogonadism and affects approximately one in 10,000 males and one in 50,000 females. Kallman's syndrome is found in all ethnic backgrounds. The incidence of KS in males is about five times greater than KS in females; the reason is not known. Turner's syndrome occurs in approximately one out of every 2,500 live births. However, all but 2 percent of fetuses affected by the disorder are miscarried. Of all the chromosomal abnormalities that result in spontaneous abortion or miscarriage, Turner's syndrome is the most common, accounting for about 20 percent of all miscarriages.

Causes and symptoms

There are a number of causes of hypogonadism, including stress, elevated prolactin levels, and several genetic disorders. Sex is determined at the moment of conception by sex chromosomes. Females have two X chromosomes, while males have one X and one Y chromosome. Male sperm cells contain either an X or a Y; if the sperm with the Y chromosome fertilizes an egg, the baby will be male. Genetic defects sometimes result in

changes in the chromosomes. If sex chromosomes are involved, there is a change in the development of sexual characteristics. Female is the default sex of the embryo, so most of the sex organ deficits at birth occur in boys. Some, but not all, are due to inadequate androgen stimulation. The penis may be small, the testicles undescended (cryptorchidism) or various degrees of "feminization" of the genitals may be present.

After birth, sexual development does not occur until puberty. Hypogonadism most often shows up as an abnormality in boys during puberty. Again, not every defect is due to inadequate hormones. Some are due to too much of the wrong ones. Female problems in puberty are usually not caused by too little estrogen. Turner's syndrome leads to failure of puberty in some girls due to the lack of estrogen and progesterone production. Female reproductive problems are usually related to complex cycling rhythms gone wrong. The most common problems with too little hormone happen during menopause, which is normal hypogonadism.

A number of adverse events can damage the gonads and result in decreased hormone levels. The childhood disease **mumps**, if acquired after puberty, can infect and destroy the testicles—a disease called viral orchitis. Ionizing radiation and **chemotherapy**, trauma, several drugs (spironolactone, a diuretic, and ketoconazole, an antifungal agent), alcohol, marijuana, heroin, methadone, and environmental toxins can all damage testicles and decrease their hormone production. Severe diseases in the liver or kidneys, certain infections, **sickle cell anemia**, and some cancers also affect gonads. To treat some male cancers, it is necessary to remove the testicles, thereby preventing the androgens from stimulating **cancer** growth. This procedure, called castration or orchiectomy, removes androgen stimulation from the whole body.

For several reasons, the pituitary gland can fail to produce hormones. It happens rarely after pregnancy. The pituitary used to be removed to treat advanced breast or prostate cancer. Sometimes the pituitary develops a tumor that destroys it. Failure of the pituitary is called hypopituitarism and, of course, leaves the gonads with no stimulation to produce hormones. Besides the tissue changes generated by hormone stimulation, the only other symptoms relate to sexual desire and function. Libido is enhanced by testosterone, and male sexual performance requires androgens. The role of female hormones in female sexual activity is less clear, although hormones strengthen tissues and promote healthy secretions, facilitating sexual activity.

XYY syndrome

XYY syndrome is a chromosome disorder that affects males. Males with this disorder have an extra Y chromosome. The error that causes the extra Y chromosome can occur in the fertilizing sperm or in the developing embryo. There are no physical abnormalities in most males with XYY syndrome. However, some males can have one or more of the following characteristics. Males who have XYY syndrome are usually normal in length at birth but have rapid growth in childhood, typically averaging in the seventy-fifth percentile (taller than 75 percent of males their same age). Many males with XYY syndrome are not overly muscular, particularly in the chest and shoulders. Individuals with XYY syndrome often have difficulties with their coordination. As a result, they can appear to be awkward or clumsy. During their teenage years, males with XYY syndrome may develop severe **acne** that may need to be treated by a dermatologist.

Men with XYY syndrome have normal, heterosexual function, and most are fertile. However, numerous cases of men with XYY syndrome presenting with infertility have been reported. Most males with XYY syndrome have normal hormones involved in their sperm production. However, a minority of males with XYY syndrome may have increased amounts of some hormones involved in sperm production. This may result in infertility due to inadequate sperm production. The actual incidence of infertility in males with XYY syndrome is unknown.

Kallman's syndrome

Kallman's syndrome is a disorder of hypogonadotropic hypogonadism, delayed puberty, and anosmia (the inability to smell). Kallman's syndrome is a birth defect in the brain that prevents release of hormones and appears as failure of male puberty. Some boys have adequate amounts of androgen in their system but fail to respond to them, a condition known as androgen resistance. Hypogonadotropic hypogonadism (HH) occurs when the body does not produce enough of two important hormones, luteinizing hormone (LH) and follicle stimulating hormone (FSH). This results in underdeveloped gonads and often infertility. Anosmia, the inability to smell, was first described with hypogonadotropic hypogonadism in 1856, but it was not until 1944 that an instance of Kallman's reported the inheritance of the two symptoms together in three separate families. Hence, the syndrome of hypogonadotropic hypogonadism and anosmia was named Kallman's syndrome (KS). Affected people usually are detected in **adolescence** when they do not undergo puberty. The most common features are

HH and anosmia, though a wide range of features can present in an affected person. Other features of KS may include a small penis or undescended testicles in males, kidney abnormalities, **cleft lip** and/or palate, **clubfoot**, hearing problems, and central nervous system problems such as synkinesia (the performance of an unintended movement when making a voluntary one), eye movement abnormalities, and visual and hearing defects.

Diabetes mellitus

Type 1 diabetes (**diabetes mellitus**) occasionally has been associated with hypogonadism. Most cases seem to be due to the hypogonadism of **malnutrition** and respond to improved control. Some specific conditions associated with diabetes mellitus, such as hemachromatosis, and the Laurence-Moon Biedl, Alstrom, and Cushing syndromes, also typically produce hypogonadism. Normal gonadal function is required for normal male development of the genital tract and for maintenance of some elements of male sexual behavior. The most clearly androgen-dependent aspects include libido, sexual activity, and spontaneous erections. In normal, young males with hypogonadism, sexual acts, fantasies, and desire are significantly diminished. Spontaneous erections also decrease by approximately 40 percent. Replacement with testosterone prevents these changes, suggesting that an intact male gonadal system is required to maintain sexual function. However, visual and possibly tactile stimulus-bound erections are not impaired in males with hypogonadism after infancy. This implies that androgen action is not required to maintain the capacity for erection.

Turner's syndrome

Turner's syndrome is a genetic disorder caused by a missing X chromosome that occurs only in females. Women with Turner's syndrome are characterized by short stature, absence of secondary sexual characteristics, infertility, and a number of other physical abnormalities. Women with Turner's syndrome are born with underdeveloped ovaries that are eventually replaced by connective tissue. Because of the resulting lack of sex hormones, these individuals do not have menstrual periods and their breasts remain undeveloped, although they may develop underarm and pubic hair. Turner's syndrome does not affect **intelligence**, although persons with the condition have poor spatial perception and mathematical aptitude, often accompanied by learning disabilities.

In girls, hypogonadism during childhood will result in lack of **menstruation** and **breast development** and short height. If hypogonadism occurs after puberty, symptoms include loss of menstruation, low libido, hot flashes, and loss of body hair. In boys, hypogonadism in childhood results in lack of muscle and beard development and growth problems. In males the usual complaints are sexual dysfunction, decreased beard and body hair, breast enlargement, and muscle loss. If a brain tumor is present (central hypogonadism) there may be headaches or visual loss or symptoms of other hormonal deficiencies (such as **hypothyroidism**). In the case of the most common pituitary tumor, prolactinoma, there may be a milky breast discharge. People with **anorexia nervosa** (excessive dieting to the point of starvation) also may have central hypogonadism.

When to call the doctor

Parents should consult a **family** physician or pediatrician if their child has any signs or symptoms of hypogonadism. Establishing the cause of hypogonadism is an important first step to getting appropriate treatment. Children may require a consultation with an endocrinologist, a physician who specializes in the hormone-producing (endocrine) glands. If the primary care physician suspects the condition is present, he or she may refer the child to an endocrinologist. Parents may also consider taking the child directly to an endocrinologist without a referral.

Diagnosis

As of the early 2000s, there are accurate blood tests for most of the hormones in the body, including those from the pituitary and even some from the hypothalamus. Chromosomes can be analyzed, and gonads can be, but rarely are, biopsied. Tests may be done that check estrogen levels (women) and testosterone levels (men) as well as FSH levels and LH levels, the pituitary hormones that stimulate the gonads. Other tests may include a thyroid level; sperm count; prolactin level (milk hormone); blood tests for anemia, chemistries, and iron; and genetic analysis. Sometimes imaging is necessary, such as a sonogram of the ovaries. If pituitary disease is suspected, a **magnetic resonance imaging** (MRI) or computerized tomography (CT) scan of the brain may be done.

Treatment

Replacement of missing body chemicals is much easier than suppressing excesses. Estrogen is recommended only to control hot flashes and sweats after menopause but is used in young women who do not produce hormones on their own. Estrogen can be taken by mouth, injection, or skin patch. It is strongly recommended that the other female hormone, progesterone, be

taken by women who have an intact uterus as well, because doing so prevents overgrowth of uterine lining and uterine cancer. Testosterone replacement is available for males who are deficient. Testosterone comes in the form of an injection, patch, or gel.

Alternative treatment

Ethinyl estradiol, an estrogen derivative, is sometimes used for the treatment of hypogonadism.

Prognosis

Many forms of hypogonadism are potentially treatable and have a good prognosis.

Prevention

People should maintain normal body weight and have healthy eating habits to prevent anorexia nervosa. Other causes may not be preventable.

Parental concerns

Adolescents with hypogonadism may have problems fitting in socially due to delayed sexual development. Testosterone replacement therapy can induce puberty, and at a slow pace in order to allow time for adjustment to body changes and new feelings. In girls with hypogonadism, complications include the social implication of failing to go through puberty with peers (if hypogonadism occurs before puberty). A supportive family that understands the diagnosis of hypogonadism is important. Adolescents may need psychological or family counseling. Support groups can help people with hypogonadism and related conditions cope with similar situations and challenges.

See also Intersex states; Turner syndrome.

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KEY TERMS

Biopsy—The surgical removal and microscopic examination of living tissue for diagnostic purposes or to follow the course of a disease. Most commonly the term refers to the collection and analysis of tissue from a suspected tumor to establish malignancy.

Diabetes mellitus—The clinical name for common diabetes. It is a chronic disease characterized by the inability of the body to produce or respond properly to insulin, a hormone required by the body to convert glucose to energy.

Embryo—In humans, the developing individual from the time of implantation to about the end of the second month after conception. From the third month to the point of delivery, the individual is called a fetus.

Endocrinologist—A physician who specializes in treating patients who have diseases of the thyroid, parathyroid, adrenal glands, and/or the pancreas.

Fetus—In humans, the developing organism from the end of the eighth week to the moment of birth. Until the end of the eighth week the developing organism is called an embryo.

Hypothalamus—A part of the forebrain that controls heartbeat, body temperature, thirst, hunger, body temperature and pressure, blood sugar levels, and other functions.

Ionizing radiation—Radiation that can damage living tissue by disrupting and destroying individual cells at the molecular level. All types of nuclear radiation–x rays, gamma rays, and beta rays–are potentially ionizing. Sound waves physically vibrate the material through which they pass, but do not ionize it.

Kallman's syndrome—A disorder of hypogonadotropic hypogonadism, delayed puberty, and anosmia. Kallman's syndrome is a birth defect in the brain that prevents release of hormones and appears as failure of male puberty.

Turner syndrome—A chromosome abnormality characterized by short stature and ovarian failure caused by an absent X chromosome. It occurs only in females.

Undescended testicle—A testicle that is still in the groin and has not made its way into the scrotum.

XYY syndrome—A chromosome disorder that affects males.

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Hypopigmentation *see* **Albinism**

Hypospadias

Definition

Hypospadias is a congenital defect of the penis in which the urinary tract opening, or urethral meatus, is abnormally located away from the tip of the penis.

Description

In males with hypospadias, the urinary opening is located on the underside of the penis. Often there is an accompanying underdevelopment of the foreskin in which the penis has a hooded appearance. Most of the foreskin is located on the top and sides of the tip of the penis. The urethral meatus may be located at any point along the penile shaft from just below the tip of the penis to closer to the body and/or near the scrotum. It may appear as a small hole in the penis or, in more severe cases, may be a longer slit-like opening. Some cases may involve chordee, a condition in which the penis bends down or away from the body during erection.

Demographics

Hypospadias is the most common anomaly of the penis affecting approximately one in 250 males born. Research has shown a doubling of the number of babies born with this anomaly. The reason for this increase is as of 2004 unknown.

Causes and symptoms

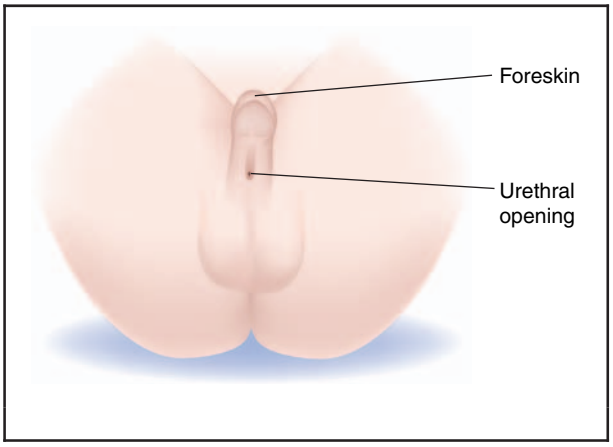
Hypospadias is a congenital anomaly resulting from incomplete closure of the tissue of the penis that forms the urethra (the tube that carries urine from the bladder to the outside of the body). The potential symptoms of hypospadias if left untreated include an abnormal direction of the urine stream, abnormal appearance of the penis, infertility if the defect is located far enough away from the tip of the penis, and an inability to have sexual intercourse in cases involving chordee.

Diagnosis

Hypospadias is diagnosed most often during the initial newborn physical examination and is classified based on where the urethral meatus is located. In rare cases infants with hypospadias occurring closer to the body and who also have undescended testicles, a karyotype or genetic screen may be performed to determine gender. Males who have hypospadias located within or near the scrotum should also have a procedure called a voiding cystogram to rule out additional urinary tract anomalies. In general, very few babies with hypospadias have other birth defects. Many males with multiple congenital anomalies, however, may also have hypospadias.

Prognosis

The prognosis for boys who have undergone hypospadias repair is excellent. Very few children experience complications. In most cases, the penis appears normal and functions normally. Less than 5 percent of children with mild hypospadias experience postoperative complications. Complications include wound infections, unexpected opening near the repair



Hypospadias, a condition in which the urethral opening is not at the tip of the penis, but rather along the penile shaft. *(Illustration by Argosy, Inc.)*

site, and rarely, meatul stenosis, a narrowing of the urinary tract opening.

Prevention

There was as of 2004 no known prevention of hypospadias.

Parental concerns

Most hypospadias cases are minor and involve few complications. Initially, parents should be sure their son is not circumcised because the foreskin is often essential in hypospadias repair surgery. Should the parents decide to allow corrective surgery, they should find a pediatric urologic surgeon with experience in performing hypospadias repairs. After surgery, care must be taken to follow all postoperative instructions and to obtain follow-up care from both the pediatrician and pediatric urologist. Parents may be concerned about the appearance and function of the penis. In most cases, following hypospadias repair surgery, the penis functions normally and is normal in appearance as well. Most males who have had a hypospadias repair are able to stand to urinate, experience normal sexual function, and normal fertility. Parents may be concerned about the physical and emotional **pain** of genital surgery. The recommended age of surgical repair is between four and 12 months. This age is ideal for many reasons including the size of the penis and the slow rate of growth of the penis at this age, the relatively low risk from anesthesia, and the fact that children at this age have not formed long-term memory and will not remember the surgery.

KEY TERMS

Anesthesia—Treatment with medicine that causes a loss of feeling, especially pain. Local anesthesia numbs only part of the body; general anesthesia causes loss of consciousness.

Chordee—An abnormal curvature of the penis.

Circumcision—A surgical procedure, usually with religious or cultural significance, where the prepuce or skin covering the tip of the penis on a boy, or the clitoris on a girl, is cut away.

Congenital—Present at birth.

Foreskin—A covering fold of skin over the tip of the penis.

Karyotype—A standard arrangement of photographic or computer-generated images of chromosome pairs from a cell in ascending numerical order, from largest to smallest.

Scrotum—The external pouch containing the male reproductive glands (testes) and part of the spermatic cord.

Urethra—A passageway from the bladder to the outside of the body for the discharge of urine. In the female this tube lies between the vagina and clitoris; in the male the urethra travels through the penis and opens at the tip. In males, seminal fluid and sperm also pass through the urethra.

Urethral meatus—The opening of the urethra on the body surface through which urine is discharged.

Voiding cystogram—A radiographic image of the mechanics of urination.

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Hypothyroidism

Definition

Hypothyroidism, or underactive thyroid, develops when the thyroid gland fails to produce or secrete as much thyroxine (T_4) and triiodothyonine (T_3) as the body needs. Because these thyroid hormones regulate such essential functions as heart rate, digestion, physical growth, and mental development, an insufficient supply of this hormone can slow metabolic processes, damage organs and tissues in every part of the body, and lead to life-threatening complications.

Description

Hypothyroidism is one of the most common chronic diseases in the United States. Symptoms may not appear until years after the thyroid has stopped functioning and often are mistaken for signs of other illnesses. Although this condition is believed to affect up to 11 million adults and children, as many as two out of every three people with hypothyroidism may not know they have the disease.

Nicknamed "Gland Central" because it influences almost every organ, tissue, and cell in the body, the thyroid is shaped like a butterfly and located just below the larynx, or Adam's apple, and in front of the trachea, or windpipe. The thyroid stores iodine that the body obtains from food, and uses this mineral to create the thyroid hormones. Low thyroid hormone levels can alter weight, appetite, **sleep** patterns, body temperature, and a variety of other physical, mental, and emotional characteristics.

Although hypothyroidism is most common in women who are middle-aged or older, the disease can occur at any age. In addition, an infant can be born with congenital hypothyroidism, i.e., without a functioning thyroid. In older children, the development of hypothyroidism may progress slowly and it may be several years before the disease is diagnosed.

Demographics

The most common cause of hypothyroidism in mid- to late-childhood and **adolescence** is Hashimoto's thyroiditis, which occurs in up to 1.2 percent of the school age population. Congenital hypothyroidism is less common. One out of every 4,000–5,000 infants is born without a properly functioning thyroid gland. Congenital hypothyroidism is twice as common in girls as in boys and about five times more common in whites than in blacks.

Causes and symptoms

Congenital hypothyroidism is a disorder that affects infants from birth, resulting from the loss of thyroid function due to the failure of the thyroid gland to develop correctly. Sometimes the thyroid gland is absent or is ectopic, i.e., in an abnormal location. This congenital defect means that the infant does not produce sufficient thyroid hormones, resulting in abnormal growth and development as well as slower mental function.

Hypothyroidism may also be caused by an abnormality of the immune system that results in damage and destruction of the thyroid gland (Hashimoto's thyroiditis). This process can result in either loss of thyroid tissue or enlargement of the thyroid. In most cases, there is no **pain** or tenderness associated with this disease, although sometimes persons affected complain of difficulty in swallowing, as if they had a lump in the throat.

Less often, hypothyroidism develops when the pituitary gland fails and does not release enough thyroid-stimulating hormone (TSH), which stimulates the thyroid to produce and secrete normal amounts of T_4 and T_3. TSH may be deficient for several reasons:

- disease of the pituitary gland (occurs rarely)
- disease of the hypothalamus (located about the pituitary), which stimulates the pituitary gland
- tumor, cyst, or other abnormal structure between the hypothalamus and pituitary gland that prevents the pituitary from receiving the stimulus to secrete TSH

Other causes of hypothyroidism include:

- Radiation. Radioactive iodine used to treat **hyperthyroidism** (overactive thyroid) or radiation treatments for head or neck cancers can destroy the thyroid gland.
- Surgery. Removal of the thyroid gland because of **cancer** or other thyroid disorders can result in hypothyroidism.
- Viruses and bacteria. Infections that depress thyroid hormone production usually cause permanent hypothyroidism.
- Medication. Nitroprusside, lithium, or iodides can induce hypothyroidism. Because patients who use these medications are closely monitored by their doctors, this side effect is very rare.
- Environmental contaminants. Certain man-made chemicals such as PCBs, found in the local environment at high levels, may also cause hypothyroidism.

Often babies with congenital hypothyroidism will appear normal at birth, which is why screening is vital.

However, some infants may have one of more of the following symptoms:

- large size (despite poor feeding habits) and increased birth weight
- puffy face and swollen tongue
- hoarse cry
- low muscle tone
- cold extremities
- persistent **constipation**, with distended abdomen
- lack of energy, sleeping most of the time and appearing tired when awake
- little or no growth

Children born with symptoms have a greater risk of **developmental delay** than children born without symptoms. The longer a child with hypothyroidism remains untreated, the greater is the loss of intellectual capacity, as measured by the standard **intelligence** testing (IQ). The ultimate IQ has been shown to be significantly higher in children whose hypothyroidism was detected and treated prior to six weeks of age, compared to those children whose hypothyroidism went untreated for six to 12 weeks.

Hypothyroidism that develops after birth is sometimes referred to as a silent disease because early symptoms may be so mild that no one realizes anything is wrong. Untreated symptoms become more noticeable and severe, and can lead to confusion and mental disorders, breathing difficulties, heart problems, fluctuations in body temperature, and death.

A child or adolescent who has hypothyroidism may have one or more of the following symptoms:

- fatigue
- decreased heart rate
- progressive hearing loss
- weight gain
- problems with memory and concentration
- depression
- goiter (enlarged thyroid gland)
- muscle pain or weakness
- numb, tingling hands
- dry skin
- swollen eyelids
- dryness or loss of hair
- extreme sensitivity to cold
- constipation
- delayed (common) or early (rare) onset of sexual development at adolescence
- irregular menstrual periods
- elevated cholesterol levels in the blood
- hoarse voice

Although hypothyroidism usually develops gradually, when the disease results from surgery or other treatment for hyperthyroidism, symptoms may appear suddenly and include severe **muscle cramps** in the arms, legs, neck, shoulders, and back.

People whose hypothyroidism remains undiagnosed and untreated may eventually develop myxedema. Symptoms of this rare, but potentially deadly, complication include enlarged tongue, swollen facial features, hoarseness, and physical and mental sluggishness. Myxedema coma can cause unresponsiveness; irregular, shallow breathing; low blood sugar; and drops in blood pressure and body temperature. The onset of this medical emergency can be sudden in children with undiagnosed hypothyroidism; it can be brought on by illness, injury, surgery, use of sedatives or anti-depressants, or exposure to very cold temperatures. Without immediate medical attention, myxedema coma can be fatal.

When to call the doctor

The doctor should be called if signs of hypothyroidism or myxedema are present. Every child who has a decrease in rate of growth in height during childhood and adolescence should be tested to determine if the growth problem is caused by hypothyroidism.

Diagnosis

In the United States, newborn infants between 24 and 72 hours old are tested for congenital thyroid deficiency (cretinism) using a test that measures the levels of thyroxine in the infant's blood. If the levels are low, the physician will likely repeat the blood test to confirm the diagnosis. The physician may take an x ray of the infant's legs. In an infant with hypothyroidism, the ends of the bones have an immature appearance. Treatment within the first few months of life can prevent **mental retardation** and physical abnormalities.

Older children who develop hypothyroidism may suddenly stop growing. If the child was above average height before the disease occurred, he or she may now be short compared to other children of the same age. Therefore, the most important feature of hypothyroidism in a child is a decrease in the rate of growth in height. If the disease is recognized early and adequately treated, the child will grow at an accelerated rate until

reaching the same growth percentile where the child measured before the onset of hypothyroidism. Diagnosis of hypothyroidism is based on the patient's observations, medical history, physical examination, and thyroid function tests. Doctors who specialize in treating thyroid disorders (endocrinologists) are most likely to recognize subtle symptoms and physical indications of hypothyroidism. A diagnostic evaluation may include a blood test known as a thyroid-stimulating hormone (TSH) assay, thyroid nuclear medicine scan, thyroid ultrasound, or needle aspiration biopsy (which is also used to provide information on thyroid masses). All patients should be sure their doctors are aware of any recent procedures involving radioactive materials or contrast media.

The blood test is extremely accurate, but some doctors doubt its ability to detect mild hypothyroidism. They advise patients to monitor their basal (resting) body temperature for below-normal readings that could indicate the presence of hypothyroidism.

Alternative treatment

Alternative treatments are primarily aimed at strengthening the thyroid but will not eliminate the need for thyroid hormone medications. Herbal remedies to improve thyroid function and relieve symptoms of hypothyroidism include bladder wrack (*Fucus vesiculosus*), which can be taken in capsule form or as a tea. The shoulder stand **yoga** position (done at least once daily for 20 minutes) is believed to improve thyroid function.

Nutritional concerns

Because the thyroid makes T_4 from iodine in food, an iodine-deficient diet can cause hypothyroidism. Adding iodine to table salt and other common foods has eliminated iodine deficiency in the United States. Some foods, including cabbage, rutabagas, radishes, peanuts, peaches, soybeans, and spinach, can interfere with thyroid hormone production. Anyone with hypothyroidism may want to avoid these foods. A high-fiber diet along with regular **exercise** is recommended to help maintain thyroid function and prevent constipation.

Prognosis

Thyroid hormone replacement therapy generally maintains normal thyroid hormone levels unless treatment is interrupted or discontinued.

KEY TERMS

Cretinism—Severe hypothyroidism that is present at birth and characterized by severe mental retardation.

Endocrine system—A group of ductless glands and parts of glands that secrete hormones directly into the bloodstream or lymphatic system to control metabolic activity. Pituitary, thyroid, adrenals, ovaries, and testes are all part of the endocrine system.

Goiter—Chronic enlargement of the thyroid gland.

Hyperthyroidism—A condition characterized by abnormal over-functioning of the thyroid glands. Patients are hypermetabolic, lose weight, are nervous, have muscular weakness and fatigue, sweat more, and have increased urination and bowel movements. Also called thyrotoxicosis.

Hypothalamus—A part of the forebrain that controls heartbeat, body temperature, thirst, hunger, body temperature and pressure, blood sugar levels, and other functions.

Myxedema—Severe hypothyroidism, characterized by swelling of the face, hands, and feet, an enlarged tongue, horseness, and physical and mental sluggishness.

Pituitary gland—The most important of the endocrine glands (glands that release hormones directly into the bloodstream), the pituitary is located at the base of the brain. Sometimes referred to as the "master gland," it regulates and controls the activities of other endocrine glands and many body processes including growth and reproductive function. Also called the hypophysis.

Thyroid-stimulating hormone (TSH)—A hormone produce by the pituitary gland that stimulates the thyroid gland to produce the hormones that regulate metabolism. Also called thyrotropin.

Thyroxine (T_4)—The thyroid hormone that regulates many essential body processes.

Triiodothyronine (T_3)—A thyroid hormone similar to thyroxine but more powerful. Preparations of triiodothyronine are used in treating hypothyroidism.

Prevention

Hypothyroidism usually cannot be prevented, but the symptoms and effects of the disease can be controlled by prompt diagnosis and treatment.

Parental concerns

Parents must ensure that medication is taken on a routine basis by making the process a part of the family's lifestyle. Taking the medication as prescribed helps assure the child's optimal growth and development.

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Endocrine Society. 4350 East West Highway, Suite 500, Bethesda, MD 20814-4410. (301) 941-0200. Web site: <www.endo- society.org>.

Thyroid Foundation of America, Inc. Ruth Sleeper Hall, RSL 350, Boston, MA 02114-2968. (800) 832-8321 or (617) 726-8500. Web site: <www.tsh.org>.

Thyroid Society for Education and Research. 7515 S. Main St., Suite 545, Houston, TX 77030. (800) THYROID or (713) 799-9909. Web site: <www.the-thyroid-society.org>.

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Hypotonia

Definition

Hypotonia, or severely decreased muscle tone, is seen primarily in children. Low-toned muscles contract very slowly in response to a stimulus and cannot maintain a contraction for as long as a normal muscle. Hypotonia is a symptom that can be caused by many different conditions.

Description

Hypotonia, also called floppy infant syndrome or infantile hypotonia, is a condition of decreased muscle tone. The low muscle tone can be caused by a variety of conditions and is often indicative of the presence of an underlying central nervous system disorder, genetic disorder, or muscle disorder. Muscle tone is the amount of tension or resistance to movement in a muscle. It is not the same as muscle weakness, which is a reduction in the strength of a muscle, but it can co-exist with muscle weakness. Muscle tone indicates the ability of a muscle to respond to a stretch. For example, if the flexed arm of a child with normal tone is quickly straightened, the flexor muscle of the arm (biceps) will quickly contract in response. Once the stimulus is removed, the muscle then relaxes and returns to its normal resting state. A child with low muscle tone has muscles that are slow to start a muscle contraction. Muscles contract very slowly in response to a stimulus and cannot maintain a contraction for as long as a normal muscle. Because low-toned muscles do not fully contract before they again relax, they remain loose and very stretchy, never achieving their full potential of sustaining a muscle contraction over time.

Hypotonic infants, therefore, have a typical "floppy" appearance. They rest with their elbows and knees loosely extended, while infants with normal muscle tone tend to have flexed elbows and knees. Head control is usually poor or absent in the floppy infant with the head falling to the side, backward, or forward. Infants with normal tone can be lifted by placing hands under their armpits, but hypotonic infants tend to slip between the hands as their arms rise unresistingly upward. While most children tend to flex their elbows and knees when resting, hypotonic children hang their arms and legs limply by their sides. Infants with this condition often lag behind in reaching the fine and gross motor developmental milestones that enable infants to hold their heads up when placed on the stomach, balance themselves, or get into a sitting position and remain seated without falling over. Hypotonia is also characterized by problems with mobility and posture, lethargy, weak ligaments and joints, and poor reflexes. Since the muscles that support the bone joints are so soft, there is a tendency for hip, jaw, and neck dislocations to occur. Some hypotonic children also have trouble feeding and are unable to suck or chew for long periods. Others may also have problems with speech or exhibit shallow breathing. Hypotonia does not, however, affect intellect.

Demographics

No demographic information as of 2004 was available for hypotonia, since it is a symptom of an underlying disorder. However, a study conducted in year 2000 by the University of Illinois provides some insights. The study followed 243 infants with hypotonia for three to seven years. By the age of three, about 30 percent had minimal problems and 46 percent had significant impairments, while 24 percent of the infants were normal. Hypotonic infants who matured into children with minimal disabilities were highly likely to have poor motor coordination at age three (78%). About 25 percent had learning problems or **language delay**; 20 percent had borderline cognition or attention deficits; and 66 percent had two or more of these characteristics.

Causes and symptoms

Hypotonias are often of unknown origin. Scientists believe that they may be caused by trauma; environmental factors; or by other genetic, muscle, or central nervous system disorders. The National Institutes of Health list the following common causes of hypotonia:

- Down syndrome: a chromosome abnormality, usually due to an extra copy of the twenty-first chromosome.
- Myasthenia gravis: a neuromuscular disorder characterized by variable weakness of voluntary muscles, which often improves with rest and worsens with activity. The condition is caused by an abnormal immune response.
- Prader-Willi syndrome: a congenital disease characterized by **obesity**, severe hypotonia, and decreased mental capacity
- Kernicterus: also called Rh incompatibility, a condition that develops when there is a difference in Rh blood type between that of the mother (Rh negative) and that of the fetus (Rh positive).
- Cerebellar ataxia: a movement disorder which with its sudden onset, often following an infectious viral disease, causes hypotonia.
- Infant **botulism**: a type of botulism, in which *Clostridium botulinum* bacteria grow within an infant's digestive tract, producing a toxin which is potentially life-threatening.
- Familial dysautonomia: also called Riley-Day syndrome, an inherited disorder that affects the function of nerves throughout the body.
- Marfan syndrome: an inherited disorder of connective tissue (tissue that adds strength to the body's structures), affecting the skeletal system, cardiovascular system, eyes, and skin.
- Muscular dystrophy: a group of disorders characterized by progressive muscle weakness and loss of muscle tissue.
- Achondroplasia: a disorder of bone growth that causes the most common type of dwarfism.
- Trisomy 13: a syndrome associated with the presence of a third number 13 chromosome.
- Sepsis: a severe, life-threatening illness caused by overwhelming infection of the bloodstream by toxin-producing bacteria.
- Aicardi syndrome: a rare genetic disorder characterized by infantile spasms (jerking), absence of the corpus callosum (the connection between the two hemispheres of the brain), **mental retardation**, and lesions of the retina of the eye or optic nerve.
- Canavan disease: an inherited metabolic disorder characterized by degeneration of the white matter of the brain.
- Congenital **hypothyroidism**: a disorder that results from decreased thyroid hormone production.
- Hypervitaminosis D: a condition that appears several months after excessive doses of vitamin D are administered.
- Krabbe disease: an inherited disorder characterized by a deficiency of the enzyme galactosylcereamidase, resulting in destruction of myelin, the fatty material that surrounds and insulates many of the nerves.
- Metachromatic leukodystrophy: an inherited disease characterized by the absence of the enzyme arylsulfatase A, which causes a material called cerebroside sulfate to accumulate in cells, which is toxic to cells, especially to the cells of the nervous system.
- Methylmalonic academia: an inherited metabolic disorder, usually diagnosed in infancy, which causes the accumulation of methylmalonic acid in the body and can lead to severe metabolic disturbances.
- Rickets: a childhood disorder involving softening and weakening of the bones, primarily caused by lack of vitamin D, calcium, or phosphate.
- Spinal muscular atrophy type 1 (Werdnig-Hoffman): a group of inherited diseases causing progressive muscle degeneration and weakness, eventually leading to death.
- Tay-Sachs disease: a genetic disorder found predominantly in Ashkenazi Jewish families results in early death.
- Vaccine reaction: any injury or condition that occurs as a result of a vaccination.

The following are common symptoms associated with hypotonia. Each child may experience different

symptoms, depending on the underlying cause of the hypotonia:

- decreased muscle tone; muscles feel soft and doughy
- ability to extend limb beyond its normal limit
- failure to acquire motor skill developmental milestones (such as holding head up without support from parent, rolling over, sitting up without support, walking)
- feeding problems (inability to suck or chew for prolonged periods)
- shallow breathing
- mouth hangs open with tongue protruding (underactive gag reflex)

When to call the doctor

Normally developing children tend to develop motor skills, posture control, and movement skills by a given age. Motor skills are divided into two categories. **Gross motor skills** include the ability of an infant to lift its head while lying on the stomach, to roll over from its back to its stomach. Normally, by a given age, a child develops the gross motor skills required to get into a sitting position and remain seated without falling over, crawl, walk, run, and jump. **Fine motor skills** include the ability to grasp, transfer an object from one hand to another, point out an object, follow a toy or a person with the eyes, or to feed oneself. Hypotonic children are slow to develop these skills, and parents should contact their pediatrician if they notice such delays or if their child appears to lack muscle control, especially if the child previously seemed to have normal muscle control.

Diagnosis

Hypotonia is normally discovered within the first few months of life. Since it is associated with many different underlying disorders, the doctor will accordingly seek to establish a **family** history as well as the child's medical history. A physical examination will be performed, usually including a detailed nervous system and muscle function examination. The latter may be performed with instruments, such as lights and reflex hammers, and usually does not cause any **pain** to the child. Most of the disorders associated with hypotonia also cause other symptoms that, when taken together, suggest a specific disorder and cause for the hypotonia. Specific diagnostic tests used will vary depending on the suspected cause of the hypotonia. Typical medical history questions include:

- When was the hypotonia first noticed?
- Was it present at birth?
- Did it start suddenly or gradually?
- Is the hypotonia always the same or does it seem worse at certain times?
- Is the child limp all over or only in certain areas?
- What other symptoms are present?

The following diagnostic tests may also be used:

- Blood tests.
- Creatine kinase (CK) test: elevated CK level in blood indicating muscles are damaged or degenerating.
- Computerized tomography scan (CT scan): a diagnostic imaging procedure that uses a combination of x-rays and computer technology to produce cross-sectional images. CT scans help physicians evaluate bone and muscle structures.
- Magnetic resonance imaging (MRI): a diagnostic procedure that uses a combination of large magnets, radio frequencies, and a computer to produce detailed images of organs and structures within the body.
- Electromyogram (EMG): a test used to evaluate nerve and muscle function.
- Electroencephalogram (EEG): a test that measures the electrical activity in the brain. An EEG measures brain waves through small button electrodes that are placed on the child's scalp.
- Spinal tap: also called lumbar puncture, measures the amount of pressure in the spinal canal and/or to remove a small amount of cerebral spinal fluid (**CSF**) for testing. Cerebral spinal fluid bathes the brain and spinal cord.
- Karyotype: a test that performs a chromosomal analysis from a blood test, used to determine whether the hypotonia is the result of a genetic disorder.
- Muscle biopsy: a sample of muscle tissue removed and examined under a microscope. Hypotonia can be assessed because muscle fibers have a smaller diameter than that of normal muscle.

Treatment

When hypotonia is caused by an underlying condition, that condition is treated first, followed by symptomatic and supportive therapy for the hypotonia. Physical therapy can improve fine motor control and overall body strength. Occupational and speech-language therapy can help breathing, speech, and swallowing difficulties. Therapy for infants and young children may also include sensory stimulation programs. Specific treatment for hypotonia is determined by the child's physician based on the following:

- the child's age, overall health, and medical history
- the extent of the condition
- the underlying cause of the condition
- the child's tolerance for specific medications, procedures, or therapies
- expectations for the course of the condition
- parent opinion or preference

No specific treatment is required to treat mild congenital hypotonia, but children with this problem may periodically need treatment for common conditions associated with hypotonia, such as recurrent joint dislocations. Treatment programs to help increase muscle strength and sensory stimulation programs are developed once the cause of the child's hypotonia is established. Such programs usually involve physical therapy through an early intervention or school-based program among other forms of therapy.

Hypotonic children are often treated by one or more of the following specialists:

- Developmental pediatrician: a pediatrician with specialized training in children's social, emotional, and intellectual development as well as health and physical growth. He or she may conduct a developmental **assessment** which will determine any delays the child has and to what extent the delay is present.
- Neurologist: a physician who has trained in the diagnosis and treatment of nervous system disorders, including diseases of the brain, spinal cord, nerves, and muscles. Neurologists perform neurological examinations of the nerves of the head and neck; muscle strength and movement; balance, ambulation, and reflexes; and sensation, memory, speech, language, and other cognitive abilities.
- Geneticist: a specialist in genetic disorders. He or she starts with the detailed history of the family's background, looks at the child's features and orders blood tests to look at the 46 chromosomes and possibly at specific genes on those chromosomes.
- Occupational therapist (OT): a professional who has specialized training in helping to develop mental or physical skills that help accomplish daily living activities, with careful attention to enhancing fine motor skills. In a developmental assessment, the occupational therapist assesses the child's fine motor skills, coordination, and age-appropriate self-help skills (eating with utensils, dressing, etc.).
- Physical therapist (PT): a professional trained in assessing and providing therapy to treat developmental delays using methods such as **exercise**, heat, light, and massage. In a developmental assessment, the physical therapist assesses the ability and quality of the child's use of legs, arms, and complete body by observing the display of specific gross motor skills as well as observing the child in play.
- Speech/language pathologist (SLP): a professional who is trained in assessing and treating problems in communication. Some SLPs are also trained to work with oral/motor problems, such as swallowing, and other feeding difficulties resulting from hypotonia.

Nutritional concerns

In some hypotonic infants, sucking is weak and in some cases not present at all. They do not act hungry or show interest in feeding. Special techniques and procedures are then required to provide adequate **nutrition**, such as special nipples, manipulation of mouth and jaw, and on rare occasions, insertion of a gastrostomy tube.

Prognosis

The outcome in any particular case depends largely on the nature of the underlying disease. Hypotonia can be life long, but in some cases, muscle tone improves over time. Children with mild hypotonia may not experience **developmental delay**, although some children acquire gross motor skills (sitting, walking, running, jumping) more slowly than most. Most hypotonic children eventually improve with therapy and time. By age five, they may not be the fastest child on the playground, but many will be there with their peers and will be holding their own. Some children are more severely affected, requiring walkers and wheelchairs and other adaptive and assistive equipment.

Prevention

As of 2004 there was no prevention for hypotonia. However, measures of prevention are increasingly possible in the early 2000s for several underlying disorders.

Parental concerns

Parents of an hypotonic child must follow the treating physician's orders for treatment of the underlying cause. They must exercise special care when lifting and carrying the hypotonic infant to avoid causing an injury to the child. If lifted under the armpits, the hypotonic infant's arms will raise with no resistance and easily slip between the hands.

A six-week-old baby girl is held horizontally by the trunk in a test for hypotonia, sometimes called "floppy infant syndrome." The girl is normal. *(Saturn Stills/ Science Photo Library/Photo Researchers, Inc.)*

Another source of concern that parents face is addressing the special needs of their hypotonic child. The world of typical children can be a difficult place for a hypotonic child, and it is tempting to isolate the child. It is not easy to go to a playgroup of toddlers when a child's latest milestone is getting from the floor into a sitting position while the other children are running across the room. There are resources for parents to help their child become as able and independent as he or she can possibly be, and the family physician is a good resource for advice.

See also Bayley Scales of Infant Development; Muscular dystrophy.

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KEY TERMS

Ataxia—A condition marked by impaired muscular coordination, most frequently resulting from disorders in the brain or spinal cord.

Biceps—The muscle in the front of the upper arm.

Biopsy—The surgical removal and microscopic examination of living tissue for diagnostic purposes or to follow the course of a disease. Most commonly the term refers to the collection and analysis of tissue from a suspected tumor to establish malignancy.

Central nervous system—Part of the nervous system consisting of the brain, cranial nerves, and spinal cord. The brain is the center of higher processes, such as thought and emotion and is responsible for the coordination and control of bodily activities and the interpretation of information from the senses. The cranial nerves and spinal cord link the brain to the peripheral nervous system, that is the nerves present in the rest of body.

Chromosome—A microscopic thread-like structure found within each cell of the human body and consisting of a complex of proteins and DNA. Humans have 46 chromosomes arranged into 23 pairs. Chromosomes contain the genetic information necessary to direct the development and functioning of all cells and systems in the body. They pass on hereditary traits from parents to child (like eye color) and determine whether the child will be male or female.

Computed tomography (CT)—An imaging technique in which cross-sectional x rays of the body are compiled to create a three-dimensional image of the body's internal structures; also called computed axial tomography.

Fine motor skill—The abilities required to control the smaller muscles of the body for writing, playing an instrument, artistic expression and craft work. The muscles required to perform fine motor skills are generally found in the hands, feet and head.

Flexor muscle—A muscle that serves to flex or bend a part of the body.

Gene—A building block of inheritance, which contains the instructions for the production of a particular protein, and is made up of a molecular sequence found on a section of DNA. Each gene is found on a precise location on a chromosome.

Genetic disease—A disease that is (partly or completely) the result of the abnormal function or expression of a gene; a disease caused by the inheritance and expression of a genetic mutation.

Gross motor skills—The abilities required to control the large muscles of the body for walking, running, sitting, crawling, and other activities. The muscles required to perform gross motor skills are generally found in the arms, legs, back, abdomen and torso.

Immune response—A physiological response of the body controlled by the immune system that involves the production of antibodies to fight off specific foreign substances or agents (antigens).

Immune system—The system of specialized organs, lymph nodes, and blood cells throughout the body that work together to defend the body against foreign invaders (bacteria, viruses, fungi, etc.).

Magnetic resonance imaging (MRI)—An imaging technique that uses a large circular magnet and radio waves to generate signals from atoms in the body. These signals are used to construct detailed images of internal body structures and organs, including the brain.

Motor control—The control of movement and posture.

Motor neuron—A nerve cell that specifically controls and stimulates voluntary muscles.

Muscle tone—Also termed tonus; the normal state of balanced tension in the tissues of the body, especially the muscles.

Muscle weakness—Reduction in the strength of one or more muscles.

Neurons—Any of the conducting cells of the nervous system that transmit signals.

Recessive disorder—Disorder that requires two copies of the predisposing gene one from each parent for the child to have the disease.

Spinal cord—The elongated nerve bundles that lie in the spinal canal and from which the spinal nerves emerge.

Underlying condition—Disorder or disease that causes the appearance of another medical disorder or condition.

ORGANIZATIONS

Child Development Institute (CDI). 3528 E. Ridgeway Road, Orange, California 92867. Web site: <www.childdevelopmentinfo.com>.

Genetic and Rare Diseases Information Center. PO Box 8126, Gaithersburg, MD 20898–8126. Web site: <www.rarediseasesinfo.nih.gov>.

March of Dimes Birth Defects Foundation. PO Box 3006, Rockville, MD 20847. Web site: <www.marchofdimes.com>.

Muscular Dystrophy Association. 3300 East Sunrise Drive, Tucson, AZ 85718–3208. Web site: <www.mdausa.org>.

National Institute of Child Health and Human Development (NICHD). 31 Center Drive, Rm. 2A32, MSC 2425, Bethesda, MD 20892–2425. Web site: <www.nichd.nih.gov>.

National Institute of Neurological Disorders and Stroke (NINDS). PO Box 5801, Bethesda, MD 20824. Web site: <www.ninds.nih.gov>.

National Organization for Rare Disorders (NORD). PO Box 1968, 55 Kenosia Avenue, Danbury, CT 06813–1968. Web site: <www.rarediseases.org>.

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I

IBS *see* **Irritable bowel syndrome**

Ibuprofen *see* **Nonsteroidal anti-inflammatory drugs**

Idiopathic thrombocytopenia purpura

Definition

Idiopathic thrombocytopenic purpura (ITP) is a bleeding disorder caused by an abnormally low level of blood platelets, small disc-shaped cells essential to blood clotting (coagulation). ITP describes both the cause and symptoms of the condition: idiopathic means that the disorder has no apparent cause; thrombocytopenia refers to a decreased number of blood platelets; and purpura refers to a purplish or reddish-brown skin rash caused by the leakage into the skin of blood from broken capillaries. ITP is as of 2004 often called immune thrombocytopenic purpura rather than idiopathic because studies in the early 2000s have shown the presence of autoimmune antibodies in the blood. Other names for ITP include purpura hemorrhagica and essential thrombocytopenia.

Description

Coagulation, or clotting, is a complex process in which specific proteins found in blood plasma combine with other blood components, including platelets, to form clots and prevent blood loss. Platelets are tiny colorless disc-shaped cells in the blood that collect (aggregate) in blood vessels to form a plug when a vessel is injured. The platelet plug then binds coagulation proteins to form a clot that stops bleeding. A deficiency in platelets or a disorder that affects platelet production can disrupt clotting and severely complicate blood loss from accidental injury, surgery, and specific diseases or conditions in which bleeding can occur. ITP affects the overall number of blood platelets rather than their function. The normal platelet level in adults is between 150,000 and 450,000/mm^3. Platelet counts below 50,000/mm^3 increase the risk of dangerous bleeding from trauma; counts below 20,000/mm^3 increase the risk of spontaneous bleeding.

ITP may be either acute or chronic. The acute form occurs in children between ages two and six. Although chronic ITP is most common in adult females, 10 to 20 percent of children with ITP have the chronic form.

Demographics

Acute ITP affects children of both sexes between the ages of two and six years. The chronic form, although most common in adult females between the ages of 20 and 40, is found in 10 to 20 percent of children with ITP. ITP does not appear to be related to race, lifestyle, climate, or environmental factors.

Causes and symptoms

In children, ITP is usually triggered by a virus infection, most often **rubella**, **chickenpox**, **measles**, cytomegalovirus (CMV), or Epstein-Barr virus (EBV). ITP usually begins about two or three weeks after the infection.

Acute ITP is characterized by bleeding into the skin or from the nose, mouth, digestive tract, or urinary tract. The onset is usually sudden. Bleeding into the skin takes the form of purpura or petechiae. Purpura, a purplish or reddish-brown rash or discoloration of the skin, and petechiae, small round pinpoint hemorrhages, are both caused by the leakage of blood from tiny capillaries under the skin. In addition to purpura and petechiae, spontaneous **bruises** may occur. In extreme cases, ITP may cause bleeding into the lungs, brain, or other vital organs.

Chronic ITP has a gradual onset and may have minimal or no external symptoms. The low **platelet count** may be discovered in the course of a routine blood test. Most parents consult a pediatrician or primary care doctor after noticing their child has the typical purpuric skin rash, frequent nosebleeds, or bleeding from the digestive or urinary tract.

In adults, ITP is considered an autoimmune disorder, which means that the body produces antibodies that damage some of its own products—in this case, blood platelets. Some adults with chronic ITP may have other autoimmune diseases such as lupus (systemic lupus erythematosus or SLE), rheumatoid arthritis, or scleroderma. Women with ITP may experience unusually heavy or lengthy menstrual periods. Risk factors for the development of chronic ITP in adults include being female, age over 10 years at onset of symptoms, bruising, and having another known autoimmune disease.

When to call the doctor

When a child bruises easily, has frequent nosebleeds, bloody stools, or develops a purplish or reddish-brown rash or tiny spots of hemorrhage, the symptoms should be reported to the pediatrician or **family** doctor, especially if they are noticed in the weeks following measles or chickenpox or a virus infection such as mononucleosis.

Diagnosis

ITP is usually considered a diagnosis of exclusion, which means that the doctor makes a diagnosis by ruling out other possible causes for the symptoms and physical findings. The presence of **fever**, for example, does not indicate ITP, whereas fever may occur in lupus and some other types of thrombocytopenia. Likewise, the doctor will look for an enlarged spleen by pressing on (palpating) the abdomen; if the spleen is noticeably enlarged, ITP is not absolutely ruled out but is a less likely diagnosis. If ITP is suspected, the doctor will examine the child's skin for bruises, purpuric areas, or petechiae. If nosebleeds or bleeding from the mouth or other parts of the body have been reported, the doctor will examine these areas for other possible causes of bleeding. Individuals with ITP usually look and feel healthy except for the bleeding. If the child has had a recent childhood illness (measles, chickenpox) or a virus, the risk for ITP is greater, and this fact will be considered along with diagnostic testing results.

Diagnostic tests will begin with a complete blood count (CBC), including a platelet count. A blood test for autoantibodies may be performed early in the diagnostic process as well as a test for antiplatelet antibodies. Specific diagnostic tests for autoimmune diseases and viruses (CMV, EBV, and rheumatoid factor or RF) may be performed. Coagulation tests, including clotting time, will be performed to determine the ability of the child's blood to form a clot. Platelet aggregation tests may be performed to evaluate platelet function, particularly if the platelet count is low.

KEY TERMS

Autoimmune disorder—One of a group of disorders, like rheumatoid arthritis and systemic lupus erythematosus, in which the immune system is overactive and has lost the ability to distinguish between self and non-self. The body's immune cells turn on the body, attacking various tissues and organs.

Clotting factors—Substances in the blood, also known as coagulation factors, that act in sequence to stop bleeding by triggering the formation of a clot. Each clotting factor is designated with a Roman numeral I through XIII.

Idiopathic—Refers to a disease or condition of unknown origin.

Petechia—Plural, petechiae. A tiny purple or red spot on the skin resulting from a hemorrhage under the skin's surface.

Platelet—A cell-like particle in the blood that plays an important role in blood clotting. Platelets are activated when an injury causes a blood vessel to break. They change shape from round to spiny, "sticking" to the broken vessel wall and to each other to begin the clotting process. In addition to physically plugging breaks in blood vessel walls, platelets also release chemicals that promote clotting.

Prednisone—A corticosteroid medication often used to treat inflammation.

Purpura—A group of disorders characterized by purplish or reddish brown areas of discoloration visible through the skin. These areas of discoloration are caused by bleeding from broken capillaries.

Splenectomy—Surgical removal of the spleen.

Thrombocytopenia—A persistent decrease in the number of blood platelets usually associated with hemorrhaging.

Children with ITP will usually have platelet counts below 20,000/mm^3 and a prolonged clotting time. The size and appearance of the platelets may be abnormal, which is observed microscopically. The red blood cell count (RBC) and white blood cell count (WBC) are usually normal, although about 10 percent of individuals with ITP are also anemic (have reduced RBCs and hemoglobin). The bone marrow test yields normal results. Detection of antiplatelet antibodies in the blood usually confirms a diagnosis of ITP.

Treatment

There is no specific treatment for ITP except to manage symptoms. In most cases, the disorder will resolve within two to six weeks without medications or surgery. Nosebleeds can be treated with ice packs when necessary. General care may include asking parents to watch for bruising, petechiae, or other signs of recurrence. Parents are also advised to avoid giving the child aspirin, ibuprofen, or other over-the-counter **pain** medications because these drugs are known to lengthen the clotting time of blood.

Children with acute ITP who are losing large amounts of blood or bleeding into their central nervous system require emergency treatment. This may include transfusions of platelets, intravenous immunoglobulins, or prednisone. Prednisone is a steroid medication that decreases the effects of antibodies on platelets and eventually lowers antibody production. If the child has been treated before for ITP and has not responded to prednisone or immunoglobulins, surgery may be required to remove the spleen (splenectomy), an organ that sometimes stores platelets and keeps them out of the general blood circulation. Splenectomy is usually avoided in children younger than five years because of the increased risk of a severe postoperative infection. In older children, however, splenectomy is recommended if the child has been treated for 12 months without improvement, if the ITP is very severe or is getting worse, or if the child begins to bleed into the head or brain.

Children with chronic ITP can be treated with prednisone, immune globulin, or large doses of intravenous gamma globulin. Although 90 percent of those with ITP respond to immunoglobulin treatment, it is an expensive treatment. Response to prednisone therapy is about 80 percent. Platelet transfusions are not recommended for routine treatment of ITP. If platelet levels do not improve within one to four months, or high doses of prednisone are required, splenectomy may be recommended. If the patient is an adolescent female with extremely heavy periods, splenectomy may also be recommended. Adults treated with splenectomy usually experience remission of chronic ITP. All medications for ITP are given either orally or intravenously; intramuscular injection is avoided because of the possibility of causing bleeding into the skin.

Prognosis

The prognosis for recovery from acute ITP is good; 80 percent of those affected recover without special treatment. The prognosis for chronic ITP is also good; most individuals experience long-term remission. In rare instances, however, ITP can cause life-threatening hemorrhage or bleeding into the central nervous system.

Prevention

Because as of 2004 the exact cause for ITP is unknown, no specific preventive measures are recommended. However, episodes of bleeding can be prevented in children with ITP by discouraging rough contact **sports** or other activities that increase the risk of trauma. To reduce the risk of ITP associated with other illnesses, children can be immunized against childhood diseases and kept away as much as possible from other children or adults with known or unidentified viruses.

Parental concerns

The sudden onset of ITP-like symptoms can be a concern, but the presence of a rash or bruising is not a signal for alarm because there are so many possible causes of these symptoms in childhood. Parents can be generally watchful, but not fearful. The symptoms to be alert for are frequent nosebleeds or frequent bruising with no specific cause, particularly if the child has had a recent illness or virus. It is helpful to remember that ITP, whether acute or chronic, has an excellent prognosis and may cause bleeding but not life-threatening hemorrhage in most cases. Parents can ask the pediatrician when in doubt and understand that simple blood and coagulation tests can be performed to rule out ITP.

See also Coagulation disorders; Infectious mononucleosis; TORCH test.

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ORGANIZATIONS

National Heart, Lung, and Blood Institute (NHLBI). 6701 Rockledge Drive, PO Box 30105, Bethesda, MD 20824–0105. Web site: <www.nhlbi.nih.gov>.

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IgA deficiency *see* Immunoglobulin deficiency syndromes

IgG subclass deficiencies *see* Immunoglobulin deficiency syndromes

Ileus

Definition

Ileus is a partial or complete non-mechanical blockage of the small and/or large intestine.

Description

There are two types of **intestinal obstructions**, mechanical and non-mechanical. Mechanical obstructions occur because the bowel is physically blocked and its contents cannot pass the point of the obstruction. This happens when the bowel twists on itself (volvulus) or as the result of hernias, impacted feces, abnormal tissue growth, or the presence of foreign bodies in the intestines. By contrast, non-mechanical obstruction, called ileus, occurs because the rhythmic contractions that move material through the bowel (called peristalsis) stop.

Demographics

The total rate of bowel obstruction due both to mechanical and non-mechanical causes is one in 1,000 people. Meconium ileus accounts for 9–33 percent of bowel obstructions in newborns.

Causes and symptoms

Ileus is most often associated with an infection of the peritoneum (the membrane lining the abdomen) or other intra-abdominal infections such as **appendicitis**. It is one of the major causes of bowel obstruction in infants and children. Another common cause of ileus is a disruption or reduction of the blood supply to the abdomen. Handling the bowel during abdominal surgery can also cause peristalsis to stop, so people who have had abdominal surgery are more likely to experience ileus.

Ileus can also be caused by kidney diseases, especially when potassium levels are decreased (a condition called hypokalemia). Narcotics and certain **chemotherapy** drugs, such as vinblastine (Velban, Velsar) and vincristine (Oncovin, Vincasar PES, Vincrex) can also cause ileus. Infants with **cystic fibrosis** are more likely to experience meconium ileus (obstruction of a dark green material in the intestine in newborns).

When the bowel stops functioning, the following symptoms occur:

- abdominal cramping
- abdominal distention (**pain** often increases as distention increases)
- nausea, **vomiting**, and/or **diarrhea**
- failure to pass gas or stool

When to call the doctor

A healthcare professional should be contacted if a child experiences persistent abdominal distention, is unable to have normal bowel movements, or exhibits other symptoms of ileus. Persistent abdominal pain and chronic or prolonged **constipation** are also reasons to call the doctor.

Diagnosis

When a doctor listens with a stethoscope to the abdomen of a child suffering from ileus, there will be few or no bowel sounds, indicating that the intestine has stopped functioning. Ileus can be confirmed by **x rays** of the abdomen, **computed tomography** scans (CT scans), or ultrasound. It may be necessary to do more invasive tests, such as a barium enema or upper GI series, if the obstruction is mechanical. Blood tests may also be useful in diagnosing ileus.

Barium studies are used in cases of mechanical obstruction but may cause problems by increasing pressure or intestinal contents if used in ileus. Also, in cases of suspected mechanical obstruction involving the gastrointestinal tract (from the small intestine downward) use of barium x rays are contraindicated, since they may contribute to the obstruction. In such cases a barium enema should always be done first.

Treatment

Patients may be treated with supervised bed rest in a hospital and bowel rest, where nothing is taken by mouth, and patients are fed intravenously or through the use of a nasogastric tube, a tube inserted through the nose, down the throat, and into the stomach. A similar tube can be inserted in the intestine. The contents are then suctioned out. In some cases, especially where there is a mechanical obstruction or death (necrosis) of intestinal tissue, surgery may be necessary.

Drug therapies that promote intestinal motility (ability of the intestine to move spontaneously), such as cisapride and vasopressin (Pitressin), are sometimes prescribed.

Alternative treatment

Alternative practitioners offer few treatment suggestions but focus on prevention by keeping the bowels healthy through eating a good diet, high in fiber and low

in fat. If the case is not a medical emergency, homeopaths and practitioners of traditional Chinese medicine can recommend therapies that may help to reinstate peristalsis.

Nutritional concerns

Following abdominal surgery, uncomplicated cases of ileus can be managed by minimizing the amount of food the patient consumes, ensuring adequate fluid intake, and correcting any electrolyte disturbances such as low potassium.

Prognosis

The outcome varies depending on the cause of ileus. When ileus results from abdominal surgery, the condition is usually temporary and lasts approximately 24–72 hours. The prognosis is less certain in cases in which death of intestinal tissue occurs; surgery becomes necessary to remove the necrotic tissue. In children with cystic fibrosis in which meconium ileus becomes evident soon after birth, the prognosis is linked with the primary disease; the median age of survival for cystic fibrosis patients is 30 years. However, new interventions in the treatment of CF are increasing the age span of people with CF every year.

Prevention

Most cases of ileus are not preventable. Surgery to remove a tumor or other mechanical obstruction may help to prevent a recurrence.

Nutritional concerns

In cases in which electrolyte imbalance is the cause of ileus, it is important to treat the underlying cause of the imbalance, which in many cases is related to chronic vomiting and/or diarrhea, poor fluid and/or food intake, or abuse of **laxatives** and diuretics (such as in individuals with **bulimia nervosa**).

Parental concerns

When their child is diagnosed with ileus, parents may be concerned about the necessity of surgery to correct the problem. Surgery, however, is considered only in medical emergencies and for patients for whom more conservative treatments have failed.

KEY TERMS

Bulimia nervosa—An eating disorder characterized by binge eating and inappropriate compensatory behavior, such as vomiting, misusing laxatives, or excessive exercise.

Computed tomography (CT)—An imaging technique in which cross-sectional x rays of the body are compiled to create a three-dimensional image of the body's internal structures; also called computed axial tomography.

Meconium—A greenish fecal material that forms the first bowel movement of an infant.

Peritoneum—The transparent membrane lining the abdominal and pelvic cavities (parietal peritoneum) and the membrane forming the outer layer of the stomach and interstines (visceral peritoneum). Between the visceral and parietal peritoneums is a potential space called the peritoneal cavity.

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Immobilization

Definition

Immobilization refers to the process of holding a joint or bone in place with a splint, cast, or brace. This is done to prevent an injured area from moving while it heals.

Purpose

Splints, casts, and braces support and protect broken bones, dislocated joints, and injured soft tissues such as tendons and ligaments. Immobilization restricts motion to allow the injured area to heal. It can help reduce **pain**, swelling, and **muscle spasms**. In some cases, splints and casts are applied after surgical procedures that repair bones, tendons, or ligaments. This allows for protection and proper alignment early in the healing process.

Description

When an arm, hand, leg, or foot requires immobilization, the cast, splint, or brace will generally extend from the joint above the injury to the joint below the injury. For example, an injury to the mid-calf requires immobilization from the knee to the ankle and foot. Injuries of the hip and upper thigh or shoulder and upper arm require a cast that encircles the body and extends down the injured leg or arm.

Casts and splints

Casts are generally used to immobilize a broken bone. Once the doctor makes sure the two broken ends of the bone are aligned, a cast is put on to keep them in place until they are rejoined through natural healing. Casts are applied by a physician, a nurse, or an assistant. They are custom-made to fit each person, and are usually made of plaster or fiberglass. Fiberglass weighs less than plaster, is more durable, and allows the skin more adequate airflow than plaster. A layer of cotton or synthetic padding is first wrapped around the skin to cover the injured area and protect the skin. The plaster or fiberglass is then applied over this and is then allowed to dry. It can take up to 24 hours for a cast to dry completely.

Most casts should be kept dry. However, some types of fiberglass casts use Gore-tex padding that is waterproof, allowing the cast to be completely immersed in water when taking a shower or bath. There are some circumstances when this type of cast material cannot be used.

A splint is often used to immobilize a dislocated joint while it heals. Splints are also often used for finger injuries, such as **fractures** or baseball finger. Baseball finger is an injury in which the tendon at the end of the finger is separated from the bone as a result of trauma. Splinting is also used to immobilize an injured arm or leg immediately after an injury. Before moving a child who has injured an arm or leg, some type of temporary splint should be applied to prevent further injury to the area. Splints may be made of acrylic, polyethylene foam, plaster of paris, or aluminum. In an emergency, a splint can be made from a piece of wood or rolled magazine.

Slings

Slings are often used to support the arm after a fracture or other injury. They are generally used along with a cast or splint, but are sometimes used alone as a means of immobilization. They can be used in an emergency to immobilize the arm until a doctor can see the child. A triangular bandage is placed under the injured arm and then tied around the neck.

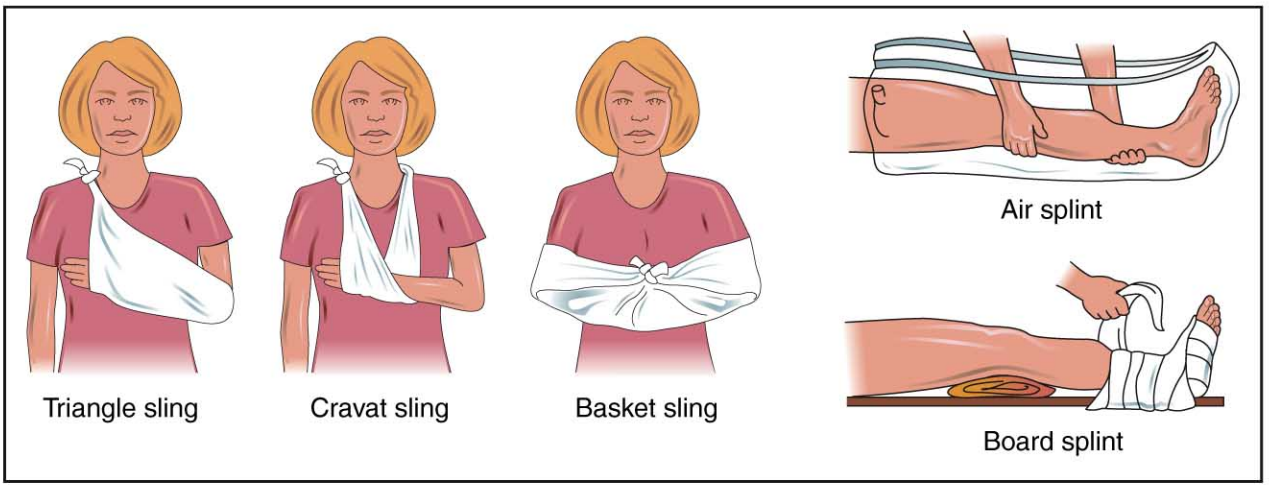
Braces

Braces are used to support, align, or hold a body part in the correct position. Braces are sometimes used after a surgical procedure is performed on an arm or a leg. They may also be used when an injury has occurred. Since some braces can be easily taken off and put back on, they are often used when the child needs physical therapy or must **exercise** the limb during the healing process. Many braces can also be adjusted to allow for a certain amount of movement.

Either a custom-made or a ready-made brace can be used. The off-the-shelf braces are made in a variety of shapes and sizes. They generally have Velcro straps that make the brace easy to adjust and to put on and take off. Both braces and splints offer less support and protection than a cast and may not be a treatment option in all circumstances.

Collars

A collar is generally used for neck injuries. A soft collar can relieve pain by restricting movement of the



These illustrations feature several types of immobilization techniques, including slings and splints. *(Illustration by Electronic Illustrators Group.)*

head and neck. Collars also transfer some of the weight of the head from the neck to the chest. Stiff collars are generally used to support the neck when there has been a fracture in one of the neck bones. Cervical collars are widely used by emergency personnel at the scene of injuries when there is a potential neck or **head injury**. The collar helps to ensure that the neck and head do not move, which could make the injury worse.

Traction

Immobilization may also be secured by traction. Traction involves using a method for applying tension to correct the alignment of two structures (e.g., two bones) and hold them in the correct position. For example, if the bone in the thigh breaks, the broken ends may have a tendency to overlap. Use of traction will hold them in the correct position for healing to occur. The strongest form of traction involves inserting a stainless steel pin through a bony prominence attached by a horseshoe-shaped bow and rope to a pulley and weights suspended over the end of the patient's bed.

Traction must be balanced by countertraction. This may be obtained by tilting the bed and allowing the patient's body to act as a counterweight. Another technique involves applying weights pulling in the opposite direction.

Traction for neck injuries may be in the form of a leather or cotton cloth halter placed around the chin and lower back of the head. For very severe neck injuries that require maximum traction, tongs that resemble ice tongs are inserted into small holes drilled in the outer skull. All traction requires careful observation and adjustment by doctors and nurses to maintain proper balance and alignment of the traction with free suspension of the weights.

Immobilization can also be secured by a form of traction called skin traction. This is a combination of a splint and traction that is applied to the arms or legs by strips of adhesive tape placed over the skin of the arm or leg. Adhesive strips, moleskin, or foam rubber traction strips are applied on the skin. This method is effective only if a moderate amount of traction is required.

Precautions

It is important to ensure that the cast is not too tight, such that blood flow is cut off and swelling is not restricted.

Preparation

There are many reasons for immobilization using splints, casts, and braces. It can be helpful if the child understands why immobilization is being done, as it may help with compliance.

Aftercare

After a cast or splint has been put on, the injured arm or leg should be elevated for 24 to 72 hours. It is recommended that the child lie or sit with the injured arm or leg raised above the level of the heart. Rest, combined with elevation, will reduce pain and speed the healing process by minimizing swelling.

Fingers or toes can be exercised as much as can be tolerated after casting. This has been found to decrease swelling and prevent stiffness. If excessive swelling is noted, the application of ice to the splint or cast may be helpful.

After the cast, splint, or brace is removed, gradual exercise is usually performed to regain muscle strength

KEY TERMS

Decubitus ulcer—A pressure sore resulting from ulceration of the skin occurring in persons confined to bed for long periods of time

Ligament—A type of tough, fibrous tissue that connects bones or cartilage and provides support and strength to joints.

Pneumonia—An infection in which the lungs become inflamed. It can be caused by nearly any class of organism known to cause human infections, including bacteria, viruses, fungi, and parasites.

Tendon—A tough cord of dense white fibrous connective tissue that connects a muscle with some other part, especially a bone, and transmits the force which the muscle exerts.

and motion. The doctor may also recommend hydrotherapy, heat treatments, and other forms of physical therapy.

Risks

For some children, such as those in traction, immobilization will require long periods of bed rest. Lying in one position in bed for an extended period can result in sores on the skin (decubitus ulcers) and skin infection. Long periods of bed rest can also cause a buildup of fluid in the lungs or an infection in the lungs (**pneumonia**). Urinary infection can also be a result of extended bed rest. Occasionally, blood clots can develop in the injured area.

People who have casts, splints, or braces on their arms or legs will generally spend several weeks not using the injured arm or leg. This lack of use can result in decreased muscle tone and shrinkage of the muscle (atrophy). This loss can usually be regained, however, through rehabilitation after the injury has healed.

Immobility can also cause psychological stress. A child restricted to a bed with a traction device may become frustrated and bored, and perhaps even depressed, irritable, and withdrawn. It may be helpful to have a bed that can be moved more easily than a traditional bed, so that the child can participate in as many **family** activities (such as dinner time and movie nights) as possible.

There is the possibility of decreased circulation if the cast, splint, or brace fits too tightly. If swelling occurs but the cast is tight enough that the swelling cannot be accommodated, serious complications can occur. Excessive pressure over a nerve can cause irritation or possible damage if not corrected. If the cast, splint, or brace breaks or malfunctions, the healing process of the bone or soft tissue can be disrupted and lead to deformity.

Parental concerns

Children can find immobilization very frustrating. Being restricted from participating in normal activities with friends because of a broken an arm or leg, or having to sit in bed for weeks if significant traction is required can be stressful. Children can be helped if parents try to provide alternate activities and engage the child in as many activities as possible with the family.

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Immune system development

Definition

The child's immune system is an intricate network of interdependent cell types, substances, and organs that collectively protect the body from bacterial, parasitic, fungal, viral infections, and tumor cells.

Description

The immune system was not recognized as a functional unit of the body until the late twentieth century,

probably because its parts are not directly connected to each other and are spread in different parts of the body.

Organs of the immune system

The immune system contains the following organs and cells: tonsils and adenoids; the thymus gland; lymph nodes; bone marrow; and white blood cells that leave blood vessels and migrate through tissues and lymphatic circulation. The spleen, appendix, and patches of lymphoid tissue in the intestinal tract are also parts of the immune system.

The essential job of this system is to distinguish self-cells from foreign substances and to recognize and take protective action against any materials that ought not to be in the body, including abnormal and damaged cells. The immune system can seek out and destroy disease germs, infected cells, and tumor cells. The immune system includes the following cells:

- T lymphocytes (T cells)
- B lymphocytes (B cells)
- natural killer cells (NK cells)
- dendritic cells
- phagocytic cells
- complement proteins

These cells develop from ''pluripotential hematopoietic stem cells'' starting from a gestational age of about five weeks. They circulate through various organs in the lymphatic system as the fetus develops. T and B lymphocytes are the only units of the immune system that have antigen-specific recognition powers; they are responsible for adaptive immunity. In other words, the T and B cells are important in the immunity that **vaccination** promotes.

How immunity works

The lymphatic system is a key participant in the body's immune actions. It is a network of vessels and nodes unified by the circulatory system. Lymph nodes occur along the course of the lymphatic vessels and filter lymph fluid before it returns to the bloodstream. The system removes tissue fluids from intercellular spaces and protects the body from bacterial invasions.

Types of immunity

Immunity is the ability of the body to resist the infecting agent. When an infectious agent enters the body, the immune system develops antibodies which can weaken or destroy the disease-producing agent or neutralize its toxins. If the body is re-introduced to the same agent at a later time, it is capable of developing antibodies at a much faster pace. As a result, the individual would likely not become sick, and immunity has developed.

Natural immunity is present when a person is immune to a disease despite not having either the disease itself nor any vaccination against it. Acquired immunity may be either active or passive. Active immunity comes from having the disease or by inoculation with antigens, such as dead organisms, weakened organisms, or toxins of organisms. The antigens introduced during vaccination produce antibodies that protect the body against the infecting agent, despite the fact that the person does not become sick. Passive immunity is relatively short lived and is acquired by transferring antibodies from mother to child in the uterus or by inoculation with serum that contains antibodies from immune persons or animals. Passive immunization is used to help a person who has been exposed or is already infected to fight off disease. Although various types of serums may be used to produce passive immunization, gamma globulin is the most frequently used source of human antibodies.

Development of the immune response

Normal infants have the capability to develop responses to antigens at birth. Infants also start life with some immunoglobulin antibodies acquired from the mother. These antibodies cross the placental barrier, but not all types are transmitted equally. In particular, infants start with antibodies to viruses and gram-positive organisms, but not to gram-negative organisms. Gram is the name of a stain that distinguishes broad classes of bacteria. Gram-negative organisms are responsible for many diseases, including gonorrhea, pertussis (whooping cough), salmonella poisoning, and cholera. *Escherichia coli (E. coli)* is another common gram-negative organism.

Immunoglobulin antibodies are divided into five classes. The capacity of the body to produce each immunoglobulin varies with age. Newborn babies (premature and full-term) begin to synthesize antibodies at an increased rate soon after birth in response to antigenic stimulation of their new environment. At about six days after birth the serum concentration of specific antibodies rises sharply, and this rise continues until adult levels are achieved by approximately the end of the first year. Maternal immunity gradually disappears during the first six to eight months of life. A concentrated level of antibodies is reached and maintained by seven to eight years of age.

Common problems

Persistent infections

One of the greatest strains on the immune system is an infection it cannot remove. Parents should pay attention to unexplained fevers; night sweats; or tender, swollen lymph nodes. These symptoms can signify a hidden infection or **cancer**. Infections of the mouth and gums as well as sexually transferred infections often go unnoticed while they drain the vitality of the immune system.

Indiscriminate use of antibiotics

When the immune system successfully controls an infection on its own, it becomes stronger and better able to handle future threats. **Antibiotics** are powerful medicines that should be given only when the immune system cannot contain a bacterial infection. Overuse of antibiotics may cause the body to breed new strains of antibiotic-resistant or more dangerous bacteria. In the long run, overuse of antibiotics weakens the immune system.

Misuse of immunosuppressive drugs

Immunosuppressive drugs used in cancer **chemotherapy** or to suppress rejection of organ transplants are necessary. Of greater concern is the widespread use of corticosteroids or steroid derivatives used to treat **allergies**, autoimmune diseases, and inflammatory conditions. Though sometimes necessary, these drugs cripple the immune response and are often misunderstood, abused, and over-prescribed.

Radiation and hazardous chemicals

Exposure to radiation and hazardous chemicals may also damage the immune system. Excessive radiation of diagnostic **x rays** of the neck and chest may damage the thymus gland behind the breastbone. The thymus gland is an integral part of the immune system.

Blood transfusions and injections of blood products

Blood transfusions and injections of blood products may broadcast viral diseases like hepatitis that stress the immune system by flooding it with foreign proteins. In an emergency it may not be possible to do without blood transfusions. Sources of blood and blood products are regulated and screened for infectious substances and were as of 2004 much safer.

Other factors

Certain factors have damaging effects on the immune system of infants. Excessive consumption of alcohol during pregnancy leads to depressive levels of vitamin B and zinc, which are essential to immune competence. **Alcoholism** can also reduce the uptake of several other important nutrients needed for neonatal immune systems. Prolonged stress during pregnant and in breastfeeding mothers reduces the effectiveness of the immune system as well as the quality of immunologic factors in breast milk.

Cigarette **smoking** raises the white blood cells count, activating the immune system; however, smoking causes low-grade chronic **bronchitis**, low birth weight infants, and weakened natural immunity in newborns. Infants and children constantly exposed to cigarette smoke have weakened immune systems.

Toxic points—areas of localized infections such as dental abscesses or infected tonsils—may disturb the normal neutralization and weaken the cellular defenses in pregnant mothers and in children.

Deficiencies of many nutrients, especially certain **vitamins** and **minerals**, may weaken the immune system. Excessive **exercise** may depress the immune system temporarily.

Autoimmunity

Autoimmunity occurs when the immune system mistakenly attacks the body's own tissues, resulting in disease that can be mild or severe. Common autoimmune disorders are rheumatoid arthritis, glomerulonephritis, **rheumatic fever**, and systemic lupus erythematosus (SLE). Autoimmune reactions may be set off by infection, tissue injury, or emotional trauma in people with a genetic tendency to them.

Parental concerns

Parents may be concerned that children with acute illnesses have compromised immune systems and are less likely to have a positive response to vaccines or may be more likely to develop adverse reaction to the vaccine than healthy children. Parents may also believe that children who are ill should not further burden an immune system already committed to fighting an infection.

Most pediatricians would agree that there should be a delay in vaccinations for children with severe illnesses until the symptoms of illness are gone. The reason for deferring immunization is to avoid superimposing a reaction to the vaccine on the underlying illness or attributing symptoms of the underlying illness to the vaccine by mistake. However, a low-grade **fever** or cold is not a contraindication for routine vaccinations.

KEY TERMS

Antibody—A special protein made by the body's immune system as a defense against foreign material (bacteria, viruses, etc.) that enters the body. It is uniquely designed to attack and neutralize the specific antigen that triggered the immune response.

Antigen—A substance (usually a protein) identified as foreign by the body's immune system, triggering the release of antibodies as part of the body's immune response.

Corticosteroids—A group of hormones produced naturally by the adrenal gland or manufactured synthetically. They are often used to treat inflammation. Examples include cortisone and prednisone.

Immune system—The system of specialized organs, lymph nodes, and blood cells throughout the body that work together to defend the body against foreign invaders (bacteria, viruses, fungi, etc.).

Immunization—A process or procedure that protects the body against an infectious disease by stimulating the production of antibodies. A vaccination is a type of immunization.

Lymphocyte—A type of white blood cell that participates in the immune response. The two main groups are the B cells that have antibody molecules on their surface and T cells that destroy antigens.

Phagocytosis—A process by which certain cells envelope and digest debris and microorganisms to remove them from the blood.

Parents may also be concerned that the many different vaccines that infants are given may overwhelm a child's immune system. However, infants have the capacity to respond to large numbers of antigens. Parents who worry about the increasing number of recommended vaccines may take comfort in knowing that children are exposed to fewer antigens in vaccines as of the early 2000s than in previous decades. Two reasons account for this decline: the worldwide elimination of smallpox and advances in protein chemistry in vaccines with fewer antigens.

Vaccines may cause temporary suppression of delayed-type hypersensitivity skin reactions or alter certain lymphocyte function tests. However, the short-lived immunosuppression caused by certain vaccines does not result in an increased risk of infections from other pathogens soon after vaccination.

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Aliene Linwood, RN, DPA

Immunization *see* **Vaccination**

Immunodeficiency

Definition

Immunodeficiency disorders are a group of disorders in which part of the immune system is missing or defective. The body's ability to fight infections is, therefore, impaired. As a result, a child with an immunodeficiency disorder has frequent infections that are generally more severe and last longer than in a healthy child.

Description

The immune system is the body's main defense against infections. Any defect in the immune system decreases a person's ability to fight infections. A person with an immunodeficiency disorder may get more frequent infections, heal more slowly, and have a higher incidence of some cancers.

The normal immune system involves a complex interaction of certain types of cells that can recognize and attack foreign invaders, such as bacteria, viruses, and fungi. It also plays a role in fighting **cancer**. The immune system has both innate and adaptive components. Innate immunity is made up of immune protections people are born with. Adaptive immunity develops

throughout life. It adapts to fight off specific invading organisms. Adaptive immunity is divided into two components: humoral immunity and cellular immunity.

The innate immune system is made up of the skin (which acts as a barrier to prevent organisms from entering the body); white blood cells called phagocytes; a system of proteins called the complement system; and chemicals called interferons. When phagocytes encounter an invading organism, they surround and engulf it in order to destroy it. The complement system also attacks bacteria. The elements in the complement system create a hole in the outer layer of the target cell, which leads to the death of the cell.

The adaptive component of the immune system is extremely complex and is as of the early 2000s still not entirely understood. Basically, it has the ability to recognize an organism or tumor cell as not being a normal part of the body and to develop a response to attempt to eliminate it.

The humoral response of adaptive immunity involves a type of cell called B lymphocytes. B lymphocytes manufacture proteins called antibodies (which are sometimes also called immunoglobulins). The terms antibody and immunoglobulin are often used interchangeably, although immunoglobulin refers to the larger classification system for antibodies. There are five types or classes of immunoglobulin that antibodies fit into, and each has a slightly different role in response against bacteria and viruses. Antibodies attach themselves to the invading foreign substance. This allows the phagocytes to begin engulfing and destroying the organism. The action of antibodies also activates the complement system. The humoral response is particularly useful for attacking bacteria.

The cellular response of adaptive immunity is useful for attacking viruses, some parasites, and possibly cancer cells. The main type of cell in the cellular response is the T lymphocyte. There are helper T lymphocytes and killer T lymphocytes. The helper T lymphocytes play a role in recognizing invading organisms, and they also help killer T lymphocytes to multiply. As the name suggests, killer T lymphocytes act to destroy the target organism.

Defects can occur in any component of the immune system or in more than one component (combined immunodeficiency). Different immunodeficiency diseases involve different components of the immune system. The defects can be inherited (congenital) or acquired.

Congenital immunodeficiency disorders

Congenital immunodeficiency is present at the time of birth and is the result of genetic defects. These immunodeficiency disorders are also called primary immunodeficiencies. Even though more than 70 different types of congenital immunodeficiency disorders have been identified, they rarely occur. Congenital immunodeficiencies may occur as a result of defects in B lymphocytes, T lymphocytes, or both. They also can occur in the innate immune system.

HUMORAL IMMUNITY DISORDERS The congenital immunodeficiency disorder, **Bruton's agammaglobulinemia**, also known as X-linked agammaglobulinemia, results in a decrease or absence of B lymphocytes and, therefore, a decreased ability to make antibodies. People with this disorder are particularly susceptible to infections of the throat, skin, middle ear, and lungs. It is seen only in males because it is caused by a genetic defect on the X chromosome. Since males have only one X chromosome, they always have the defect if the gene is present. Females can have the defective gene, but since they have two X chromosomes, there will be a normal gene on the other X chromosome to counter it. Women may pass the defective gene on to their sons.

B LYMPHOCYTE DEFICIENCIES If there is an abnormality in either the development or function of B lymphocytes, the ability to make antibodies will be impaired. This deficit makes the body susceptible to recurrent infections.

A type of B lymphocyte deficiency involves a group of disorders called selective **immunoglobulin deficiency syndromes**. The five different types of immunoglobulins are called IgA, IgG, IgM, IgD, and IgE. The most common type of immunoglobulin deficiency is selective IgA deficiency, occurring in about one in every 500 white persons. The amounts of the other antibody types are normal. Some patients with selective IgA deficiency experience no symptoms, while others have occasional lung infections and **diarrhea**. In another immunoglobulin disorder, IgG and IgA antibodies are deficient, and there is increased IgM. People with this disorder tend to get severe bacterial infections.

Common variable immunodeficiency (CVID) is another type of B lymphocyte deficiency. In this disorder, the production of one or more of the immunoglobulin types is decreased, and the antibody response to infections is impaired. It generally develops in people between the ages of ten and 20. The symptoms vary among affected people. Most people with this disorder have frequent infections, and some also experience autoimmune phenomena, such as autoimmune hemolytic

anemia or rheumatoid arthritis. Persons with CVID develop cancer at a higher rate than the general population, particularly lymphomas.

T LYMPHOCYTE DEFICIENCIES Severe defects in the ability of T lymphocytes to mature result in impaired immune responses to infections with viruses, fungi, and certain types of bacteria. These infections are usually severe and can be fatal.

DiGeorge syndrome is a genetic syndrome most frequently associated with a chromosomal deletion (22q11.2). This syndrome is often associated with T lymphocyte deficiencies. Children with DiGeorge syndrome either do not have a thymus or have an underdeveloped thymus. Since the thymus is a major organ that directs the production of T lymphocytes, these patients have low numbers of T lymphocytes. If the T cell count is very low the patients are susceptible to recurrent infections. The syndrome can be associated with other physical abnormalities. For example, these individuals may have distinctive facial features such as thin upper lip and flattened nasal bridge, and they may have low calcium from hypoparathyroidism or cardiac defects. If the entire syndrome is not present (as is the usual case), the syndrome is called incomplete DiGeorge, and if all elements are present and the thymus is absent, the syndrome is called complete. Children with complete DiGeorge are particularly susceptible to viral and fungal infections.

In some cases, no treatment is required for DiGeorge syndrome because T lymphocyte production improves. Either an underdeveloped thymus begins to produce more T lymphocytes, or organ sites other than the thymus compensate by producing more T lymphocytes.

COMBINED IMMUNODEFICIENCIES Some types of immunodeficiency disorders affect both B lymphocytes and T lymphocytes. For example, **severe combined immunodeficiency** disease (SCID) is caused by the defective development or function of these two types of lymphocytes. It results in impaired humoral and cellular immune responses. SCID usually is recognized during the first year of life. It tends to cause fungal infections, including severe thrush that does not respond to usual treatment; severe diarrhea; and serious bacterial infections. If the deficiency is not treated (usually by bone marrow transplant), a person with SCID usually dies from infection before the age of two years. The most common form of SCID is X-linked, i.e. the defect is on the X chromosome and, therefore, occurs only in boys. In the early 2000s new genetic defects leading to SCID are being identified each year.

DISORDERS OF INNATE IMMUNITY Disorders of innate immunity affect phagocytes or the complement system. These disorders also result in recurrent infections.

Acquired immunodeficiency disorders

Acquired immunodeficiency is more common than congenital immunodeficiency. It is the result of an infectious process or other disease. For example, the human immunodeficiency virus (HIV) is the virus that causes acquired immunodeficiency syndrome (**AIDS**). HIV, however, is not the most common cause of acquired immunodeficiency.

Acquired immunodeficiency often occurs as a complication of other conditions and diseases. For example, the most common causes of acquired immunodeficiency are **malnutrition**, some types of cancer, and infections. People who weigh less than 70 percent of the average weight of persons of the same age and gender are considered to be malnourished. Examples of types of infections that can lead to immunodeficiency are **chickenpox**, cytomegalovirus, German **measles** (**rubella**), measles, **tuberculosis**, **infectious mononucleosis** (Epstein-Barr virus), chronic hepatitis, lupus, and bacterial and fungal infections.

In some cases, acquired immunodeficiency is brought on by drugs used to treat another condition. For example, patients who have an organ transplant are given drugs to suppress the immune system so the body will not reject the organ. Also, some **chemotherapy** drugs that are given to treat cancer have the side effect of killing cells of the immune system. During the period of time that these drugs are being taken, the risk of infection increases. It usually returns to normal after the person stops taking the drugs.

Demographics

About 50,000 new cases of congenital immunodeficiencies are diagnosed in the United States each year. The frequency of severe combined immunodeficiency is estimated to be one out of every 50,000 to 500,000 births, and of combined variable immunodeficiency, one out of every 10,000 to 50,000 births. As of 2004 HIV is estimated to affect approximately 4.4 million children worldwide.

Causes and symptoms

Congenital immunodeficiency is caused by genetic defects that generally occur while the fetus is developing in the uterus. These defects affect the development and/or function of one or more of the components of the immune system. Acquired immunodeficiency is the

result of a disease process, and it occurs later in life. The causes can be diseases, infections, or the side effects of drugs given to treat other conditions.

People with an immunodeficiency disorder tend to become infected by organisms that do not usually cause disease in healthy persons. The major symptoms of most immunodeficiency disorders are repeated infections that heal slowly. These chronic infections cause symptoms that persist for long periods of time. People with chronic infection tend to be pale and thin. They may have skin **rashes**. Their lymph nodes may be absent or larger than usual, and in some types of immune deficiency the spleen and liver may be enlarged. (The lymph nodes are small organs that house antibodies and lymphocytes.) This can result in black-and-blue marks in the skin. The person may lose hair from their head. Sometimes, a red inflammation of the lining of the eye (**conjunctivitis**) is present. They may have a crusty appearance in and on the nose from chronic nasal dripping.

When to call the doctor

In an undiagnosed child, parents should inquire about immune deficiency if there are frequent infections, prolonged infections, unusual infections, unusual complications of usual infections, or if there is a **family** history of immune deficiency. If a child is known to have an immunodeficiency disorder, a healthcare provider should be contacted if the child shows signs of having an infection, such as **fever**, **vomiting**, diarrhea, swelling of the lymph nodes, or unusual fatigue.

Diagnosis

Usually, the first sign that individuals may have an immunodeficiency disorder is that they do not improve rapidly when given **antibiotics** to treat an infection. An immunodeficiency disorder is likely to be present when rare diseases occur or the patient gets ill from organisms that do not normally cause diseases, especially if the patient gets repeatedly infected. When this happens in very young children, a genetic defect may be causing an immunodeficiency disorder. When this situation occurs in older children or young adults, their medical history may indicate that childhood diseases may have caused an immunodeficiency disorder. Other possibilities also exist, such as recently acquired infections (e.g. HIV, hepatitis, tuberculosis, etc.).

Laboratory tests are used to determine the exact nature of the immunodeficiency. Most tests are performed on blood samples. Blood contains antibodies, lymphocytes, phagocytes, and complement components, all of the major immune components that might cause immunodeficiency. A blood cell count determines if the number of phagocytic cells or lymphocytes is below normal. Lower than normal counts of either of these two cell types correlates with immunodeficiency. The blood cells also are checked for their appearance. Sometimes a person may have normal cell counts, but the cells are structurally defective. If the lymphocyte cell count is low, further testing is usually done to determine whether any particular type of lymphocyte is lower than normal. A lymphocyte proliferation test is done to determine if the lymphocytes can respond to stimuli. The failure to respond to stimulants correlates with immunodeficiency. Antibody levels can be measured. Complement levels can be determined by immunodiagnostic tests.

Treatment

There is no cure for congenital immunodeficiency disorders. Therapy is aimed at controlling infections (such as with antibiotics) and, for some disorders, replacing defective or absent components.

Patients with Bruton's agammaglobulinemia must be given periodic infusions of pooled immunoglobulin from multiple donors. The product is called intravenous immunoglobulin (IVIG). The infusions are given approximately once a month for life to compensate for the patients' inability to make these proteins.

Common variable immunodeficiency also is treated with periodic infusions of IVIG throughout life. Additionally, antibiotics are given when necessary to treat infections.

Patients with selective IgA deficiency usually do not require any treatment. Antibiotics can be given for frequent infections.

In some cases, no treatment is required for DiGeorge syndrome because T lymphocyte production improves on its own. In some severe cases, a bone marrow transplant or thymus transplant can be performed to correct the problem.

For most patients with SCID, bone marrow transplantation is necessary. In this procedure, healthy bone marrow from a donor who has a similar type of tissue (usually a relative, such as a brother or sister) is removed. The bone marrow, a substance that is found in the cavity of bones, is the factory that produces blood cells, including some of the white blood cells that make up the immune system. The bone marrow of the person receiving the transplant is destroyed and is then replaced with marrow from the donor. One type of SCID called adenosine deaminase (ADA) deficiency is treated with infusion of the deficient enzyme on a regular basis, and

another type of SCID due to an absence of an interleukin, a protein that is important in directing the immune response, is also treated by infusions of the missing protein.

Treatment of the **HIV infection** that causes AIDS consists of drugs called antiretrovirals. These drugs interrupt the virus replication cycle and, therefore, spare the T cells. Several of these drugs used in various combinations with one another can treat but not cure the disease. Decreasing the viral in the blood to very low levels allows the immune system to remain in tact. Other treatments for people with AIDS are aimed at the particular infections and conditions that arise as a result of the impaired immune system.

For people being treated for cancer, periodic relief from chemotherapy drugs can restore the function of the immune system. In some cases, IVIG is utilized to boost the immune system.

Alternative treatment

For some individuals, alternative treatments such as acupuncture therapy to ease infection-related symptoms or homeopathic medicines to boost immunity may be used in conjunction with traditional medicine as part of a patient's treatment plan.

Nutritional concerns

In most cases, immunodeficiency caused by malnutrition is reversible. The health of the immune system is directly linked to the nutritional status of the patient. Among the essential nutrients required by the immune system are proteins, **vitamins**, iron, and zinc.

Prognosis

The prognosis depends on the type of immunodeficiency disorder. People with Bruton's agammaglobulinemia who are given IVIG infusions generally live into their 30s or 40s. They often die from chronic infections, usually of the lung. People with selective IgA deficiency generally live normal lives. They may experience problems if given a blood transfusion, and therefore they should wear a Medic Alert bracelet or have some other way of alerting any physician who treats them that they have this disorder.

SCID is the most serious of the immunodeficiency disorders. If a bone marrow transplant is not successfully performed, the child usually may not live beyond two years of age.

People with HIV/AIDS are living longer than in the past because of the antiretroviral drugs that became available in the mid-1990s. In the early 2000s HIV is still a potentially fatal illness, but medications have changed the face of the disease for those who have access to them. If medical treatment is timely and successful, T cells do not become depleted and opportunistic infections do not occur.

Prevention

Primary or congenital immunodeficiencies are genetic and are not preventable by avoidance of exposures or by dietary measures. However, someone with a congenital immunodeficiency disorder might want to consider getting genetic counseling before having children in order to find out if there is a chance they will pass the defect on to their children.

Some of the infections associated with acquired immunodeficiency can be prevented or treated before they cause problems. For example, there are effective treatments for tuberculosis and most bacterial and fungal infections. HIV infection can be prevented by practicing safe sex (e.g. using a **condom**) and by not using illegal intravenous drugs. These are the primary routes of transmitting the virus.

Nutritional concerns

In general, people with immunodeficiency disorders should maintain a healthy diet because malnutrition can aggravate immunodeficiencies. People can prevent malnutrition by getting adequate **nutrition**. They also should avoid being near others who have colds or are sick because they can easily acquire new infections. For the same reason, they should practice good personal hygiene, especially dental care. People with immunodeficiency disorders also should avoid eating undercooked food because it might contain bacteria that could cause infection. While this food might not cause infection in others, it is a potential source of infectious organisms for someone with an immunodeficiency.

Parental concerns

If a child has been diagnosed with an immunodeficiency disorder, the parents may be instructed to refrain from having normal childhood vaccinations that contain live viruses, since even weakened versions of the virus may cause serious disease. In some cases, the immunodeficient child needs to be encouraged to wear a mask when in public or around family members who are sick in order to reduce the risk of developing an infection.

KEY TERMS

Agammaglobulinemia—The lack of gamma globulins in the blood, associated with an increased susceptibility to infection.

B lymphocytes—Specialized blood cells that manufacture proteins called antibodies that attach themselves to invading foreign substances.

Chromosome—A microscopic thread-like structure found within each cell of the human body and consisting of a complex of proteins and DNA. Humans have 46 chromosomes arranged into 23 pairs. Chromosomes contain the genetic information necessary to direct the development and functioning of all cells and systems in the body. They pass on hereditary traits from parents to child (like eye color) and determine whether the child will be male or female.

T lymphocytes—Specialized blood cells that recognize invading organisms (helper T lymphocytes) and destroy them (killer T lymphocytes).

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Immunoglobulin deficiency syndromes

Definition

Immunoglobulin deficiency syndromes are a group of disorders that involve defects of any component of the immune system or a defect of another system that affects the immune system, leading to an increased incidence or severity of infection. In these disorders, specific disease-fighting antibodies (immunoglobulins such as IgG, IgA, and IgM) are either missing or are present in reduced levels. Children who have **immunodeficiency** syndromes may be subject to infection, diseases, disorders, or allergic reactions to a greater extent than individuals with fully functioning immune systems.

Description

Immunodeficiency is a defect of any component of the immune system or a defect of another system that affects the immune system leading to an increased incidence or severity of infection. Immunoglobulin deficiencies refer to missing or reduced levels of immunoglobulin (IgG, IgA, IgM) associated with an inability to make adequate specific antibody. These antibodies are specific proteins (immunoglobulins) produced by the immune system to respond to bacteria, viruses, fungi, parasites, or toxins that invade the body. Each class of antibody binds to corresponding molecules (antigens) on the cell surfaces of certain foreign organisms or substances, attempting to protect the body against reactions or illness. When the immune system is challenged by invading organisms, the antibodies may each play a protective role:

- Immunoglobulin G (IgG) is the most abundant class of immunoglobulins, directed toward viruses, bacterial organisms, and toxins. It is found in most tissues and in plasma, the clear portion of blood.
- Immunoglobulin M (IgM) is the first antibody produced in an immune response to any invading organism or toxic substance.
- Immunoglobulin A (IgA) is activated early in response to invasion by bacteria and viruses. It is found in saliva, tears, and all other mucus secretions.
- As of the early 2000s, IgD activity is not well understood.
- Immunoglobulin E (IgE) is found in respiratory secretions and is directed toward invasion of the body by parasites and in allergic reactions such as hay fever, **atopic dermatitis**, and allergic **asthma**.

Immunoglobulins are made by white blood cells known as B cells (B lymphocytes). Any disease that harms the development or function of B cells will, therefore, affect the production of immunoglobulin antibodies. T cells, another type of white blood cell, may also be involved in immunodeficiency disorders. About 70 percent of immunoglobulin deficiencies involve B lymphocytes and 20–30 percent involve T lymphocytes. Another 10 percent may involve both B and T lymphocytes.

Many of the infections that occur in children with immunoglobulin deficiency syndromes are caused by bacterial organisms or microbes. Certain of these invasive organisms form capsules when they enter the body, a mechanism used to confuse the immune system. In a healthy body with an adequately functioning immune system, immunoglobulin antibodies bind to the capsule and overcome the bacteria's defenses. Streptococci, meningococci, and *Haemophilus influenzae*, organisms that cause diseases such as **otitis media**, **sinusitis**, **pneumonia**, **meningitis**, osteomyelitis, septic arthritis, and sepsis, all make capsules. Children with immunoglobulin deficiencies are also prone to viral infections, including echovirus, enterovirus, and **hepatitis B**. They may also develop infection after receiving live (attenuated) **polio vaccine**. This is one of the reasons that live **polio** vaccine is no longer used routinely in the United States.

There are two types of immunodeficiency diseases: primary and secondary. Immunoglobulin deficiency syndromes are primary immunodeficiency diseases. They account for 50 percent of all primary immunodeficiencies and are the largest group of immunodeficiency disorders. Some are well defined and some are not fully understood. Secondary disorders occur in normally healthy people who are suffering from an underlying disease that weakens the immune system. Successful treatment of the disease usually reverses the immunodeficiency.

Examples of well defined immunoglobulin deficiency disorders include the following:

- X-linked agammaglobulinemia is an inherited disease stemming from a defect on the X chromosome, consequently affecting more males than females. Defect results in absence or reduced numbers of B cells that do not mature and perform normal function. Mature B cells are capable of making antibodies and developing memory, a feature in which the B cell will rapidly recognize and respond to an infectious agent the next time it is encountered. All classes of immunoglobulin antibodies are decreased in agammaglobulinemia.
- Immunoglobulin heavy chain deletion, a form of agammaglobulinemia, is a genetic disorder in which part of the antibody molecule is absent. This condition results in the loss of several antibody classes and subclasses, including most IgG antibodies and all IgA and IgE antibodies. The disease occurs because part of the gene for the heavy chain has been lost.
- X-linked hypogammaglobulinemia can occur in combination with growth hormone (GH) deficiency, producing short stature and delayed **puberty**, primarily in boys but also occurring in girls.
- Transient hypogammaglobulinemia of infancy is a temporary disease of unknown cause. It is believed to be caused by a defect in the development of T helper cells (cells that recognize foreign antigens and activate T and B cells in an immune response). As the child ages, the number and condition of T helper cells improves, and this situation corrects itself.

Hypogammaglobulinemia is characterized by low levels of gammaglobulin antibodies in the blood. During the disease period, children may have decreased levels of IgG and IgA antibodies. In some infants with this disorder, laboratory tests are able to show that the antibodies present do not react properly with infectious bacteria.

- IgG subclass deficiency is a disorder associated with a poor ability to respond and make antibody against polysaccharide antigens, primarily pneumococcus.
- Selective IgA deficiency is an inherited disease characterized by a failure of B cells to switch from making IgM to IgA antibodies. The amount of IgA produced is limited in either serum or the mucosal linings of organs. This condition may result in more infections of mucosal surfaces, such as the nose, throat, lungs, and intestines. However, most persons with this abnormality are asymptomatic.
- IgM deficiency is characterized by the absence or low level of total IgM antibodies, the body's first defense against infection. This condition results in slow response to infective organisms and slow response to treatment.
- IgG deficiency with hyper-IgM is a disorder that results when B-cells fail to switch from making IgM to IgG. This condition produces an increase in the amount of IgM antibodies present and a decrease in the amount of IgG and IgA antibodies. This disorder is the result of a genetic mutation.
- **Severe combined immunodeficiency** (SVID) is not precisely an immunoglobulin deficiency, but a combined deficiency resulting from a T-cell disorder. The T-cell dysfunction can either be X-linked, affecting more males than females and characterized by the absence of T lymphocytes, or it can occur through autosomal inheritance (not sex linked), resulting in an absence of both T and B lymphocytes and a deficient thymus gland, the lymphoid organ that produces T-cell lymphocytes.
- **Common variable immunodeficiency** (CVID) is a primary immunodeficiency with onset of symptoms typically occurring in the second or third decade of life. It is never diagnosed before two years of age and is diagnosed only after drug toxicity and other primary immune deficiencies have been ruled out. IgG and IgA and/or IgM will be measured at about two standard deviations below normal. The individual will typically not make antibodies against protein or polysaccharide antigens and will not make IgM antibodies against incompatible blood group antigens (hemagluttinins). T-cell dysfunction is the variable in this disorder. Children who have this disorder are subject to recurring infections and may not respond appropriately to immunization.

Demographics

Primary immunoglobulin deficiency syndromes occur only rarely. Those that are X-linked occur more in males than females; other immunoglobulin deficiencies occur equally in both sexes. Detection of the syndromes usually occurs in childhood. Numbers of new cases of specific syndromes are difficult to estimate because many deficiencies go undiagnosed. Among the syndromes for which incidence rates are available are IgA deficiency (one in 500–700), agammaglobulinemia (one in 50,000–100,000), severe combined immunodeficiency or SCID (one in 100,000–500,000), and common variable immunodeficiency or CVID (one in 50,000–200,000).

Causes and symptoms

Primary immunoglobulin deficiencies are primarily the result of congenital defects that affect the development and function of B lymphocytes (B cells), the white cells that fight infection and disease. Defects can occur at two main points in the development of B-cells. First, B cells can fail to develop into antibody-producing cells. X-linked agammaglobulinemia is an example of this disease. Secondly, B cells can fail to make a particular type of antibody or fail to switch classes during maturation. Initially, when B cells start making antibodies for the first time, they make IgM. As they mature and develop memory, they switch to one of the other immunoglobulin classes. Failure to switch or failure to make a subclass can lead to immunoglobulin deficiency diseases. Defects in the thymus gland that manufactures T lymphocytes or defects in the T lymphocytes themselves can also result in reduced production of immunoglobulins.

Symptoms are frequent and so are persistent infections, particularly of the respiratory system. Frequent digestive disturbances and **diarrhea** may lead to malabsorption of essential nutrients and **failure to thrive**. Children with primary immunoglobulin deficiency syndromes will exhibit some of the following characteristics:

- signs of infection in the first days or weeks of life
- a slow response to treatment
- infection suppressed by appropriate treatment but not cured
- common bacterial or viral organisms causing increasingly acute recurring infections

- uncommon bacterial or viral organisms causing infection
- multiple simultaneous infections at more than one site
- delays in growth and development
- development of unexpected complications such as **anemias** and chronic diseases

When to call the doctor

Parents should seek medical care from a pediatrician or **family** practitioner if their young child or teenager has frequent or persistent infections such as upper respiratory infections, or chronic **cough**, ear infections, sinusitis, asthma, or pneumonia. Sores that do not heal or recurring or long-lasting skin irritations may also be signs of reduced immune system functioning.

Diagnosis

An immunodeficiency disease is suspected when children become ill frequently, especially repeat illness caused by the same organisms. Diagnosis will begin with a detailed history of the child's illnesses (dates, duration, and infection site) and review of all prior medications and immunizations and results of diagnostic tests performed. Determining which immunoglobulins are present and which are absent or present in reduced amounts is critical for diagnosis. Diagnostic testing may include routine blood tests such as a complete blood count (CBC) and differential (peripheral blood smear) to evaluate overall health and determine the type and number of red cells, white cells, and platelets present in the blood. Tests or cultures may be performed to determine the type of bacteria or virus causing recurring infections. B lymphocytes and T lymphocytes may be quantified. When immunodeficiency is suspected, levels of the classes of immunoglobulins are measured in blood serum by using a clinical laboratory procedure called electrophoresis. This procedure both quantifies the amount of each antibody present and identifies the various classes and subclasses of antibodies. Deficiencies may be noted in one class or subclass or in combinations of antibodies. Genetic testing may be done to help identify the type of immunodeficiency disease.

Treatment

Immunoglobulin deficiency diseases cannot be cured, but treatment that replaces or boosts specific immunoglobulins can help support immune function in affected children. Immune serum, obtained from donated blood that contains adequate levels of IgG antibodies, may sometimes be transfused as a source of antibodies to boost the immune response, even though it may not contain all antibodies needed and may lack antibodies specific for some of the recurring infections. The preferred treatment is to give specific immunoglobulins intravenously (immunoglobulin intravenous therapy or IVIG) or subcutaneously. No replacement therapy is available for treating IgA deficiencies.

Treatment will also focus on controlling infections in immunodeficient children. Immunization against frequent infection can be achieved in some children by administering polysaccaride-protein conjugate vaccines shown to improve immune response in certain types of infection. **Antibiotics** are used routinely at the first sign of an infection to help eliminate infectious organisms. Antifungal drug therapy may be administered to treat fungus infections. Few drugs are effective against viral diseases, and each viral illness will be evaluated and treated differently, depending on the virus and the overall health of the child. Bone marrow transplantation may correct immunodefiency in some cases.

Alternative treatment

Several nutritional supplements are reported to help build the immune system. These include garlic (contains the essential trace element germanium), essential fatty acids (found in flax seed oil, evening primrose oil, and fish oils), sea vegetables such as kelp, acidophilus to supply natural bacteria in the digestive tract, and **vitamins** A and C, both powerful antioxidants that improve immune function and increase resistance to infection. Zinc is another nutrient essential to immune system functioning. Green drinks made with young barley are believed to cleanse the blood and supply chlorophyll and nutrients for tissue repair. Alcohol, certain prescription and over-the-counter drugs, and coffee and other **caffeine** drinks should be avoided. Stress is known to produce biochemicals that reduce white blood cell functioning, making it important to get sufficient **sleep** and reduce stress to help keep the immune system functioning. Therapeutic massage, **yoga**, and other types of stress reduction programs are available in most communities.

Nutritional concerns

Immune system function requires having certain essential nutrients and avoiding things that depress immunity. A diet that improves immune system functioning includes nutrients obtained as much as possible from whole foods such as fresh fruits and vegetables, whole grain breads and cereals, and brown rice and whole grain pasta for essential vitamins, **minerals**, and fiber. Such a diet also limits or eliminates refined foods.

KEY TERMS

Antibody—A special protein made by the body's immune system as a defense against foreign material (bacteria, viruses, etc.) that enters the body. It is uniquely designed to attack and neutralize the specific antigen that triggered the immune response.

Antigen—A substance (usually a protein) identified as foreign by the body's immune system, triggering the release of antibodies as part of the body's immune response.

Autosomal inheritance—Inheritance involving any of the autosomes (22 pairs) and not involving sex-linked chromosomes X and Y.

Bacteria—Singular, bacterium; tiny, one-celled forms of life that cause many diseases and infections.

Immunization—A process or procedure that protects the body against an infectious disease by stimulating the production of antibodies. A vaccination is a type of immunization.

Immunoglobulin G (IgG)—Immunoglobulin type gamma, the most common type found in the blood and tissue fluids.

Thymus gland—An endocrine gland located in the upper chest just below the neck that functions as part of the lymphatic system. It coordinates the development of the immune system.

Virus—A small infectious agent consisting of a core of genetic material (DNA or RNA) surrounded by a shell of protein. A virus needs a living cell to reproduce.

Fish, fowl, and lean meats can be consumed in moderation. Sweets should be reduced or avoided.

Prognosis

Regular medical observation, treatment of symptoms, and appropriate immune system boosting usually produces a good result in children with immunodeficiencies. Prognosis is related to the immune system's ability to produce the specific antibodies that are missing or present in reduced amounts. Individuals with immunodeficiency syndromes may have a normal life span although a variety of complications can occur, including autoimmune, gastrointestinal, granulomatous, and malignant conditions as a result of progressive immune deficiency disorders and/or repeat infections.

Prevention

Immunodeficiency cannot be prevented; however, challenges to the immune system can be reduced and infections avoided in immunodeficient individuals. Immunoglobulin deficiencies require impeccable health maintenance and care, paying particular attention to good hygiene, balanced **nutrition**, sufficient rest, regular check ups and immunizations, and optimal dental care, as well as avoiding crowds and contact with other children or relatives with bacterial or virus infections.

Nutritional concerns

A healthy immune system can be maintained by providing essential nutrients through a good diet and regular supplementation. Parents can help assure that their children and teens have three nutritious, low-fat, high-fiber, whole-food meals a day (limiting or eliminating altogether refined or prepared foods and fast foods) and healthy snacks in between, such as nuts, fresh fruit, popcorn, raw veggies, and whole grain crackers with nut butters (if no **allergies** to peanuts or other nuts), naturally sweetened jellies, and low-fat cheeses. Vitamin supplements should include vitamins A, C, and E, valuable parts of the body's defense system that help to increase production of healthy white blood cells and to fight infection.

Parental concerns

Parents with immunoglobulin deficient children and teenagers will likely be concerned that their children are in frequent contact with schoolmates and friends, the common route to infection. When infection occurs frequently, it is important to remember that the pediatrician or family practitioner will have specific criteria and diagnostic tests for evaluating the child, identifying the immunodeficiency, and determining appropriate therapy. Meanwhile, parents can help keep their children away from crowds and avoid contact with other children or relatives with bacterial or virus infections.

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Immunotherapy *see* **Allergy shots**

Impetigo

Definition

Impetigo refers to a very localized bacterial infection of the skin. There are two types, bullous and epidemic.

Description

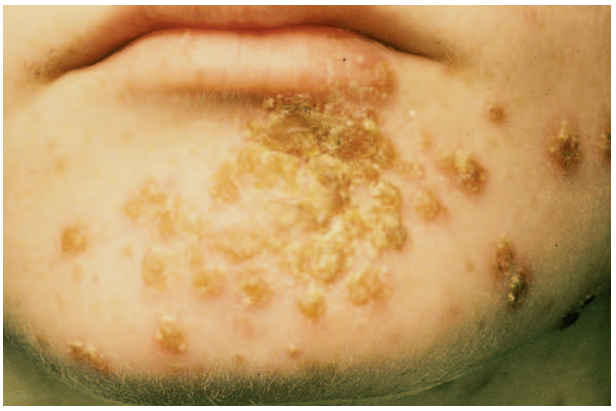
Impetigo is a skin infection that tends primarily to afflict children. Impetigo caused by the bacterium *Staphylococcus aureus* (also known as staph) affects children of all ages. Impetigo caused by the bacteria called group A streptococci (also known as strep) are most common in children ages two to five.

The bacteria that cause impetigo are very contagious. They can be spread by a child from one part of his or her body to another by scratching or contact with a towel, clothing, or stuffed animal. These same methods can pass the bacteria on from one person to another.

Impetigo tends to develop in areas of the skin that have already been damaged through some other mechanism (a cut or scrape, burn, insect bite, or vesicle from **chickenpox**).

Demographics

About 10 percent of all skin problems in children are ultimately diagnosed as impetigo.



Impetigo is a contagious bacterial skin infection that has affected the area around this patient's nose and mouth. *(© NMSB/Custom Stock Medical Photo, Inc.)*

Causes and symptoms

The first sign of bullous impetigo is a large bump on the skin with a clear, fluid-filled top (a vesicle). The bump develops a scab-like, honey-colored crust. There is usually no redness or **pain**, although the area may be quite itchy. Ultimately, the skin in this area will become dry and flake away. Bullous impetigo is usually caused by staph bacteria.

Epidemic impetigo can be caused by staph or strep bacteria and (as the name implies) is very easily passed among children. Certain factors, such as heat and humidity, crowded conditions, and poor hygiene increase the chance that this type of impetigo will spread rapidly among large groups of children. This type of impetigo involves the formation of a small vesicle surrounded by a circle of reddened skin. The vesicles appear first on the face and legs. When a child has several of these vesicles close together, they may spread to one another. The skin surface may become eaten away (ulcerated), leaving irritated pits. When there are many of these deep, pitting ulcers, with pus in the center and brownish-black scabs, the condition is called ecthyma. If left untreated, the type of bacteria causing this type of impetigo has the potential to cause a serious kidney disease (glomerulonephritis). Even when impetigo is initially caused by strep bacteria, the vesicles are frequently secondarily infected with staph bacteria.

Impetigo is usually an uncomplicated skin condition. Left untreated, however, it may develop into a serious disease, including osteomyelitis (bone infection), septic arthritis (joint infection), or **pneumonia**. If large quantities of bacteria are present and begin circulating in the bloodstream, the child is in danger of developing an overwhelming systemic infection known as sepsis.

KEY TERMS

Systemic—Relating to an entire body system or the body in general.

Ulcer—A site of damage to the skin or mucous membrane that is characterized by the formation of pus, death of tissue, and is frequently accompanied by an inflammatory reaction.

Vesicle—A bump on the skin filled with fluid.

Diagnosis

Characteristic appearance of the skin is the usual method of diagnosis, although fluid from the vesicles can be cultured and then examined in an attempt to identify the causative bacteria.

Treatment

Uncomplicated impetigo is usually treated with a topical antibiotic cream called mupirocin. In more serious, widespread cases of impetigo, or when the child has a **fever** or swollen glands, **antibiotics** may be given by mouth or even through a needle placed in a vein (intravenously).

Prognosis

Prognosis for a child with impetigo is excellent. The vast majority of children recover quickly, completely, and uneventfully.

Prevention

Prevention involves good hygiene. Hand washing; never sharing towels, clothing, or stuffed animals; and keeping fingernails well-trimmed are easy precautions to take to avoid spreading the infection from one person to another.

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Impulse control disorders

Definition

Impulse control disorders are a relatively new class of **personality disorders** characterized by an ongoing inability to resist impulses to perform actions that are harmful to oneself or others. The most common of these are **intermittent explosive disorder**, kleptomania, pyromania, compulsive gambling disorder, and **trichotillomania**.

Description

Impulse control disorders include five conditions that involve a recurrent failure to resist impulsive behaviors that harm one's self or others: intermittent explosive disorder, pyromania, kleptomania, trichotillomania, and compulsive gambling disorder. Intermittent explosive disorder involves unusually aggressive and violent outbursts. Pyromania is characterized by repetitive and purposeful fire-setting. Kleptomania involves urges to steal and repetitive acts of unnecessary theft. Trichotillomania is recurrent pulling out of hair. Compulsive gambling disorder is maladaptive, repetitive gambling.

Repetitive **self-mutilation** is considered by some experts to be a type of impulse control disorder. In this condition, people cause intentional harm to themselves through burning, cutting, or scratching.

Demographics

The incidence of impulse control disorders in children and adolescents is difficult to determine. In general, intermittent explosive disorder, pyromania, and compulsive gambling disorder are more common in boys, while kleptomania is more common in girls.

Causes and symptoms

Exact causes of impulse control disorders are unknown, but may be linked to genetics, **family** environment, and/or neurological factors. Some research suggests that impulse control disorders are linked to certain hormones, abnormal nerve impulses, and variations in brain chemistry and function. Children and adolescents who have had a severe **head injury** and who have epilepsy may be at greater risk of developing these disorders. In children and adolescents, impulse control disorders often occur along with other psychological conditions, such as attention-deficit hyperactivity disorder (ADHD).

Intermittent explosive disorder is characterized by episodes of aggressive and violent outbursts and loss and lack of control of anger. Often, explosive episodes result in destruction of property, domestic violence, and physical assault, which, in turn, have legal ramifications. The degree of aggressiveness during each episode is grossly out of proportion to any stresses.

Pyromania is the repetitive, deliberate, and purposeful setting of fires. Children and adolescents with pyromania are often aroused by fire-setting, and/or feel pleasure, relief, or gratification when setting fires or witnessing the consequences of fire. In addition, pyromaniacs are fascinated and attracted to fire and related accessories (e.g., matches, lighters), and are unnaturally curious about its use and consequences. Fire-setting is not performed for any other reasons, such as for financial gain, to express anger, to conceal a criminal act, or to express sociopolitical views.

Kleptomania is an inability to resist impulses to repetitively steal objects that are not necessary for personal use or monetary value. Children and adolescents with kleptomania experience a growing sense of tension just before **stealing**, followed by pleasure, relief, or gratification during or just after stealing. Career thieves, those who steal out of need or to support substance abuse, and those who steal because they have no regard for society's laws, are not considered to have kleptomania. Individuals with kleptomania do not want to steal and feel guilty about it.

Trichotillomania is characterized by recurrent pulling out of one's hair to produce noticeable hair loss. Children and adolescents with trichotillomania experience a growing sense of tension or stress just before pulling hair out or when trying to resist hair pulling. They experience pleasure, relief, or gratification when pulling out the hair.

Compulsive gambling disorder, also called pathological gambling, is recurrent and persistent gambling behavior characterized by five or more of the following:

- having a preoccupation with gambling
- needing to gamble with increasingly larger amounts of money to achieve the same excitement
- having repeated unsuccessful efforts to control gambling
- telling lies to family members, therapists, and others to conceal extent of gambling involvement
- committing forgery, fraud, or theft to finance gambling
- being restless or irritable when trying to stop gambling
- gambling to escape problems or to relieve tension or other feelings
- jeopardizing or losing a significant job, relationship, or other opportunity due to gambling
- relying on others to provide financial support due to financial difficulties caused by gambling

When to call the doctor

Parents of children and adolescents who exhibit problems with impulse control should see a physician as soon as possible. Usually, a referral to a psychologist, psychiatrist, or therapist will be given.

Diagnosis

Impulse control disorders are diagnosed by psychological and psychiatric evaluations, interviews with family members, teachers, and caregivers, and observation and interviews with the child or adolescent. Diagnosis is based on clinical criteria defined in the American Psychiatric Association's *Diagnostic and Statistical Manual of Mental Disorders, Fourth Edition, Text Revision C (DSM-IV-TR)*.

Impulse control disorders often have characteristics in common with other psychological disorders and often occur in conjunction with other conditions, such as ADHD or **conduct disorder**. Therefore, diagnosis of impulse control disorders may be difficult, and they are usually diagnosed after exclusion of other disorders. For example, intermittent explosive disorder is diagnosed if the aggressive episodes cannot be better explained by another psychological disorder, such as **antisocial personality disorder**; a manic episode; ADHD; or by substance abuse or medical conditions such as head trauma. Pyromania is diagnosed when fire-setting is not better explained by conduct disorder, antisocial personality disorder, mental impairment, delusions or hallucinations, or intoxication. Kleptomania is diagnosed when repetitive

stealing is not better explained by anger or vengeance, **peer pressure**, delusions or hallucinations, conduct disorder, a manic episode, or antisocial personality disorder. Trichotillomania is diagnosed when pulling out of hair is not better explained by another mental disorder or a dermatological or medical condition, and when this practice causes clinically significant social or occupational dysfunction or impairment. Compulsive gambling disorder is diagnosed when the behavior cannot be better explained by a manic episode, conduct disorder, or peer pressure.

Treatment

Impulse control disorders are treated with medication, psychotherapy, and behavior modification. If these disorders are occurring in conjunction with another condition, such as ADHD, medication and therapy for that condition often helps alleviate the impulse control disorder. Depression is often an underlying factor in some impulse control disorders, particularly compulsive gambling disorder and trichotillomania. Therefore, treatment with **antidepressants** may be helpful.

Long-term counseling and psychotherapy is usually necessary as well. Therapy methods to help with impulse control generally involve behavior modification, anger and stress management, and psychoanalysis. Therapy can occur in residential or day treatment facilities, or on an outpatient basis. Support groups, such as Gamblers Anonymous, may also help.

Prognosis

Prognosis depends on the severity of the disorder and the commitment of the individual to seek therapy. Impulse control disorders can affect social, academic, and occupational functioning, as well as result in legal problems. Long-term participation in individual counseling and group therapy can improve prognosis.

Prevention

Impulse control disorders cannot be prevented.

Parental concerns

Children and adolescents with impulse control disorders may have difficulties in school and at home. In some cases, impulse control disorders can result in expulsion from school. Parents should investigate **alternative school** settings that may be able to provide counseling and group therapy integrated with academics. **Family therapy** may help alleviate stressful family situations and help other family members understand the impulse control disorder.

KEY TERMS

Alternative school—An educational setting designed to accommodate educational, behavioral, and/or medical needs of children and adolescents that cannot be adequately addressed in a traditional school environment.

Antisocial personality disorder—A disorder characterized by a behavior pattern that disregards for the rights of others. People with this disorder often deceive and manipulate, or their behavior might include aggression to people or animals or property destruction, for example. This disorder has also been called sociopathy or psychopathy.

Attention deficit hyperactivity disorder (ADHD)—A condition in which a person (usually a child) has an unusually high activity level and a short attention span. People with the disorder may act impulsively and may have learning and behavioral problems.

Conduct disorder—A behavioral and emotional disorder of childhood and adolescence. Children with a conduct disorder act inappropriately, infringe on the rights of others, and violate societal norms.

Manic episode—A distinct period of abnormally and persistently elevated, expansive, or irritable mood, lasting at least one week, characterized by inflated sense of self-importance, decreased need for sleep, extreme talkativeness, racing thoughts, and excessive participation in pleasure-seeking activities.

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(202) 966-7300. Fax: 202-966-2891. Web site: <www.aacap.org>.

American Psychiatric Association. 1000 Wilson Boulevard, Suite 1825, Arlington, Va. 22209-3901. (703) 907-7300 Web site: <www.psych.org>.

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Jennifer E. Sisk, M.A.

Inclusion conjunctivitis

Definition

Inclusion **conjunctivitis** is an inflammation of the conjunctiva, or white of the eye. In the neonate this condition is part of a larger group of eye diseases called neonatal conjunctivitis. Inclusion conjunctivitis is also called a chlamydial conjunctivitis.

Description

Chlamydiae are similar to bacteria but cannot produce their own energy and thus live in the cells of other organisms. Once inside the host cell, chlamydiae replicate and form inclusion bodies. They then replace and finally destroy the cell membrane of the host, releasing more chlamydiae to continue the infection process. The life cycle of chlamydia is 72 hours. Chlamydiae are found in parts of the body with a mucosal membrane, which are the eye, the respiratory tract, and the genitourinary tract.

Transmission

Neonatal inclusion conjunctivitis develops within five to 12 days after birth and is contracted as the child passes through the mother's cervix. Two-thirds of those females with a chlamydial infection pass the infection on to the child during **childbirth**.

Adult inclusion conjunctivitis, which can affect sexually active adolescents, is usually transmitted sexually and develops when the eye is infected by the urogenital secretions of an individual infected with chlamydia, but it can be transmitted by eye-to-eye contact. Symptoms do not always exist with chlamydial infections, and thus it is often transmitted unknowingly. Up to 80 percent of female adults and adolescents with inclusion conjunctivitis are asymptomatic, and almost half of those with adult inclusion conjunctivitis do not have a systemic infection of chlamydia.

Demographics

The exact number of individuals with adult inclusion conjunctivitis is not known. But adult inclusion conjunctivitis, which is seen only if one is infected with chlamydia, affects 3 million annually in the United States. It is seen most often in sexually active 15 to 30 years olds, and most of these infections are reported in women 15 to 19. Forty-six percent of new cases of chlamydia fall within this group. Up to 10 percent of pregnant women harbor the chlamydial parasite. Twenty-five percent of those men with chlamydia infections are not aware of their infection.

Up to 6 percent of newborns develop neonatal inclusion conjunctivitis. Forty percent of neonatal conjunctivitis is due to chlamydia. Between 35 and 50 percent of newborns infected with chlamydia develop neonatal inclusion conjunctivitis. Neonatal chlamydial or inclusion conjunctivitis is 10 times more common than neonatal gonorrheal conjunctivitis.

Causes and symptoms

Inclusion conjunctivitis is caused by an intracellular organism called *Chlamydia trachomatis*.

The signs and symptoms of adult inclusion conjunctivitis appear two to 19 days after contact with an individual who harbors the chlamydia parasite. The symptoms of adult inclusion conjunctivitis are a foreign body sensation, watery eyes, and eyelids that stick together upon awakening. Large follicles may be seen if the lower lid is pulled down. The lymph nodes near the ears, called the preauricular nodes, may be swollen. Because the symptoms of chlamydia wax and wane and because the

adolescent or adult may be asymptomatic, proper diagnosis may be delayed.

The signs of neonatal inclusion conjunctivitis appear five to 14 days after birth. Since the lymphatic system of the newborn is not well developed, follicles will not usually be present, and the lymph nodes will not be enlarged, but the eye of the neonate with chlamydia will be red and inflamed. The infant will be tearing and have a purulent ocular discharge, and the eyelids will be swollen. Other accompanying symptoms in the infant include a **cough** and **rhinitis**.

When to call the doctor

Any red eye, with or without discharge, should be examined by an appropriate healthcare practitioner. There are many causes of eye problems, and appropriate treatment should be instituted as soon as possible if inclusion conjunctivitis is the cause of the ocular problem.

The children of mothers who give birth outside the traditional hospital setting should contact their healthcare provider regarding the necessity of prophylactic antibiotic drops. A healthcare provider should be informed if the mother or father of a newborn has an untreated sexually transmitted disease.

Diagnosis

The diagnosis of inclusion conjunctivitis cannot be made definitively without laboratory testing, but the signs of inclusion conjunctivitis can be seen by the eye care provider, even if a patient is not symptomatic. Follicles can seen on the inside inferior eyelids and occasionally under the superior eyelid of the patient with adult inclusion conjunctivitis, and if treatment has been delayed, scarring of the interior of the eyelids may be present as well as kerititis, an inflammation of the cornea, and neovascularization, or new blood vessel formation of the cornea. Upon questioning the individual may report a history of a genitourinary infection.

The laboratory testing for inclusion conjunctivitis begins with swabbing a sample from the inside of the eyelids to test for the presence of the characteristic inclusion bodies made only by chlamydia. The Giemsa stain is used often to diagnose neonatal inclusion conjunctivitis. This technique has a high rate of false positives for the adult with inclusion conjunctivitis. Immunofluorescence monoclonal antibody testing is very sensitive technique that gives a rapid diagnosis of inclusion conjunctivitis. Other techniques used to diagnose a chlamydial infection are enzyme immunoassays, serum antibody tests, and DNA probes.

Since inclusion conjunctivitis can mimic other diseases, it is important to rule out other types of conjunctivitis, such as those of viral etiology or allergy or those caused by gonorrhea.

Treatment

Neonatal inclusion conjunctivitis may resolve spontaneously within nine months without treatment. But the standard treatment for an infant younger than four months of age is oral erythromycin, four times a day for two weeks. The eye may be irrigated with saline to help remove the mucus discharge. The parents of the infant are treated as well.

Doxycycline, tetracycline, ocufloxacin, and erythromycin are sometimes prescribed. Tetracycline is not given to children under eight years of age, and ocufloxacin is not given to those under 18 years of age. Neither drug is given to pregnant or nursing women because of side effects. **Topical antibiotics** are not required if systemic or oral medication is prescribed, but if there is a co-existing inflammation in the eye, then topical steroids may be given. Finally, the sexual partners of individuals with inclusion conjunctivitis must also be receive antibiotic treatment.

Prognosis

Usually adult inclusion conjunctivitis resolves within two to four weeks with treatment. Rarely does inclusion conjunctivitis lead to blindness, unless it has been left untreated for months or longer. If untreated, a chlamydial infection can lead to pelvic inflammatory disease and scarring of fallopian tubes in women, causing infertility or ectopic pregnancies. In males, urethritis may result.

Ten to 20 percent of infants infected with chlamydia develop **pneumonia** during the first six months of life. In the infant, inclusion conjunctivitis may persist for several years.

Prevention

Since in the United States adult inclusion conjunctivitis is primarily a sexually transmitted disease, the incidence of inclusion conjunctivitis can be decreased either through abstinence or through the use of condoms. Pregnant women with a chlamydial infection should talk to their doctor about treatment of the infection. Antibiotic eye drops only may not be sufficient to prevent inclusion conjunctivitis in the newborn if the mother is infected with chlamydia.

KEY TERMS

Chlamydia—The most common bacterial sexually transmitted disease in the United States. It often accompanies gonorrhea and is known for its lack of evident symptoms in the majority of women.

Conjunctiva—Plural, conjunctivae. The mucous membrane that covers the white part of the eyes (sclera) and lines the eyelids.

Cornea—The clear, dome-shaped outer covering of the eye that lies in front of the iris and pupil. The cornea lets light into the eye.

Gonorrhea—A sexually transmitted disease that causes infection in the genital organs and may cause disease in other parts of the body.

Trachoma—A type of chlamydia that causes blindness.

The incidence of neonatal conjunctivitis can be reduced by applying erythromycin ointment to the newborn's eyes shortly after delivery. Silver nitrate, which may be instilled at some institutions at birth (instead of erythromycin), is not effective against chlamydia.

Parental concerns

In the newborn, inclusion conjunctivitis may resolve spontaneously, but there are chlamydial infections which can cause blindness if not treated. So, any eye problem in the newborn needs to be diagnosed properly and treated as indicated by the pediatrician or eye-care provider.

When inclusion conjunctivitis is diagnosed in an adolescent, it is almost always has been contracted through sexual activity. Sixty-five percent of adolescents have had sexual intercourse by age 16. Only 40 percent of young adults between 18 and 21 years of age used a **condom** during their most recent sexual encounter, and 45 percent of these individuals have already had more than three sexual partners. Because **peer pressure** makes it difficult to resist sex and because adolescents have difficulty understanding they are risks of contracting **sexually transmitted diseases**, including chlamydial infections, parents should be involved in helping the adolescent understand these risks.

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Martha Reilly, OD

Infant massage

Definition

Infant massage is the process of rubbing an infant's muscles and stroking the infant in a manner specifically designed for them. Although there are professionally trained and certified infant massage therapists, the obvious first choice to massage the baby is the mother, father, grandparent, or guardian. Equally important are the people who care for children outside the home such as, nurses on neonatal intensive care units (NICU) that work with premature babies and those who work with the disabled. The benefits derived from massage are applicable and advantageous for all of these groups.

Purpose

Infant massage provides many benefits for the infant. A caring touch is good for everyone, but especially for infants who are new to the world and need the reassurance of someone special being there for them. However, there are some major benefits for the massage givers as well. They gain an increased awareness of the baby and his or her needs while enhancing the **bonding** process between care giver and baby. Research from experiments conducted at the Touch Research Institutes at the University of Miami School of Medicine and Nova Southeastern University has been cited for the clinical benefits massage has on infants and children. Touch therapy triggers many physiological changes that help infants and children grow and develop.

Studies have shown that infant massage alleviates the stress that newborns experience as a result of the enormous change that birth creates. They have just spent nine months in a home that fed them; kept them warm; brought them the oxygen they needed; took care of waste products; and provided a gentle rocking motion to soothe them. Now, the outside world has taken over, and things are not as simple as they were. Massage enables a smoother transition from the comfortable womb to that of humankind. The benefits of massage for the infant include:

- It helps baby learn to relax.
- It improves immune system.
- It promotes bonding and communication.
- It promotes positive body image.
- It decreases the production of stress hormones.
- It promotes sounder and longer sleep.
- It helps to regulate digestive, respiratory, and circulatory systems.
- It helps relieve discomfort from gas and **colic**, congestion, and teething.

The benefits of massage for parents include:

- It improves parent-infant communication.
- It helps parents to understand and respond appropriately to baby's nonverbal cues.
- It eases stress of parent who must be separated from child during the day.
- It promotes feelings of competence and confidence in caring for baby.
- It provides a special focused time that helps deepen bonding.
- It increases parents' ability to help child relax in times of stress.
- It is fun and relaxing for parents to massage their children.

There are additional benefits that can be derived from infant massage to elicit positive outcomes for premature infants and disadvantaged mothers. They include:

- Cross-cultural studies show that babies who are held, massaged, carried, rocked, and breast fed grow into less aggressive and violent adults who demonstrate a greater degree of compassion and cooperation.
- Recent research demonstrates benefits for premature infants, children with **asthma**, diabetes, and certain skin disorders.
- Mothers with postpartum depression have shown improvement after starting infant massage.
- Teenage mothers have shown improved bonding behavior and interactions with their infants.

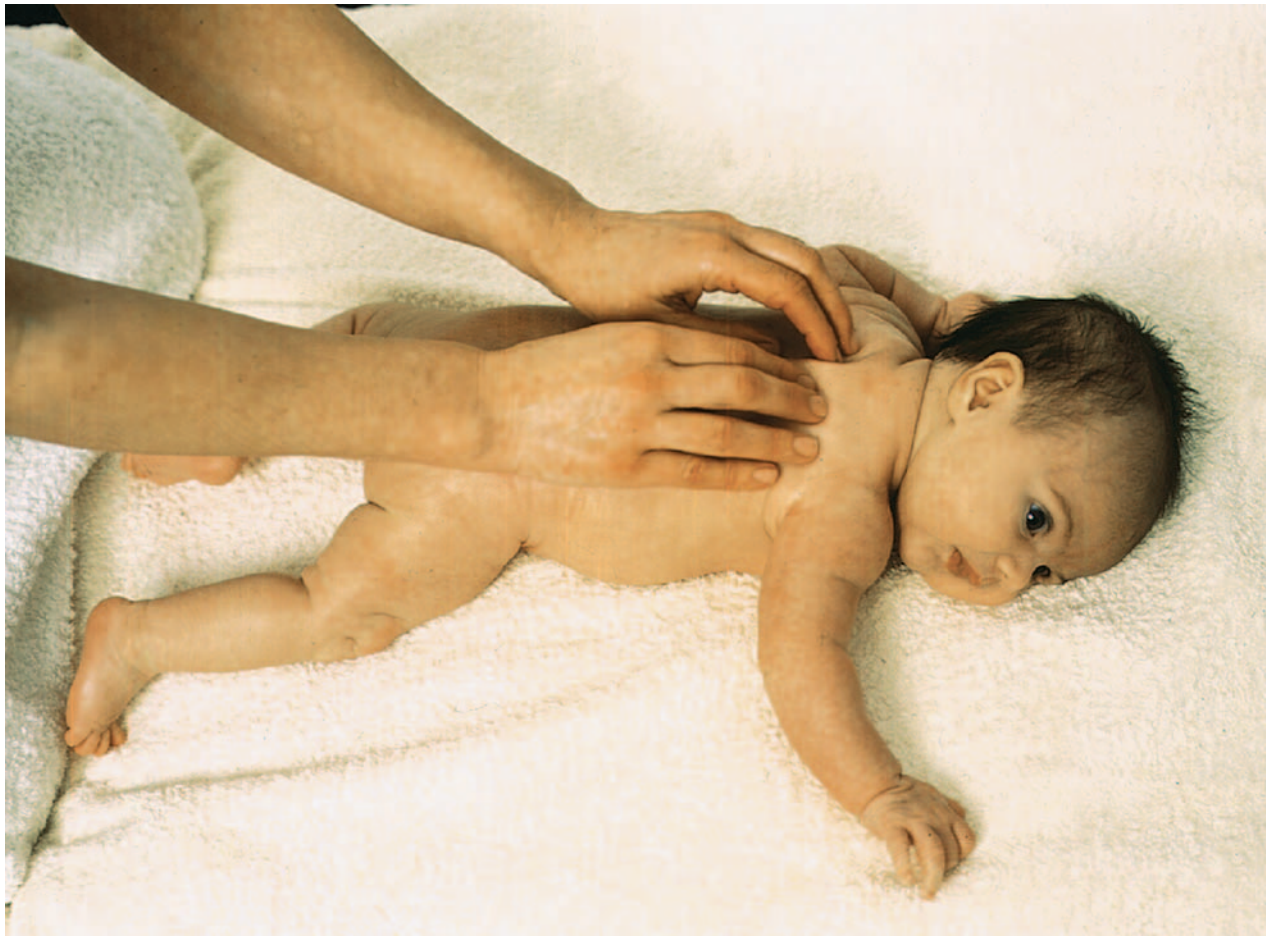
Description

Origins

Infant massage is an ancient practice used primarily in Asian and Pacific Island cultures because touch in these cultures is considered healthful both physically and spiritually. For example, the inclusion of infant massage into regular bath time is typical of the Maoris and Hawaiians. With the introduction of infant massage in the West in the late 1970s, it was tested to prove or disprove its efficacy. Dr. Frederick Leboyer, a French physician who advocated natural **childbirth**, supported the interest in infant massage with the publication of his photojournalistic book on the Indian art of baby massage. He believed that touch is the child's first language and that understanding spoken language comes long after understanding touch.

Infant massage was introduced formally into the United States in 1978 when Vimala Schneider McClure, a **yoga** practitioner who served in an orphanage in Northern India, developed a training program for instructors at the request of childbirth educators. An early research study by R. Rice in 1976 had shown that premature babies who were massaged surged ahead in weight gain and neurological development over those who were not massaged. McClure's practice in India, her knowledge of Swedish massage and reflexology along with her knowledge of yoga postures, which she had already adapted for babies, served to make her the foremost authority on infant massage. The International Association of Infant Massage (IAIM) had its origins in 1980 and was incorporated in 1986 by McClure and her original seven trainers. As of 2004, there were over 30 countries that have chapters of IAIM and over 15,000 certified instructors have been trained in the United States.

Various techniques are used in infant massage, with the different strokes specific to a particular therapy. Special handling is used for treating a baby with gas and colic. Some of the strokes are known as Indian milking, which is a gentle stroking of the child's legs; and the twist and squeeze stroke, a gentle squeeze of the muscles in the thigh and calf. The light strokes often employed in regular Swedish massage are applied at the end of a massage. The procedure is not unlike certain forms of adult massage, but with extra care taken for the fragility of the infant.



Infant receiving a massage. *(© Photo Researchers, Inc.)*

There are also specific Chinese techniques of pediatric massage, including massage of children with special needs. In China, these forms of massage can be given by medical professionals, but parents are often taught how to do the simpler forms for home treatment of their children.

Preparation

It is good to get a baby into a routine for massage. The time can be early in the morning, after a bath, or just before bedtime—the caregiver and baby know what is best and the time can be determined by the response. The room needs to be warm because the baby's clothes will be removed and infants have a difficult time regulating their body temperature. This is especially true for premature babies. It is preferable to have the room not be too bright with electrical light or sunlight shining on the baby's face. Research has shown that babies prefer to be massaged with oil such as a vegetable or plant oil. Traditional baby oils are mineral based, which are not readily absorbed. The two oils preferred by most massage therapists are grape seed oil and sweet almond oil. A caregiver can try both and see which is the most desirable. Generally, removing the diaper permits greater freedom of movement for the baby. For protection, the baby can by placed on a thick towel.

Precautions

It is necessary to use caution when performing infant massage in order not to injure the infant. Strokes are made with the greatest delicacy, and appropriate techniques are taught by licensed massage therapists to ensure that the infant is treated with accepted physical touch. Anyone who is unfamiliar with handling a baby should receive appropriate instruction before beginning infant massage.

Risks

No adverse side effects have been reported when infant massage is done properly after careful instruction, or by a licensed massage therapist who specializes in infant care.

Normal results

Many studies have been mentioned relating to the benefits of massage and there has been research published as early as 1969 relating to the topic. Hundreds of individual projects have been conducted throughout the world focusing on infant massage. Many of the studies are related to the benefits of massage and touch for premature infants and others born with such risk factors as drug dependence or **cerebral palsy**. Needless to say, the benefits are overwhelmingly positive and the research indicates that infant massage is increasingly recognized as a legitimate health care treatment.

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Infant mortality

Definition

The infant mortality rate is the number of deaths of infants under one year of age per 1,000 live births in a given population. In 2002, the United States' infant mortality rate varied widely by race of the mother from 14.3 for infants of black mothers to 5.9 for infants of Hispanic mothers to 5.8 for infants of white mothers. As can be noted, the mortality rate for black infants is more than twice that of white infants. The overall infant mortality rate in 2002 for all races was 7.0 per 1,000 live births, which was a slight increase over the previous year.

Description

Infant mortality rate is one of the key indicators of a nation's health status. When the rate increases, as it did from 2001 to 2002, the factors that precipitated this change need to be assessed and scrutinized. The U.S. infant mortality rate is of great concern because the United States has fallen to the twenty-second nd place among industrialized nations in infant mortality rankings. Therefore, healthcare professionals and the public have stressed the need for better prenatal care, coordination of health services, and the provision of comprehensive maternal-child services.

Infant mortality rates have typically been the highest for the babies of adolescent mothers and lowest for women in their late 20s and early 30s. The rates have also been high for women in their forties and older. In general, infant mortality rates decrease with increasing maternal educational levels. Similarly the infant mortality rate for unmarried mothers is often more than 83 percent higher than the mortality rate for married women. Likewise, the infant mortality rate is characteristically higher for the infants of mothers who smoke than for those of nonsmokers.

The leading cause of infant mortality is congenital malformations, deformations and chromosomal abnormalities with a rate of 20.2 percent. Disorders related to short gestation and low birth weight was the second leading cause of death for all infants at 16.4 percent of all deaths. **Sudden infant death syndrome** (SIDS) is the third leading cause of infant death. Its incidence decreased by about

KEY TERMS

Perinatal mortality—The number of late fetal deaths, 28 weeks or more gestation, and neonatal deaths that occur in the first seven days.

9 percent, which it has been doing since 1988. The fourth leading cause of death comes under the heading of newborn affected by maternal complications of pregnancy. This rate actually increased from 2001 to 2002 from 37.2 per 100,000 live births to 42.9 per 100,000 in 2002.

An analysis of the data established that the rise in the infant mortality rate was concentrated in the neonatal period (less than 28 days) and primarily in the first week of life where more than half of all infants' deaths occur. Final birth data for 2002 made it apparent that two key predictors of infant health, the percentage of infants born preterm (less than 37 weeks gestation) and low birth weight (less than 2,500 grams) rose during this time frame. This has been a continuing long-term upward trend. The **cesarean section** rate for 2002 rose to 26.1, which is the highest ever recorded in the US. The primary cesarean rate was 7 percent higher than the previous year, and the rate of vaginal birth after cesarean (VBAC) experienced a sharp decline. The cesarean rate increase could be due to nonmedical factors as demographics, physician practice patterns, and maternal choice. Other contributing factors may be the use of continuous **electronic fetal monitoring** and inductions before 41 weeks gestation. Unnecessary interventions can contribute to a rise in cesarean rates. On the other hand, the perinatal mortality rate (the number of late fetal deaths [28 weeks or more gestation] and early neonatal deaths [less than 7 days] per 1,000 live births) remain unchanged.

Common problems

The infant mortality rate increased in the United States in 2002 for the first time since 1958, which indicates a need to examine what factors contributed to this raise. Is there a difference in mortality rates among racial groups? That is obvious—the rate for blacks is 14.2 and the rate for whites is 5.8. Experts associate this difference with the availability of prenatal care to minorities. It is expensive, and over 40 million Americans do not have health insurance. The mother's socioeconomic status is a possible contributing factor because the leading cause of death was related to congenital malformations, which in some cases can be eliminated with appropriate nutritional intake and prenatal **vitamins**. Lack of prenatal care could also contribute to the fourth largest cause of infant death, which is maternal complications. Many other industrialized countries have a socialized system of health care, which offers universal access to prenatal care and helps lower country-wide infant mortality rates.

Parental concerns

Recent data showed good news for parents of teenagers. The teen birth rate declined by 30 percent over the past decade to a historic low and the rate for black teens was down by more than 40 percent. For young black teens (15 to 17 years) the results were even more striking—the rate was cut in half since 1991. The average age at first birth was 25.1 years in 2002, an all-time high in the United States. Birth rates for women 35–39 (41 births per 1,000 women) and 40–44 (eight per 1,000) were the highest in more than three decades. The rate for women ages 20–24 (104 births per 1,000 women) was on the decline and the rate for those 25–29 was stable, but still the highest of all age groups, at 114 per 1,000 women. In contrast, the rate for teens was 43 per 1,000. In addition, just over one in 10 women smoked during pregnancy in 2002, a decline of 42 percent since 1989.

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Infant respiratory distress syndrome *see* **Respiratory distress syndrome**

Infantile eczema *see* **Atopic dermatitis**

Infantile paralysis *see* **Polio**

Infectious hepatitis *see* **Hepatitis A**

Infectious mononucleosis

Definition

Infectious mononucleosis is a contagious illness caused by the Epstein-Barr virus that can affect the liver, lymph nodes, and oral cavity. While mononucleosis is not usually a serious disease, its primary symptoms of fatigue and lack of energy can linger for several months.

Description

Infectious mononucleosis, frequently called "mono" or the "kissing disease," is caused by the Epstein-Barr virus (EBV) found in saliva and mucus. The virus affects a type of white blood cell called the B lymphocyte, producing characteristic atypical lymphocytes that may be useful in the diagnosis of the disease.

The disease typically runs its course in four to six weeks in people with normally functioning immune systems. People with weakened or suppressed immune systems, such as **AIDS** patients or those who have had organ transplants, are particularly vulnerable to the potentially serious complications of infectious mononucleosis.

Demographics

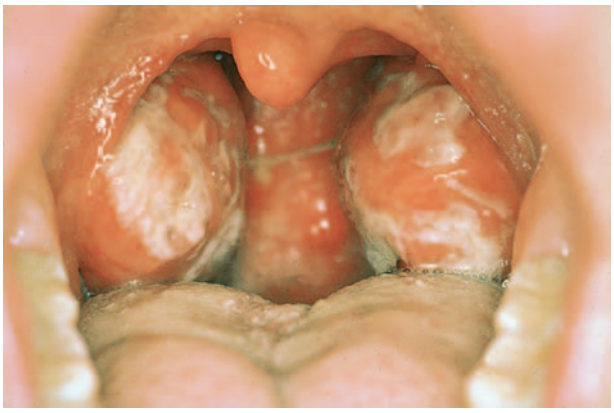
While anyone, even young children, can develop mononucleosis, it occurs most often in young adults between the ages of 15 and 35 and is especially common in teenagers. The mononucleosis infection rate among college students who have not previously been exposed to EBV has been estimated to be about 15 percent. In younger children, the illness may not be recognized.

Causes and symptoms

The EBV that causes mononucleosis is related to a group of herpes viruses, including those that cause cold sores, **chickenpox**, and shingles. Most people are exposed to EBV at some point during their lives. Mononucleosis is most commonly spread by contact with virus-infected saliva through coughing, sneezing, kissing, or sharing drinking glasses or eating utensils.

In addition to general weakness and fatigue, symptoms of mononucleosis may include any or all of the following:

- **sore throat** and/or swollen tonsils
- fever and chills
- nausea and **vomiting**, or decreased appetite
- swollen lymph nodes in the neck and armpits



Sore throat and swollen tonsils caused by infectious mononucleosis, frequently called mono or the kissing disease. *(Photograph by Dr. P. Marazzi. Science Photo Library/Photo Researchers, Inc.)*

- headaches or joint **pain**
- enlarged spleen
- jaundice
- skin rash

Complications that can occur with mononucleosis include a temporarily enlarged spleen or inflamed liver. In rare instances, the spleen may rupture, producing sharp pain on the left side of the abdomen, a symptom that warrants immediate medical attention. Additional symptoms of a ruptured spleen include light-headedness, rapidly beating heart, and difficulty breathing. Other rare, but potentially life-threatening, complications may involve the heart or brain. The infection may also cause significant destruction of the body's red blood cells or platelets.

Symptoms do not usually appear until four to seven weeks after exposure to EBV. An infected person can be contagious during this incubation time period and for as many as five months after the disappearance of symptoms. Also, the virus will be excreted in the saliva intermittently for the rest of their lives, although the individual will experience no symptoms. Contrary to popular belief, the EBV is not highly contagious. As a result, individuals living in a household or college dormitory with someone who has mononucleosis have a very small risk of being infected unless they have direct contact with the person's saliva.

Diagnosis

If symptoms associated with a cold persist longer than two weeks, mononucleosis is a possibility; however, a variety of other conditions can produce similar symptoms. If mononucleosis is suspected, a physician will

KEY TERMS

Antibody—A special protein made by the body's immune system as a defense against foreign material (bacteria, viruses, etc.) that enters the body. It is uniquely designed to attack and neutralize the specific antigen that triggered the immune response.

Herpes virus—A family of viruses including herpes simplex types 1 and 2, and herpes zoster (also called varicella zoster). Herpes viruses cause several infections, all characterized by blisters and ulcers, including chickenpox, shingles, genital herpes, and cold sores or fever blisters.

Reye's syndrome—A serious, life-threatening illness in children, usually developing after a bout of flu or chickenpox, and often associated with the use of aspirin. Symptoms include uncontrollable vomiting, often with lethargy, memory loss, disorientation, or delirium. Swelling of the brain may cause seizures, coma, and in severe cases, death.

typically conduct a physical examination, including a "Monospot" antibody blood test that can indicate the presence of proteins or antibodies produced in response to infection with the EBV. These antibodies may not be detectable, however, until the second or third weeks of the illness. Occasionally, when this test is inconclusive, other blood tests may be conducted.

Treatment

The most effective treatment for infectious mononucleosis is rest and a gradual return to regular activities. Individuals with mild cases may not require bed rest but should limit their activities. Any strenuous activity, athletic endeavors, or heavy lifting should be avoided until the symptoms completely subside, since excessive activity may cause the spleen to rupture.

The sore throat and **dehydration** that usually accompany mononucleosis may be relieved by drinking water and fruit juices. Gargling salt water or taking throat lozenges may also relieve discomfort. In addition, taking over-the-counter medications, such as **acetaminophen** or ibuprofen, may relieve symptoms, but aspirin should be avoided because mononucleosis has been associated with **Reye's syndrome**, a serious illness aggravated by aspirin.

While **antibiotics** do not affect EBV, the sore throat accompanying mononucleosis can be complicated by a streptococcal infection, which can be treated with antibiotics. Cortisone anti-inflammatory medications are also occasionally prescribed for the treatment of severely swollen tonsils or throat tissues.

Prognosis

While the severity and length of illness varies, most people diagnosed with mononucleosis are able to return to their normal daily routines within two to three weeks, particularly if they rest during this time period. It may take two to three months before a person's usual energy levels return. One of the most common problems in treating mononucleosis, particularly in teenagers, is that people return to their usual activities too quickly and then experience a relapse of symptoms. Once the disease has completely run its course, the person cannot be reinfected.

Prevention

Although there is no way to avoid becoming infected with EBV, paying general attention to good hygiene and avoiding sharing beverage glasses or having close contact with people who have mononucleosis or cold symptoms can help prevent infection.

Parental concerns

The main concern for parents of children with mononucleosis is to keep the child resting until he or she fully recovers from the illness. Parents should also be aware of the symptoms of more serious complications of the liver and spleen, and should seek medical attention for a child who complains of severe abdominal pain, light-headedness, rapid heartbeat, or difficulty breathing.

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National Institute of Allergy and Infectious Disease. Building 31, Room 7A-50, 31 Center Drive MSC 2520, Bethesda,

MD 20892–2520. Web site: <www.niaid.nih.gov/default.htm>.

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Influenza

Definition

Usually referred to as the flu or grippe, influenza is a highly infectious respiratory disease. The disease is caused by certain strains of the influenza virus. When the virus is inhaled, it attacks cells in the upper respiratory tract, causing typical flu symptoms such as fatigue, **fever** and chills, a hacking **cough**, and body aches. Influenza victims are also susceptible to potentially life-threatening secondary infections. Although the stomach or intestinal "flu" is commonly blamed for stomach upsets and **diarrhea**, the influenza virus rarely causes gastrointestinal symptoms. Such symptoms are most likely due to other organisms such as rotavirus, *Salmonella*, *Shigella*, or *Escherichia coli*.

Description

The flu is considerably more debilitating than the **common cold**. Influenza outbreaks occur suddenly, and infection spreads rapidly. In the 1918–19 Spanish flu pandemic, the death toll reached a staggering 20 to 40 million worldwide. Approximately 500,000 of these fatalities occurred in the United States.

Influenza outbreaks occur on a regular basis. The most serious outbreaks are pandemics, which affect millions of people worldwide and last for several months. The 1918–19 influenza outbreak serves as the primary example of an influenza pandemic. Pandemics also occurred in 1957 and 1968 with the Asian flu and Hong Kong flu, respectively. The Asian flu was responsible for 70,000 deaths in the United States, while the Hong Kong flu killed 34,000.

Epidemics are widespread regional outbreaks that occur every two to three years and affect 5–10 percent of the population. The Russian flu in the winter of 1977 is an example of an epidemic. A regional epidemic is shorter lived than a pandemic, lasting only several weeks. Finally, there are smaller outbreaks each winter that are confined to specific locales.

The earliest existing descriptions of influenza were written nearly 2,500 years ago by the ancient Greek physician Hippocrates. Historically, influenza was ascribed to a number of different agents, including "bad air" and several different bacteria. In fact, its name comes from the Italian word for "influence," because people in eighteenth-century Europe thought that the disease was caused by the influence of bad weather. It was not until 1933 that the causative agent was identified as a virus.

There are three types of influenza viruses, identified as A, B, and C. Influenza A can infect a range of animal species, including humans, pigs, horses, and birds, but only humans are infected by types B and C. Influenza A is responsible for most flu cases, while infection with types B and C virus are less common and cause a milder illness.

Demographics

The annual death toll attributable to influenza and its complications averages 20,000 in the United States alone. In the United States, 90 percent of all deaths from influenza occur among persons older than 65. Flu-related deaths have increased substantially in the United States since the 1970s, largely because of the aging of the American population. In addition, elderly persons are vulnerable because they are often reluctant to be vaccinated against flu.

Hospitalization due to complications of influenza are common in children. Among children with chronic illnesses, about 500 children per every 100,000 between the ages of birth and age four are hospitalized annually due to influenza, while about 100 children per 100,000 without chronic illnesses are hospitalized annually. Among those with underlying high-risk conditions, infants younger than six months have the highest hospitalization rates (approximately 10–40 per 100,000 population).

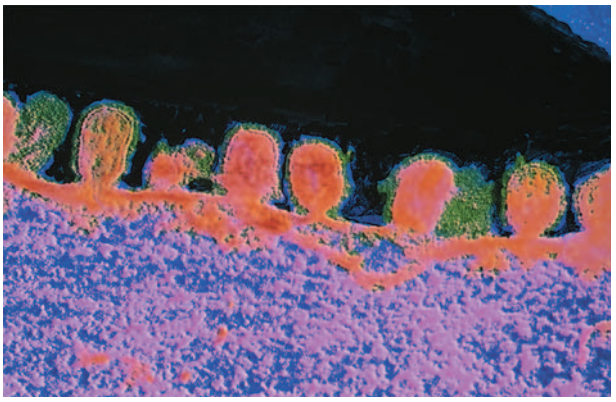
Causes and symptoms

Approximately one to four days after infection with the influenza virus, the victim is hit with an array of symptoms. "Hit" is an appropriate term, because symptoms are sudden, harsh, and unmistakable. Typical influenza symptoms include the abrupt onset of a **headache**, dry cough, and chills, rapidly followed by overall achiness and a fever that may run as high as 104°F (40°C). As the fever subsides, nasal congestion and a **sore throat** become noticeable. Flu victims feel extremely tired and weak and may not return to their normal energy levels for several days or even a couple of weeks.

Influenza complications usually arise from bacterial infections of the lower respiratory tract. Signs of a

secondary respiratory infection often appear just as the victim seems to be recovering. These signs include high fever, intense chills, chest pains associated with breathing, and a productive cough with thick yellowish green sputum. If these symptoms appear, medical treatment is necessary. Other secondary infections, such as sinus or ear infections may also require medical intervention. Children with heart and lung problems, as well as other chronic diseases, are at higher risk for complications from influenza.

With children and teenagers, it is advisable to be alert for symptoms of **Reye's syndrome**, a rare, but serious complication. Symptoms of Reye's syndrome are **nausea and vomiting**, and more seriously, neurological problems such as confusion or delirium. The syndrome has been associated with the use of aspirin to relieve flu symptoms.



A transmission electron microscopy (TEM) image of influenza viruses budding from the surface of an infected cell. *(CNRI/Science Photo Library, National Audubon Society Collection/Photo Researchers, Inc.)*

Diagnosis

Although there are specific viral culture tests available to identify the flu virus strain from respiratory samples, results can take several days. Therefore, doctors typically rely on a set of symptoms and the presence of influenza in the community for diagnosis. Specific tests are useful to determine the type of flu in the community, but they do little for individual treatment. Doctors may administer tests, such as throat cultures, to identify secondary infections.

Several rapid (30-minute) diagnostic tests for flu have become commercially available. These tests appear to be especially useful in diagnosing flu in children, allowing doctors to make more accurate treatment decisions in less time.

Treatment

Essentially, a bout of influenza must be allowed to run its course. Symptoms can be relieved with bed rest and by keeping well hydrated. A steam vaporizer may make breathing easier, and **pain** relievers can mask the aches and pain. Food may not seem very appetizing, but an effort should be made to consume nourishing food. Recovery should not be pushed. Returning to normal activities too quickly invites a possible relapse or complications.

Drugs

Since influenza is a viral infection, **antibiotics** are useless in treating it. However, antibiotics are frequently used to treat secondary infections.

Over-the-counter medications are used to treat flu symptoms, but it is not necessary to purchase a medication marketed specifically for flu symptoms. Any medication that is designed to relieve symptoms, such as pain and coughing, will provide some relief. Medications containing alcohol, however, should be avoided because of the dehydrating effects of alcohol. The best medicine for symptoms is simply an analgesic, such as **acetaminophen** or naproxen. (Without a doctor's approval, aspirin is generally not recommended for people under 18 owing to its association with Reye's syndrome, a rare aspirin-associated complication seen in children recovering from the flu. To be on the safe side, children should receive acetaminophen or ibuprofen to treat their symptoms.)

As of 2004, there were a number of **antiviral drugs** marketed for treating influenza. To be effective, treatment should begin no later than two days after symptoms appear. These medications are useful for decreasing the severity and duration of symptoms. Antivirals may be useful in treating patients who have weakened immune systems or who are at risk for developing serious complications. They include amantadine (Symmetrel, Symadine) and rimantadine (Flumandine), which work against Type A influenza, and zanamavir (Relenza) and oseltamavir phosphate (Tamiflu), which work against both Types A and B influenza. Amantadine and rimantadine can cause side effects such as nervousness, **anxiety**, lightheadedness, and **nausea**. Severe side effects include seizures, delirium, and hallucination, but are rare and are nearly always limited to people who have kidney problems, seizure disorders, or psychiatric disorders. Zanamavir and oseltamavir phosphate can cause **dizziness**, jitters, and insomnia.

KEY TERMS

Common cold—A mild illness caused by upper respiratory viruses. Usual symptoms include nasal congestion, coughing, sneezing, throat irritation, and a low-grade fever.

Epidemic—Refers to a situation in which a particular disease rapidly spreads among many people in the same geographical region in a relatively short period of time.

Guillain-Barré syndrome—Progressive and usually reversible paralysis or weakness of multiple muscles usually starting in the lower extremities and often ascending to the muscles involved in respiration. The syndrome is due to inflammation and loss of the myelin covering of the nerve fibers, often associated with an acute infection. Also called acute idiopathic polyneuritis.

Pandemic—A disease that occurs throughout a regional group, the population of a country, or the world.

Prognosis

Following proper treatment guidelines, healthy people under the age of 65 usually suffer no long-term consequences associated with flu infection. The elderly and the chronically ill are at greater risk for secondary infection and other complications, but they can also enjoy a complete recovery.

Most people recover fully from an influenza infection, but it should not be viewed complacently. Influenza is a serious disease, and approximately one in 1,000 cases proves fatal.

Prevention

The Centers for Disease Control and Prevention recommends that people get an influenza vaccine injection each year before flu season starts. In the United States, flu season typically runs from late December to early March. Vaccines should be received two to six weeks prior to the onset of flu season to allow the body enough time to establish immunity. Adults only need one dose of the yearly vaccine, but children under nine years of age who have not previously been immunized should receive two doses with a month between each dose.

Each season's **flu vaccine** contains three virus strains that are the most likely to be encountered in the coming flu season. When there is a good match between the anticipated flu strains and the strains used in the vaccine, the vaccine is 70–90 percent effective in people under 65. Because immune response diminishes somewhat with age, people over 65 may not receive the same level of protection from the vaccine, but even if they do contract the flu, the vaccine diminishes the severity and helps prevent complications.

The virus strains used to make the vaccine are inactivated and will not cause the flu. In the second half of the twentieth century, flu symptoms were associated with vaccine preparations that were not as highly purified as modern vaccines, not to the virus itself. In 1976, there was a slightly increased risk of developing Guillain-Barré syndrome, a very rare disorder, associated with the swine flu vaccine. This association occurred only with the 1976 swine flu vaccine preparation and as of 2004 had not recurred.

Serious side effects with modern vaccines are extremely unusual. Some people experience a slight soreness at the point of injection, which resolves within a day or two. People who have never been exposed to influenza, particularly children, may experience one to two days of a slight fever, tiredness, and muscle aches. These symptoms start within six to 12 hours after the **vaccination**.

It should be noted that certain people should not receive an influenza vaccine. Infants six months and younger have immature immune systems and will not benefit from the vaccine. Since the vaccines are prepared using hen eggs, people who have severe **allergies** to eggs or other vaccine components should not receive the influenza vaccine. As an alternative, they may receive a course of amantadine or rimantadine, which are also used as a protective measure against influenza. Other people who might receive these drugs are those that have been immunized after the flu season has started or who are immunocompromised, such as people with advanced HIV disease. Amantadine and rimantadine are 70–90 percent effective in preventing influenza.

Certain groups are strongly advised to be vaccinated because they are at increased risk for influenza-related complications. These groups are:

- children under age two
- all people 65 years and older
- residents of nursing homes and chronic-care facilities, regardless of age
- adults and children who have chronic heart or lung problems, such as asthma
- adults and children who have chronic metabolic diseases, such as diabetes and renal dysfunction, as well as severe anemia or inherited hemoglobin disorders

- children and teenagers who are on long-term aspirin therapy
- women who will be in their second or third trimester during flu season or women who are nursing
- anyone who is immunocompromised, including HIV-infected persons, **cancer** patients, organ transplant recipients, and patients receiving steroids, and those receiving **chemotherapy** or radiation therapy
- anyone in contact with the above groups, such as teachers, care givers, healthcare personnel, and **family** members
- travelers to foreign countries

A person need not be in one of the at-risk categories listed above, however, to receive a flu vaccination. Anyone who wants to forego the discomfort and inconvenience of an influenza attack may receive the vaccine.

Parental concerns

Parents should make sure that their children who fall into any of the risk categories should be vaccinated against the flu. Pregnant women in the second or third trimesters should also be vaccinated. Flu vaccines are available through pediatricians or local public health departments. Parents should also make sure kids follow good hygiene practices, including regular hand washing, and covering the mouth when sneezing or coughing. Children may acquire secondary infections, such as ear infections or sinus infections, so parents should call the pediatrician if a child develops a high fever, sudden pain in the ears or sinuses, or develops a productive cough with thick yellow-green phlegm.

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Influenza vaccine *see* **Flu vaccine**
Inner ear infection *see* **Labrynthitis**
Insect bites *see* **Bites and stings**

Insect sting allergy

Definition

Many children experience insect **stings** every year. For most of them, these stings only cause mild **pain** and discomfort lasting for just a period of hours. Symptoms might include swelling, **itching**, and redness at the sting site. However, some children are allergic to insect stings. When they are stung by an insect to which they are allergic, their bodies produce an antibody called immunoglobulin E (IgE), which reacts with the insect venom and triggers the release of various chemicals, including histamine, that cause the allergic reaction. Stings may be life threatening for a small number of children. These severe allergic reactions may develop quickly and can involve several body organs. This type of reaction is called **anaphylaxis** and can be fatal.

Description

The majority of insect stings in the United States are from wasps, hornets, bees, yellow jackets, and fire ants. The class of insects capable of injecting venom into a person is called Hymenoptera. With the exception of fire ants, all of these insects are found throughout the United States. Fire ants are found primarily in the southeastern region of the country but have also been noted in some western states.

Insect venom is made up of proteins and other substances that usually only cause itching, pain, and swelling in those who are stung. This local reaction is usually confined to the site of the sting. Sometimes the redness and swelling may extend from the sting site and cover a larger area of the body. These large, local, non-allergic reactions can persist for days. Occasionally the site may become infected, requiring antibiotic treatment. Although most local reactions are not serious, if they are near the face or neck, swelling can block the airway and cause serious problems.

Some children may have a venom allergy, and more serious reactions can result if they are stung. It is important to note that allergic reactions to stings normally do not occur after the initial sting. A reaction may take place after two or more stings that have happened over an extended period of time. Therefore, it is essential to be aware of the possibility for allergic symptoms in children, even if they have been stung previously and had no reaction.

Demographics

It is estimated that over 2 million Americans are allergic to stinging insects. Up to one million hospital emergency room visits occur annually because of insect stings. Between 50 and 150 Americans die each year as a result of insect sting-induced anaphylaxis. It is possible that this number may be markedly underestimated. Bee, wasp, and insect stings cause more deaths in the United States than any other kind of injection of venom. Most deaths occur in people 35 to 45 years of age. About one out of 100 children has a systemic allergic reaction from the sting of an insect. Fifty percent of deaths occur within 30 minutes of the sting.

Causes and symptoms

Allergic reactions to insect stings result from an overreaction of a child's immune system to the venom injected by the insect. After the first sting, the child's body produces an allergic substance called immunoglobulin E (IgE) antibody, which reacts with the insect venom. If the child is stung again by the same type of insect or by one from a similar species, the insect venom will interact with the IgE antibody produced in response to the previous sting. This in turn causes the release of histamine and several other chemicals that cause allergic symptoms.

The sting of an insect may only cause a local response, where pain, redness, itching, and swelling are confined to the site of the sting. This type of reaction is considered normal. The normal reaction to fire ant stings is different. Clear blisters usually form within several hours then become cloudy within 24 hours. (The reaction usually presents in a ring or cluster, since a fire ant pivots and repeatedly stings. Also, fire ants travel in groups and a child may receive multiple stings from many ants.)

Larger allergic reactions often affect almost the entire arm, leg, foot, hand, or other area of the sting. Swelling occurs, and may last as long as seven to 10 days. The child may also experience a low-grade **fever**, fatigue, and **nausea**.

Some children experience a more severe allergic reaction. For a small percentage of these individuals, the stings may be life threatening. Severe allergic reactions can involve multiple body organs and may progress rapidly. This reaction is called anaphylaxis. Anaphylaxis is considered a medical emergency and may be fatal. The symptoms of anaphylaxis include the following:

- wheezing
- difficulty breathing
- itching and **hives** over large areas of the body
- swelling in the tongue or throat
- **dizziness**, chest pain, racing heartbeat, or fainting
- stomach cramps, nausea, or **diarrhea**

In severe cases, a rapid fall in blood pressure may result in shock and loss of consciousness. (This is less common in children than adults.) The progression of these symptoms may only take a few minutes.

When to call the doctor

For the majority of insect stings, home care is all that is necessary. However, in many cases medical attention is warranted. If any of the following are true, parents should seek professional assistance promptly.

- Symptoms progress beyond the site of the sting.
- Swelling becomes extensive and painful.
- The sting is located on the head or neck area.
- The child has had severe large reactions in the past.
- There is evidence of infection, such as increased pain, redness, warmth, and swelling at the sting site.

If a child develops hives, has difficulty breathing or swallowing, swelling of the lips or face, fainting, or dizziness, he or she should be transported to an emergency department immediately.

Diagnosis

An allergy to insect stings is determined by the doctor, who takes a thorough history from the patient and his or her parents. The history will usually show that the child has been stung previously. The doctor will also note the presence of the various symptoms common to insect sting allergic reactions. Skin testing may be performed by an allergist to determine the specific sensitivities the child may have.

Treatment

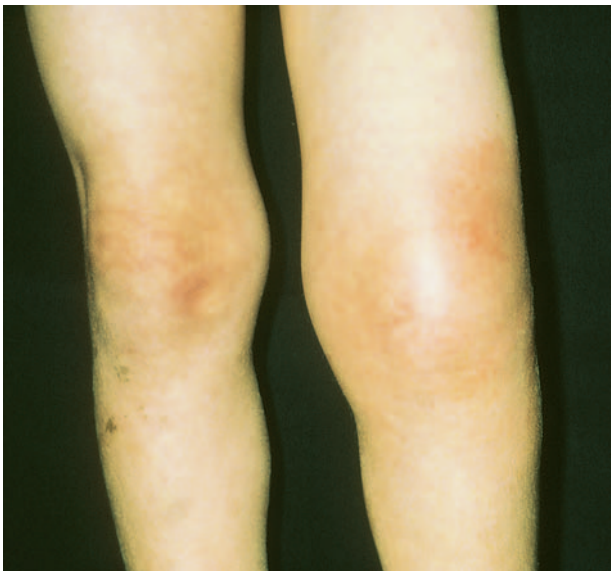
If a child has been stung by an insect that has left its stinger, it should be removed by flicking the fingers at it. Avoid squeezing the venom sac, as this can force more venom into the skin. If fire ants have stung the child, they should be carefully brushed off to prevent repeated stings.

Local treatment is normally all that is needed for small skin reactions. The affected arm or leg should be elevated and an ice pack applied to the area to reduce swelling and pain. Over-the-counter products can also be used to decrease the pain and itching. These include the following:

- products with a numbing effect, including topical anesthetics like benzocaine and phenol
- hydrocortisone products, which may decrease inflammation and swelling
- skin protectants, such as calamine lotion and zinc oxide, which have astringent, cooling, and antibacterial affects
- diphenhydramine, an antihistamine, which will help to control itching, and will counter some of the substances produced as part of the reaction
- ibuprofen or **acetaminophen** for pain relief

It is important to keep the area of the sting clean. The site should be gently cleansed with mild soap and water. Avoid breaking any blisters, as this can increase the chances of a secondary infection.

Any symptoms that progress beyond the local area of the sting require immediate attention. Allergic reactions to insect stings are considered medical emergencies. The physician will treat the child with epinephrine (adrenaline), which is usually given as an injection into the arm. An antihistamine such as diphenhydramine is usually given by mouth or injection to diminish the histamine reaction. Gluococorticoids, such as prednisone or methylprednisolone, are often given to decrease any swelling and to suppress the immune response. The physician may write prescriptions for both **antihistamines** and steroids to take after the child leaves the hospital.



Swelling in the patient's left knee from an allergic reaction to a wasp sting. *(© Dr. P. Marazzi/Photo Researchers, Inc.)*

After a child has experienced a severe allergic reaction and received emergency treatment, the doctor may write a prescription for a self-injecting epinephrine device. This device should be carried by the parent or child at all times, especially when the child is out of reach of medical care, such as on an airplane or in the woods. However, sometimes epinephrine is not enough, and other treatment may be needed. Whenever children with a known severe insect sting allergy are stung, they should receive prompt medical attention, even if they have received an epinephrine injection.

Prognosis

Prompt treatment normally prevents immediate complications, but a delay in the treatment of a severe allergic reaction can result in rapid deterioration and even death. The long-term prognosis is usually good, with the rare exception of possible local infections. If a child develops anaphylaxis after an insect sting, that child is at an increased risk of developing anaphylaxis if stung again.

Prevention

Obviously the best way to avoid an allergic reaction from an insect sting is to avoid getting stung in the first

KEY TERMS

Allergy shots—Injections given by an allergy specialist to desensitize an allergic person. Also known as immunotherapy treatment.

Anaphylaxis—Also called anaphylactic shock; a severe allergic reaction characterized by airway constriction, tissue swelling, and lowered blood pressure.

place. One way to do this is to be able to identify stinging insects and where they live.

- Honeybees have a fuzzy, rounded body with dark brown coloring and yellow markings. After stinging, the honeybee normally leaves its barbed stinger in its victim, and then the bee dies. Honeybees are usually not aggressive and will only sting if provoked. However, the so-called "killer bees," or Africanized honeybees, are far more aggressive and may sting in swarms. Wild honeybees live in honeycombs or colonies in cavities of buildings or in hollow trees. Africanized honeybees may nest in old tires or holes in the ground, in house frames, or between fence posts.
- Yellow jackets are black with yellow markings. Their nests have a papier-mâché appearance and are usually located underground. However, they can also be found in woodpiles, in the walls of frame buildings, or in masonry cracks.
- Paper wasps have slender, elongated bodies and are black, red, or brown with yellow markings. Their nests are also made of a paper-like substance that opens downward, in a circular comb of cells. Their nests are often located behind shutters, in shrubs or woodpiles, or under eaves.
- Fire ants are reddish brown to black stinging insects. They build nests of dirt in the ground that may be quite tall. Fire ants attack with little warning.

A variety of precautionary measures will decrease the chances of a child getting stung.

- Avoid walking barefoot on lawns and wear closed-toe shoes. The majority of honeybee stings occur on the bottom of the foot when a child steps on the bee.
- Hire an exterminator to destroy nests and hives around the home.
- The smell of food attracts insects, so be careful when eating, drinking, or cooking outdoors. Keep food covered.
- Remain calm and quiet if flying insects are noted and move away slowly. Do not attempt to swat them.
- Avoid using highly scented perfumes, colognes, or hair sprays.
- Avoid wearing brightly colored clothing.
- Do not wear loose fitting garments that can trap insects between the material and skin.
- Keep the areas around trash containers clean and at some distance away from where children are playing.

Allergy shots

Allergy shots for insect stings, also known as venom immunotherapy, can be an effective treatment for children who experience a severe reaction to insect stings. Any child who has had a significant reaction to an insect sting should be evaluated by an allergy specialist. Not all children who have had a reaction will get allergy shots, but many should. It was once believed that most children would outgrow insect sting **allergies** and that allergy shots were not needed. However, as of 2004, it is known that about one in five will remain allergic into adulthood. Because of this pattern, it is recommended that immunotherapy should be used for the approximately 40 percent of children who experience moderate-to-severe systemic reactions to insect stings.

Venom immunotherapy is a highly effective **vaccination** program that actually prevents future sting reactions in most patients who receive them. The child is initially tested to determine their individual sensitivities. The treatment normally involves twice weekly injections of venom in dosages that are gradually increased over about 10 to 20 weeks. At this point, a maintenance dosage is administered about every one to two months. Allergy shots given in childhood can protect the child for 10 to 20 years.

Nutritional concerns

For children who have a known allergy to the venom of honeybees, parents need to use caution (and consult with a physician) before using any honeybee products.

Parental concerns

Parents should be aware of the potential risks of insect stings and should teach their children to take measures to avoid being stung. If their child does get stung, parents need to begin treatment immediately and watch the child closely for any signs of allergic reaction. If these do occur, parents should transport their child immediately to a hospital emergency department.

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Insomnia *see* **Sleep disorders**

Insulin shock *see* **Hypoglycemia**

Intelligence

Definition

Intelligence is an abstract concept whose definition continually evolves and often depends upon current social values as much as scientific ideas. Modern definitions refer to a variety of mental capabilities, including the ability to reason, plan, solve problems, think abstractly, comprehend complex ideas, learn quickly, and learn from experience, as well as the potential to do these things.

Description

Several theories about intelligence emerged in the twentieth century and with them debate about the nature of intelligence and whether it determined by hereditary factors, the environment, or both. As methods developed to assess intelligence, experts theorized about the measurability of intelligence, its accuracy, and the field known as psychometrics, a branch of psychology dealing with the measurement of mental traits, capacities, and processes. Publication in 1994 of *The Bell Curve: Intelligence and Class Structure in American Life* by Richard J. Herrnstein and Charles Murray stirred the controversy. Their findings pointed to links between social class, race, and intelligence quotient (IQ) scores, despite questions by many about the validity of IQ tests as a measurement of intelligence or a predictor of achievement and success.

Part of the problem regarding intelligence stems from the fact that nobody has adequately defined what intelligence really means. In everyday life, people have a general understanding that some people are "smart," but when they try to define "smart" precisely, they often have difficulty because a person can be gifted in one area and average or below in another. To explain this phenomenon, some psychologists have developed theories to include multiple components of intelligence.

Since about 1970, psychologists have expanded the notion of what constitutes intelligence. Newer definitions of intelligence encompass more diverse aspects of thought and reasoning. For example, American psychologist Robert Sternberg developed a three-part theory of intelligence which states that behaviors must be viewed within the context of a particular culture; that a person's experiences impact the expression of intelligence; and that certain cognitive processes control all intelligent behavior. When all these aspects of intelligence are viewed together, the importance of how people use their intelligence becomes more important than the question of "how much" intelligence a person has. Sternberg has suggested that some intelligence tests focus too much on what a person has already learned rather than on how well a person acquires new skills or knowledge.

Another multifaceted approach to intelligence is Howard Gardner's proposal that people have eight intelligences:

- Musical: Children with musical intelligence are always singing or tapping out a beat. They are aware of sounds others miss. Musical children are discriminating listeners.
- Linguistic: Children with linguistic intelligence excel at reading, writing, telling stories, and doing crossword or other word puzzles.
- Logical-Mathematical: Children with this type of intelligence are interested in patterns, categories, and

relationships. They are good at mathematic problems, science, strategy games, and experiments.

- Bodily-Kinesthetic: These children process knowledge through their senses. They usually excel at athletics and **sports**, dance, and crafts.
- Spatial: These children think in images and pictures. They are generally good at mazes and jigsaw puzzles. They often spend lots of time drawing, building (with blocks, Legos, or erector sets), and daydreaming.
- Interpersonal: This type of intelligence fosters children who are leaders among their peers, are good communicators, and understand the feelings and motives of others.
- Intrapersonal: These children are shy, very aware of their own feelings, and are self-motivated.
- Naturalist: This type of intelligence allows children to distinguish among, classify, and use features of the environment. These children are likely to make good farmers, gardeners, botanists, geologists, florists, and archaeologists. Naturalist adolescents can often name and describe the features of every make of car around them.

Intelligence tests

There are many different types of intelligence tests, and they all do not measure the same abilities. Although the tests often have aspects that are related with each other, one should not expect that scores from one intelligence test that measures a single factor will be similar to scores on another intelligence test that measures a variety of factors. Many people are under the false assumption that intelligence tests measure a person's inborn or biological intelligence. Intelligence tests are based on an individual's interaction with the environment and never exclusively measure inborn intelligence. Intelligence tests have been associated with categorizing and stereotyping people. Additionally, knowledge of one's performance on an intelligence test may affect a person's aspirations and motivation to obtain goals. Intelligence tests can be culturally biased against certain groups.

STANFORD-BINET INTELLIGENCE SCALES Consisting of questions and short tasks arranged from easy to difficult, the Stanford-Binet measures a wide variety of verbal and nonverbal skills. Its fifteen tests are divided into the following four cognitive areas: verbal reasoning (vocabulary, comprehension, absurdities, verbal relations); quantitative reasoning (math, number series, equation building); abstract/visual reasoning (pattern analysis, matrices, paper folding and cutting, copying); and short-term memory (memory for sentences, digits, and objects, and bead memory). A formula is used to arrive at the intelligence quotient, or IQ. An IQ of 100 means that the child's chronological and mental ages match. Traditionally, IQ scores of 90–109 are considered average; scores below 70 indicate **mental retardation**. Gifted children achieve scores of 140 or above. Revised in 1986, the Stanford-Binet intelligence test can be used with children starting at age two. The test is widely used to assess **cognitive development** and often to determine placement in **special education** classes.

WECHSLER INTELLIGENCE SCALES The Wechsler intelligence scales are divided into two sections: verbal and nonverbal, with separate scores for each. Verbal intelligence, the component most often associated with academic success, implies the ability to think in abstract terms using either words or mathematical symbols. Performance intelligence suggests the ability to perceive relationships and fit separate parts together logically into a whole. The inclusion of the performance section in the Wechsler scales is especially helpful in assessing the cognitive ability of children with speech and **language disorders** or whose first language is not English. The test can be of particular value to school psychologists screening for specific learning disabilities because of the number of specific subtests that make up each section.

KAUFMAN ASSESSMENT BATTERY FOR CHILDREN The Kaufman **Assessment** Battery for Children (KABC) is an intelligence and achievement test for children ages 2.5–12.5 years. It consists of 16 subtests, not all of which are used for every age group. A distinctive feature of the KABC is that it defines intelligence as problem-solving ability rather than knowledge of facts, which it considers achievement. This distinction is evident in the test's division into two parts—intelligence and achievement—which are scored separately and together. The test's strong emphasis on memory and lesser attention to verbal expression are intended to offset cultural disparities between black and white children. In addition, the test may be given to non-native speakers in their first language and to hearing impaired children using American Sign Language.

Infancy

Babies were once thought to enter the world with minds that were blank slates that developed through a lifetime of experiences. It is as of the early 2000s known that newborns have brains as sophisticated as the most powerful supercomputers, pre-wired with a large capacity for learning and knowledge. In the first few months of life, a baby's brain develops at an amazing rate. At birth, infants have the senses of sight, sound, and touch. At about three or four months, infants begin to develop memory, and it expands quickly. Modern brain imaging

techniques have confirmed that children's intelligence is not just hereditary but is also affected greatly by environment. Babies' brains develop faster during their first year than at any other time. By three months, babies can follow moving objects with their eyes, are extremely interested in their surroundings, and can recognize familiar sounds, especially their parents' voices. At six months, infants begin to remember familiar objects, react to unfamiliar people or situations, and realize that objects are permanent. At seven months, babies can recognize their own name. Parents can help their infants develop their intelligence by talking and reading to them, playing with them, and encouraging them to **play** with a variety of age-appropriate **toys**.

Toddlerhood

Toddlers' lives generally revolve around experimenting with and exploring the environment around them. The primary source of learning for toddlers is their families. During their third year, toddlers should be able to sort and group similar objects by their appearance, shape, and function. They also start to understand how some things work, and their memory continues to improve rapidly. They are able to remember and seek out objects that are hidden or moved to a different location. Toddlers should be able to follow two-step instructions and understand contrasting ideas, such as large and small, inside and outside, opened and closed, and more and less. Toddlers also develop a basic understanding of time in relation to their regular activities, such as meals and bedtime.

Preschool

At age three, preschoolers can say short sentences, have a vocabulary of about 900 words, show great growth in communication, tell simple stories, use words as tools of thought, want to understand their environment, and answer questions. At age four, children can use complete sentences, have a 1,500-word vocabulary, frequently ask questions, and learn to generalize. They are highly imaginative, dramatic, and can draw recognizable simple objects. Preschoolers also should be able to understand basics concepts such as size, numbers, days of the week, and time. They should have an attention span of at least 20 minutes. Children this age are still learning the difference between reality and fantasy. Their curiosity about themselves and the world around them continues to increase.

School age

At age five, children should have a vocabulary of more than 2,000 words. They should be able to tell long stories, carry out directions well, read their own name, count to ten, ask the meaning of words, know colors, begin to know the difference between fact and fiction, and become interested in their surrounding environment, neighborhood, and community. Between the ages of seven and 12, children begin to reason logically and organize their thoughts coherently. However, generally, they can only think about actual physical objects; they cannot handle abstract reasoning. They also begin to lose their self-centered way of thinking. During this age range, children can master most types of conservation experiments and begin to understand that some things can be changed or undone. Early school-age children can coordinate two dimensions of an object simultaneously, arrange structures in sequence, change places or reverse the normal order of items in a series, and take something such as a story, incident, or play out of its usual setting or time and relocate it in another.

Gifted 11-year-old jazz pianist Matt Savage. Many gifted musicians have above-average intelligence. *(© Rick Friedman/Corbis.)*

Starting at about age 12, adolescents can formulate hypotheses and systematically test them to arrive at an answer to a problem. For example, they can formulate hypotheses based on the phrase "what if." They can think abstractly and understand the form or structure of a mathematical problem. Another characteristic of the later school-age years is the ability to reason contrary to fact. That is, if they are given a statement and asked to use it as the basis of an argument, they are capable of accomplishing the task. Until they reach the age of 15 or 16, adolescents are generally not capable of reasoning as an adult. High school-age adolescents continue to gain

KEY TERMS

Autism—A developmental disability that appears early in life, in which normal brain development is disrupted and social and communication skills are retarded, sometimes severely.

Down syndrome—A chromosomal disorder caused by an extra copy or a rearrangement of chromosome 21. Children with Down syndrome have varying degrees of mental retardation and may have heart defects.

Fragile X syndrome—A genetic condition related to the X chromosome that affects mental, physical, and sensory development. It is the most common form of inherited mental retardation.

Intelligence quotient (IQ)—A measure of somebody's intelligence, obtained through a series of aptitude tests concentrating on different aspects of intellectual functioning.

Kaufman Assessment Battery for Children—An intelligence and achievement test for children ages 2.5 to 12.5 years.

Psychometrics—The development, administration, and interpretation of tests to measure mental or psychological abilities. Psychometric tests convert an individual's psychological traits and attributes into a numerical estimation or evaluation.

Stanford-Binet intelligence scales—A device designed to measure somebody's intelligence, obtained through a series of aptitude tests concentrating on different aspects of intellectual functioning. An IQ score of 100 represents "average" intelligence.

Wechsler intelligence scales—A test that measures verbal and non-verbal intelligence.

cognitive and study skills. They can adapt language to different contexts, master abstract thinking, explore and prepare for future careers and roles, set goals based on feelings of personal needs and priorities, and are likely to reject goals set by others.

Common problems

Autism

Autism is a profound mental disorder marked by an inability to communicate and interact with others. The condition's characteristics include language abnormalities, restricted and repetitive interests, and the appearance of these characteristics in early childhood. As many as two-thirds of children with autistic symptoms are mentally deficient. However, individuals with autism can also be highly intelligent. Autistic individuals typically are limited in their ability to communicate nonverbally and verbally. About half of all autistic people never learn to speak. They are likely to fail in developing social relationships with peers, have limited ability to initiate conversation if they do learn how to talk, and show a need for routine and ritual. Various abnormalities in the autistic brain have been documented. These include variations in the frontal lobes of the brain that focus on control and planning and in the limbic system, a group of structures in the brain that are linked to emotion, behavior, smell, and other functions. Autistic individuals may suffer from a limited development of the limbic system. This would explain some of the difficulties faced by autistic individuals in processing information.

Mental retardation

Mental retardation usually refers to people with an IQ below 70. According to the American Psychiatric Association, a mentally retarded person is significantly limited in at least two of the following areas: self-care, communication, home living, social-interpersonal skills, self-direction, use of community resources, functional academic skills, work, leisure, health, and **safety**. Mental retardation affects roughly 1 percent of the U.S. population. According to the U.S. Department of Education, about 11 percent of school-aged children were enrolled in special education programs for students with mental retardation. There are four categories of mental retardation: mild, moderate, severe, and profound. There are many different causes of mental retardation, both biological and environmental. In about 5 percent of cases, retardation is transmitted genetically, usually through abnormalities in chromosomes, such as **Down syndrome** or **fragile X syndrome**. Children with Down syndrome have both mental and motor retardation. Most are severely retarded, with IQs between 20 and 49. Fragile X syndrome, in which a segment of the chromosome that determines gender is abnormal, primarily affects males.

Parental concerns

Autism symptoms begins in infancy, but typically the condition is diagnosed between the ages of two to five. The symptoms of mental retardation are usually evident by a child's first or second year. In the case of Down syndrome, which involves distinctive physical characteristics, a diagnosis can usually be made shortly after birth. Mentally retarded children lag behind their peers in developmental milestones such as sitting up,

smiling, walking, and talking. They often demonstrate lower than normal levels of interest in their environment and less responsiveness to others, and they are slower than other children in reacting to visual or auditory stimulation. By the time a child reaches the age of two or three, retardation can be determined using physical and **psychological tests**. Testing is important at this age if a child shows signs of possible retardation because alternate causes, such as impaired hearing, may be found and treated. There is no cure for autism or mental retardation.

When to call the doctor

Parents should consult a healthcare professional if their child's intellectual development appears to be significantly slower than their peers. Children suspected of having intelligence development problems should undergo a comprehensive evaluation to identify their difficulties as well as their strengths. Since no specialist has all the necessary skills, many professionals might be involved. General medical tests as well as tests in areas such as neurology (the nervous system), psychology, psychiatry, special education, hearing, speech and vision, and physical therapy may be needed. A pediatrician or a child and adolescent psychiatrist often coordinates these tests.

Parents should pay close attention to possible symptoms in their children. Autism is diagnosed by observing the child's behavior, **communication skills**, and social interactions. Medical tests should rule out other possible causes of autistic symptoms. Criteria that mental health experts use to diagnose autism include problems developing friendships, problems with make-believe or social play, endless repetition of words or phrases, difficulty in carrying on a conversation, obsessions with rituals or restricted patterns, and preoccupation with parts of objects. A diagnosis of mental retardation is made if an individual has an intellectual functioning level well below average and significant limitations in two or more adaptive skill areas. If mental retardation is suspected, a comprehensive physical examination and medical history should be done immediately to discover any organic cause of symptoms. If a neurological cause such as brain injury is suspected, the child may be referred to a neurologist or neuropsychologist for testing.

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Intermittent explosive disorder

Definition

Intermittent explosive disorder (IED) is a mental disturbance that is characterized by specific episodes of

violent and **aggressive behavior** that may involve harm to others or destruction of property. IED is discussed in the *Diagnostic and Statistical Manual of Mental Disorders*, fourth edition *(DSM-IV)* under the heading of "Impulse-Control Disorders Not Elsewhere Classified." As such, it is grouped together with kleptomania, pyromania, and pathological gambling.

A person must meet certain specific criteria to be diagnosed with IED:

- There must be several separate episodes of failure to restrain aggressive impulses that result in serious assaults against others or property destruction.
- The degree of aggression expressed must be out of proportion to any provocation or other stressor prior to the incidents.
- The behavior cannot be accounted for by another mental disorder, substance abuse, medication side effects, or such general medical conditions as epilepsy or head injuries.

Description

People diagnosed with IED sometimes describe strong impulses to act aggressively prior to the specific incidents reported to the doctor and/or the police. They may experience racing thoughts or a heightened energy level during the aggressive episode, with fatigue and depression developing shortly afterward. Some report various physical sensations, including tightness in the chest, **tingling** sensations, tremor, hearing echoes, or a feeling of pressure inside the head.

Many people diagnosed with IED appear to have general problems with anger or other impulsive behaviors between explosive episodes. Some are able to control aggressive impulses without acting on them while others act out in less destructive ways, such as screaming at someone rather than attacking them physically.

DSM-IV's classification of IED is not universally accepted. Many psychiatrists do not place intermittent explosive disorder into a separate clinical category but consider it a symptom of other psychiatric and mental disorders. In many cases individuals diagnosed with IED do in fact have a dual psychiatric diagnosis. IED is frequently associated with mood and **anxiety** disorders; substance abuse; eating disorders; and narcissistic, paranoid, and antisocial **personality disorders**.

One culturally specific psychiatric syndrome resembling IED is amok, which was first reported in Malaysia. As the English phrase "running amok" implies, the syndrome is characterized by sudden outbursts of indiscriminate aggression or murderous rage that are completely unprovoked or that are triggered by trivial slights.

Demographics

Although the editors of *DSM-IV* stated in 2000 that IED "is apparently rare," a group of researchers in Chicago reported in 2004 that it is more common than previously thought. They estimate that 1.4 million persons in the United States meet the criteria for IED, with a total of 10 million meeting the lifetime criteria for the disorder.

The symptoms of IED can appear at any time from late childhood through the early 20s, although the disorder is not usually diagnosed in children. The onset may be abrupt, without any warning in the form of a period of gradual change in the child or adolescent's behavior. IED appears to be more common in people from families with a history of **mood disorders** or substance abuse. The severity of the disorder appears to peak in people in their thirties and to decline rapidly in people over 50.

With regard to gender, 80 percent of individuals diagnosed with IED in the United States are adolescent and adult males; amok is a syndrome that almost always involves males. Women do experience IED, however, and have reported it as part of **premenstrual syndrome** (PMS).

Causes and symptoms

Causes

As with other impulse-control disorders, the cause of IED has not been determined. As of 2004, researchers disagreed as to whether it is learned behavior, the result of biochemical or neurological abnormalities, or a combination of factors. Some scientists have reported abnormally low levels of serotonin, a neurotransmitter that affects mood, in the cerebrospinal fluid of some anger-prone persons, but the relationship of this finding to IED is not clear. Similarly, some individuals diagnosed with IED have a medical history that includes migraine headaches, seizures, attention-deficit hyperactivity disorder, or developmental problems of various types, but it is not clear that these cause IED, as most persons with migraines, learning problems, or other neurological disorders do not develop IED.

Symptoms

Some psychiatrists who take a cognitive approach to mental disorders believe that IED results from rigid beliefs and a tendency to misinterpret other people's behavior in accordance with these beliefs. According to

Aaron Beck, a pioneer in the application of cognitive therapy to violence-prone individuals, most people diagnosed with IED believe that other people are basically hostile and untrustworthy, that physical force is the only way to obtain respect from others, and that life in general is a battlefield. Beck also identifies certain characteristic errors in thinking that go along with these beliefs:

- Personalizing: The person interprets others' behavior as directed specifically against him.
- Selective perception: The person notices only those features of situations or interactions that fit his negative view of the world rather than taking in all available information.
- Misinterpreting the motives of others: The person tends to see neutral or even friendly behavior as either malicious or manipulative.
- Denial: The person blames others for provoking his violence while denying or minimizing his own role in the fight or other outburst.

When to call the doctor

Parents should seek help for any older child or adolescent who has had more than one episode of irrationally angry or destructive behavior—if possible before the individual causes serious injury to others, has his education cut short, or gets into trouble with the law.

Diagnosis

The diagnosis of IED is basically a diagnosis of exclusion, which means that the doctor will eliminate such other possibilities as neurological disorders, mood or substance abuse disorders, anxiety syndromes, and personality disorders before deciding that the patient meets the *DSM-IV* criteria for IED. In addition to taking a history and performing a physical examination to rule out general medical conditions, the doctor may administer one or more psychiatric inventories or screening tests to determine whether the person meets the criteria for other mental disorders.

In some cases the doctor may order imaging studies or refer the person to a neurologist to rule out brain tumors, traumatic injuries of the nervous system, epilepsy, or similar physical conditions.

Treatment

Emergency room treatment

A person brought to a hospital emergency room by **family** members, police, or other emergency personnel after an explosive episode will be evaluated by a psychiatrist to see whether he can safely be released after any necessary medical treatment. If the patient appears to be a danger to self or others, he or she may be committed for further treatment. In terms of legal issues, a physician is required by law to notify the specific individuals as well as the police if the patient threatens to harm particular persons. In most states, the doctor is also required by law to report suspected abuse of children, the elderly, or other vulnerable family members.

The doctor will perform a thorough medical examination to determine whether the explosive outburst was related to substance abuse, withdrawal from drugs, head trauma, delirium, or other physical conditions. If the patient becomes violent inside the hospital, he or she may be placed in restraints or given a tranquilizer (usually either lorazepam [Ativan] or diazepam [Valium]), most often by injection. In addition to the physical examination, the doctor will obtain as detailed a history as possible from the family members or others who accompanied the patient.

Medications

Medications that have been shown to be beneficial in treating IED in nonemergency situations include lithium, carbamazepine (Tegretol), propranolol (Inderal), and such selective serotonin reuptake inhibitors as fluoxetine (Prozac) and sertraline (Zoloft). Adolescents diagnosed with IED have been reported to respond well to clozapine (Clozaril), a drug normally used to treat **schizophrenia** and other psychotic disorders.

Psychotherapy

Some persons with IED benefit from cognitive therapy in addition to medications, particularly if they are concerned about the impact of their disorder on their education, employment, or interpersonal relationships. Psychoanalytic approaches are not useful in treating IED.

Alternative treatment

Some patients diagnosed with IED have reported being helped by biofeedback, mindfulness meditation, and various forms of martial arts. Mind/body therapies appear to be helpful in gaining greater self-control, while martial arts workouts help to channel the person's physical energy or muscular tension.

Prognosis

The prognosis of IED depends on several factors that include the individual's socioeconomic status, the stability of the immediate family, the values of the

KEY TERMS

Amok—A culture-specific psychiatric syndrome first described among the Malays, in which adolescent or adult males are overcome by a sudden fit of murderous fury provoked by a perceived insult or slight. Some researchers consider amok to be a variant of intermittent explosive disorder.

Cognitive therapy—Psychological treatment aimed at changing a person's way of thinking in order to change his or her behavior and emotional state.

Delirium—Sudden confusion with a decreased or fluctuating level of consciousness.

Kleptomania—An impulse control disorder in which one steals objects that are of little or no value.

Neurotransmitter—A chemical messenger that transmits an impulse from one nerve cell to the next.

Pyromania—An impulse control disorder characterized by fire setting.

Serotonin—A widely distributed neurotransmitter that is found in blood platelets, the lining of the digestive tract, and the brain, and that works in combination with norepinephrine. It causes very powerful contractions of smooth muscle and is associated with mood, attention, emotions, and sleep. Low levels of serotonin are associated with depression.

surrounding neighborhood, and his or her motivation to change. One reason why the Chicago researchers think that IED is more common than previously thought is that most people who meet the criteria for the disorder do not seek help for the problems in their lives that result from it. The researchers found that although 88 percent of the 253 individuals with IED whom they studied were upset by the results of their explosive outbursts, only 13 percent had ever asked for treatment in dealing with it.

Prevention

Since the cause(s) of IED are not fully understood as of the early 2000s, preventive strategies should focus on treatment of young children who may be at risk for IED before they enter **adolescence**.

Parental concerns

An adolescent or young adult diagnosed with IED can cause severe disruption to family life in many different areas, ranging from the economic costs of property damage or accidents to emotional problems in other family members to serious legal penalties. It is important for the person's family to know that they do not have to tolerate violent behavior, destruction of property, harm to pets, or abuse of smaller or weaker family members. Depending on the specific situation and the pattern of previous explosive episodes, family members of adolescents or young adults may decide to leave the immediate situation, call the police or other emergency help, or take out a restraining order.

Another important dimension of IED is the damage done to the person's own life. One reason for seeking treatment for IED is to get help before the person establishes a record of school suspensions, arrests or other legal problems, hospitalizations for injuries sustained in fights or automobile accidents, or repeated firings from jobs. A history of such issues can lead to a self-fulfilling prophecy in which the person with IED continues to have episodes of uncontrolled aggression because of the belief that he or she cannot overcome the past.

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Janie F. Franz

Intersex states

Definition

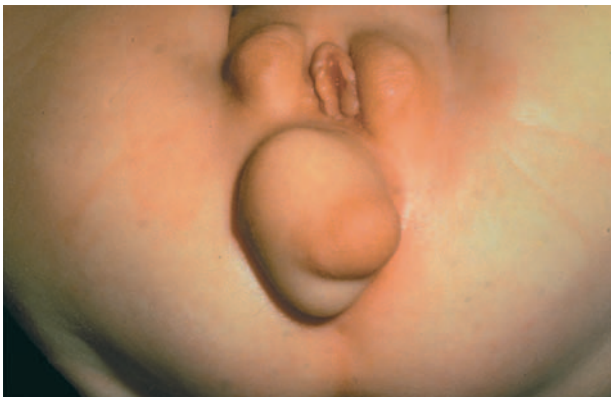
Intersex states are conditions where a newborn's sex organs (genitals) look unusual, making it impossible to identify the gender of the baby from its outward appearance.

Description

All developing babies start out with external sex organs that look female. If the baby is male, the internal sex organs mature and begin to produce the male hormone testosterone. If the hormones reach the tissues correctly, the external genitals that looked female change into the scrotum and penis. Sometimes, the genetic sex (as indicated by chromosomes) may not match the appearance of the external sex organs.

Persons with intersex states can be classified as a true hermaphrodite, a female pseudohermaphrodite, or a male pseudohermaphrodite. This is determined by examining the internal and external structures of the child.

True hermaphrodites are born with both ovaries and testicles. They also have mixed male and female external genitals. This condition is extremely rare.



This infant was born with female and male genitalia. *(Photograph by Mike Peres. Custom Medical Stock Photo, Inc.)*

A female pseudohermaphrodite is a genetic female. However, the external sex organs have been masculinized and look like a penis. This may occur if the mother takes the hormone progesterone to prevent a miscarriage, but more often it is caused by an overproduction of certain hormones.

A male pseudohermaphrodite is a genetic male. However, the external sex organs fail to develop normally. Intersex males may have testes and a female-like vulva, or a very small penis.

Demographics

About one in every 2,000 births results in a baby whose sex organs look unusual. True hermaphrodites are extremely rare.

Any abnormality in chromosomes or sex hormones, or in the unborn baby's response to the hormones, can lead to an intersex state in a newborn. Intersex states may also be caused by a condition called **congenital adrenal hyperplasia**, which occurs in about one out of every 5,000 newborns. This disease blocks the baby's metabolism and can cause a range of symptoms, including abnormal genitals.

Common problems

When doctors are uncertain about a newborn's sex, a specialist in infant hormonal problems is consulted as soon as possible. Ultrasound can locate a uterus behind the bladder and can determine if there is a cervix or uterine canal. Blood tests can check the levels of sex hormones in the baby's blood, and chromosome analysis (called karyotyping) can determine sex. Explorative

KEY TERMS

Chromosome—A microscopic thread-like structure found within each cell of the human body and consisting of a complex of proteins and DNA. Humans have 46 chromosomes arranged into 23 pairs. Chromosomes contain the genetic information necessary to direct the development and functioning of all cells and systems in the body. They pass on hereditary traits from parents to child (like eye color) and determine whether the child will be male or female.

surgery or a biopsy of reproductive tissue may be necessary. Only after thorough testing can a correct diagnosis and determination of gender be made.

Parental concerns

Treatment of intersex states is controversial. Traditional treatment assigns sex according to test results. Most doctors believe this gives the child the potential to identify with a sex. Treatment may then include reconstructive surgery followed by hormone therapy. Babies born with congenital adrenal hyperplasia can be treated with cortisone-type drugs and sometimes surgery.

Counseling should be given to the entire **family** of an intersex newborn. Families should explore all available medical and surgical options. Counseling should also be provided to the child when he or she is old enough.

Since the mid-1950s, doctors have typically assigned a sex to an intersex infant based on how easy reconstructive surgery would be. The American Academy of Pediatrics states that children with these types of genitals can be raised successfully as members of either sex, and recommends surgery within the first 15 months of life.

Some people are critical of this approach, including intersex adults who were operated on as children. The remolded genitals do not function sexually and can be the source of lifelong **pain**. They suggest that surgery be delayed until the individual can make informed choices about surgery and intervention.

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American Academy of Family Physicians. 11400 Tomahawk Creek Parkway, Leawood, KS 66211-2672. (913) 906-6000. E-mail: fp@aafp.org. Web site: <http://www.aafp.org/>.

American Academy of Pediatrics. 141 Northwest Point Boulevard, Elk Grove Village, IL 60007-1098. (847) 434-4000, Fax: (847) 434-8000. E-mail: kidsdoc@aap.org. Web site: <http://www.aap.org/default.htm>.

American Board of Obstetrics and Gynecology. 2915 Vine Street Suite 300, Dallas TX. 75204. (214) 871-1619. Fax: (214) 871-1943. E-mail: info@abog.org. Web site: <http://www.abog.org>.

American College of Obstetricians and Gynecologists. 409 12th St., S.W., PO Box 96920, Washington, D.C. 20090-6920. Web site: <http://www.acog.org>.

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Intestinal obstructions

Definition

Intestinal obstructions are a partial or complete blockage of the small or large intestine, resulting in failure of the contents of the intestine to pass through the bowel normally.

Description

Intestinal obstructions can occur in children as a result of congenital defects, with symptoms appearing any time between birth and adulthood. Abdominal **pain** and **vomiting** are the most frequent symptoms and a common cause of admission to emergency rooms. It is difficult for doctors to predict at birth which infants will suffer intestinal obstructions.

Intestinal obstructions can be mechanical or non-mechanical. Mechanical obstruction is the physical blockage of the intestine by a tumor, scar tissue, or another type of blockage that prevents intestinal contents from getting past the point of obstruction. One type of mechanical obstruction is caused by the bowel twisting on itself (volvulus) or telescoping into itself (intussusception). Mechanical obstruction can also result from hernias, fecal impaction, abnormal tissue growth, the presence of foreign bodies in the intestines, or inflammatory bowel disease (Crohn's disease). Non-mechanical obstruction occurs when the normal wavelike muscular contractions of the intestinal walls (peristalsis), which ordinarily move the waste products of digestion through the digestive tract, are disrupted (as in spastic **ileus**, dysmotility syndrome, or psuedo-obstruction) or stopped altogether as in paralysis of the bowel walls (paralytic ileus).

Mechanical obstruction in infants under one year of age can be caused by meconium ileus, volvulus, intussusception, and hernias. Meconium ileus is a disorder that occurs in newborns in which the meconium, the neonate's first fecal excretion after birth, is abnormally thick and stringy, rather than the collection of mucus and bile that is normally passed. The abnormal meconium blocks the intestines and must be removed with an enema or through surgery. This condition is due to a deficiency of the enzyme trypsin and other digestive enzymes produced in the pancreas. It can be an early clue that the infant may have **cystic fibrosis**. Intussusception commonly follows an infection that causes increased lymph node size in the gut, which acts as the point of folding for the intussusception.

Hirschsprung's disease (congenital megacolon), which may involve meconium ileus, is a motility disorder that is responsible for 25 percent of newborn non-mechanical intestinal obstructions, though symptoms may not develop until late in infancy or in childhood, delaying diagnosis. Children diagnosed with Hirschsprung's disease lack nerve cells (ganglia) in the large intestine, severely affecting the wavelike movements that propel material through the colon. In most affected infants, the first sign is failure to pass a stool (meconium) within 24 to 48 hours after birth. Between birth and age two, these children will likely develop other symptoms, such as chronic **constipation**, small watery stools, a distended abdomen, vomiting, poor appetite, slow weight gain, and **failure to thrive**. Most children will require surgery to remove the affected part of the colon. Surgery can be performed at age six months or as soon as diagnosed in an older infant or child. Symptoms can be removed in at least 90 percent of children born with Hirschsprung's disease. The disease is sometimes associated with other congenital conditions, such as **Down syndrome**.

Volvulus is the twisting of the small or large bowel around itself (malrotation). Volvulus of the large bowel is rare in infants and children; when it does occur it is usually in the sigmoid (sigmoid volvulus) in the lower colon. Duodenal volvulus occurs when the duodenum, the portion of small intestine that connects the stomach and jejunum, is twisted. Twisting of any portion of the intestines may cut off the supply of blood to a loop of bowel (strangulation), reducing the flow of oxygen to bowel tissue (ischemia) and leading to tissue death (gangrene). Strangulation occurs in about 25 percent of bowel obstruction cases and is a serious condition that can progress to gangrene within six to 12 hours.

Intussusception is a condition in which the bowel telescopes into itself like a radio antenna folding up. Intussusception is the most common cause of intestinal obstruction in children between the ages of three months and six years. Boys are twice as likely as girls to suffer intussusception.

Hernias are weaknesses in the abdominal wall that can trap a portion of intestine (incarceration) and cut off

the passage of food and waste through the digestive tract. In 1–5 percent of children, a **hernia** results when a feature of fetal anatomy in the inguinal area of the groin (processus vaginalis, the space through which the testis or ovaries descend) fails to close normally after birth. These inguinal hernias easily become incarcerated, trapping the bowel and causing obstruction. They are sometimes found on both sides (bilateral hernia) and they occur nine times more often in boys than girls. Parents may see a bulge in the groin area when an inguinal hernia is present. Incarceration occurs only rarely after eight years of age. In most cases, the incarcerated hernias are corrected manually rather than surgically by pushing the incarcerated bowel back up into the abdominal cavity.

Congenital adhesions or post-surgical adhesions can also cause intestinal obstruction in children. Adhesions are bands of fibrous tissue that can bind the loops of intestine to each other or to abdominal organs, narrowing the space between the intestinal walls or pulling sections of the intestines out of place, blocking the passage of food and waste. In adults, adhesions are most often caused by repeat surgery; children who have a history of abdominal surgery can also develop adhesions that can obstruct the intestines. It is not known precisely what causes the abnormal growth of fibrous tissue in congenital adhesions.

Demographics

Each year, one in 1,000 individuals of all ages are diagnosed with intestinal obstruction. Adhesions are responsible for 50–70 percent of cases. In children, 2.4 cases of intussusception are reported among 1,000 live births annually in the United States. Inguinal hernias occur in 1–5 percent of infants, with a male to female ratio of nine to one. Volvulus occurs in older children (mean age seven years) with a male to female ratio of 3.5 to one.

Causes and symptoms

The causes of small bowel obstruction in children are most often volvulus, intussusception, adhesions, or abdominal hernia, a weakness in the abdominal wall that traps a portion of intestine. The most frequent causes of large-bowel obstruction are tumors, volvulus, or small pouches that form on the intestinal wall (diverticula) that can fill with waste and expand to block the intestines. Motility disorders such as Hirschsprung's disease and psuedo-obstruction may cause blockages by retarding peristalsis, the intestinal muscle contractions that move food and waste.

Meconium ileus in newborns is caused by increased viscosity of waste products in the intestinal tract, and is sometimes secondary to cystic fibrosis. Its primary symptom will be failure of the infant to eliminate the meconium within the first two days of life.

One of the earliest signs of mechanical intestinal obstruction is abdominal pain or cramps that come and go in waves. Infants typically pull up their legs and cry in pain, then stop crying suddenly. They may behave normally for as long as 15–30 minutes between episodes, only to start crying again when the next cramp begins. The cramping results from the inability of the muscular contractions of the bowel to push the digested food past the obstruction. A classic symptom is passage of "current jelly stool" (i.e. blood) by infants after a crying fit during intussusception. Some children with intussusception may appear lethargic or have altered mental status, believed by physicians to be related to ischemia of the bowel and a decreased level of consciousness.

Vomiting is another typical symptom of intestinal obstruction. The speed of its onset is a clue to the location of the obstruction. Vomiting follows shortly after the pain if the obstruction is in the small intestine, but is delayed if it is in the large intestine. The vomited material may be green from bile or fecal in character. If the blockage is complete, the individual will not pass any gas or feces. If the blockage is only partial, however, **diarrhea** can occur. Initially there is little or no **fever**.

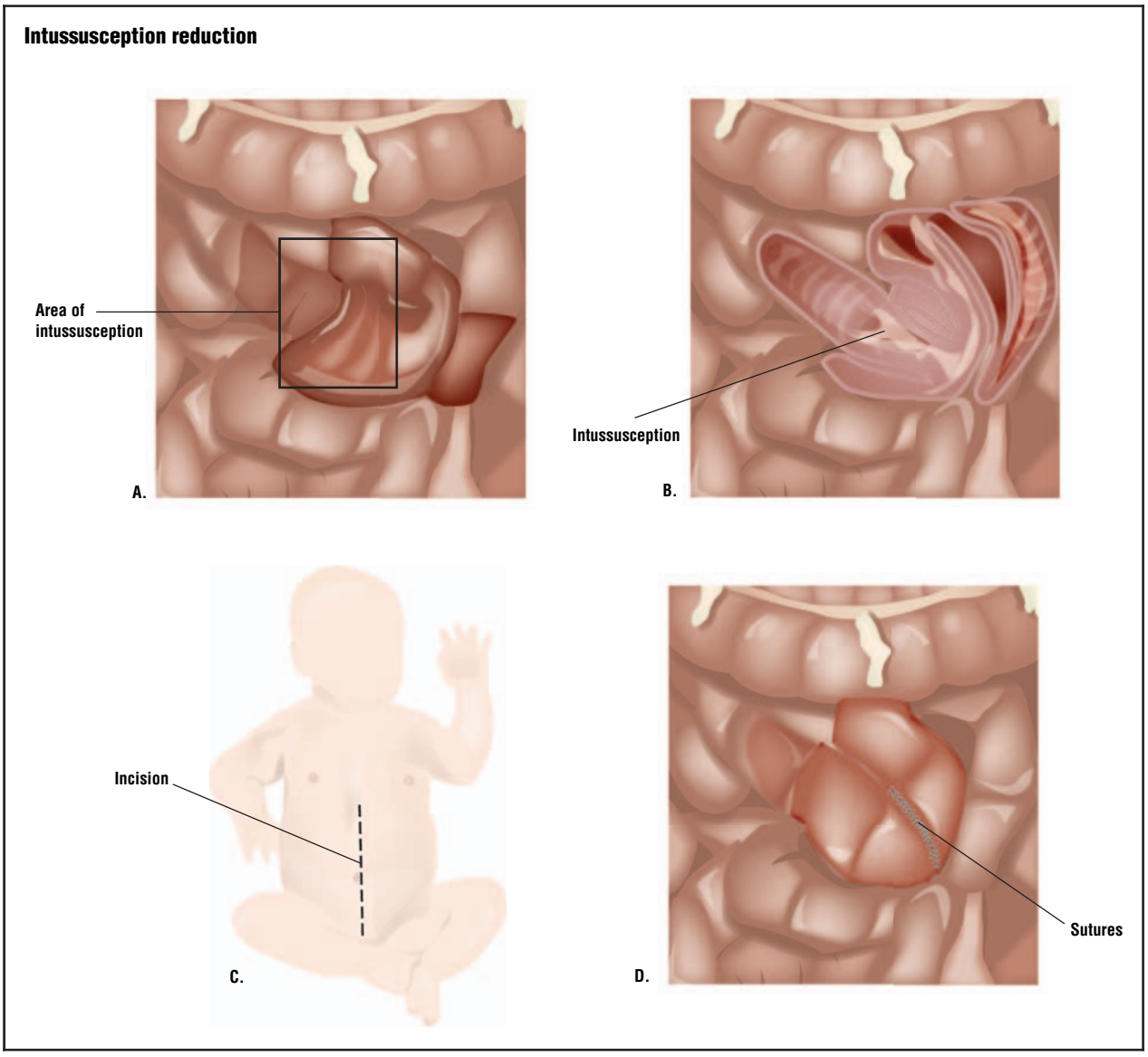
When the material in the bowel cannot move past the obstruction, the body reabsorbs large amounts of fluid and the abdomen becomes sore to the touch and swollen (distended). Persistent vomiting can result in **dehydration**. Fluid imbalances upset the balance of certain important chemicals (electrolytes) in the blood, which can cause complications such as irregular heartbeat and, without correction of the electrolyte imbalance, shock. Kidney failure is a serious complication that can occur as a result of severe dehydration and/or systemic infection from perforation of the bowel.

When to call the doctor

Medical attention is needed early in intestinal obstruction and should be sought as soon as symptoms suggest abdominal distress. Symptoms may begin with abdominal pain or cramping that may cause a toddler or older child to double over in pain. Infants will periodically cry in pain and pull their legs up to their chest. Fever may or may not be present. Vomiting may occur along with pain. If pain and crying occur every 15 or 30 minutes, it is critical to see the pediatrician or go to the emergency room so that early diagnosis can be made and treatment begun.

Diagnosis

If the doctor suspects intestinal obstruction based on the child's symptoms and the physical examination, imaging studies will be ordered that may include abdominal



Intussusception of the bowel results in the bowel telescoping onto itself (A and B). To repair it, an incision is made in the baby's abdomen to expose the bowel (C). If the surgeon cannot manipulate the bowel into a normal shape manually, the area of intussusception will be removed and remaining bowel sutured together (D). *(Illustration by GGS Information Services.)*

x rays, **computed tomography** (CT scan), or an ultrasound evaluation of the abdomen. Abdominal ultrasound is able to effectively visualize and diagnose most obstructions. The x ray images may be enhanced by giving the child a barium enema, a form of contrast or opaque media that allows more detail to be seen in x rays and MRI or CT scans. In a barium enema, barium sulfate is infused through the rectum and the intestinal area is scanned. With contrast enhancement, the exact location of the obstruction can be pinpointed in the scans or x ray film. Sometimes a lighted, flexible fiber optic instrument (sigmoidoscope) may be inserted rectally in conjunction with a barium enema to visualize the bowel. It may not possible to determine if an obstruction is simple or strangulated on scanning, and this will only be determined by performing abdominal surgery.

Diagnostic testing will include a complete blood count (CBC), electrolytes (sodium, potassium, chloride) and other blood chemistries, blood urea nitrogen (BUN), and urinalysis. Coagulation tests may be performed if the child requires surgery.

KEY TERMS

Bowel—The intestine; a tube-like structure that extends from the stomach to the anus. Some digestive processes are carried out in the bowel before food passes out of the body as waste.

Dysmotility—Abnormally slow or fast rhythmic movement of the stomach or intestine.

Electrolytes—Salts and minerals that produce electrically charged particles (ions) in body fluids. Common human electrolytes are sodium chloride, potassium, calcium, and sodium bicarbonate. Electrolytes control the fluid balance of the body and are important in muscle contraction, energy generation, and almost all major biochemical reactions in the body.

Gangrene—Decay or death of body tissue because the blood supply is cut off. Tissues that have died in this way must be surgically removed.

Ileus—An obstruction of the intestines usually caused by the absence of peristalsis.

Intussusception—The slipping or telescoping of one part of the intestine into the section next to it.

Ischemia—A decrease in the blood supply to an area of the body caused by obstruction or constriction of blood vessels.

Meconium—A greenish fecal material that forms the first bowel movement of an infant.

Motility—The movement or capacity for movement of an organism or body organ. Indigestion is sometimes caused by abnormal patterns in the motility of the stomach.

Peristalsis—Slow, rhythmic contractions of the muscles in a tubular organ, such as the intestines, that move the contents along.

Shock—A medical emergency in which the organs and tissues of the body are not receiving an adequate flow of blood. This deprives the organs and tissues of oxygen and allows the build-up of waste products. Shock can be caused by certain diseases, serious injury, or blood loss.

Sigmoid colon—The final portion of the large intestine that empties into the rectum.

Strangulated obstruction—An obstruction in which a loop of the intestine has its blood supply cut off.

Volvulus—A twisting of the intestine that causes an obstruction.

Treatment

Children with suspected intestinal obstruction will be hospitalized after the initial diagnostic evaluation. Treatment will likely begin immediately and proceed rapidly to avoid strangulation, which can be fatal. The first step in treatment is inserting a nasogastric tube to suction out the contents of the stomach and intestines. Intravenous fluids will be infused to prevent dehydration and to correct electrolyte imbalances that may have already occurred. Surgery can be avoided in some cases. With volvulus, for example, it may be possible to guide a rectal tube into the intestines to straighten the twisted bowels. In infants, a barium enema may reverse intussusception in 50–90 percent of cases. Another newer contrast agent, gastrografin, may be used; it is believed to have therapeutic properties as well as its ability to enhance scans. An air enema is sometimes used instead of a barium or gastrografin enema. This treatment successfully relieves partial obstruction in many infants. Children usually remain hospitalized for observation for two to three days after these procedures.

Surgical treatment will be necessary if other efforts are unsuccessful in correcting or removing the blockage. Generally, complete obstructions require surgery while partial obstructions do not. Strangulated obstructions require emergency surgery. The obstructed area is removed and part of the bowel is cut away (bowel resection). If the obstruction is caused by tumors, polyps, or scar tissue, they will be surgically removed. Hernias, if present, are repaired to correct the obstruction. **Antibiotics** may be given pre- or post-operatively to avoid the threat of infection at the site of the obstruction. Fluid replacement is given intravenously as needed.

Alternative treatment

Immediate surgery is often the only means of correcting intestinal obstruction. Alternative practitioners may recommend a high fiber diet to encourage proper stool formation; however, simple constipation is not the cause of intestinal obstruction.

Prognosis

Most intestinal obstructions can be corrected with prompt treatment and the affected child will recover without complications. Untreated intestinal obstructions can be fatal, however. The bowel either strangulates or perforates, causing massive infection. Recurrence is likely in as many as 80 percent of those in whom volvulus is treated medically rather than surgically. Recurrences in infants with intussusception are most likely to happen during the first

36 hours after the blockage has been cleared. The mortality rate for unsuccessfully treated infants is 1–2 percent.

Prevention

Most cases of intestinal obstruction are not preventable. Surgery to remove tumors or polyps in the intestines helps prevent recurrences, although adhesions can form after surgery, which can be another cause of obstruction.

Nutritional concerns

Preventing certain types of intestinal problems that may lead to intestinal obstruction may include making sure that the diet includes sufficient fiber to help encourage proper stool formation and regular elimination. High-fiber foods include whole grain breads and cereals; apples and other fresh fruits; dried fruits such as prunes; pumpkin and squash; fresh raw vegetables; beans and lentils; and nuts and seeds.

Parental concerns

Diagnosis of intestinal obstruction in a child is dependent on recognizing related symptoms. Parents may be concerned about preventing intestinal problems in their children and about missing the symptoms of possible intestinal obstruction. It is important to remember that a healthy diet with plenty of whole fruits, vegetables, and grains, and drinking a sufficient amount of water each day, will help keep the intestines healthy and elimination regular. Parents should be aware of the child's bowel habits and report constipation, diarrhea, abdominal pain, and vomiting to the pediatrician when it occurs. Diagnosed early, intestinal obstruction can be corrected without complications.

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ORGANIZATIONS

*American Association of Family Physicians.*11400 Tomahawk Creek Parkway, Leawood, KS 66211-2672. (800) 274-2237. Web site: <http://www.aafp.org>.

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Intrauterine growth retardation

Definition

The term intrauterine growth retardation (also known as intrauterine growth restriction)(IUGR) is generically defined as a fetus who is at or below the tenth percentile in weight for its gestational age. There are two factors necessary to define an IUGR fetus: first, the fetal weight is at or below the tenth percentile for gestational age and sex; second, there is a pathological process present that prevents expression of normal growth potential. If the baby is small given its in utero age, then it is said to be small for gestational age (SGA).

Description

There are standards or averages in weight for unborn babies according to their gestational age in weeks; however, using a fetal growth curve derived from one population and applying it to another can result in over- or underestimation of the true incidence of SGA. For example, the lowest mean birth weight has been noted in Africa (New Guinea Lumi's tribe: mean birth weight = 2,400 grams); whereas the largest mean birth weight has been noted in the Caribbean (Aguilla: mean birth weight = 3,880 g). A population of smaller individuals will have smaller babies, so the difference lies in the genetic growth potential.

The normal intrauterine growth pattern occurs in three stages. In the first stage, four to 20 weeks gestation, rapid cell division and multiplication (hyperplasia) occurs as the embryo grows into a fetus. In the second stage, 20–28 weeks gestation, cell division (hyperplasia) declines and the cells increase in size (hypertrophy). In stage three, 28–40 weeks, there is a rapid increase in cell size, rapid accumulation of fat, muscle, and connective tissue. Ninety-five percent of fetal weight gain occurs during the last 20 weeks of gestation. If the delicate process of development and weight gain is disturbed or interrupted, the baby can suffer from restricted growth.

IUGR is usually classified as symmetrical or asymmetrical. Growth inhibition during the first stage produces an undersized fetus with fewer cells, but normal cell size, causing symmetric IUGR. In symmetrical

IUGR weight, head and length are all below the tenth percentile and the baby's head and body are proportionately small. Conditions associated with symmetric IUGR include: genetic (constitutional, chromosomal and single gene defects, and deletion disorders and inborn errors of metabolism), congenital anomalies, intrauterine infections, and therapeutic irradiation. Substance abuse and cigarette **smoking**, depending on dose and timing, can cause either symmetrical or asymmetrical IUGR.

Growth inhibition during stage two and three causes a decrease of cell size and fetal weight with less effect on total cell number and fetal length and head circumference, causing asymmetric IUGR. Conditions associated with asymmetric IUGR include: uteroplacental insufficiency, which is usually caused by chronic **hypertension** or preeclampsia; chronic renal disease; cyanotic heart disease; hemoglobinopathies; placental infarcts; abruptio placenta; multiple gestation; velamentous insertion of the umbilical cord and circumvallate placenta; and high altitude. In asymmetrical IUGR, weight is below the tenth percentile, and head and length are preserved. The brain can weigh five or six times more than the liver, whereas in a normal infant, the brain weighs about three times more than the liver.

Intrauterine growth retardation (IUGR)

Conditions associated with IUGR

Maternal history	Alcohol use Cocaine use Smoking Malnutrition Use of prescription drugs warfarin (Coumadin, Panwarfarin) and phenytoin (Dilantin) Prior history of IUGR pregnancy Residing at altitude over 5,000 ft (1,500 m)
Medical conditions (of mother)	Chronic hypertension Preeclampia early in gestation Diabetes mellitus Systemic lupus erythematosus Chronic kidney disease Inflammatory bowel disease Severe lung disease
Infectious diseases	Syphilis Cytomegalovirus Toxoplasmosis Rubella Hepatitis B Herpes simplex virus 1 or 2 HIV-1
Congenital disorders (of fetus)	Trisomy 21 (Down syndrome) Trisomy 18 (Edwards syndrome) Trisomy 13 (Patau syndrome) Turner's syndrome

(Table by GGS Information Services.)

Causes and symptoms

The two types of IUGR described previously contribute to IUGR according to the development at that stage. Symmetrical IUGR may occur when the unborn baby experiences a problem during early development. Asymmetrical IUGR may occur when the unborn baby experiences a problem during later development. In general, most physicians believe that IUGR is the consequence of a disease process within one or more of the three partitions that maintain and regulate fetal growth, i.e., the maternal compartment, the placenta, or the fetus.

In consideration of risk factors uteroplacental insufficiency contributes to 80 percent of IUGR due to the following maternal causes:

- deficient supply of nutrients
- smoking
- malnutrition
- anemia
- drug abuse
- vascular diseases, i.e., high blood pressure
- chronic kidney disease
- severe diabetes
- multiple gestation
- antigen/antibody reactions, i.e., lupus, antiphospholipid antibody syndrome (APA)
- primary placental causes, i.e., extensive placental infarctions, chronic placental separation, placenta previa

Primary fetal causes contribute to 20 percent of IUGR and include the following:

- exposure to an infection, i.e., **rubella** (German **measles**), cytomegalovirus, syphilis, or toxoplasmosis
- birth defects, i.e., **congenital heart disease**, genitourinary anomalies, central nervous system defects
- chromosomal abnormalities, i.e., trisomy 13, 18, or 21
- primary bone or cartilage disorder
- decreased intrinsic growth, symmetrical IUGR

Diagnosis

IUGR can be difficult to diagnose but it begins with taking an extensive maternal history. A mother with a previous IUGR is at risk of having another during a subsequent pregnancy. Maternal renal and cardiopulmonary disease and multiple gestation are factors in IUGR.

KEY TERMS

Abruptio placentae—Premature separation of the placenta from the uterine wall. It occurs late in pregnancy and results in bleeding that may or may not establish an obstetrical emergency.

Circumvallate placenta—The existence of a thick, round, white, opaque ring around the periphery of the placenta that limits the expansion of the fetal vessels.

Fundus—The inside of an organ. In the eye, fundus refers to the back area that can be seen with the ophthalmoscope.

Hemoglobinopathies—Abnormal hemoglobins in the blood, i.e., sickle cell disease, thalessemia.

Hyperplasia—A condition where cells, such as those making up the prostate gland, rapidly divide abnormally and cause the organ to become enlarged.

Hypertrophy—An increase in the size of a tissue or organ brought about by the enlargement of its cells rather than cell multiplication.

Hypoplasia—An underdeveloped or incomplete tissue or organ usually due to a decrease in the number of cells.

Placenta—The organ that provides oxygen and nutrition from the mother to the unborn baby during pregnancy. The placenta is attached to the wall of the uterus and leads to the unborn baby via the umbilical cord.

Placenta previa—A condition in which the placenta totally or partially covers the cervix, preventing vaginal delivery.

Placental infarction—An area of dead tissue in the placenta that is due to an obstruction of circulation in the area.

Preeclampsia—A condition that develops after the twentieth week of pregnancy and results in high blood pressure, fluid retention that doesn't go away, and large amounts of protein in the urine. Without treatment, it can progress to a dangerous condition called eclampsia, in which a woman goes into convulsions.

Uteroplacental insufficiency—Designates the lack of blood flow from the uterus to the placenta, resulting in decreased nourishment and oxygen to the fetus.

Velamentous insertion of the umbilical cord—The attachment of the umbilical cord close to the membranes (bag of water) or in the membranes.

A positive rollover test is predictive of IUGR and is defined as a rise in diastolic blood pressure of 20 mm Hg or more with a position change from the left lateral recumbent to supine, which is an early indication of possible pregnancy-induced hypertension. Determination of an accurate due date is essential in order for the doctor to know if development and weight gain are appropriate. Checking the mother's weight and abdominal measurements can help diagnose cases in which there are no other risk factors present. During each prenatal visit, the healthcare provider uses a tape measure to record the uterine fundal height (measured from the top of the pubic area to the top of the uterus in centimeters). As the pregnancy continues and the baby grows, the uterus stretches upward in the direction of the mother's head. Between 20 and 34 weeks gestation, the uterine fundal height, in centimeters, equals the weeks of gestation. If the uterine fundal height is more than three to four below normal, IUGR is suspected. Ultrasound is used to evaluate the growth of the baby and the ratio of the head circumference (HC) to the abdominal circumference (AC) is a good predictor of asymmetric IUGR. Decreased amniotic fluid (oligohydramnios) is associated with IUGR as the fetus may have a decreased cardiac output and thus decreased renal flow to produce less urine. The grading of the placenta indicates whether it may be inappropriately aging, but this is not as accurate as fetal measurements. Doppler flow studies of the uterine artery provide information regarding the blood flow to the uterus. Doppler studies on the umbilical cord artery and the middle cerebral artery also provide information regarding fetal growth. Biochemical tests on hormone levels of estriol and human placental lactogen provide minimal information.

Treatment

Maternal bed rest is the initial approach for the treatment of IUGR. The benefit of bed rest is that it results in increased blood flow to the uterus. Studies on the efficacy of bed rest have been inconclusive, although in the early 2000s it is still used in combination with intravenous fluid therapy and oxygen therapy. The use of baby aspirin therapy remains controversial but theoretically it serves to preserve or improve blood flow to the placenta. If the baby does not grow well in utero after conservative treatment, an obstetrician may suggest inducing labor in the mother a few weeks early, or delivering the baby by **cesarean section**.

Prognosis

Outcomes for infants with IUGR differ depending on whether the condition is symmetrical or not and

whether there are other sequelae from the underlying cause of the IUGR. Outcomes include an increased risk of long-term neurologic and behavioral handicaps, possible intrauterine demise, low blood sugar (**hypoglycemia**), and low body temperature (hypothermia). These risks increase with the severity of the growth restriction. The growth that occurs after birth cannot be predicted with certainty based on the size of the baby when it is born. Infants with asymmetrical IUGR are more likely to catch up in growth after birth than are infants who suffer from prolonged symmetrical IUGR. Delayed eruption of teeth and enamel hypoplasia and an increased incidence of postnatal infections are frequently seen. Each case is unique. Some infants who have IUGR develop normally, while others have complications of the nervous system or intellectual problems such as **learning disorders**. If IUGR is related to a disease or a genetic defect, the future of the infant is related to the severity and the nature of that disorder.

Parental concerns

Pregnant women who have been diagnosed with a fetus with IUGR should talk to their obstetrician about options for treatment. If the woman is uncomfortable with options presented by the doctor, she should seek a qualified second opinion. All pregnant women are well-advised to eat properly, get enough rest, and refrain from alcohol, drugs, and tobacco use for the health of the infant. In addition, pregnant women should be especially careful to keep themselves healthy by following good hygiene practices and receiving a **flu vaccine** if one is recommended.

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ORGANIZATIONS

American College of Obstetricians and Gynecologists. 409 12th Street, SW, PO Box 96920, Washington, DC 20090. Web site: <www.acog.org>.

Association of Women's Health, Obstetric and Neonatal Nursing. 2000 L Street, NW Suite 740, Washington, DC 20036. Web site: <www.awhonn.org.

Linda K. Bennington

Intravenous rehydration

Definition

Intravenous rehydration is the process by which sterile water solutions containing small amounts of salt or sugar are injected into the body through a tube attached to a needle which is inserted into a vein.

Purpose

Intravenous rehydration is used to restore the fluid and electrolyte balance of the body due to illness, surgery, or accident. Electrolytes are salts (sodium, potassium, chloride, calcium, magnesium, phosphate, sulfate, and bicarbonate) that become ions when mixed with fluids in the body and blood and have the ability to conduct electricity. The body uses electrolytes to carry electrical impulses from cell to cell. Moderate to severe **dehydration** can interfere with the body's normal functioning. Restoration of fluids and electrolytes through intravenous means is the swiftest means to achieve fluid balance.

Description

Fever, **vomiting**, and **diarrhea** can cause a child to become dehydrated fairly quickly. Infants and children are especially vulnerable to dehydration. Athletes who have over-exerted themselves in hot weather may also require rehydration with IV (intravenous) fluids. An IV for rehydration can be in place for several hours to several days and is generally used if a patient cannot drink fluids.

Basic IV solutions are made of sterile water with small amounts of sodium (an ingredient in table salt) or dextrose (sugar) supplied in bottles or thick plastic bags that can hang on a stand mounted next to the patient's bed. Additional mineral salts such as potassium and calcium, **vitamins**, or medications can be added to the IV solution by injecting them into the bottle or bag with a needle or injected directly into the IV line.

Precautions

Patients receiving IV therapy need to be monitored to ensure that the IV solutions are providing the correct amounts of fluids and **minerals** needed. People with kidney and heart disease are at increased risk for **overhydration**, so they must be carefully monitored when receiving IV therapy.

A child receiving fluids through an intravenous (IV) bag. *(© Tom Stewart/Corbis.)*

Preparation

The doctor orders the IV solution and any additional nutrients or medications to be added to it. The doctor also specifies the rate at which the IV will dispense the solution.

The IV solutions are prepared under the supervision of a pharmaceutical company, using sanitary techniques that prevent bacterial contamination, and come prepackaged. Additions to the IV solutions are supervised by a doctor or nurse. Just like a prescription, the IV is clearly labeled to show its contents and the amounts of any additives.

The skin around the area where the needle for the IV catheter is inserted is cleaned and disinfected. Once the IV catheter is in place, it is taped to the skin to prevent it from being dislodged. The IV line is then attached to the IV catheter. Any other IV lines can be added to the IV catheter.

Aftercare

Patients need to take fluids by mouth before an IV solution is discontinued. After the IV needle is removed, the site should be inspected for any signs of bleeding or infection.

Risks

There is a small risk of infection at the injection site that is usually treated topically. It is also possible that the IV solution may not provide all of the nutrients needed, leading to a deficiency or an imbalance, which would need to be corrected.

If the needle becomes dislodged, the solution can flow into tissues around the injection site rather than into the vein. This is called extravasation, or infiltration, and occurs in about half of pediatric IVs. In most cases, the patient reports a burning or stinging sensation at the site of the needle or IV catheter, especially when new IV fluids are started or the speed of the IV drip is increased. The tissues usually swell and become discolored, looking like a bruise. Usually, the IV catheter is removed and reinserted at another site.

If an IV has been in place for a long time or the child has had a medical condition that weakens the veins, the

KEY TERMS

Dextrose—A sugar solution used in intravenous drips.

Electrolytes—Salts and minerals that produce electrically charged particles (ions) in body fluids. Common human electrolytes are sodium chloride, potassium, calcium, and sodium bicarbonate. Electrolytes control the fluid balance of the body and are important in muscle contraction, energy generation, and almost all major biochemical reactions in the body.

Extravasation—To pass from a blood vessel into the surrounding tissue.

Sodium—An element; sodium is the most common electrolyte found in animal blood serum.

Thrombosis—The formation of a blood clot in a vein or artery that may obstruct local blood flow or may dislodge, travel downstream, and obstruct blood flow at a remote location. The clot or thrombus may lead to infarction, or death of tissue, due to a blocked blood supply.

child may experience vein collapse. This occurs when the vein is not able to receive anymore intravenous fluid and forces the IV solution into the surrounding tissues. It can also occur if a thrombosis, or blood clot, forms in the vein at the IV catheter site.

A collapsed vein feels and looks much like a dislodged IV catheter. This can sometimes happen when the nurse has inserted a needle or IV catheter that is too big for the size of the vein. This isn't a misjudgement on the nurse's part. Standard sized needles are used and only rarely are extremely thin needles necessary. They may be needed in adult patients as well as children. If vein collapse occurs, the IV catheter should be removed and reinserted into a different vein, usually in another part of the body. For example, if a vein in the left arm collapses, the nurse can put a new IV catheter into the right arm.

Treatment for an extravasation or a collapsed vein are similar. A warm compress is usually applied to the injection site to reduce swelling. If there is sufficient injury at the injection site, general wound care is done to prevent infection and speed healing.

Parental concerns

Usually intravenous rehydration is very effective, allowing the child's body to return to its normal fluid equilibrium. Once the child can keep fluids down orally and urine output has returned to normal, then intravenous rehydration is discontinued. Most children don't relapse once they are home if they can continue taking fluids by mouth.

Intravenous rehydration should only be used for moderate to severe dehydration. It is not recommended for mild dehydration from stomach upset or flu. Oral products that restore fluid and electrolytes balance are better for hydrating a sick child who has been vomiting during an illness or after strenuous activity in extremely hot weather. Oral rehydration also does not require a hospital stay.

To reduce the risk of displacement of the needle when the IV catheter or needle is inserted, parents should help the child keep still. The child should also be careful when moving about so as not to dislodge the IV catheter, especially at night. Parents can also look at the injection site and report any discoloration they see. They should also encourage the child to report any burning or stinging around the IV catheter to the nursing staff.

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Janie Franz
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Intussusception *see* **Intestinal obstructions**

Iron deficiency anemia

Definition

Iron deficiency anemia refers to anemia that is caused by lower than normal levels of iron. This type of anemia is caused by deficient erythropoiesis, the ongoing process of the bone marrow to produce healthy red blood cells (RBCs). It is characterized by the production of small (microcytic) RBCs. When examined under a microscope, the RBCs also appear pale or light colored from the absence of heme, the major component of hemoglobin, which is the iron-bearing protein and coloring pigment in RBCs. Anemia resulting from a deficiency of iron is also called microcytic anemia.

Description

Anemia is a blood disorder characterized by abnormally low levels of healthy RBCs or reduced levels of hemoglobin (Hgb), the iron-bearing protein in RBCs that delivers oxygen to tissues throughout the body. Blood cell volume (hematocrit) may also be reduced in some **anemias**, but not necessarily in iron deficiency anemia. The reduction of any or all of these blood parameters reduces the essential delivery of oxygen through the bloodstream to the organs of the body. Iron is a mineral found in the bloodstream that is essential for growth, enzyme development and function, a healthy immune system, energy levels, and muscle strength. It is an important component of hemoglobin and myoglobin, the type of hemoglobin in muscle tissue.

Iron deficiency anemia is the most common type of anemia throughout the world, although it occurs to a lesser extent in the United States because of the higher consumption of iron-rich red meat and the practice of food fortification (addition of iron to foods by manufacturers). In developing countries in tropical climates, the most common cause of iron deficiency anemia is infestation with hookworm.

The onset of iron deficiency anemia is gradual and may not have early symptoms. The deficiency begins when the body's store of iron is depleted and more iron is being lost through bleeding or malabsorption than is derived from food and other sources. Because depleted iron stores cannot meet the red blood cells' needs, fewer red blood cells develop. In this early stage of anemia, the red blood cells look normal, but they are reduced in number. Eventually the body tries to compensate for the iron deficiency by producing more red blood cells, which are characteristically small in size (spherocytosis). Symptoms of anemia, especially weakness and fatigue, develop at this stage.

Causes and symptoms

Iron is an essential component of the production of healthy RBCs, and iron stores must be maintained for the ongoing production of RBCs by the bone marrow. Iron deficiency anemia can develop as a result of depleted iron stores from chronic blood loss, increased demands for iron as seen in periods of growth (e.g., in infancy and **adolescence**), or malabsorption of iron even when foods or supplements are supplying adequate amounts. It is accepted that iron is hard to absorb; this, in combination with diets that may not meet daily requirements, is a common route to iron deficiency and iron deficiency anemia. Iron can also be lost through strenuous **exercise** and heavy perspiration, poor digestion, frequent consumption of antacids, long-term illness, heavy menstrual cycles, and other causes.

Infancy is a period of increased risk for iron deficiency because dietary iron may not be adequate for the rapid growth of the child in the first two years of life, an example of increased demand. The human infant is born with a built-in supply of iron, which can be tapped during periods of drinking low-iron milk or formula. Both human milk and cow milk contain rather low levels of iron (0.5–1.0 mg iron/liter). However, the iron in the mother's breast milk is about 50 percent absorbed by the infant, while the iron of cow milk is only 10 percent absorbed. During the first six months of life, growth of the infant is made possible by the milk in the diet and by the infant's built-in supply of iron. However, premature infants have a lower supply of iron and, for this reason, it is recommended that pre-term infants, beginning at two months of age, be given oral supplements of 7 mg iron/day, as ferrous sulfate. Iron deficiency can be provoked where infants are fed formulas based on unfortified cow milk. For example, unfortified cow milk is given free of charge to mothers in Chile. This practice has the fortunate result of preventing general **malnutrition**, but the unfortunate result of allowing the development of mild iron deficiency.

Children have a great need for iron as they grow, and in most cases, the diet will provide replacement iron for the iron used in growth. Children seem to stay in balance unless a bleeding disorder of some kind exists, either hereditary (**hemophilia** or von Willebrand's) or related to hookworm infection or another illness. In adolescence, girls have an increased requirement for iron because of increased growth and the start of **menstruation**. Adolescent boys also experience a major growth spurt that demands more iron; iron stores are worn thin especially when healthy red cell function is needed for adequate oxygenation of exercising muscles and developing organs. Teenagers are also not noted for making

healthy food choices; often they are losing iron stores and not replenishing iron through diet.

Iron deficiency occurs most often through chronic blood loss, more often in adults than in children, although the sources of bleeding can apply to people of all ages. Blood losses from gastrointestinal bleeding, excessive menstrual bleeding, and infection with hookworm can deplete iron and lead to iron deficiency anemia. In hookworm infection, a parasitic worm that thrives in warm climates, including in the southern United States, enters the body through the skin, such as through bare feet. The hookworm then migrates to the small intestines where it attaches itself to small sausage-shaped structures in the intestines (villi) that help with the absorption of all nutrients. The hookworm damages the villi, resulting in blood loss; they simultaneously produce anti-coagulants that promote continued bleeding. Each worm can initiate losses of up to 0.25 ml of blood per day.

Chronic blood losses through gradual bleeding in the gastrointestinal tract can be provoked by other conditions such as hemorrhoids, bleeding ulcers, anal fissures, **irritable bowel syndrome**, aspirin-induced bleeding, blood clotting disorders, and diverticulosis (a condition caused by an abnormal opening from the intestine or bladder). Several genetic diseases also lead to bleeding disorders. These include the **coagulation disorders** hemophilia A and hemophilia B, and von Willebrand's disease, a bleeding disorder caused by a deficiency in von Willebrand factor, an essential component of the coagulation system. All three genetic diseases can produce symptoms and be diagnosed in childhood.

The symptoms of iron deficiency anemia appear slowly and typically include weakness and fatigue. These symptoms result because of the reduced oxygen carrying capacity of RBCs and the reduced ability of the RBCs to carry iron to working muscles. Iron deficiency can also affect other tissues, including the tongue and fingernails. Prolonged iron deficiency can result in a smooth, shiny, and reddened tongue, a condition called glossitis. The fingernails may grow abnormally and acquire a spoon-shaped appearance.

When to call the doctor

Weakness, **dizziness**, listlessness, or fatigue may be the first signs of iron deficiency anemia. A compulsion to chew on ice cubes or to eat soil is also an indication of iron deficiency. The pediatrician should be consulted if the child is extremely pale, with little or no color in the gums, nail beds, creases of the palm, or lining of the eyelids.

Demographics

In the United States, iron deficiency anemia affects thousands of toddlers between one and two years of age and more than 3 million women of childbearing age. This condition is less common in older children and in adults over 50 and rarely occurs in teenage boys and young men.

Diagnosis

Diagnosing iron deficiency anemia begins with the pediatrician taking a careful history, including the child's age, symptoms, illnesses, general state of health, and a **family** history of anemias. Symptoms noticed in children by their parents may include fatigue, weight loss, inability to concentrate, loss of appetite, and light-headedness when standing up. The physical examination may reveal paleness, and lack of color in the creases of the palms, in gums, and in the linings of the eyelids.

Diagnostic testing starts with a complete blood count (CBC) and differential, counting RBCs, white blood cells (WBCs) and measuring hemoglobin (Hgb), hematocrit (Hct), and other factors. In iron deficiency anemia, the RBC count can be normal or elevated and hemoglobin will be abnormally low. In infants, iron deficiency anemia is defined as having a hemoglobin level below 109 mg/ml when measured in whole blood, and a hematocrit of less than 33 percent. In the microscopic examination of a stained blood smear (differential), red cells may appear smaller than normal. The mean corpuscular volume (MCV) will be measured to compare the size of RBCs with normal RBCs. A reticulocyte (young RBCs) count will help determine if anemia is caused by impaired RBC production, as in iron deficiency anemia, or increased RBC destruction as in some other types of anemia. Iron, vitamin C or vitamin B_{12}, and folate levels will be measured in blood serum to identify possible deficiencies. In addition to measuring iron itself (Fe) and total iron-binding capacity (TIBC), transferrin and transferrin saturation tests may be performed to evaluate iron metabolism. Different types of hemoglobin may be measured by a diagnostic testing method called hemoglobin electrophoresis. Protoporphyrin IX, a component of hemoglobin, may be measured to help confirm a diagnosis of iron deficiency anemia. Confirmation may also be obtained by taking a bone marrow sample (bone marrow biopsy) for microscopic examination. Kidney function tests, coagulation tests, and stool examinations for occult (hidden) blood may also be performed.

The presence of occult blood found in a stool examination may indicate gastrointestinal bleeding or other causes of bleeding such as aspirin-induced or a bleeding

ulcer. The physician then needs to examine the gastrointestinal tract to determine the cause and location of bleeding. In this case, a diagnosis of iron deficiency anemia may include examination with a sigmoidoscope, a flexible, tube-like instrument with a light source that permits examination of the colon. A barium-enhanced x ray of the intestines may also be used to detect abnormalities that can cause bleeding.

The diagnosis of iron deficiency anemia may include a test for oral iron absorption, especially when evidence suggests that oral iron supplements have failed to raise hemoglobin. The oral iron absorption test is conducted by injecting 64 mg iron (325 mg ferrous sulfate) in a single dose. Blood samples are then taken after two hours and four hours. The iron content of the blood serum is then measured. The concentration of iron should rise by about 22 micromolar, when iron absorption is normal. Lesser increases in concentration mean that iron absorption is abnormal and that more effective therapy may involve injections or infusions of iron.

Treatment

The goal of treatment for iron deficiency anemia is to restore iron levels and the production of healthy RBCs and increase the essential flow of oxygen to tissues. Iron preparations may be given by injection or, in older children, as oral supplements. Taking vitamin C along with oral iron supplementation is accepted as a way to achieve better absorption of the iron. Taking iron supplements can result in **constipation**, **diarrhea**, cramps, or **vomiting** in sensitive individuals. Injections and infusions of iron can be given to individuals with poor iron absorption. Treatment of iron deficiency anemia sometimes requires more than iron supplementation. When iron deficiency is provoked by hemorrhoids or gastrointestinal bleeding, for example, surgery may be required to prevent recurrent iron deficiency anemia. When iron deficiency is provoked by bleeding due to aspirin ingestion, aspirin is discontinued. Iron deficiency caused by hookworm infection requires drug therapy to eliminate the parasite; prevention includes wearing shoes when walking in soil known to be infested with hookworms.

Alternative treatment

Vitamin C is noted for helping to absorb iron in the diet and iron supplements. Cooking in a cast iron skillet may leach small amounts of absorbable iron into the diet. Besides the iron found in eggs, fish, liver, meat, poultry, green leafy vegetables, whole grains, and enriched or whole grain breads and cereals, good food sources of iron include blackstrap molasses, brewer's yeast, and certain types of sea vegetable (e.g., hijiki, kelp, dulse). Herbal supplements that benefit individuals who have iron deficiency anemia include alfalfa, burdock root, dandelion, dong quai, mullein, nettle, raspberry leaf, shepherd's purse, and yellow dock. Herbs are available as tinctures and teas or in capsules.

Nutritional concerns

Decreased dietary iron intake is a contributing factor in iron deficiency and iron deficiency anemia. Deciding how to add enough iron to the diet, however, depends not just on which foods contain it, but in which foods iron is most available for absorption and use by the body. Bioavailability describes the percent of dietary iron that is successfully absorbed via the gastrointestinal tract to the bloodstream. Non-absorbed iron is lost in the feces. Generally, iron bioavailability in fruits, vegetables, and grains is lower than the iron availability of meat. The availability of iron in plants ranges from only 1 to 10 percent, with some exceptions, while that in meat, fish, chicken, and liver is consistently 20–30 percent. In the following list, the iron content is given parenthetically for each food.

- cabbage (1.6 mg/kg)
- spinach (33 mg/kg)
- lima beans (15 mg/kg)
- potatoes (14 mg/kg)
- tomatoes (3 mg/kg)
- apples (1.5 mg/kg)
- peanut butter (6.0 mg/kg)
- raisins (20 mg/kg)
- whole wheat bread (43 mg/kg)
- eggs (20 mg/kg)
- canned tuna (13 mg/kg)
- chicken (11 mg/kg)
- beef (28 mg/kg)

It is easy to see that apples, tomatoes, and peanut butter are relatively low in iron, while spinach, whole wheat bread, and beef are relatively high in iron. Red meat sources reliably replace the heme component of red blood cells, raising hemoglobin levels and helping to correct iron deficiency. For infants and toddlers, the most available source of iron is human milk (50% availability).

The assessment of whether a food is low or high in iron can also be made by comparing the amount of that food eaten per day with the recommended dietary allowance (RDA) for iron. The RDA for iron for the adult male is 10 mg/day, while that for the adult woman is

KEY TERMS

Erythropoiesis—The process through which new red blood cells are created; it begins in the bone marrow.

Hematocrit—A measure of the percentage of red blood cells in the total volume of blood in the human body.

Heme—The iron-containing molecule in hemoglobin that serves as the site for oxygen binding.

Hemoglobin—An iron-containing pigment of red blood cells composed of four amino acid chains (alpha, beta, gamma, delta) that delivers oxygen from the lungs to the cells of the body and carries carbon dioxide from the cells to the lungs.

Protoporphyrin IX—A protein the measurement of which is useful for the assessment of iron status. Hemoglobin consists of a complex of a protein plus heme. Heme consists of iron plus protoporphyrin IX. Normally, during the course of red blood cell formation, protoporphyrin IX acquires iron, to generate heme, and the heme becomes incorporated into hemoglobin. However, in iron deficiency, protophoryrin IX builds up.

Recommended Dietary Allowance (RDA)—The Recommended Dietary Allowances (RDAs) are quantities of nutrients in the diet that are required to maintain good health in people. RDAs are established by the Food and Nutrition Board of the National Academy of Sciences, and may be revised every few years. A separate RDA value exists for each nutrient. The RDA values refer to the amount of nutrient expected to maintain good health in people. The actual amounts of each nutrient required to maintain good health in specific individuals differ from person to person.

15 mg/day. The RDA during pregnancy is 30 mg/day. The RDA for infants five months of age or younger is 6 mg/day, while that for infants of five months to one year of age is 10 mg/day. RDA values are based on the assumption that people eat a mixture of plant and animal foods.

Prognosis

The prognosis for treating and curing iron deficiency anemia is excellent, particularly when those affected take iron supplements as advised and are able to assimilate the iron. A number of studies have shown that iron deficiency anemia in infancy can result in reduced **intelligence**, when intelligence was measured in early childhood. It is not certain if iron supplementation of children with reduced intelligence, due to iron-deficiency anemia in infancy, has any influence in allowing a "catch-up" in intellectual development.

Prevention

In the healthy population, mineral deficiencies can be prevented by the consumption of inorganic nutrients at levels defined by the RDA. Iron deficiency anemia in infants and young children can be prevented by breastfeeding, consuming good dietary sources of iron, and using fortified foods. Liquid cow milk-based infant formulas are generally supplemented with iron (12 mg/L). The iron in liquid formulas is added as ferrous sulfate or ferrous gluconate. Commercial infant cereals are also fortified with iron, adding small particles of elemental iron. The levels used are about 0.5 gram iron/kg dry cereal. This amount of iron is about 10-fold greater than that of the iron naturally present in the cereal. Iron supplementation is not recommended for all infants, and children and pediatricians should be consulted before giving supplements. Vitamin C is recommended to improve the assimilation of iron in the body, especially when iron is obtained from non-food sources.

Nutritional concerns

The average diet in the United States contains about 6 mg of iron per calorie of food, which is sufficient for maintaining iron stores. Only 1 mg of iron, however, is absorbed for every 10 mg. consumed, which mean sources of iron must be carefully chosen. The bioavailability of iron in foods varies, influencing the amounts that can be absorbed through the intestines. Absorption is best when the food contains heme, just as in human red cells. That makes meat the best choice as a source of iron and iron-rich vegetables and fruits such as spinach and apricots the next best choice. Certain other plant foods that contain fiber, such as bran, actually reduce the absorption of non-heme iron; so do antacid medications, often taken to relieve the upset stomach associated with taking oral iron supplements. Additionally, food interactions reduce bioavailability. Ascorbic acid (vitamin C) is the only food constituent known to increase the availability of non-heme iron, such as in vegetables and also in food supplements.

Parental concerns

Understanding iron metabolism and the ways to ensure that iron deficiency anemia in infants and children can be successfully treated and prevented from

recurring may be concerns of parents. It is important to remember that although iron deficiency anemia is common in infants and toddlers, it is easily corrected by feeding infants mother's milk or iron-fortified formulas. In older children, the diet usually balances iron usage and replacement. In teenage years, when demands for iron increase for rapid growth and to compensate for menstruation in girls, parents will need to pay attention once again to providing adequate food sources. However, supplementation of iron should only be done with a doctor's recommendation.

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ORGANIZATIONS

National Heart, Lung, and Blood Institute (NHLBI). 6701 Rockledge Drive, PO Box 30105, Bethesda, MD 20824–0105. Web site: <www.nhlbi.nih.gov>.

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Irritable bowel syndrome

Definition

Irritable bowel syndrome (IBS) is a common gastrointestinal condition characterized by abdominal **pain** and cramps; changes in bowel movements (**diarrhea**, **constipation**, or both); gassiness; bloating; **nausea**; and other symptoms. There is no cure for IBS; however, dietary changes, stress management, and sometimes medications are often able to eliminate or substantially reduce its symptoms.

Description

IBS is the name people use today for a condition that was once called—among other things—spastic colitis, mucous colitis, spastic colon, nervous colon, spastic bowel, and functional bowel disorder. Some of these names reflected the now outdated belief that IBS is a purely psychological disorder, a product of the patient's imagination. Although modern medicine recognizes that stress, **anxiety** and depression can trigger IBS attacks, medical specialists agree that IBS is a genuine physical disorder—or group of disorders—with specific identifiable characteristics. IBS is considered a functional disorder because it is thought to result from changes in the activity of the major part of the large intestine (the colon).

Demographics

IBS is one of the most common functional gastrointestinal disorders, affecting 10-20 percent of adults in the United States. Research has demonstrated that symptoms compatible with IBS are about as common in school-age children as in adults. IBS normally makes its first appearance during young adulthood, and symptoms usually begin at about age 20. Women with IBS represent over 70 percent of IBS sufferers. IBS is responsible for more time lost from school and work than any medical problem—other than the **common cold**. It accounts for a substantial proportion of the patients seen by specialists in diseases of the digestive system (gastroenterologists).

A community-based study of 507 middle school and high school students by Hyams, et al, found that 6-14 percent of the adolescent population had IBS symptoms. Anxiety and depression scores were significantly higher for this group. Eight percent of all the students in the study had seen a physician for abdominal pain in the previous year.

Causes and symptoms

Causes

Although the exact cause or causes of IBS are unknown, research suggests that people with IBS may have a colon that is more sensitive and reactive to certain foods and stress.

After food is digested by the stomach and small intestine, the undigested material passes in liquid form into the colon, which absorbs water, nutrients and salts. Normally, the colon is quiet during most of that period except after meals, when its muscles contract in a series of wavelike movements called peristalsis. Peristalsis helps absorption by bringing the undigested material into contact with the colon wall. It also pushes undigested material that has been converted into solid or semisolid feces toward the rectum, where it remains until a bowel movement occurs.

In IBS, however, the normal rhythm and intensity of peristalsis is disrupted. Sometimes there is too little

peristalsis, which can slow the passage of undigested material through the colon and cause constipation. Sometimes there is too much, which has the opposite effect and causes diarrhea. In other cases, peristalsis can be spasmodic, causing sudden strong muscle contractions that come and go.

DIET Some foods and beverages appear to play a key role in triggering IBS attacks. Certain foods and drinks may disrupt peristalsis in IBS patients, which may explain why IBS attacks often occur shortly after meals. Some of the chief culprits include:

- chocolate
- dairy products
- **caffeine** (in coffee, tea, colas, and other drinks)
- carbonated beverages (colas, pop, soda)
- wheat
- rye
- barley
- excess alcohol

Other foods also have been identified as problems, and the pattern of what can and cannot be tolerated is different for each person.

STRESS Stress—feeling mentally or emotionally tense, troubled, angry or overwhelmed—stimulates colon spasms in people with IBS since there is a close nervous system connection between the brain and the intestines. A large network of nerves control the normal rhythmic contractions of the colon. Although researchers do not yet understand all of the links between changes in the nervous system and IBS, they point out the similarities between mild digestive upsets and IBS. Just as healthy people can feel nauseated or have an upset stomach when under stress, people with IBS react the same way, but to a greater degree.

MENSTRUATION IBS symptoms sometimes intensify during **menstruation**, suggesting female reproductive hormones may trigger the condition.

Symptoms

The symptoms of IBS tend to rise and fall in intensity, rather than grow steadily worse over time. Symptoms always include:

- abdominal pain, which may be relieved by defecation
- diarrhea
- constipation
- diarrhea alternating with constipation

Other symptoms, which vary from person to person, include:

- cramps
- gassiness
- bloating
- nausea
- passage of mucus during bowel movements
- abnormal stool frequency—defined as greater than three bowel movements per day or less than three bowel movements per week
- abnormal stool form (lumpy, hard, loose, or watery stool)
- abnormal stool passage (straining, urgency, or feeling of incomplete bowel movement)

In general, symptoms are not present all the time and do not interfere with school and other normal activities. IBS symptoms rarely occur at night and disrupt the patient's **sleep**. Moderate IBS occasionally disrupts normal activities.

When to call the doctor

If a child has the following symptoms, the parent should contact the child's pediatrician or gastroenterologist:

- abdominal pain or diarrhea that wakes the child during the night
- persistent or severe abdominal pain
- unexplained weight loss
- rectal bleeding
- fever
- **family** history of irritable bowel disease

Diagnosis

The Rome II criteria are the accepted diagnostic criteria for IBS. These criteria were developed by an international group of pediatric gastroenterologists and include:

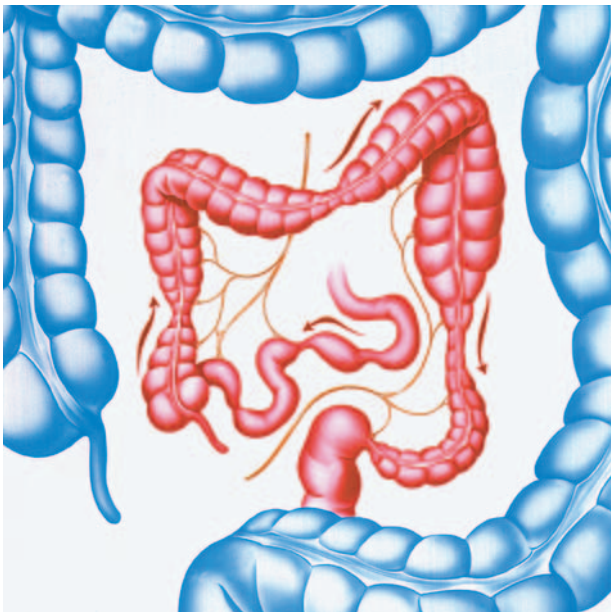
- Continuous or recurrent abdominal discomfort or pain for at least three months that is: a) Relieved with defecation and/or b) Associated with a change in frequency and/or c) Associated with a change in appearance of stool. Two or three of these features are present with an IBS diagnosis.
- No structural or metabolic abnormalities are present that may be responsible for the IBS symptoms.

The diagnosis of IBS is further supported by the presence of the symptoms listed previously. In addition, the

primary pediatrician or gastroenterologist may confirm the diagnosis of IBS after questioning the child (if old enough to provide an accurate history of symptoms) or parent about his or her physical and mental health (the medical history), performing a physical examination, and ordering laboratory tests to rule out other conditions that resemble IBS, such as Crohn's disease and ulcerative colitis.

Diagnostic tests may include stool or blood tests, hydrogen breath test, or an x ray of the bowel, called a barium enema. When symptoms continue even after treatment, endoscopic tests such as a colonoscopy or sigmoidoscopy may be performed. An endoscopic test is an internal examination of the colon using a flexible instrument (a sigmoidoscope or colonoscope) that is inserted through the anus.

A nutritional **assessment** performed by a registered dietitian may be included in the child's diagnostic evaluation. The nutritional assessment includes a review of the child's fiber intake as well as his or her usual consumption of sugars such as sorbitol and fructose—common culprits of diarrhea.



Normal and diseased (center) colons. Areas of constriction in the colon cause constipation, while areas of distention cause diarrhea. *(© 1995 John Bavosi/Science Photo Library. Custom Medical Stock Photo, Inc.)*

Treatment

Dietary changes and sometimes medications are considered the keys to successful treatment. Psychosocial difficulties are also addressed and treated with therapy or counseling as needed. Treatment requires a long-term commitment; six months or more may be needed before the child notices substantial improvement.

Alternative treatment

Alternative and complementary therapies include approaches that are considered to be outside the mainstream of traditional health care. Alternative and traditional approaches to IBS treatment overlap to a certain extent. Like traditional doctors, alternative practitioners advise a high-fiber diet to reduce digestive system irritation. They also suggest avoiding caffeine and fatty, gassy, or spicy foods, as well as alcohol. Recommended stress management techniques include **yoga**, meditation, guided imagery, hypnosis, biofeedback, and reflexology. Reflexology is a foot massage technique that is thought to relieve diarrhea, constipation, and other IBS symptoms.

The list of alternative treatments for IBS is quite long. It includes aromatherapy, homeopathy, hydrotherapy, juice therapy, acupuncture, chiropractic, osteopathy, naturopathic medicine, and Chinese traditional herbal medicine.

Before learning or practicing any particular technique, it is important for the parent/caregiver and child to learn about the therapy, its safety and effectiveness, potential side effects, and the expertise and qualifications of the practitioner. Although some practices are beneficial, others may be harmful to certain patients.

Relaxation techniques and dietary supplements should not be used as a substitute for medical therapies prescribed by a doctor. Parents should discuss these alternative treatments with the child's doctor to determine the techniques and remedies that may be beneficial for the child.

Nutritional concerns

Dietary changes, including a low-fat, high-fiber diet, may help decrease IBS symptoms. The addition of wheat bran or other fiber may be suggested to decrease symptoms. The formula for determining the recommended fiber intake for children, as advised by the American Dietetic Association, is to take the child's age plus five to equal the grams of dietary fiber the child should consume daily. Fiber should be added gradually to the child's diet.

The doctor may recommend a lactose-free diet for two or three weeks to determine if **lactose intolerance** is causing the symptoms. Lactose is the milk sugar found

KEY TERMS

Anus—The opening at the end of the intestine through which solid waste (stool) passes as it leaves the body.

Barium enema—An x ray of the bowel using a liquid called barium to enhance the image of the bowel. This test is also called a lower GI (gastrointestinal) series.

Colonoscopy—An examination of the lining of the colon performed with a colonoscope.

Constipation—Difficult bowel movements caused by the infrequent production of hard stools.

Crohn's disease—A chronic, inflammatory disease, primarily involving the small and large intestine, but which can affect other parts of the digestive system as well.

Defecation—The act of having a bowel movement or the passage of feces through the anus.

Diarrhea—A loose, watery stool.

Endoscopy—Visual examination of an organ or body cavity using an endoscope, a thin, tubular instrument containing a camera and light source. Many endoscopes also allow the retrieval of a small sample (biopsy) of the area being examined, in order to more closely view the tissue under a microscope.

Feces—The solid waste, also called stool, that is left after food is digested. Feces form in the intestines and pass out of the body through the anus.

Gastroenterologist—A physician who specializes in diseases of the digestive system.

Hydrogen breath test—A test used to determine if a person is lactose intolerant or if abnormal bacteria are present in the colon.

Lactose—A sugar found in milk and milk products.

Peristalsis—Slow, rhythmic contractions of the muscles in a tubular organ, such as the intestines, that move the contents along.

Sigmoidoscopy—A procedure in which a thin, flexible, lighted instrument, called a sigmoidoscope, is used to visually examine the lower part of the large intestine. Colonoscopy examines the entire large intestine using the same techniques.

Ulcerative colitis—A form of inflammatory bowel disease characterized by inflammation of the mucous lining of the colon, ulcerated areas of tissue, and bloody diarrhea.

in dairy products. Lactose intolerance is a common condition in up to 40% of patients with IBS. During the lactose-free period, the child should avoid all products containing lactose. The parent and child are asked to record the intake of all foods and beverages and note when symptoms occur after eating or drinking.

To identify other problem-causing foods or beverages, it is helpful for the parent and child to keep a diary of symptoms for two or three weeks, including daily activities, meals, symptoms and emotions. The doctor can then review the diary with the parent and child to identify possible problem areas.

In addition to lactose, known problem-causing substances include caffeine, beans, onions, cabbage, cucumbers, broccoli, fatty foods, alcohol, and certain medications. Once the specific substances that trigger symptoms are identified, they should be avoided. A registered dietitian can help the parent and child make specific dietary changes.

If lactose intolerance is a problem, the child may need to take calcium supplements or choose other foods high in calcium to meet the recommended daily requirement. If lactose intolerance is not a problem, the child can still have milk or milk products.

Medications

Medications affect each child differently, and no one medication works for every child with IBS. The child and parent will need to work with the doctor to find the best combination of medicine, diet, counseling and support to manage symptoms.

Stool softeners such as polyethelene glycol (Miralax) or an over-the-counter laxative may be recommended for constipation. Mineral oil also may be helpful. However, it is important not to use over-the-counter remedies without first consulting with the child's doctor.

Tricyclic **antidepressants** in low doses may be prescribed for pain relief. Antidepressants work by blocking pain transmission from the nervous system. Antispasmodic medications can slow bowel contractions and decrease diarrhea. Anticholinergics may help control intestinal cramping. Keep in mind that the effectiveness of these drugs to treat IBS has not been studied extensively in children.

Counseling and support

Psychological counseling or behavioral therapy may be recommended for some patients to reduce anxiety and stress and to learn to cope with the symptoms of IBS. Biofeedback, guided imagery, relaxation therapy, hyp-

nosis, and cognitive-behavioral therapy are examples of behavioral therapy. An ongoing and supportive doctor-patient relationship is also very important. The child and family must be reassured that although IBS causes symptoms that are uncomfortable and sometimes painful, it is not a harmful condition and does indicate a serious problem.

Prognosis

IBS is not a life-threatening condition. It is not an anatomical or structural defect, nor an identifiable physical or chemical disorder. IBS does not cause intestinal bleeding or inflammation, nor does it cause other gastrointestinal diseases or **cancer**. Although IBS can last a lifetime, in up to 30% of cases the symptoms eventually disappear. Even if the symptoms cannot be eliminated, with appropriate treatment they usually can be managed enough so IBS becomes merely an occasional inconvenience.

Prevention

Nutritional concerns

To help prevent or decrease the child's symptoms, parents can:

- help the child identify and avoid problematic foods
- work with a registered dietitian to facilitate specific dietary changes
- incorporate changes in the child's diet gradually so his or her body has time to adjust
- establish set times for meals; not allowing the child to skip a meal
- encourage the child to drink at least eight 8-ounce glasses of water per day
- serve small portions during meals
- teach the child to eat slowly, to avoid swallowing too much air that can produce excess gas
- try offering smaller, more frequent meals
- keep a regular schedule for bathroom visits

Parental concerns

Parents should reinforce with the child that IBS is not a life-threatening condition and that dietary changes and stress reduction can help reduce symptoms. Remind the child that six months or more may be needed before he or she notices substantial improvement in symptoms.

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American College of Gastroenterology (ACG). P.O. Box 3099, Alexandria, VA 22302. (703) 820-7400. Web site: <http://www.acg.gi.org/patientinfo/cgp/cgpvol2.html>.

American Gastroenterological Association. 4930 Del Ray Ave., Bethesda, MD 20814. (301) 654-2055. Web site: <http://www.gastro.org/clinicalRes/brochures/ibs.html>.

International Foundation for Functional Gastrointestinal Disorders (IFFGD). P.O. Box 170864, Milwaukee, WI 53217-8076. (888) 964-2001. E-mail: iffgd@iffgd.org. Web site: <http://www.iffgd.org>.

Irritable Bowel Syndrome (IBS) Association. 1440 Whalley Ave., #145, New Haven, CT 06515. E-mail: ibsa@ibsassociation.org. Web site: <http://www.ibsassociation.org>.

Irritable Bowel Syndrome Self Help and Support Group. 1440 Whalley Ave., #145 New Haven, CT 06515. E-mail: ibs@ibsgroup.org. Web site: <http://www.ibsgroup.org>.

National Digestive Diseases Information Clearinghouse (NDDIC). 2 Information Way, Bethesda, MD 20892-3570. (800) 891-5389. E-mail: nddic@info.niddk.nih.gov. Web site: <http://www.niddk.nih.gov/health/digest/nddic.htm>.

WEB SITES

About IBS. Available online at <http://www.aboutibs.org.>

Howard Baker

Itching

Definition

Itching is an intense, distracting irritation or tickling sensation that may be felt all over the skin's surface or confined to just one area. The medical term for itching is pruritus.

Description

Itching instinctively leads most people to scratch the affected area. Different people can tolerate different amounts of itching, and the threshold of tolerance can change due to stress, emotions, and other factors. In general, itching is more severe if the skin is warm and if there are few distractions. This is why people tend to notice itching more at night.

Demographics

It is common for children to be itchy occasionally. Prolonged itching in a specific location and generalized itching in many different areas of the body are less common.

Causes and symptoms

The reason for the sensation of itching is not well understood. While itching is the most noticeable symptom in many skin diseases, it does not necessarily mean that a person who feels itchy has a disease.

Stress and emotional upset can make itching worse, no matter what the underlying cause. Itching is often worse at night or at times when there are no distractions. If emotional problems are the primary reason for the itch, the condition is known as psychogenic itching. Some people become convinced that their itch is caused by a parasite; this conviction is often linked to burning sensations in the tongue and may be caused by a major psychiatric disorder.

Generalized itching

Itching that occurs all over the body may indicate a medical condition such as **diabetes mellitus**, liver disease, kidney failure, **jaundice**, thyroid disorders, or rarely, **cancer**. Blood disorders such as leukemia and lymphatic conditions such as Hodgkin's disease may sometimes cause itching as well.

Some children may develop an itch without a rash when they take certain drugs such as aspirin or codeine. Others may develop an itchy red drug rash or **hives** because of an allergy to a specific drug such as penicillin.

Itching also may be caused when any of the family of hookworm larvae penetrate the skin. This includes swimmer's itch and creeping eruption caused by cat or dog hookworm and ground itch caused by the true hookworm.

Many skin conditions cause an itchy rash. These include:

- atopic **dermatitis**
- **contact dermatitis**
- dermatitis herpetiformis (occasionally)
- eczema
- fungus infections (such as athlete's foot)
- hives (urticaria)
- insect **bites**
- lice
- lichen planus
- neurodermatitis (lichen simplex chronicus)
- psoriasis (occasionally)
- scabies

Itching all over the body can be caused by something as simple as bathing too often, which removes the skin's natural oils and may make the skin too dry.

Localized itching

Specific itchy areas may occur if a person comes in contact with soap, detergents, or wool or other rough-textured, scratchy material. Adults who have hemorrhoids, anal fissure, or persistent **diarrhea** may notice itching around the anus (called pruritus ani). When children itch in this area, the cause is most likely **pinworms**.

Intense itching in the external genitalia in women (pruritus vulvae) may be due to **candidiasis** (yeast), hormonal changes, or the use of certain spermicides or vaginal suppositories, ointments, or deodorants.

When to call the doctor

If the child is itchy all over or has a localized itch in combination with a rash, **fever**, infection, or is acting sick, the doctor should be contacted.

Diagnosis

Itching is a symptom that is obvious to its victim. Because itching can be caused by such a wide variety of triggers, a complete physical examination and medical history will help diagnose the underlying problem.

KEY TERMS

Atopic dermatitis—An intensely itchy inflammation often found on the face, in the bend of the elbow, and behind the knees of people prone to allergies. In infants and young children, this condition is called infantile eczema.

Creeping eruption—Itchy, irregular, wandering red lines on the foot made by burrowing larvae of the hookworm family and some roundworms.

Dermatitis herpetiformis—A chronic, very itchy skin disease with groups of red lesions that leave spots behind when they heal.

Eczema—A superficial type of inflammation of the skin that may be very itchy and weeping in the early stages; later, the affected skin becomes crusted, scaly, and thick.

Hodgkin's disease—One of two general types of lymphoma (cancers that arise in the the lymphatic system and can invade other organs), Hodgkin's disease is characterized by lymph node enlargement and the presence of a large polyploid cells called Reed-Sternberg cells.

Lichen planus—A noncancerous, chronic itchy skin disease that causes small, flat purple plaques on wrists, forearm, ankles.

Neurodermatitis—An itchy skin disease (also called lichen simplex chronicus) found in nervous, anxious people.

Psoriasis—A chronic, noncontagious skin disease that is marked by dry, scaly, and silvery patches of skin that appear in a variety of sizes and locations on the body.

Scabies—A contagious parasitic skin disease caused by a tiny mite and characterized by intense itching.

Swimmer's itch—An allergic skin inflammation caused by a sensitivity to flatworms that die under the skin, resulting in an itchy rash.

A variety of blood and stool tests may be needed to help the doctor to determine the cause of the itch.

Treatment

Antihistamines such as diphenhydramine (Benadryl) can help relieve itching caused by hives but will not relieve itching from other causes. Most antihistamines also make people sleepy, which can help children **sleep** who would otherwise be awakened by the itch.

Specific treatment of itching depends on the underlying condition that causes it. In general, itchy skin should be treated very gently. While scratching may temporarily ease the itch, in the long run scratching makes the itch worse and can lead to an endless cycle in which scratching an itch makes it more itchy.

To avoid the urge to scratch, a cooling or soothing lotion or cold compress can be applied when the urge to scratch occurs. Soaps are often irritating to the skin and can make an itch worse; they should be avoided or used only when necessary.

Creams or ointments containing cortisone may help control the itch from insect bites, contact dermatitis, or eczema. Cortisone cream should not be applied to the face unless prescribed by a doctor.

Probably the most common cause of itching is dry skin. There are a number of simple things that can be done to ease the annoying itch:

- Do not wear tight clothes.
- Avoid synthetic fabrics.
- Take shorter baths.
- Wash the area in lukewarm water with a little baking soda.
- For generalized itching, take a lukewarm shower or oatmeal (or Aveeno) bath.
- Apply bath oil or lotion (without added colors or scents) right after bathing.

Children who itch as a result of mental problems or stress may benefit from seeing a mental health expert.

Prognosis

Most cases of itching resolve successfully when the underlying cause is treated.

Prevention

There are certain things people can do to avoid itchy skin. Children who tend toward itchy skin should take the following steps:

- Avoid a daily bath.
- Use only lukewarm water when bathing.
- Use only gentle soap.
- Pat dry, not rub dry, after bathing, leaving a bit of water on the skin.

- Apply a moisture-holding ointment or cream after the bath.
- Use a humidifier in the home.

Children who are allergic to certain substances, medications, or foods can avoid the resulting itch if they avoid contact with the allergen. Avoiding insect bites, bee **stings**, **poison ivy**, and similar plants can prevent the resulting itch. Treating sensitive skin carefully, avoiding overdrying of the skin, and protecting against diseases that cause itchy **rashes** are all good ways to avoid itching.

Parental concerns

Children who are itchy should have their finger nails cut short to help ensure that they do not scratch the itchy area enough to create breaks in the skin. Scratching until the skin is broken can lead to infection. Itching can be very frustrating, and, if it is severe, it can interfere with normal activities such as studying or sleeping. Itching is a symptom of many common childhood ailments such as **chickenpox** and contact with poison ivy, as well as of some more serious conditions.

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J

Jaundice

Definition

Jaundice is a yellowing of the skin and/or whites of the eyes caused by high levels of bilirubin—a dark yellow-green or orange-red pigment—in the blood.

Description

Jaundice, also called icterus or hyperbilirubinemia, is a very common condition in newborns. Newborn or **neonatal jaundice**, sometimes referred to as physiologic or physiological jaundice, affects more than half of all full-term newborns and 80 percent of premature newborns within the first few days of life. It commonly lasts for one to two weeks. Jaundice that is present at birth or that lasts more than a couple of weeks may be abnormal jaundice and a symptom of an underlying problem. Jaundice in older children or adults is a symptom of hepatitis (inflammation of the liver) or some other liver disorder.

Jaundice results from higher than normal levels of bilirubin in the blood. Bilirubin is a breakdown product of red blood cells. Red blood cells normally are removed and broken down in the spleen after about 120 days in circulation. Heme (component of hemoglobin in red blood cells that carries oxygen throughout the body) is broken down into bilirubin, which moves to the liver where it is processed and added to bile, a digestive fluid. The bile travels through the bile ducts to the intestine and is excreted in the stool.

Infants are born with excess red blood cells that are rapidly recycled by the spleen and liver, releasing bilirubin. This pigment gives a newborn's stools their yellow color. If more bilirubin is produced than can be processed by the liver, blood levels of bilirubin rise, and the excess is deposited in tissues causing the skin to appear yellow.

Demographics

Although jaundice affects the majority of newborns, it often is more severe in Asian or Native American children. It also is more common in infants who are not breastfeeding efficiently, resulting in low fluid intake.

In 2001 the U.S. Centers for Disease Control and Prevention (CDC) reported that cases of brain damage associated with hyperbilirubinemia (called neonatal encephalopathy, bilirubin-induced brain injury, or kernicterus) had been increasing since about 1990, perhaps due to shorter hospital stays following birth. One cause of hyperbilirubinemia in seemingly healthy full-term or near-term infants is **biliary atresia**, an obstruction or inflammation of the bile ducts. This condition occurs in about one in every 15,000 live births, and girls are slightly more at risk than boys.

Causes and symptoms

Neonatal jaundice

Prior to birth the mother's liver processes bilirubin for the fetus. At birth, particularly with preterm births, an infant's immature liver may not be able to process all of the bilirubin formed as red blood cells are removed from circulation. The excess bilirubin causes jaundice by the third or fourth day after birth. The jaundice usually appears first on the face and progresses downward to the chest, abdomen, legs, and feet. If newborn feeding is delayed for any reason, such as illness, a digestive tract problem, or low fluid intake due to inefficient breastfeeding; the infant produces fewer stools, resulting in critically high blood levels of bilirubin and severe jaundice.

Most full-term babies with neonatal jaundice have no other symptoms. However, if bilirubin levels continue to rise, other symptoms may include:

- sleepiness
- lethargy
- slow or reluctant feeding

Risk factors for hyperbilirubinemia include:

- birth more than two weeks before the due date
- jaundice within the first 24 hours after birth
- significant bruising or bleeding under the scalp caused by labor and delivery
- high bilirubin levels prior to hospital discharge
- difficulty breastfeeding, resulting in low fluid intake
- a parent or sibling who had high bilirubin levels at birth

Abnormal jaundice in newborns

Jaundice at birth or within the first 24 hours after birth can be a sign of abnormal jaundice. Abnormal jaundice can be dangerous, particularly in preterm or unhealthy newborns. Depending on the cause and extent of the jaundice, it also may be harmful in full-term infants.

The most common cause of abnormal jaundice is an ABO blood type incompatibility between mother and child. If the mother has O-type blood and the infant has either A or B blood type, or if the mother has A-type blood and the child has B-type or vice versa, the mother's antibodies circulating in the baby's blood attack the child's foreign blood type, causing damage to and destruction of the baby's red blood cells. This process, called hemolysis, is accompanied by the release of excess amounts of bilirubin.

In the past Rhesus (Rh) blood factor incompatibility between the mother and child was a major cause of kernicterus. An Rh-negative mother who was exposed to her fetus's Rh-positive blood during a previous pregnancy or delivery or who has accidentally received an Rh-positive blood transfusion has antibodies against Rh-positive blood cells. These antibodies can circulate in her Rh-positive newborn, initiating hemolysis and causing severe abnormal jaundice.

Rare causes of severe neonatal jaundice

Jaundice can result from a congenital (present at birth) malformation of the liver, bile ducts, or gall bladder. Jaundice resulting from a congenital defect usually does not appear until the baby is at least ten days old. Biliary atresia—the underdevelopment, inflammation, or obstruction of the bile ducts that carry bile from the liver to the gall bladder and small intestine—causes bile to build up in the liver and forces the bilirubin into the blood. The cause of biliary atresia was as of 2004 unknown, and jaundice may not appear until the infant is two to six weeks old. Other symptoms of biliary atresia include:

- itching
- dark brown urine due to excess bilirubin excreted in the urine
- light-gray or chalky-colored stools from lack of bilirubin excreted by the intestines

Jaundice that develops or persists after the second week of life also can be due to the following:

- breast milk jaundice (prolonged jaundice resulting from breastfeeding) that occurs when a chemical in the mother's breast milk interferes with the infant liver's ability to process bilirubin
- liver malfunction or damaged liver cells
- an enzyme deficiency
- an abnormality of the red blood cells such as anemia
- blood hemorrhaging
- a blood infection (sepsis)
- a liver infection such as hepatitis virus
- toxoplasmosis, an infection caused by an animal parasite and transmitted to the fetus via an infected mother (House cats can be carriers of toxoplasmosis.)
- an infection anywhere in the body that impairs the efficiency of the liver, including neonatal **herpes simplex** or salmonella

Such infections may be congenital, having been passed from the mother to the fetus, or may occur after birth.

Other causes of jaundice

There are numerous other causes of neonatal and childhood jaundice, including the following:

- liver cell damage resulting from a variety of conditions such as a viral infection, an adverse drug reaction, or drugs or other chemicals that damage the liver (Jaundice can be a late symptom of hepatitis in an older baby or child.)
- hemolytic jaundice caused by hemolytic anemia, in which red blood cells are turned over faster than usual
- Hodgkin's disease in teenagers

Symptoms accompanying jaundice caused by liver cell damage may include:

- nausea
- vomiting
- abdominal pain
- swollen abdomen

When to call the doctor

A doctor should be consulted any time a child develops jaundice. Infants who are discharged from the hospital before bilirubin levels begin to rise, about three days after birth, should have their bilirubin level tested within a few days, particularly if they were preterm infants. Infants who become lethargic or reluctant to feed should be examined immediately, because symptoms can be signs of severe hyperbilirubinemia that can cause brain damage.

Diagnosis

Newborns are examined under good light for signs of jaundice. A simple blood test, with a few drops of blood taken from the infant's heel, measures bilirubin levels in the blood. The test may be repeated frequently in a jaundiced newborn to assure that bilirubin levels are dropping. An instrument called a bilirubinometer can be held against the baby's skin to assess the level of jaundice. The Minolta/Hill-Rom Air-Shields Transcutaneous Jaundice Meter accurately measures bilirubin levels by shining lights of different colors through the skin and measuring the reflection, eliminating the need for blood tests via heel pricks.

If there is reason to believe that the newborn is suffering from an abnormal jaundice, additional tests must be performed. These include:

- blood cell counts to detect anemia
- tests for blood clotting function
- tests for excess destruction of red blood cells
- blood tests to assess liver function
- a liver biopsy, in which liver cells are removed and examined under a microscope to look for liver disease
- urine and stool samples to check for signs of bacterial or viral infection

Breast milk jaundice due to a reaction with a breast milk component is suspected when the more common causes of jaundice have been ruled out.

Biliary atresia must be detected before two months of age to prevent further liver damage. Diagnoses of biliary atresia and other liver conditions are made by imaging techniques, including the following:

- ultrasound scanning, which uses sound waves to obtain images of the liver, gallbladder, and biliary tract (Abdominal ultrasound can distinguish between jaundice caused by biliary atresia and jaundice caused by liver malfunction.)
- magnetic resonance imaging (MRI) of the liver
- computed tomography (CT) or computed axial tomography (CAT) scans, which use a thin, rotating x-ray beam to obtain an image
- endoscopic retrograde cholangiopancreatography (ERCP), in which a radiopaque dye that is visible on **x rays** is inserted into the upper portion of the small intestine so that it flows back up the biliary tract
- liver scans using radioactive dyes

Treatment

Neonatal jaundice usually requires only observation. The infant may stay in the hospital for an extra day or return within the next few days for an examination. However, jaundice in a preterm baby may require intensive care. As the infant's liver matures and the excess blood cells are removed, the jaundice disappears. The child may be given additional fluids, possibly intravenously, to help remove the bilirubin. Frequent feedings lead to more frequent stools, which reduces the reabsorption of bilirubin from the intestines into the blood. Breast milk usually is considered superior to water or formula for relieving jaundice because breast milk produces stool with every feeding, thereby excreting bilirubin. Breastfeeding should not be discontinued because of neonatal jaundice.

If an infant's bilirubin levels are quite high or rising rapidly, phototherapy can prevent complications. The child is undressed and placed in a lighted incubator to stay warm. A high-intensity, cool, blue-fluorescent light is absorbed by the bilirubin and converts it into a harmless form than can be excreted in the bile and urine. An eye shield protects the baby's eyes. The infant is removed from the incubator for feeding. Other phototherapy methods—such as a fiber optic bilirubin blanket—incorporate the light into a blanket so that the child can be breastfed during treatment or treated at home. Phototherapy is continued until bilirubin levels have returned to normal, usually within a few days.

Side effects of phototherapy may include:

- loose stools
- rash
- dehydration
- sleepiness
- disinterest in breastfeeding

If bilirubin approaches a dangerous level, an exchange blood transfusion is used to rapidly lower it. A catheter is placed into the umbilical vein at the cut surface of the umbilical cord, and the newborn's blood is replaced with an equal volume of new blood. Rh incompatibility also may be treated by exchange transfusion.

Antibiotics may be used to prevent or treat a suspected infection in jaundiced infants. Babies with very severe jaundice have their hearing tested and are monitored for several months.

Surgery for biliary atresia must be performed within the first few weeks of an infant's life to prevent fatal liver damage. About 40–50 percent of infants with biliary atresia are candidates for replacement bile ducts leading from the liver into the intestine. Called the Kasai procedure or hepatoportoenterostomy, the obstructed ducts are replaced with sections from the infant's intestines. Infants with a duct obstruction within the liver itself usually require a liver transplant by the age of two.

Prolonged breast-milk jaundice may require breastfeeding to be halted for a few days until bilirubin levels drop. The breasts should be pumped in the interim so that the mother does not stop producing milk and breastfeeding can be resumed.

Prognosis

Neonatal jaundice disappears after one to two weeks. It may last slightly longer in breastfed infants. The jaundice does not harm the infant in any way, and breastfeeding should not be discontinued.

Severe untreated jaundice leading to kernicterus may result in the following:

- mental retardation
- cerebral palsy
- deafness
- death

Untreated biliary atresia leads to biliary cirrhosis, a progressive, irreversible scarring of the liver, by about two months of age. About 50 percent of bile duct replacement surgeries are successful, and the jaundice usually disappears within several weeks. Despite this success, the liver damage often progresses on to cirrhosis.

Breast-milk jaundice, resulting from a reaction to a breast milk component, is not dangerous. The baby's liver soon adapts to the problem and the jaundice disappears.

Prevention

In 2004 the American Academy of Pediatrics issued revised guidelines for identifying and managing neonatal jaundice. They recommend:

- that all newborns be assessed for their risk of developing severe jaundice, including measuring bilirubin levels before hospital discharge
- a follow-up visit occur within three to five days after birth when bilirubin levels are likely to peak

KEY TERMS

Antibody—A special protein made by the body's immune system as a defense against foreign material (bacteria, viruses, etc.) that enters the body. It is uniquely designed to attack and neutralize the specific antigen that triggered the immune response.

Bile—A bitter yellow-green substance produced by the liver. Bile breaks down fats in the small intestine so that they can be used by the body. It is stored in the gallbladder and passes from the gallbladder through the common bile duct to the top of the small intestine (duodenum) as needed to digest fat.

Bile ducts—Tubes that carry bile, a thick yellow-green fluid that is made by the liver, stored in the gallbladder, and helps the body digest fats.

Biliary atresia—An obstruction or inflammation of a bile duct that causes bilirubin to back up into the liver.

Bilirubin—A reddish yellow pigment formed from the breakdown of red blood cells, and metabolized by the liver. When levels are abnormally high, it causes the yellowish tint to eyes and skin known as jaundice. Levels of bilirubin in the blood increase in patients with liver disease, blockage of the bile ducts, and other conditions.

Hemolysis—The process of breaking down of red blood cells. As the cells are destroyed, hemoglobin, the component of red blood cells which carries the oxygen, is liberated.

Hyperbilirubinemia—A condition characterized by a high level of bilirubin in the blood. Bilirubin is a natural byproduct of the breakdown of red blood cells, however, a high level of bilirubin may indicate a problem with the liver.

Kernicterus—A potentially lethal disease of newborns caused by excessive accumulation of the bile pigment bilirubin in tissues of the central nervous system.

Phototherapy—Another name for light therapy in mainstream medical practice.

- breastfeeding a newborn at least eight to 12 times per day, since effective breastfeeding significantly reduces the risk of hyperbilirubinemia
- that parents be provided with written and oral information about the risks of neonatal jaundice

In cases of known Rh incompatibility, the mother is given an injection of RhoGAM, an immune globulin preparation, at about 28 weeks of pregnancy and again immediately after the child's birth. This destroys any Rh-positive fetal blood cells in the mother's circulation before her immune system can produce antibodies against them.

Parental concerns

Parents should examine their infant in natural daylight and under fluorescent lighting for signs of jaundice. Jaundice may be harder to see in infants with darker skin. However, when a child's nose and forehead are pressed gently, the skin is white in healthy babies of all races, but yellowish if jaundice is present. If the skin appears yellow, the test should be repeated on the chest or abdomen. Parents also should be aware of symptoms that may accompany jaundice, including fussiness, unusual sleepiness, or difficulty feeding.

Mothers who are having difficulty breastfeeding should seek help. Although breast milk is an effective treatment for jaundice, breastfed babies may receive fewer calories than formula-fed babies during the first days of life, causing bilirubin levels to rise.

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Margaret Alic, PhD

Jaundice test *see* **Bilirubin test**

Jock itch *see* **Ringworm**

Juvenile arthritis

Definition

Juvenile arthritis (JA) refers to a number of different conditions, all of which strike children and all of which have joint inflammation as their major manifestation. The condition is also referred to as juvenile rheumatoid arthritis.

Description

The skeletal system of the body is made up of different types of the strong, fibrous tissue known as connective tissue. Bone, cartilage, ligaments, and tendons are all forms of connective tissue which have different compositions and different characteristics.

The joints are structures that hold two or more bones together. Some joints (synovial joints) allow for movement between the bones being joined (called articulating bones). The simplest model of a synovial joint involves two bones, separated by a slight gap called the joint cavity. The ends of each articular bone are covered by a layer of cartilage. Both articular bones and the joint cavity are surrounded by a tough tissue called the articular capsule. The articular capsule has two components: the

fibrous membrane on the outside and the synovial membrane (or synovium) on the inside. The fibrous membrane may include tough bands of fibrous tissue called ligaments, which are responsible for providing support to the joints. The synovial membrane has special cells and many capillaries (tiny blood vessels). This membrane produces a supply of synovial fluid which fills the joint cavity, lubricates it, and helps the articular bones move smoothly about the joint.

In JA, the synovial membrane becomes intensely inflamed. Usually thin and delicate, the synovium becomes thick and stiff, with numerous infoldings on its surface. The membrane becomes invaded by white blood cells, which produce a variety of destructive chemicals. The cartilage along the articular surfaces of the bones may be attacked and destroyed, and the bone, articular capsule, and ligaments may begin to be worn away (eroded). These processes severely interfere with movement in the joint.

JA specifically refers to chronic arthritic conditions which affect a child under the age of 16 years and which last for a minimum of three to six months. JA is often characterized by a waxing and waning course, with flares separated by periods during which no symptoms are noted (remission). Some literature refers to JA as juvenile rheumatoid arthritis, although most types of JA differ significantly from the adult disease called rheumatoid arthritis, in terms of symptoms, progression, and prognosis.

Demographics

Between five and 18 of every 100,000 children develop juvenile rheumatoid arthritis each year; the overall prevalence is approximately 30–150 per 100,000. More than 65,000 young people in the United States develop it each year. It can affect children as young as two years of age.

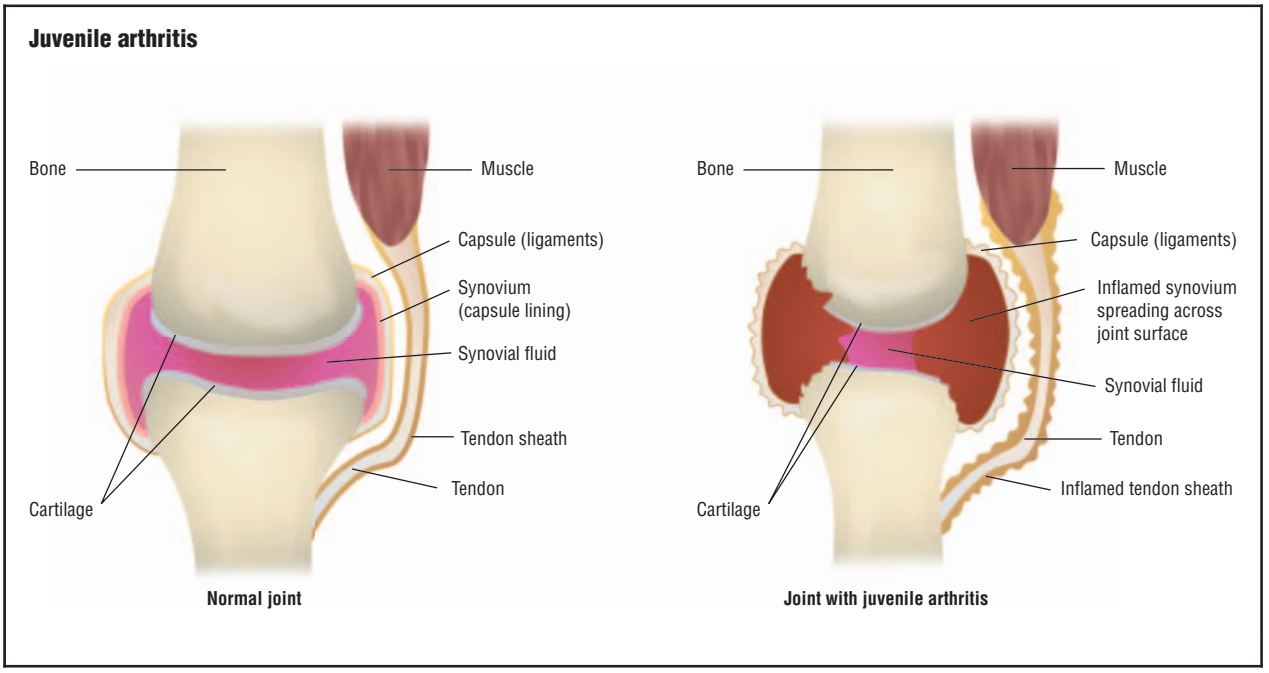
Causes and symptoms

A number of different causes have been sought to explain the onset of JA. There seems to be some genetic link, based on the fact that the tendency to develop JA sometimes runs in a particular **family** and based on the fact that certain genetic markers are more frequently found in patients with JA and other related diseases. Many researchers have looked for some infectious cause for JA, but no clear connection to a particular organism had been made as of 2004. JA is considered by some to be an autoimmune disorder. Autoimmune disorders occur when the body's immune system mistakenly identifies the body's own tissue as foreign and goes about attacking those tissues, as if trying to rid the body of an invader (such as a bacteria, virus, or fungi). While an autoimmune mechanism is strongly suspected, certain markers of such a mechanism (such as rheumatoid factor, often present in adults with such disorders) are rarely present in children with JA.

Joint symptoms of arthritis may include stiffness, **pain**, redness and warmth of the joint, and swelling. Bone in the area of an affected joint may grow too quickly or too slowly resulting in limbs which are of different lengths. When the child tries to avoid moving a painful joint, the muscle may begin to shorten from disuse. This condition is called a contracture.

Symptoms of JA depend on the particular subtype. JA is classified by the symptoms which appear within the first six months of the disorder:

- Pauciarticular JA: The most common and the least severe type of JA affects about 40–60 percent of all JA patients. This type of JA affects fewer than four joints, usually the knee, ankle, wrist, and/or elbow. Other more general (systemic) symptoms are usually absent, and the child's growth usually remains normal. Very few children (fewer than 15 percent) with pauciarticular JA end up with deformed joints. Some children with this form of JA experience painless swelling of the joint. Some children with JA have a serious inflammation of structures within the eye, which if left undiagnosed and untreated could even lead to blindness. While many children have cycles of flares and remissions, in some children the disease completely and permanently resolves within a few years of diagnosis.
- Polyarticular JA: About 40 percent of all cases of JA are of this type. More girls than boys are diagnosed with this form of JA, and it is most common in children up to age three or after the age of ten. Polyarticular JA affects five or more joints simultaneously. This type of JA usually affects the small joints of both hands and both feet, although other large joints may be affected as well. Some patients with arthritis in their knees will experience a different rate of growth in each leg. Ultimately, one leg will grow longer than the other. About half of all patients with polyarticular JA have arthritis of the spine and/or hip. Some patients with polyarticular JA will have other symptoms of a systemic illness, including anemia (low red blood cell count), decreased growth rate, low appetite, low-grade **fever**, and a slight rash. The disease is most severe in those children who are diagnosed in early **adolescence**. Some of these children test positive for a marker present in other autoimmune disorders, called rheumatoid factor (RF), which is found in adults who have rheuma-



Normal knee joint (left) and one affected by juvenile arthritis, which shows damaged cartilage and inflammation of the synovial fluid and tendon sheath. *(Illustration by GGS Information Services.)*

toid arthritis. Children who are positive for RF tend to have a more severe course, with a disabling form of arthritis that destroys and deforms the joints. This type of arthritis is thought to be the adult form of rheumatoid arthritis occurring at a very early age.

- Systemic onset JA: Sometimes called Still disease (after a physician who originally described it), this type of JA occurs in about 10–20 percent off all patients with JA. Boys and girls are equally affected, and diagnosis is usually made between the ages of five and 10 years. The initial symptoms are not usually related to the joints. Instead, these children have high fevers; a rash; decreased appetite and weight loss; severe joint and muscle pain; swollen lymph nodes, spleen, and liver; and serious anemia. Some children experience other complications, including inflammation of the sac containing the heart (pericarditis); inflammation of the tissue lining the chest cavity and lungs (pleuritis); and inflammation of the heart muscle (myocarditis). The eye inflammation often seen in pauciarticular JA is uncommon in systemic onset JA. Symptoms of actual arthritis begin later in the course of systemic onset JA, and they often involve the wrists and ankles. Many of these children continue to have periodic flares of fever and systemic symptoms throughout childhood. Some children go on to develop a polyarticular type of JA.
- Spondyloarthropathy: This type of JA most commonly affects boys older than eight years of age. The arthritis occurs in the knees and ankles, moving over time to include the hips and lower spine. Inflammation of the eye may occur occasionally but usually resolves without permanent damage.
- Psoriatic JA: This type of arthritis usually shows up in fewer than four joints but goes on to include multiple joints (appearing similar to polyarticular JA). Hips, back, fingers, and toes are frequently affected. A skin condition called **psoriasis** accompanies this type of arthritis. Children with this type of JA often have pits or ridges in their fingernails. The arthritis usually progresses to become a serious, disabling problem.

When to call the doctor

A pediatrician, family physician, or other primary care doctor frequently manages the treatment of a child with JA, often with the help of other doctors. Depending on the patient's and parents' wishes and the severity of the disease, the team of doctors may include pediatric rheumatologists (doctors specializing in childhood arthritis), ophthalmologists (eye doctors), orthopaedic surgeons (bone specialists), and physiatrists (rehabilitation specialists), as well as physical and occupational therapists. The main goals of treatment are to preserve a high level of physical and social functioning and

maintain a good quality of life. To achieve these goals, doctors recommend treatments to reduce swelling; maintain full movement in the affected joints; relieve pain; and identify, treat, and prevent complications. Most children with JA need medication and physical therapy to reach these goals.

Diagnosis

Diagnosis of JA is often made on the basis of the child's collection of symptoms. Laboratory tests often show normal results. Some nonspecific indicators of inflammation may be elevated, including white blood cell count, erythrocyte sedimentation rate, and a marker called C-reactive protein. As with any chronic disease, anemia may be noted. Children with an extraordinarily early onset of the adult type of rheumatoid arthritis have a positive test for rheumatoid factor.

Treatment

Treating JA involves efforts to decrease the amount of inflammation in order to preserve movement. Medications which can be used for this include nonsteroidal anti-inflammatory agents (such as ibuprofen and naproxen). Oral (by mouth) steroid medications are effective but have many serious side effects with long-term use. Injections of steroids into an affected joint can be helpful. Steroid eye drops are used to treat eye inflammation. Other drugs that have been used to treat JA include methotrexate, sulfasalazine, penicillamine, and hydroxychloroquine. Physical therapy and exercises are often recommended in order to improve joint mobility and to strengthen supporting muscles. Occasionally, splints are used to rest painful joints and to try to prevent or reduce deformities.

Alternative treatment

Alternative treatments that have been suggested for arthritis include juice therapy, which can work to detoxify the body, helping to reduce JA symptoms. Some recommended fruits and vegetables to include in the juice are carrots, celery, cabbage, potatoes, cherries, lemons, beets, cucumbers, radishes, and garlic. However, for children with osteoarthritis, citrus fruits and vegetables from the nightshade family, including potatoes, tomatoes, peppers, and eggplant, should be avoided since they can promote swelling. As an adjunct therapy, aromatherapy preparations use cypress, fennel, and lemon. Massage oils include rosemary, benzoin, chamomile, camphor, juniper, and lavender. Other types of therapy that have been used for JA include acupuncture, acupressure, and bodywork. Nutritional supplements that may be beneficial include large amounts of antioxidants (**vitamins** C, A, E, zinc, selenium, and flavenoids), as well as B vitamins and a full complement of **minerals** (including boron, copper, and manganese). Other nutrients that assist in detoxifying the body, including methionine, cysteine, and other amino acids, may also be helpful. A number of autoimmune disorders, including JA, seem to have a relationship to **food allergies**. Identification and elimination of reactive foods may result in a decrease in JA symptoms. Constitutional homeopathy can also work to quiet the symptoms of JA and bring about balance to the whole person.

A young girl with juvenile arthritis uses braces to help rest painful joints and prevent deformities. *(© John Moss/Photo Researchers, Inc.)*

Nutritional concerns

When female patients were compared to age-matched normals, greater than one serving per week of grilled (broiled) or baked fish (other than tuna) was

associated with a decreased risk of disease. This inverse association was stronger than that for omega-3 fatty acid intake alone. In another study, patients with active disease randomly received treatment with or without a diet in which the energy consumption was adjusted to normal standards for body-weight, and the intake of fish and antioxidants was increased. After six months, those following the diet demonstrated significant improvement in the duration of morning stiffness, the number of swollen joints, and pain status.

Several studies, some of which were controlled, have found beneficial effects from a vegetarian or vegan diet. In one study, for example, patients randomly received either an uncooked vegan diet or a control diet for three months. Those on the vegan diet experienced subjective relief of rheumatic stiffness and joint swellings and an improvement in general wellbeing. After returning to the meat diet, these symptoms worsened. The degree of tender joints remained unchanged in controls, but in the diet group their number decreased significantly.

Prognosis

The prognosis for pauciarticular JA is quite good, as is the prognosis for spondyloarthropathy. Polyarticular JA carries a slightly worse prognosis. RF-positive polyarticular JA carries a difficult prognosis, often with progressive, destructive arthritis and joint deformities. Systemic onset JA has a variable prognosis, depending on the organ systems affected and the progression to polyarticular JA. About 1–5 percent of all JA patients die of such complications as infection, inflammation of the heart, or kidney disease.

Prevention

Because so little is known about what causes JA, there are no recommendations available for ways to avoid developing it.

Parental concerns

JA affects the entire family who must cope with the special challenges of this disease. JA can strain a child's participation in social and after-school activities and make schoolwork more difficult. There are several things that family members can do to help the child do well physically and emotionally. These include the following:

- Ensure that the child receives appropriate medical care and follows the doctor's instructions. Many treatment options are available, and because JA is different in each child, what works for one may not work for another. If the medications that the doctor prescribes do not relieve symptoms or if they cause unpleasant side effects, patients and parents should discuss other choices with their doctor. A person with JA can be more active when symptoms are controlled.
- Encourage **exercise** and physical therapy for the child. For many young people, exercise and physical therapy play important roles in treating JA. Parents can arrange for children to participate in activities that the doctor recommends. During symptom-free periods, many doctors suggest playing team **sports** or doing other activities to help keep the joints strong and flexible and to provide play time with other children and encourage appropriate social development.
- Work closely with the school to develop a suitable lesson plan for the child and to educate the teacher and the child's classmates about JA. Some children with JA may be absent from school for prolonged periods and need to have the teacher send assignments home. Some minor changes such as an extra set of books or leaving class a few minutes early to get to the next class on time can be a great help. With proper attention, most children progress normally through school.
- Treat the child as normally as possible. Explain to the child that getting JA is nobody's fault. Some children believe that JA is a punishment for something they did. Consider joining a support group. The American Juvenile Arthritis Organization runs support groups for people with JA and their families. Support group meetings provide the chance to talk to other young people and parents of children with JA and may help a child and the family cope with the condition.

KEY TERMS

Articular bones—Two or more bones that are connected to each other via a joint.

Joint—The connection point where two bones meet.

Synovial joint—A fully moveable joint in which a synovial cavity is present between two articulating bones. Also called a diarthrosis.

Synovial membrane—The membrane that lines the inside of the articular capsule of a joint, and produces a lubricating fluid called synovial fluid.

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K

Kawasaki syndrome

Definition

Kawasaki syndrome is a potentially fatal inflammatory disease that affects several organ systems in the body, including the heart, circulatory system, mucous membranes, skin, and immune system. As of 2004 its cause was unknown.

Description

In the 1960s, Tomisaku Kawasaki noted a characteristic cluster of symptoms in Japanese schoolchildren. Ultimately named for Kawasaki, the disorder was subsequently found worldwide. Kawasaki syndrome, also called mucocutaneous lymph node syndrome (MLNS), is an inflammatory disorder with potentially fatal complications affecting the heart and its larger arteries.

Demographics

Kawasaki syndrome occurs primarily in infants and children; about 80 percent of diagnosed patients are under the age of five. On rare occasions, the disorder has been diagnosed in teenagers or adults. Nearly twice as many males are affected as females. Although persons of Asian descent are affected more frequently than either black or white individuals, there does not appear to be a distinctive geographic pattern of occurrence. Although the disease usually appears in individuals, it sometimes affects several members of the same **family** and occasionally occurs in small epidemics. About 3,000 cases are diagnosed annually in the United States.

Causes and symptoms

The specific cause of Kawasaki syndrome was as of 2004 unknown, although the disease resembles an infectious illness in many ways. It has been suggested that Kawasaki syndrome represents an allergic reaction or other unusual response to certain types of infections. Some researchers think that the syndrome may be caused by the interaction of an immune cell, called the T cell, with certain poisons (toxins) secreted by bacteria.

Kawasaki syndrome has an abrupt onset, with **fever** as high as 104°F (40°C) and a rash that spreads over the patient's chest and genital area. The fever is followed by a characteristic peeling of the skin beginning at the fingertips and toenails. In addition to the body rash, the patient's lips become very red, with the tongue developing a "strawberry" appearance. The palms, soles, and mucous membranes that line the eyelids and cover the exposed portion of the eyeball (conjuntivae) become purplish-red and swollen. The lymph nodes in the patient's neck may also become swollen. These symptoms may last from two weeks to three months, with relapses in some patients.

In addition to the major symptoms, about 30 percent of patients develop joint **pain** or arthritis, usually in the large joints of the body. Others develop **pneumonia**, **diarrhea**, dry or cracked lips, **jaundice**, or an inflammation of the membranes covering the brain and spinal cord (**meningitis**). A few patients develop symptoms of inflammation in the liver (hepatitis), gallbladder, lungs, or tonsils.

About 20 percent of patients with Kawasaki syndrome develop complications of the cardiovascular system. These complications include inflammation of the heart tissue (myocarditis), disturbances in heartbeat rhythm (arrhythmias), and areas of blood vessel dilation (aneurysms) in the coronary arteries. Other patients may develop inflammation of an artery (arteritis) in their arms or legs. Complications of the heart or arteries begin to develop around the tenth day after the illness begins, when the fever and rash begin to subside. A few patients may develop gangrene (the death of soft tissue) in their hands and feet. The specific causes of these complications were as of 2004 not known.

Diagnosis

Because Kawasaki syndrome is primarily a disease of infants and young children, the disease is most likely to be diagnosed by a pediatrician. The physician will first consider the possible involvement of other diseases that cause fever and skin **rashes**, including **scarlet fever**, **measles**, **Rocky Mountain spotted fever**, **toxoplasmosis** (a disease carried by cats), juvenile rheumatoid arthritis, and a blistering and inflammation of the skin caused by reactions to certain medications (Stevens-Johnson syndrome).

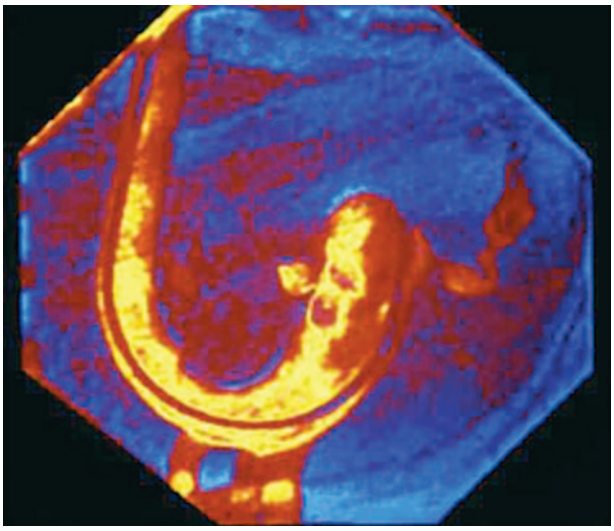
Once other diseases have been ruled out, the patient's symptoms will be compared with a set of diagnostic criteria. The patient must have a fever lasting five days or longer that does not respond to **antibiotics**, together with four of the following five symptoms:

- inflammation of the conjunctivae of both eyes with no discharge
- at least one of the following changes in the mucous membranes of the mouth and throat: "strawberry" tongue, cracked lips, or swollen throat tissues
- at least one of the following changes in the hands or feet: swelling caused by excess fluid in the tissues, peeling of the skin, or abnormal redness of the skin
- a skin eruption or rash associated with fever (exanthem) on the patient's trunk
- swelling of the lymph nodes in the neck to a size greater than 1.5 cm

Given the unknown cause of this syndrome, there are no laboratory tests that can confirm the diagnosis. The following test results, however, are associated with the disease:

- Blood tests show a high white blood cell count, high **platelet count**, a high level of protein in the blood serum, and mild anemia.
- A chest x ray may show enlargement of the heart (cardiomegaly).
- Urine may show the presence of pus or an abnormally high level of protein.
- An electrocardiogram may show changes in the heartbeat rhythm.

In addition to these tests, it is important to take a series of echocardiograms during the course of the illness because 20 percent of Kawasaki patients develop coronary aneurysms or arteritis that will not appear during the first examination.



An angiogram showing abnormal coronary arteries in a child suffering from Kawasaki's disease. The coronary arteries bulge into balloon shapes, called aneurysms, along their lengths. *(Photograph by Mehau Kulyk. Photo Researchers, Inc.)*

Treatment

Kawasaki syndrome is usually treated with a combination of aspirin, to control the patient's fever and skin inflammation, and high doses of intravenous immune globulin to reduce the possibility of coronary artery complications. Some patients with heart complications may be treated with drugs that reduce blood clotting or may receive corrective surgery.

Follow-up care includes two to three months of monitoring with chest **x rays**, electrocardiography, and echocardiography. Treatment with aspirin is often continued for several months.

Prognosis

Most patients with Kawasaki syndrome will recover completely, but about 1–2 percent die as a result of blood clots forming in the coronary arteries or as a result of a heart attack. Deaths are sudden and unpredictable. Almost 95 percent of fatalities occur within six months of infection, but some have been reported as long as 10 years afterward. Long-term follow-up of patients with aneurysms indicates that about half show some healing of the aneurysm. The remaining half has a high risk of heart complications in later life.

Parental concerns

It is important that parents of children diagnosed with Kawasaki syndrome follow recommended treatments and follow-up care for the disease, because of the

KEY TERMS

Aneurysm—A weakened area in the wall of a blood vessel which causes an outpouching or bulge. Aneurysms may be fatal if these weak areas burst, resulting in uncontrollable bleeding.

Arrhythmia—Any deviation from a normal heart beat.

Arteritis—Inflammation of an artery.

Cardiomegaly—An enlarged heart.

Conjunctiva—Plural, conjunctivae. The mucous membrane that covers the white part of the eyes (sclera) and lines the eyelids.

Exanthem—A skin eruption associated with a disease, usually one accompanied by fever as in Kawasaki syndrome.

Gangrene—Decay or death of body tissue because the blood supply is cut off. Tissues that have died in this way must be surgically removed.

Hepatitis—An inflammation of the liver, with accompanying liver cell damage or cell death, caused most frequently by viral infection, but also by certain drugs, chemicals, or poisons. May be either acute (of limited duration) or chronic (continuing). Symptoms include jaundice, nausea, vomiting, loss of appetite, tenderness in the right upper abdomen, aching muscles, and joint pain. In severe cases, liver failure may result.

Meningitis—An infection or inflammation of the membranes that cover the brain and spinal cord. It is usually caused by bacteria or a virus.

Mucocutaneous lymph node syndrome (MLNS)—Another name for Kawasaki syndrome. The name comes from the key symptoms of the disease, which involve the mucous membranes of the mouth and throat, the skin, and the lymph nodes. MLNS is a potentially fatal inflammatory disease of unknown cause.

Myocarditis—Inflammation of the heart muscle (myocardium).

Stevens-Johnson syndrome—A severe inflammatory skin eruption that occurs as a result of an allergic reaction or respiratory infection.

T cell—A type of white blood cell that is produced in the bone marrow and matured in the thymus gland. It helps to regulate the immune system's response to infections or malignancy.

risk of potentially serious complications. Any worsening or unexplained new symptoms should be reported to the treating pediatrician.

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Klinefelter syndrome

Definition

Klinefelter syndrome is a chromosome disorder in males that results in **hypogonadism** (small penis and small firm testicles). People with this condition are born with at least one extra X chromosome.

Description

Klinefelter syndrome is a condition where one or more extra X-chromosomes are present in a male. Boys with this condition appear normal at birth, but the defect becomes apparent in **puberty** when secondary sexual characteristics fail to develop, or develop later. By mid-puberty, boys with Klinefelter syndrome have low levels of testosterone, resulting in small testicles and the inability to make sperm. Affected males may also have learning disabilities and behavior problems such as **shyness** and immaturity and at an increased risk for certain other health problems such as pulmonary disease, varicose veins, breast **cancer**, extragonadal germ cell tumor (a rare tumor), and osteoporosis. Some mild cases may be

undetected, with no abnormalities present except infertility.

Demographics

Klinefelter syndrome is one of the most common chromosomal abnormalities. About 1 in every 500 to 800 males is born with this disorder. Approximately 3% of the infertile male population have Klinefelter syndrome.

Causes and symptoms

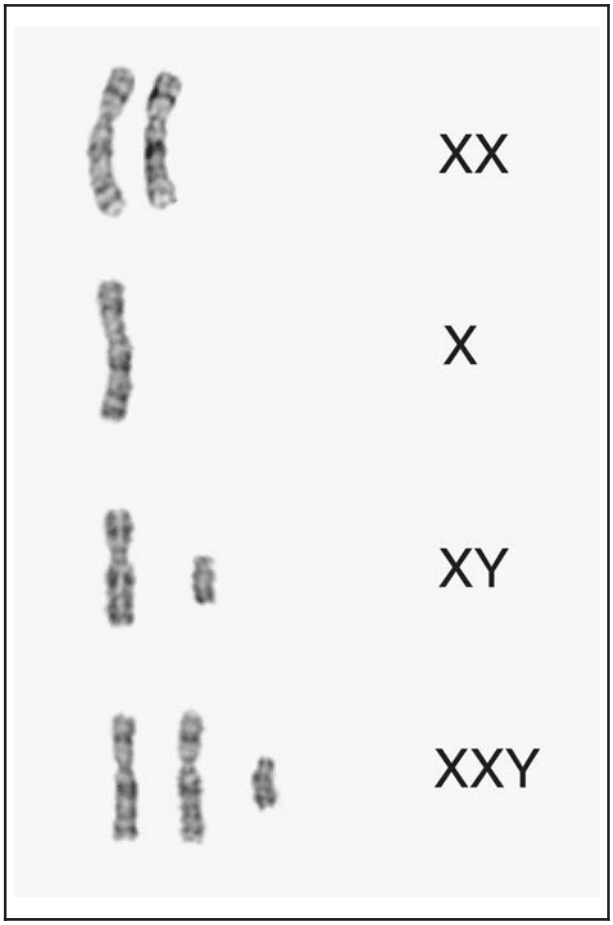
Normally, a person has a total of 46 chromosomes in each cell, two of which are responsible for determining that individual's sex. These two sex chromosomes are called X and Y. The combination of these two types of chromosomes determines the sex of a child. Females have two X chromosomes (the XX combination); males have one X and one Y chromosome (the XY combination).

In Klinefelter syndrome, a problem very early in fetal development results in an abnormal number of chromosomes. Most commonly, a male with Klinefelter syndrome will be born with 47 chromosomes in each cell, rather than the normal number of 46. The extra chromosome is an X chromosome. This means that rather than having the normal XY combination, the male has an XXY combination. Because people with Klinefelter syndrome have a Y chromosome, they are all male.

Approximately 1/3 of all males with Klinefelter syndrome have other chromosome changes involving an extra X chromosome. Mosaic Klinefelter syndrome occurs when some of the cells in the body have an extra X chromosome and the other have normal male chromosomes. These males can have the same or milder symptoms than those with non-mosaic Klinefelter syndrome. Males with more than one additional extra X chromosome, such as 48,XXXY, are usually more severely affected than males with 47,XXY.

Klinefelter syndrome is not considered an inherited condition. The risk of Klinefelter syndrome reoccurring in another pregnancy for a woman who had a son with Klinefelter syndrome is not increased above the risk of the general population.

The symptoms of Klinefelter syndrome are variable; not every affected person will have all of the features of the condition. Males with Klinefelter syndrome appear normal at birth and have normal male genitalia. From childhood, males with Klinefelter syndrome are taller than average with long limbs. Approximately 20–50% have a mild intention tremor, an uncontrolled shaking. Many males with Klinefelter syndrome have poor upper body strength and can be clumsy. Klinefelter syndrome does not cause **homosexuality**. Approximately 1/3 of males with Klinefelter syndrome have breast growth, some requiring breast reduction surgery.



Sex chromosomes showing different phenotypes: XX–normal female; X–female with Turner syndrome; XY–normal male; and XXY–male with Klinefelter. *(Lerner & Lerner, LLC.)*

Most boys enter puberty normally, though some can be delayed. The Leydig cells in the testicles usually produce testosterone. With Klinefelter syndrome, the Leydig cells fail to work properly causing the testosterone production to slow. By mid-puberty, testosterone production is decreased to approximately half of normal. This can lead to decreased facial and pubic hair growth. The decreased testosterone also causes an increase in two other hormones, follicle stimulating hormone (FSH) and luteinizing hormone (LH). Normally, FSH and LH help the immature sperm cells grown and develop. In Klinefelter syndrome, there are few or no sperm cells. The increased amount of FSH and LH cause

hyalinization and fibrosis, the growth of excess fibrous tissue, in the seminiferous tubules, where the sperm are normally located. As a result, the testicles appear smaller and firmer than normal. With rare exception, men with Klinefelter syndrome are infertile because they can not make sperm.

While it was once believed that all boys with Klinefelter syndrome were mentally retarded, doctors now know that the disorder can exist without retardation. However, children with Klinefelter syndrome frequently have difficulty with language, including learning to speak, read, and write. Approximately 50% of males with Klinefelter syndrome are dyslexic. They may also have attention deficient hyperactivity disorder.

Some people with Klinefelter syndrome have difficulty with social skills and tend to be more shy, anxious, or immature than their peers. They can also have poor judgement and do not handle stressful situations well. As a result, they often do not feel comfortable in large social gatherings. Some people with Klinefelter syndrome can also have **anxiety**, nervousness and/or depression.

Taurodontism, an enlargement of the pulp of the teeth with surface thinning, is very common in Klinefelter syndrome and can be diagnosed with dental x rays.

The greater the number of X chromosomes present, the greater the disability. Boys with several extra X chromosomes have distinctive facial features, more severe retardation, deformities of bony structures, and even more disordered development of male features.

When to call the doctor

Parents should call a doctor if their son fails to develop secondary sexual characteristics at puberty.

Diagnosis

During a physical examination, a doctor will look for a simian crease (a single crease in the palm). A rectal exam may show an enlarged prostate. A single testicle may be present in the scrotum, indicating a probable undescended testicle. A semen examination will show low sperm count, while other tests will show decreased serum testosterone levels, increased levels of serum luteining and serum follicle stimulating hormones and increased serum estradiol levels (a type of estrogen).

Diagnosis of Klinefelter syndrome is confirmed by examining chromosomes for evidence of more than one X chromosome present in a male. This can be done in pregnancy with prenatal testing such as chorionic villus sampling or **amniocentesis**. Chorionic villus sampling is a procedure done early in pregnancy (approximately 10–12 weeks) to obtain a small sample of the placenta for testing. An amniocentesis is done further along in pregnancy (from approximately 16–18 weeks) to obtain a sample of fluid surrounding the baby for testing. Both procedures have a risk of miscarriage. Usually these procedures are done for a reason other than diagnosing Klinefelter syndrome. For example, a prenatal diagnostic procedure may be done on an older woman to determine if her baby has **Down syndrome**. If the diagnosis of Klinefelter syndrome is suspected in a young boy or adult male, chromosome testing can also be on a small blood or skin sample after birth.

Treatment

There is no treatment available to change chromosomal makeup. Children with Klinefelter syndrome may benefit from a speech therapist for speech problems or other educational intervention for learning disabilities. Testosterone injections started around the time of puberty and continued for life may help to produce more normal development, including more muscle mass, hair growth and increased sex drive. Testosterone supplementation will not increase testicular size, decrease breast growth or correct infertility.

Prognosis

While many men with Klinefelter syndrome live normal lives, nearly 100 percent of these men will be sterile (unable to produce a child). However, a few men with Klinefelter syndrome have been reported who have fathered a child through the use of assisted fertility services. Males with Klinefelter syndrome have an increased risk of several conditions such as osteoporosis, pulmonary disease, varicose veins, autoimmune disorders such as lupus and arthritis, diabetes, breast cancer and extragonadal germ cell tumor (a rare tumor).

Prevention

Klinefelter syndrome usually is not inherited but occurs during fetal development, so there is no means of preventing the disease. However, one risk factor for this condition is the mother giving birth at an older age; therefore, genetic counseling and testing is recommended.

Parental concerns

Families may wish to seek counseling regarding the effects of the syndrome on relationships within the **family**. Many people respond with guilt, **fear**, or

KEY TERMS

Follicle-stimulating hormone (FSH)—A pituitary hormone that in females stimulates the ovary to mature egg capsules (follicles) and in males stimulates sperm production.

Luteinizing hormone—A hormone secreted by the pituitary gland that regulates the menstrual cycle and triggers ovulation in females. In males it stimulates the testes to produce testosterone.

Testosterone—Male hormone produced by the testes and (in small amounts) in the ovaries. Testosterone is responsible for some masculine secondary sex characteristics such as growth of body hair and deepening voice. It also is sometimes given as part of hormone replacement therapy to women whose ovaries have been removed.

blame when a genetic disorder is diagnosed in the family, or they may overprotect the affected member. Support groups are often good sources of information about Klinefelter syndrome; they can offer helpful suggestions about living with it as well as emotional support.

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American Association for Klinefelter Syndrome Information and Support. 2945 West Farwell Avenue, Chicago, IL 60645-2925. (773) 761-5298, (800) 466-5747. <http://www.aaksis.org>

Klinefelter's Organisation. 234 Turton Road, Bolton, BL2 3EE, United Kingdom. <http://www.klinefelter.org.uk/>

Klinefelter Syndrome and Associates, Inc. PO Box 119, Roseville, CA 95678-0119. (916) 773-2999 or (888) 999-9428. Fax: (916) 773-1449. <http://www.genetic.org/ks>.

WEB SITES

Klinefelter Syndrome Support Group Home Page. <http://klinefeltersyndrome.org/index.html>

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Carin Lea Beltz, MS

Kwashiorkor *see* **Protein-energy malnutrition**

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INFANCY THROUGH ADOLESCENCE

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Labor and delivery *see* **Childbirth**

Labyrinthitis

Definition

Labyrinthitis is an inflammation of the inner ear that is often a complication of infection of the middle ear (**otitis media**). It is usually caused by the spread of bacterial or viral infections from the head or respiratory tract into the inner ear.

Description

The labyrinth is a group of interconnected canals chambers located in the inner ear. It is made up of the cochlea and the semicircular canals. The cochlea is involved in transmitting sounds to the brain. The semicircular canals send information to the brain about the head's position and how it is moving. The brain uses this information to maintain balance. Labyrinthitis is caused by the inflammation of the labyrinth. Its most frequent symptom is vertigo (**dizziness**), because the information that the semicircular canals send to the brain about the position of the head is affected.

Demographics

Labyrinthitis is rare and is more likely to occur after middle ear infections, **meningitis**, or upper respiratory infection. It may also occur after trauma, because of a tumor, or after the ingesting of toxic substances. It is thought to be more common in females than in males.

Causes and symptoms

When a disease agent causes labyrinthitis, the disease agent usually reaches the inner ear by one of three routes:

- Bacteria may be carried from the middle ear or the membranes that cover the brain.
- Viruses, such as those that cause **mumps**, **measles**, **influenza**, and colds may reach the inner ear following an upper respiratory infection.
- The **rubella** virus can cause labyrinthitis in infants prior to birth.

Labyrinthitis can also be caused by toxins, by a tumor in the ear, by trauma to the ear, and sometimes high doses of medications or **allergies**.

The primary symptoms of labyrinthitis are vertigo and hearing loss, along with a sensation of ringing in the ears called tinnitus. Vertigo occurs because the inner ear controls the sense of balance, as well as hearing. Some individuals also experience **nausea and vomiting** and spontaneous eye movements in the direction of the unaffected ear. Bacterial labyrinthitis may produce a discharge from the infected ear.

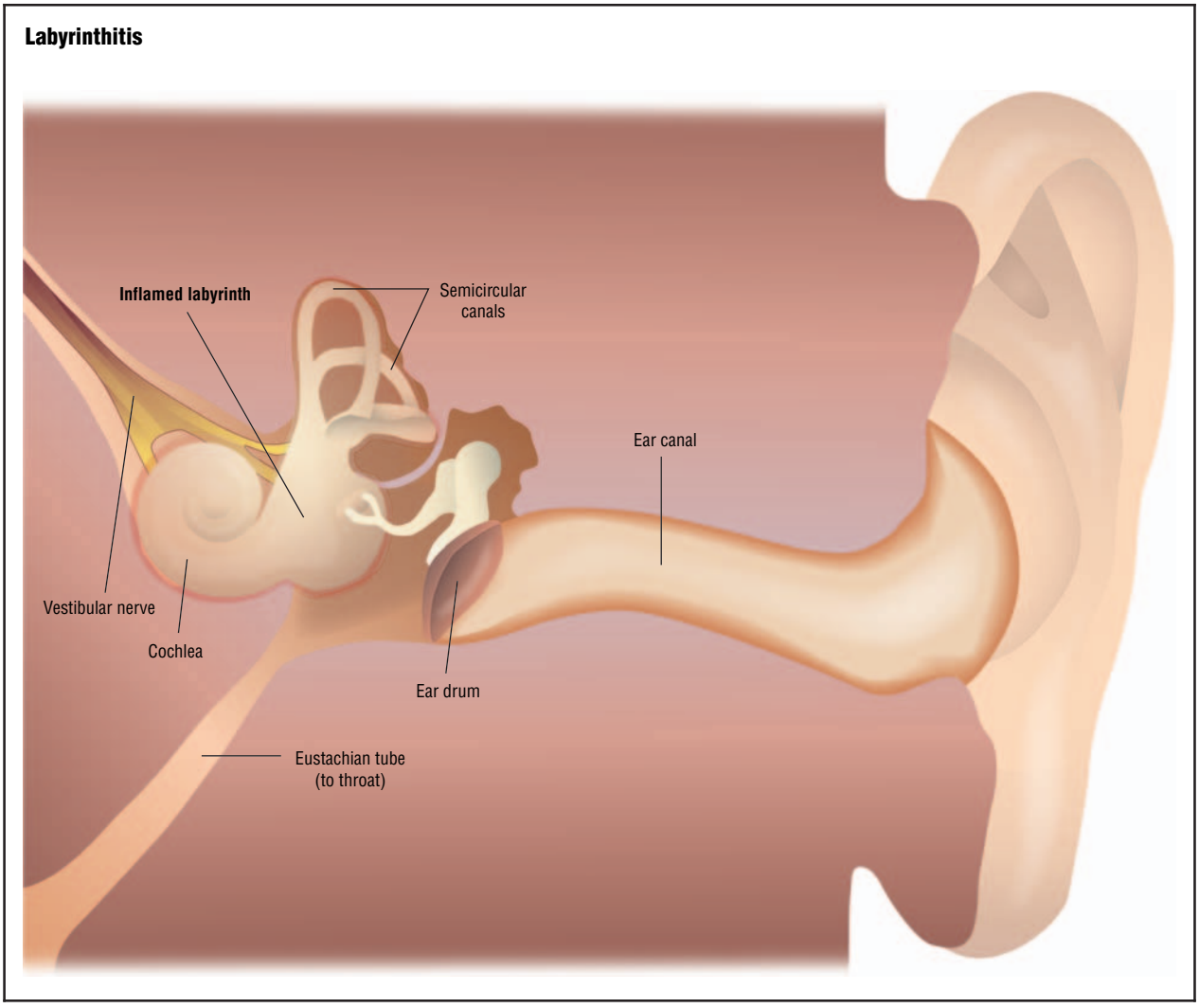
When to call the doctor

If a child has vertigo, especially along with **nausea**, **vomiting**, or hearing loss, the doctor should be called.

Diagnosis

Diagnosis of labyrinthitis is based on a combination of the individual's symptoms and history, especially a history of a recent upper respiratory infection. The doctor will test the child's hearing and order a laboratory culture to identify the organism if the patient has a discharge.

If there is no history of a recent infection, the doctor will order tests such as a commuted topography (CT) scan or a **magnetic resonance imaging** (MRI) scan to help rule out other possible causes of vertigo, such as tumors. If it is believed a bacterium is causing the labyrinthitis, blood tests may be done, or any fluid draining from the ear may be analyzed to help determine what type of bacteria is present.



Labyrinthitis, or inner ear infection, causes the labyrinth area of the ear to become inflamed. *(Illustration by GGS Information Services.)*

Treatment

If a bacterial agent is found to be the cause, the individual is given **antibiotics** to clear up the infection. Antibiotics cannot cure viral infections. Some patients may require surgery to drain the inner and middle ear. If an underlying condition such as a tumor is found to be the cause of the labyrinthitis, treatment will depend on the underlying condition.

Because most labyrinthitis resolves on its own, most treatment is focused on controlling the symptoms. Medications may be prescribed to help reduce vertigo and nausea. If vomiting cannot be controlled, so that fluids cannot be kept down, fluids may be administered intravenously to prevent **dehydration**.

Individuals with labyrinthitis should rest in bed until the acute dizziness subsides. Some experts believe that recovery is aided by moving around once the most acute symptoms are no longer present. This can be difficult, however, because moving often makes symptoms worse.

Prognosis

Most people who have labyrinthitis recover completely, although it often takes five to six weeks for the vertigo to disappear entirely and the individual's hearing to return to normal. In a few cases, the hearing loss may be permanent. Permanent hearing loss is more common in cases of labyrinthitis that are caused by bacteria. For

KEY TERMS

Labyrinth—The bony cavity of the inner ear.

Otitis media—Inflammation or infection of the middle ear space behind the eardrum. It commonly occurs in early childhood and is characterized by ear pain, fever, and hearing problems.

Vertigo—A feeling of dizziness together with a sensation of movement and a feeling of rotating in space.

some individuals, episodes of dizziness may still occur months after the main episode is over.

Prevention

The most effective preventive strategy includes prompt treatment of middle ear infections, as well as monitoring of patients with mumps, measles, influenza, or colds for signs of dizziness or hearing problems.

Parental concerns

Labyrinthitis generally resolves by itself; however, in some cases permanent hearing loss can result. Labyrinthitis may cause repeated episodes of vertigo even after the main symptoms have gone away. If the episodes occur when the head is moved suddenly, this can make it difficult for a child to engage in some physical activities or **sports**.

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American Academy of Pediatrics. 141 Northwest Point Boulevard, Elk Grove Village, IL 60007–1098. Web site: <www.aap.org>.

Helen Davidson

Lactation

Definition

Lactation refers to the formation of milk in the breasts during the period following **childbirth**. Breastfeeding is the process of the infant obtaining milk by suckling at the breast.

Description

Although **breast development** begins around **puberty**, development of mammary function is only completed in pregnancy. During the first half of pregnancy the mammary ducts proliferate and group together to form large lobules. During the second half of pregnancy, secretory activity increases and the alveoli become distended by accumulating colostrum. After 16 weeks of pregnancy, lactation occurs even if the pregnancy does not progress.

The ability of the mammary gland to secrete milk during later pregnancy is called lactogenesis, stage 1. During this time, breast size increases and fat droplets accumulate in the secretory cells. The onset of copious milk secretions after birth is lactogenesis, stage 2, and usually occurs from day two or three to eight days postpartum. During this time, the milk goes through a maturation process to match the infant's needs. Without the hormone prolactin, lactation would not occur. During pregnancy prolactin helps to increase breast mass but does not cause lactation because it is inhibited by the hormone progesterone, which is made by the placenta. The inhibiting influence of progesterone is so strong that lactation is delayed if any of the placenta is retained after birth. Prolactin levels rise and fall in direct proportion to the frequency, intensity, and duration of nipple stimulation from the infant's suckling. During the first week after birth, prolactin levels in breastfeeding women fall about 50 percent. If a mother does not breastfeed,

prolactin levels usually reach the levels of the nonpregnant state by seven days postpartum. After milk "comes in" or rapidly increases in volume, lactation is no longer driven by the hormone prolactin. It shifts control to a milk removal driven process, i.e., sucking stimulus. Thus, the initiation of lactation is not driven by breastfeeding, but breastfeeding is necessary for the continuation of lactation.

The breast is not a passive container of milk. It is an organ that actively produces milk due to the stimulus of the infant's sucking; the removal of milk from the breasts causes continued milk production. It is a supply and demand response that regulates the production of milk to match the intake of the infant. The composition of breast milk changes to meet the specific needs of the growing infant. In response to suckling, the hormone oxytocin causes the milk ejection reflex or "let-down" reflex to occur. Milk ejection is the forceful expulsion of milk from the alveoli openings. Oxytocin secretion is also nature's way of causing a woman's uterus to contract after birth to control postpartum bleeding and assist in uterine involution. These contractions can continue for up to 20 minutes after feeding and may be painful during the first few days. The benefit of this, however, is that uterine discharge diminishes faster and the uterine involution occurs more quickly.

Colostrum is thick and creamy yellow as compared with mature milk, which is thin and bluish-white. Compared with mature milk, colostrum is richer in protein and **minerals** and lower in carbohydrates, fat, and some **vitamins**. The high concentration of total protein and minerals in colostrum gradually changes to meet the infant's needs over the first two to three weeks until lactation is established. The key component in colostrum and breast milk is immunoglobulins or antibodies that serve to protect the infant against infections or viruses. Breast milk also facilitates the development of the infant's own immune system to mature faster. As a result, breast-fed babies have fewer ear infections, **diarrhea**, **rashes**, **allergies**, and other medical problems than bottle-fed babies. Human milk is rich in proteins, lipids, carbohydrates, vitamins, minerals, hormones, enzymes, growth factors, and many types of protective agents. It contains about 10 percent solids for energy and growth and the rest is water, which is essential to maintain hydration. This is also why a breastfed baby does not need additional water. Infants can digest breast milk much more rapidly than formula and, therefore, do not get constipated. On average, it takes about 30 minutes longer to digest formula as opposed to breast milk. Breastfed babies have better cheekbone development and better jaw alignment.

Besides the benefits of the contracting uterus, the process of producing milk burns calories, which helps the mother to lose excess weight gained during pregnancy. After all, that is why pregnant women put on extra fat during pregnancy—energy storage for milk production. Breastfeeding is also related to a lower risk of breast **cancer** and ovarian cancer. For every year of life spent breastfeeding, a woman's risk of developing breast cancer drops by 4.3 percent and this is on top of the 7 percent reduction she enjoys for every baby to whom she gives birth.

Additionally, there is the convenience. Breast milk is always with the mother. Mothers do not have to store it. It is always at the right temperature. It is free. It does not require sterilization. In fact, it prevents diseases and has protective factors resulting in healthier babies and decreased healthcare costs. It saves money as there is no need to buy formula, bottles, and nipples.

Procedure

It is best to begin breastfeeding immediately after birth as it is an infant's natural instinct to nurse then. Regardless of the baby's initial suckling behavior, this interaction stimulates uterine contractions, promotes colonization of harmless bacteria on the nipple, and helps to protect the infant from pathogenic bacteria. It is an important time to nuzzle. Women breastfeed for a longer duration if feedings are started early. The first several feedings have an imprinting effect. It is recommended to continue feeding about every two to three hours. It is important to remember that all babies are different; some need to nurse almost constantly at first, while others can go much longer between feedings. There are babies and mothers who have no trouble breastfeeding, while others may need some assistance. Once the baby begins to suck, the mother makes sure that the entire dark area around the nipple (areola) is in the baby's mouth. This helps stimulate milk flow and allows the baby to get enough milk. Nipple soreness can be a result of the infant not getting a good grasp of the entire areola. A newborn needs to be fed at least eight to 12 times in 24 hours. Since breast milk is so easily digested, a baby may be hungry again as soon as one and one-half hours after the last feeding.

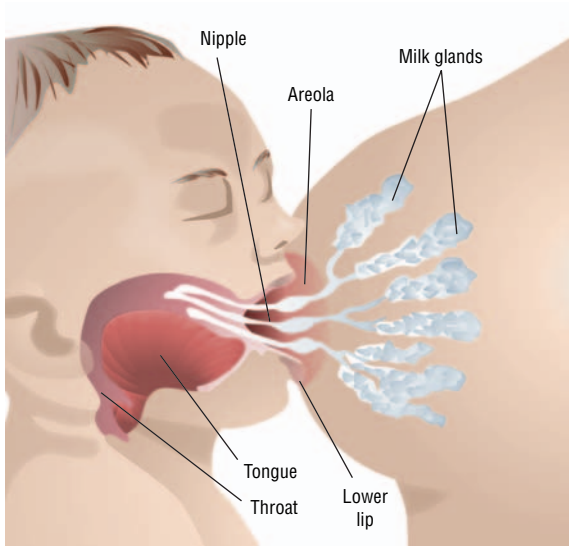
Mothers need to be comfortable when nursing; therefore, loose, front-opening clothes and a good nursing bra are essential. They need to explore different positions for breastfeeding to determine what is best for them. The cradle hold works well in bed or sitting in a comfortable chair. The football hold is excellent if the woman had a **cesarean section**. The mother can use pillows to support the baby and a footstool to flatten her

Lactation

Signs of good breastfeeding progress	Warning signs
Eight to 12 feeding per 24 hours	Fewer than eight feedings in 24 hours; baby sleeps four to six hours at time
Baby nurses every 1.5 to three hours	Baby nurses every hour or more, but never seems satisfied
Six to eight wet diapers every 24 hours after the third day	Fewer than six wet diapers after the third day
Soft yellow stools, about 1 tablespoon or larger	Dark black, green, or brown stools after the third day
After the third day, four to 10 stools per day	Fewer than three or four stools per day after the third day
Average daily weight gain of 15 to 30 g once milk comes in	Baby does not regain birth weight by 10 days of age
Milk comes in; breasts are full and warm and may leak milk	Milk does not seem to come in by the fifth day
Intermittent periods of rhythmic sucking and audible swallows	Milk comes in, but sucking or swallowing is not audible
Breasts are tender and may be slightly painful or sore	Sore and painful nipples throughout most feedings; scabbed or cracked nipples
Breasts soften after a feed	Severe engorgement; breast remain very hard after a feed

(Table by GGS Information Services.)

Lactation



When an infant is properly latched onto the breast, the baby's nose touches (or nearly touches) the breast. He or she takes the entire areola into the mouth, facilitating the intake of milk far back into the throat. *(Illustration by GGS Information Services.)*

lap. The mother can position the baby's head by snuggling it in one arm and supporting her breast with the other hand by keeping her thumb well above the areola and the rest of the fingers below and under the breast (sometimes called the C-hold). In this position, the mother can lift her breast and guide her nipple in any direction as she helps the baby to take in more of the areola.

For early feedings, the infant should be offered both breasts at each feeding as this stimulates the need-supply response. The length of the feeding is up to the mother. The general rule is to watch the baby, not the clock. If, however, it is a first time mother, 20 to 30 minutes on the first side can be suggested. If the baby falls asleep at the breast, the next feeding should begin with the breast that was not nursed. Mothers can tell if the baby is getting enough milk by checking diapers; a baby who is wetting between four to six disposable diapers (six to eight cloth) and who has three or four bowel movements in 24 hours is getting enough milk.

Common problems

New mothers may experience nursing problems, including the following:

- Engorgement: Breasts that are too full can prevent the baby from suckling because they cannot be grasped. Expressing milk manually or with a breast pump can alleviate this problem.
- Sore nipples: Transient soreness can occur during the first week postpartum and is usually temporary. Air drying the nipples and rubbing colostrum or breast milk into them provides relief. Prolonged, abnormal soreness lasts longer than a week postpartum. Discontinuing use of soap on breasts while bathing and applying purified lanolin to nipples and air drying them helps.
- Infection: Soreness and inflammation on the breast surface or a fever in the mother may be an indication of breast infection (mastitis). If it is just starting, the mother should drink lots of water and nurse frequently on the affected breast. **Antibiotics** may be necessary if the infection persists.

Lactation consultants work at almost every hospital where babies are delivered. First-time mothers can request the lactation consultant to visit her. The mother should make a note of the lactation consultant's phone number should problems be encountered after mother and infant go home.

There are no rules about when to stop breastfeeding. A baby needs breast milk for at least the first year of life and it is preferred that no solid food be given for at least

KEY TERMS

Alveoli—The tiny air sacs clustered at the ends of the bronchioles in the lungs in which oxygen-carbon dioxide exchange takes place.

Bromocriptine—Also known as Parlodel, it is a dopamine receptor agonist used to treat galactorrhea by reducing levels of the hormone prolactin and is also used to treat Parkinson's disease.

Colostrum—Milk secreted for a few days after birth and characterized by high protein and antibody content.

Ergotamine—A drug used to prevent or treat migraine headaches. It can cause vomiting, diarrhea, and convulsions in infants and should not be taken by women who are nursing.

Involution—The return of a large organ to normal size.

Lactogenesis—The initiation of milk secretion.

Lithium—A medication prescribed to treat the manic (excited) phases of bipolar disorder.

Mammary—Relating to the breast.

Methotrexate—A drug that interferes with cell growth and is used to treat rheumatoid arthritis as well as various types of cancer. Side-effects may include mouth sores, digestive upsets, skin rashes, and hair loss. Since this drug can supress an infant's immune system, it should not be taken by nursing mothers.

Placenta—The organ that provides oxygen and nutrition from the mother to the unborn baby during pregnancy. The placenta is attached to the wall of the uterus and leads to the unborn baby via the umbilical cord.

Postpartum—After childbirth.

Progesterone—The hormone produced by the ovary after ovulation that prepares the uterine lining for a fertilized egg.

Prolactin—A hormone that helps the breast prepare for milk production during pregnancy.

the first six months to prevent allergies. As long as a baby eats age-appropriate solid food, the mother may nurse for several years.

Parental concerns

The majority of illnesses are not transmitted via breast milk; in fact, breast milk prevents many illnesses. However, some viruses, including HIV (the virus that causes **AIDS** can be passed in breast milk; for this reason, women who are HIV-positive should not breastfeed unless they are living in a country that does not have clean water to make formula. A lack of clean water to make formula could result in an infant dying from diarrhea.

Many medications have not been tested in nursing women, so it is not certain what drugs can affect a breastfed child. A nursing woman should always check with her doctor or lactation consultant before taking any medications, including over-the-counter drugs. The mother can usually take antibiotics without discontinuing breastfeeding.

The following drugs are not safe for a mother to take while she is nursing:

- radioactive drugs for some diagnostic tests
- chemotherapy drugs for cancer
- bromocriptine
- ergotamine
- lithium
- methotrexate
- street drugs (including marijuana, heroin, amphetamines)
- tobacco

Resources

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National Alliance for Breastfeeding Advocacy. 9684 Oak Hill Drive; Ellicott City, MD 21042. Web site: <www.healthfinder.gov/orgs/HR2952.htm>.

Linda K. Bennington

Lactose intolerance

Definition

Lactose intolerance refers to the inability of the body to digest lactose.

Description

Lactose is the predominant form of sugar present in milk. The enzyme lactase, which is normally produced by cells lining the small intestine, breaks down lactose into substances that can be absorbed into the bloodstream. When dairy products are ingested, the lactose reaches the digestive system and is broken down by lactase into the simpler sugars glucose and galactose, which can then be absorbed into the bloodstream. Lactose intolerance occurs when, due to a deficiency of lactase, lactose is not completely broken down and consequently blood sugar levels do not rise. While not usually a dangerous condition, lactose intolerance can cause severe discomfort.

Lactose intolerance is also referred to as lactase deficiency, milk intolerance, dairy product intolerance, or disaccharidase deficiency.

Demographics

From 30 to 50 million Americans suffer from the symptoms of lactose intolerance by the age of 20. People from cultures in which adult consumption of milk and milk products occurred earliest are less likely to be lactose intolerant than people from areas where dairy farming began more recently. The prevalence of deficiency in production of the lactase enzyme, therefore, varies among different ethnic groups. Among Asian populations it is almost 100 percent, with symptoms occurring around the age of five; among Native Americans it is 80 percent; among blacks it is 70 percent, with symptoms appearing by the age of 10; and among American Caucasians, the prevalence of lactose intolerance is only 20 percent. However, individuals who are mildly or moderately deficient in the production of the lactase enzyme may not exhibit symptoms of lactose intolerance.

Causes and symptoms

Lactose intolerance can be caused by some diseases of the digestive system (for example, celiac sprue and **gastroenteritis**) and by injuries to the small intestine that result in a decreased production of lactase. While rare, some children are also born unable to produce the enzyme. For most people, however, lactase deficiency develops naturally because, after about two years of age, the body produces less lactase. Before humans became dairy farmers, they usually did not continue to drink milk, so their bodies did not produce lactase after early childhood.

Symptoms of lactose intolerance include **nausea**, cramps, **diarrhea**, floating and foul-smelling stools, bloating, and intestinal gas. The symptoms usually occur between 30 minutes to two hours after eating or drinking lactose-containing foods. A child may also exhibit weight loss, slow growth, and **malnutrition**.

When to call the doctor

If a child develops symptoms of lactose intolerance, the doctor should be consulted concerning dietary substitutions.

Diagnosis

To diagnosis lactose intolerance, usually healthcare professionals measure the absorption of lactose in the digestive system by using the lactose tolerance test, the hydrogen breath test, or the stool acidity test. Each of these can be performed as an outpatient in a hospital, clinic, or doctor's office.

Children who are to take the lactose tolerance test must fast before being tested. They then drink a lactose-containing liquid for the test; medical personnel take blood samples during the next two hours to measure the children's blood glucose level. The blood glucose level, or blood sugar level, indicates how well the body is digesting the lactose. A diagnosis of lactose intolerance is confirmed when blood glucose level does not rise. This test is not administered to infants and very young children because of the risk of **dehydration** from drinking the lactose-containing liquid, which can cause diarrhea in those who are lactose intolerant, resulting in dehydration.

Hydrogen is usually detected only in small amounts in the breath. However, when undigested lactose found in the colon is fermented by bacteria, hydrogen in the breath is produced in greater quantities. The hydrogen is exhaled after being absorbed from the intestines and carried through the bloodstream to the lungs. The hydrogen breath test involves having the child drink a lactose-containing beverage. Healthcare professionals monitor the breath at regular intervals to see if the hydrogen levels rise, which indicates improper lactose digestion. Children taking the test who have had certain foods, medications, or cigarettes before the test may get inaccurate results. While the test is useful for children and adults, infants and young children should not take it because of the risk of dehydration from diarrhea in those who are lactose intolerant.

The stool acidity test measures the amount of acid in the stool. This is a safe test for newborns and young children. The test detects lactic acid and other short-chain fatty acids from undigested lactose fermented by bacteria in the colon. Glucose may also be found in the stool sample, resulting from unabsorbed lactose in the colon.

Some parents may try to self-diagnose lactose intolerance in their child by using an **elimination diet**, a diet that eliminates obvious milk and milk products. However, because there are so many food products that may contain hidden sources of milk, such a diet should be supervised by a dietician or developed by following a guide to a lactose-eliminating diet. A simpler way to self-diagnose lactose intolerance is by a milk challenge. The child fasts overnight, drinks a glass of milk in the morning, and then fasts for the next three to five hours. If the child is lactose intolerant, the child should experience symptoms within several hours. If symptoms do occur, the child should be evaluated by a healthcare professional to rule out the possibility of a milk allergy. However, milk **allergies** are rare and usually only occurs in infants and young children.

Treatment

Since there is no treatment that can improve the body's ability to produce lactase, treatment for lactose deficiency is focused on controlling the diet.

Most children affected by lactose intolerance do well if they limit their intake of lactose-containing food and drinks. Individuals differ in the amounts they can handle before experiencing symptoms. Many children may only need to eliminate major milk-containing products from their diet, while others who are intolerant to even small amounts of lactose may be required to follow severe dietary restrictions.

Foods that contain lactose include milk, low-fat milk, skim milk, chocolate milk, buttermilk, sweetened condensed milk, dried whole milk, instant nonfat dry milk, low-fat yogurts, frozen yogurt, ice cream, ice milk, sherbet, cheese, cottage cheese, low-fat cottage cheese, cream, and butter. Other foods that may contain hidden lactose are: nondairy creamers, powdered artificial sweeteners, foods containing milk power or nonfat milk solids, bread, cake, margarine, creamed soups, pancakes, waffles, processed breakfast cereals, salad dressings, lunch meats, puddings, custards, confections, and some meat products. Lactose is also used as the base for more than 20 percent of prescription drugs and 6 percent of over-the-counter drugs.

For infants younger than two years of age, soy formulas are adequate substitutes for milk. Toddlers may drink rice or soymilk, while older children who are sensitive to lactose can take lactase enzymes, which are available without a prescription. Using the liquid form of lactase enzymes, children can add a few drops in their milk, put the milk in the refrigerator and drink it after 24 hours, when the lactase enzymes have reduced the lactose content by 70 percent. If the milk is heated first and double the amount of lactase liquid enzymes is added, the milk will be 90 percent lactose-free. Supermarkets also carry lactose-reduced milk and other products, which contain nutrients found in the regular products but without the lactose.

In the early 2000s, researchers have developed a chewable lactase enzyme tablet. Taking three to six tablets just before eating helps some children digest lactose-containing solid foods.

Nutritional concerns

Eliminating milk from the diet can result in deficiencies of calcium, vitamin D, riboflavin, and protein. Milk substitutes for children are a necessity, as other sources of calcium are required. Fermented milk products such as yogurt are often tolerated. Buttermilk and cheeses have less lactose than milk. Goat's milk can sometimes be tolerated but should be consumed with meals.

Prognosis

Lactose intolerance is easy to manage and is not considered dangerous. People of all ages, but especially children, have to replace the calcium that is lost by cutting back on milk products; this can be accomplished by taking supplements and eating calcium-rich foods, such as broccoli, kale, canned salmon with bones, calcium-fortified foods, and tofu. They may also add lactase enzymes to dairy products to reduce lactose content as

KEY TERMS

Galactose—One of the two simple sugars (glucose is the other one) that makes up the protein, lactose, found in milk. Galactose can be toxic in high levels.

Glucose—A simple sugar that serves as the body's main source of energy.

Lactase—The enzyme produced by cells that line the small intestine that allows the body to break down lactose.

Lactose—A sugar found in milk and milk products.

well as use lactose-reduced dairy products. Many children who suffer with lactose intolerance are able to continue eating some milk products.

Prevention

Often lactose intolerance is a natural occurrence that cannot be avoided. However, people can prevent symptoms by managing the condition with diet and lactase supplements.

Parental concerns

Parents must guard the health of a child who is lactose intolerant by carefully managing the child's diet to avoid foods that will result in symptoms while providing foods that contain necessary nutrients for the child's health and growth.

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Judith Sims
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Langerhans cell histiocytosis *see* **Histiocytosis X**

Language delay

Definition

A language delay is **language development** that is significantly below the norm for a child of a specified age.

Description

Language delay is a communication disorder, a category that includes a wide variety of speech, language, and hearing impairments. The milestones of language development, including the onset of babbling and a child's first words and sentences, normally occur within approximate age ranges. However, individual children vary enormously regarding the exact age at which each milestone is reached. There also are different styles of language development. Most children have acquired good verbal communication by the age of three. But one child may be wordless until the age of two and a half and then immediately start talking in three-word sentences. Another child might have several words at ten months but add very few additional words over the following year. Other children start talking at about 12 months and progress steadily.

Language delay usually becomes apparent during infancy or early childhood. Any delay in general development usually causes language delay. Children with language delay may acquire language skills in the usual progression but at a much slower rate, so that their language development may be equivalent to a normally developing child of a much younger chronological age. Maturation delay, also called developmental language delay, is one of the most common types of language delay. Children with a maturation delay may be referred to as "late talkers" or "late bloomers." Maturation delays frequently run in families.

Demographics

Speech/language delay is the most common developmental disorder in children aged three to 16 years, affecting approximately 3 to 10 percent of children. It is three to four times more common in boys than in girls.

Causes and symptoms

Environmental causes

Common nonphysical causes of language delay include circumstances in which the following are the case:

- The child is concentrating on some other skill, such as walking perfectly, rather than on language.
- The child has a twin or sibling very close in age and thus may not receive as much individual attention.
- The child has older siblings who interpret so well that the child has no need to speak or whose talk is so continuous that the child lacks the opportunity to speak.
- The child is in a daycare situation with too few adults to provide individual attention.
- The child is under the care of a non-English speaker.
- The child is bilingual or multilingual, learning two or more languages simultaneously but at a slower speed; the child's combined comprehension of the languages is normal for that age.
- The child suffers from psychosocial deprivation such as poverty, **malnutrition**, poor housing, neglect, inadequate linguistic stimulation, emotional stress.
- The child is abused; abusive parents are more likely to neglect their children and less likely to communicate with them verbally.

Physical causes

Language delay may result from a variety of underlying disorders, including the following:

- **mental retardation**
- maturation delay (This delay in the maturation of the central neurological processes required to produce speech is often the cause of late talking.)
- **hearing impairment**
- **dyslexia**, a specific reading disorder which may cause language delay in preschoolers
- a learning disability
- cerebral palsy, in which numerous factors may contribute to language delay
- autism, a developmental disorder in which, among other things, children do not use language or use it abnormally
- congenital blindness, even in the absence of other neurological impairment
- brain damage
- Klinefelter syndrome, a disorder in which males are born with an extra X chromosome
- receptive aphasia or receptive language disorder, a deficit in spoken language comprehension or in the ability to respond to spoken language, resulting from brain damage
- expressive aphasia, an inability to speak or write, although comprehension is normal; caused by malnutrition, brain damage, or hereditary factors
- childhood apraxia of speech, a nervous system disorder

Mental retardation accounts for more than 50 percent of language delays. Language delay is usually more severe than other developmental delays in retarded children, and it is often the first noticeable symptom of mental retardation. Mental retardation causes global language delay, including delayed auditory comprehension and use of gestures.

Impaired hearing is one of the most common causes of language delay. Any child who does not hear speech in a clear and consistent manner will have language delay. Even a minor hearing impairment can significantly affect language development. In general the more severe the impairment, the more serious the language delay. Children with congenital (present at birth) hearing impairment or hearing loss that occurs within the first two years of life (known as prelingual hearing loss) experience serious language delay, even when the impairment is diagnosed and treated at an early age. However, deaf children born to parents who use sign language develop infant babble and a fully expressive sign language at the same rate as hearing children.

Symptoms of language delay

Symptoms of language delay include the following:

- failure to meet the developmental milestones for language development
- language development that lags behind other children of the same age by at least one year
- inability to follow directions
- slow or incomprehensible speech after three years of age
- serious difficulties with syntax (placing words in a sentence in the correct order)
- serious difficulties with articulation, including the substitution, omission, or distortion of certain sounds

Language delays resulting from underlying conditions may have symptoms specific to the condition. Nonetheless, specific symptoms of language delay may include the following:

- not babbling by 12 to 15 months of age
- not understanding simple commands by 18 months of age
- not talking by two years of age
- not using sentences by three years of age

- not being able to tell a simple story by four or five years of age

Symptoms of language delay with mental retardation

Mentally impaired children usually babble during their first year and may speak their first words within the normal age range. However, they often cannot do the following:

- put words together
- speak in complete sentences
- acquire a larger, more varied vocabulary
- develop grammatically

Mentally impaired children in conversation may be repetitive and routine, exhibiting little **creativity**. Nevertheless vocabulary and grammatical development appear to proceed by very similar processes in mentally retarded and developmentally normal children.

In general the severity of language delay depends on the severity of the mental retardation. Levels of retardation and language skill are ranked as follows:

- mild retardation (**intelligence** quotient [IQ] range of 52–68): usually eventually develop language skills
- moderate retardation (IQ range of 36–51): usually learn to talk and communicate
- severe retardation (IQ range of 20–35): have limited language but can speak a few words

Language delays among mentally retarded children vary greatly. Some severely mentally impaired children who also have **hydrocephalus** or **Williams syndrome** may acquire exceptional conversational language skills, sometimes called the "chatterbox syndrome." Some children (called savants) test as mentally retarded but learn their native language, as well as foreign languages, very easily. With **Down syndrome** and some other disorders, language delay is more severe than other mental impairments. This factor may be due to the characteristic facial abnormalities and relatively large tongues of Down-syndrome children. Children with Down syndrome also are at higher risk for hearing impairment and ear infections that cause hearing loss.

Symptoms of language delay with other disorders

Symptoms of language delay in a hearing-impaired child include the following:

- babbling at an older-than-normal age
- babbling that is less varied and less sustained
- first words at age two or older
- only two-word sentences by age four or five in a profoundly deaf child

Dyslexic children have difficulty separating parts of words and single words within a group of words. Symptoms of dyslexia may include:

- poor articulation
- difficulties identifying sounds within words, blending sounds, or rhyming
- difficulty putting sounds in the correct order
- hesitation in choosing words

A learning-disabled child usually exhibits an uneven pattern of language development. In addition, about 50 percent of autistic children never learn to speak. Those who do speak often have severe language delay and may use words in unusual ways. They rarely participate in interactive dialogue and often speak with an unusual rhythm or pitch. The speech of some autistic children has an atonic or sing-song quality.

Children with congenital blindness average about an eight-month delay in speaking words. Although blind children develop language in much the same way as sighted children, they may rely more on conversational formulas.

The speech of children with receptive aphasia is both delayed and sparse, ungrammatical, and poorly articulated. Children with expressive aphasia fail to speak at the usual age although they have normal speech comprehension and articulation. Children with defined lesions in language areas on either side of the brain have initial but quite variable language delays. Usually their language catches up by the age of two or three without noticeable deficits.

Apraxia affects the ability to sequence and vocalize sounds, syllables, and words. Children with apraxia know what they want to say, but their brains do not send the correct signals to the lips, jaw, and tongue to form the words. In addition to language delay, apraxia often causes other expressive **language disorders**.

When to call the doctor

Children who are not talking at all by the age of two should have a complete developmental **assessment**. Children who are not progressing in word-learning skills by the end of the first grade should be tested for dyslexia.

Diagnosis

Diagnosis of language delay requires a complete physical examination and a thorough developmental history, with special attention to language milestones. In young children it may be very difficult to distinguish between a late talker and a developmental expressive disorder. The diagnosis often is made by a speech/language pathologist. Language performance of bilingual children must be compared to that of other bilingual children of a similar cultural and linguistic background. Generalized delay in all developmental milestones suggests mental retardation.

Numerous tests are used to screen for language delay and assess language development. Some of these are described below:

- The Denver Developmental Screening Test is the most popular test in clinical use for children from birth to six years of age. Since language delay is the most common early symptom of global intellectual impairment, the test provides a comprehensive developmental assessment.
- The Early Language Milestone Scale is a simple tool for assessing language development in children under the age of three. It relies on parents' reports and a very short test focusing on visual, receptive, and expressive language.
- The Mullen Scales of Early Learning is a comprehensive assessment of language, motor, and perceptual abilities in children from birth to five years eight months of age.
- The Peabody Picture Vocabulary Test, for children aged two-and-a-half to 18 years, is a useful screening instrument for word comprehension.
- The Receptive One-Word Picture Vocabulary Test provides information about a child's ability to understand language.

Other tests for language delay include:

- Early Speech Perception Test
- Assessing Prelinguistic and Early Linguistic Behaviors in Developmentally Young Children
- Joliet 3-Minute **Preschool** Speech and Language Screen
- Fluharty Preschool Speech and Language Screening Test
- Assessment of Fluency in School-Age Children
- Children's Articulation Test
- Clinical Evaluation of Language Fundamentals
- Phonological Assessment of Child Speech (Initial assessment may indicate the need for additional testing to identify underlying physical conditions.)

Treatment

About 60 percent of language delays in children under age three resolve spontaneously. Early detection and intervention for language delay can help prevent social, cognitive, and emotional problems. Treatment of language delay is individualized for each child with the primary goal of teaching the child strategies for comprehending spoken language and producing appropriate linguistic or communicative behavior. Depending on the type and cause of the delay, the healthcare team may include a physician, a speech/language pathologist, an audiologist, a psychologist, an occupational therapist, and a social worker. Psychotherapy may be recommended if the language delay is accompanied by **anxiety** or depression. Speech therapy is used to help mentally impaired children develop intelligible language. Behavior therapy may help autistic children progress in speech acquisition.

Hearing-impaired children who are identified and receive early intervention before six months of age develop significantly better language skills than children identified after six months of age. Early, consistent, and conscious use of visual communication modes such as sign language, finger spelling, and cued speech, and/or hearing amplification and oral training can reduce the language delay. Since only about 10 percent of deaf children are born to deaf parents, hearing parents can promote their deaf child's language development by learning and using sign language. Many types of hearing aids are available for children as young as three months. **Cochlear implants** may be used for profoundly deaf children aged two to six. These children usually develop better language skills than those with hearing aids or other devices.

Prognosis

With appropriate intervention language-delayed children usually catch up with their peers. Children with maturation delay usually have normal language development by the time they enter school. Although a bilingual home environment can cause a temporary language delay, most children become proficient in both languages before the age of five. Nevertheless, early language delays may cause problems with behavior and social interactions. A language delay can lead to elective **mutism**, a condition in which children choose not to speak.

KEY TERMS

Apraxia—Impairment of the ability to make purposeful movements, but not paralysis or loss of sensation.

Expressive aphasia—A developmental disorder in which a child has lower-than-normal proficiency in vocabulary, production of complex sentences, and word recall, although language comprehension is normal.

Maturation delay—Developmental language delay; a language delay caused by the slow maturation of speech centers in the brain; often causes late talking.

Receptive aphasia—A developmental disorder in which a child has difficulty comprehending spoken and written language.

Such children typically speak when they are on their own, with their friends, and sometimes with their parents; however, they will not speak in school, in public situations, or with strangers.

Most mentally retarded children eventually develop at least some degree of language. But frustration and anger at their inability to communicate effectively can lead to numerous social and behavioral problems. An adolescent with Down syndrome and an IQ of about 50 may speak at the grammatical level of an unaffected three-year-old, with short, repetitive, simple sentences. Nevertheless, Down-syndrome children often are very sociable and interested in conversational language.

Children who receive early intervention for hearing impairments can develop at nearly the same rate as other children. However, depending on the severity of their hearing loss, they may continue to have difficulties with articulation and speech quality as well as with written language. Children who lose their hearing after the first few years of life have far fewer language delays than children who are deaf from birth or who lose their hearing within the first year.

Most children with receptive aphasia gradually acquire a language of their own, understood only by those close to them. Children with expressive aphasia will not develop normal language skills without intervention and are at risk for language-based learning disabilities.

Prevention

There are no known preventions for most language delays. Prenatal care and good **nutrition** during pregnancy and early childhood may help prevent some expressive language delays. Hearing-impaired children who use sign language within their families usually have no signed-language delay.

Parental concerns

A speech/language pathologist can teach parents methods for encouraging and enhancing their child's language development. Special programs also are available for helping language-delayed children and their parents. Parents, caregivers, and teachers of children with language delay should take the following steps:

- adjust their speech to the child's level
- use consistent language
- use meaningful language
- repeat words, phrases, sentences, and stories
- use small-group instruction

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Language development

Definition

Language development is the process by which children come to understand and communicate language during early childhood.

Description

From birth up to the age of five, children develop language at a very rapid pace. The stages of language development are universal among humans. However, the age and the pace at which a child reaches each milestone of language development vary greatly among children. Thus, language development in an individual child must be compared with norms rather than with other individual children. In general girls develop language at a faster rate than boys. More than any other aspect of development, language development reflects the growth and maturation of the brain. After the age of five it becomes much more difficult for most children to learn language.

Receptive language development (the ability to comprehend language) usually develops faster than expressive language (the ability to communicate). Two different styles of language development are recognized. In referential language development, children first speak single words and then join words together, first into two-word sentences and then into three-word sentences. In expressive language development, children first speak in long unintelligible babbles that mimic the cadence and rhythm of adult speech. Most children use a combination these styles.

Infancy

Language development begins before birth. Towards the end of pregnancy, a fetus begins to hear sounds and speech coming from outside the mother's body. Infants are acutely attuned to the human voice and prefer it to other sounds. In particular they prefer the higher pitch characteristic of female voices. They also are very attentive to the human face, especially when the face is talking. Although crying is a child's primary means of communication at birth, language immediately begins to develop via repetition and imitation.

Between birth and three months of age, most infants acquire the following abilities:

- seem to recognize their mother's voice
- quiet down or smile when spoken to
- turn toward familiar voices and sounds
- make sounds indicating pleasure
- cry differently to express different needs
- grunt, chuckle, whimper, and gurgle
- begin to coo (repeating the same sounds frequently) in response to voices
- make vowel-like sounds such as "ooh" and "ah"

Between three and six months, most infants can do the following:

- turn their head toward a speaker
- watch a speaker's mouth movements
- respond to changes in a tone of voice
- make louder sounds including screeches
- vocalize excitement, pleasure, and displeasure
- cry differently out of **pain** or hunger
- laugh, squeal, and sigh
- sputter loudly and blow bubbles
- shape their mouths to change sounds
- vocalize different sounds for different needs
- communicate desires with gestures
- babble for attention
- mimic sounds, inflections, and gestures
- make many new sounds, including "p," "b," and "m," that may sound almost speech-like

The sounds and babblings of this stage of language development are identical in babies throughout the world, even among those who are profoundly deaf. Thus all babies are born with the capacity to learn any language. Social interaction determines which language they eventually learn.

Six to 12 months is a crucial age for receptive language development. Between six and nine months babies begin to do the following:

- search for sources of sound
- listen intently to speech and other sounds
- take an active interest in conversation even if it is not directed at them
- recognize "dada," "mama," "bye-bye"
- consistently respond to their names
- respond appropriately to friendly and angry tones
- express their moods by sound and body language
- **play** with sounds
- make long, more varied sounds
- babble random combinations of consonants and vowels
- babble in singsong with as many as 12 different sounds
- experiment with pitch, intonation, and volume
- use their tongues to change sounds
- repeat syllables
- imitate intonation and speech sounds

Between nine and 12 months babies may begin to do the following:

- listen when spoken to
- recognize words for common objects and names of **family** members
- respond to simple requests
- understand "no"
- understand gestures
- associate voices and names with people
- know their own names
- babble both short and long groups of sounds and two-to-three-syllable repeated sounds (The babble begins to have characteristic sounds of their native language.)
- use sounds other than crying to get attention
- use "mama" and "dada" for any person
- shout and scream
- repeat sounds
- use most consonant and vowel sounds
- practice inflections
- engage in much vocal play

Toddlerhood

During the second year of life language development proceeds at very different rates in different children. By the age of 12 months, most children use "mama/dada" appropriately. They add new words each month and temporarily lose words. Between 12 and 15 months children begin to do the following:

- recognize names
- understand and follow one-step directions
- laugh appropriately
- use four to six intelligible words, usually those starting with "b," "c," "d," and "g," although less than 20 percent of their language is comprehensible to outsiders
- use partial words
- gesture and speak "no"
- ask for help with gestures and sounds

At 15 to 18 months of age children usually do the following:

- understand "up," "down," "hot," "off"
- use 10 to 20 intelligible words, mostly nouns
- use complete words
- put two short words together to form sentences
- chatter and imitate, use some echolalia (repetitions of words and phrases)
- have 20 to 25 percent of their speech understood by outsiders

At 18 to 24 months of age toddlers come to understand that there are words for everything and their language development gains momentum. About 50 of a child's first words are universal: names of foods, animals, family members, **toys**, vehicles, and clothing. Usually children first learn general nouns, such as "flower" instead of "dandelion," and they may overgeneralize words, such as calling all toys "balls." Some children learn words for social situations, greetings, and expressions of love more readily than others. At this age children usually have 20 to 50 intelligible words and can do the following:

- follow two-step directions
- point to parts of the body
- attempt multi-syllable words
- speak three-word sentences

- ask two-word questions
- enjoy challenge words such as "helicopter"
- hum and sing
- express pain verbally
- have 50 to 70 percent of their speech understood by outsiders

After several months of slower development, children often have a "word spurt" (an explosion of new words). Between the ages of two and 18 years, it is estimated that children add nine new words per day. Between two and three years of age children acquire:

- a 400-word vocabulary including names
- a word for most everything
- the use of pronouns
- three to five-word sentences
- the ability to describe what they just saw or experienced
- the use of the past tense and plurals
- names for body parts, colors, toys, people, and objects
- the ability to repeat rhymes, songs, and stories
- the ability to answer "what" questions

Children constantly produce sentences that they have not heard before, creating rather than imitating. This **creativity** is based on the general principles and rules of language that they have mastered. By the time a child is three years of age, most of a child's speech can be understood. However, like adults, children vary greatly in how much they choose to talk.

Preschool

Three to four-year-olds usually can do the following:

- understand most of what they hear
- converse
- have 900 to 1,000-word vocabularies, with verbs starting to predominate
- usually talk without repeating syllables or words
- use pronouns correctly
- use three to six-word sentences
- ask questions
- relate experiences and activities
- tell stories (Occasional **stuttering** and stammering is normal in preschoolers.)

Language skills usually blossom between four and five years of age. Children of this age can do the following:

- verbalize extensively
- communicate easily with other children and adults
- articulate most English sounds correctly
- know 1,500 to 2,500 words
- use detailed six to eight-word sentences
- can repeat four-syllable words
- use at least four prepositions
- tell stories that stay on topic
- can answer questions about stories

School age

At age five most children can do the following:

- follow three consecutive commands
- talk constantly
- ask innumerable questions
- use descriptive words and compound and complex sentences
- know all the vowels and consonants
- use generally correct grammar

Six-year-olds usually can correct their own grammar and mispronunciations. Most children double their vocabularies between six and eight years of age and begin reading at about age seven. A major leap in reading comprehension occurs at about nine. Ten-year-olds begin to understand figurative word meanings.

Adolescents generally speak in an adult manner, gaining language maturity throughout high school.

Common problems

Language delay is the most common **developmental delay** in children. There are many causes for language delay, both environmental and physical. About 60 percent of language delays in children under age three resolve spontaneously. Early intervention often helps other children to catch up to their age group.

Common circumstances that can result in language delay include:

- concentration on developing skills other than language
- siblings who are very close in age or older siblings who interpret for the younger child

- inadequate language stimulation and one-on-one attention
- bilingualism, in which a child's combined comprehension of two languages usually is equivalent to other children's comprehension of one language
- psychosocial deprivation

Language delay can result from a variety of physical disorders, including the following:

- mental retardation
- maturation delay (the slower-than-usual development of the speech centers of the brain), a common cause of late talking
- a hearing impairment
- a learning disability
- cerebral palsy
- autism (a developmental disorder in which, among other things, children do not use language or use it abnormally)
- congenital blindness, even in the absence of other neurological impairment
- Klinefelter syndrome, a disorder in which males are born with an extra X chromosome

Brain damage or disorders of the central nervous system can cause the following:

- receptive aphasia or receptive language disorder, a deficit in spoken language comprehension or in the ability to respond to spoken language
- expressive aphasia, an inability to speak or write despite normal language comprehension
- childhood apraxia of speech, in which a sound is substituted for the desired syllable or word

Parental concerns

Language development is enriched by verbal interactions with other children and adults. Parents and caregivers can have a significant impact on early language development. Studies have shown that children of talkative parents have twice the vocabulary as those of quiet parents. A study from the National Institute of Child Health and Human Development (NICHD) found that children in high-quality childcare environments have larger vocabularies and more complex language skills than children in lower-quality situations. In addition language-based interactions appear to increase a child's capacity to learn. Recommendations for encouraging language development in infants include:

- talking to them as much as possible and giving them opportunities to respond, perhaps with a smile; short periods of silence help teach the give-and-take of conversation
- talking to infants in a singsong, high-pitched speech, called "parentese" or "motherese" (This is a universal method for enhancing language development.)
- using one- or two-syllable words and two to three-word sentences
- using proper words rather than baby words
- speaking slowly, drawing-out vowels, and exaggerating main syllables
- avoiding pronouns and articles
- using animated gestures along with words
- addressing the baby by name
- talking about on-going activities
- asking questions
- singing songs
- commenting on sounds in the environment
- encouraging the baby to make vowel-like and consonant-vowel sounds such as "ma," "da," and "ba"
- repeating recognizable syllables and repeating words that contain the syllable

When babies reach six to 12 months-of-age, parents should play word games with them, label objects with words, and allow the baby to listen and participate in conversations. Parents of toddlers should do the following:

- talk to the child in simple sentences and ask questions
- expand on the toddler's single words
- use gestures that reinforce words
- put words to the child's gestures
- name colors
- count items
- gently repeat correctly any words that the child has mispronounced, rather than criticizing the child

Parents of two to three-year-olds should do the following:

- talk about what the child and parent are doing each day
- encourage the child to use new words
- repeat and expand on what the child says
- ask the child yes-or-no questions and questions that require a simple choice

Language development

Age	Activity
Two months	Cries, coos, and grunts.
Four months	Begins babbling. Makes most vowel sounds and about half of consonant sounds.
Six months	Vocalizes with intonation. Responds to own name.
Eight months	Combines syllables when babbling, such "Ba-ba."
Eleven months	Says one word (or fragment of a word) with meaning.
Twelve months	Says two or three words with meaning. Practices inflection, such as raising pitch of voice at the end of a question.
Eighteen months	Has a vocabulary between five and 20 words, mostly nouns. Repeats word or phrase over and over. May start to join two words together.
Two years	Has a vocabulary of 150–300 words. Uses I, me, and you. Uses at least two prepositions (in, on, under). Combines words in short sentences. About two-thirds of what is spoken is understandable.
Three years	Has a vocabulary of 900–1000 words. Uses more verbs, some past tenses, and some plural nouns. Easily handles three-word sentences. Can give own name, sex, and age. About 90% of speech is understandable.
Four years	Can use at least four prepositions. Can usually repeat words of four syllables. Knows some colors and numbers. Has most vowels and diphthongs and consonants p, b, m, w, and n established. Talks a lot and repeats often.
Five years	Can count to ten. Speech is completely understandable, although articulation might not be perfect. Should have all vowels and consonants m, p, b, h, w, k, g, t, d, n, ng, y. Can repeat sentences as long as nine words. Speech is mostly grammatically correct.
Six years	Should have all vowels and consonants listed above, has added, f, v, sh, zh, th, l. Should be able to tell a connected story about a picture.
Seven years	Should have consonants s–z, r, voiceless th, ch, wh, and soft g. Should be able to do simple reading and print many words.
Eight years	All speech sounds established. Carries on conversation at a more adult level. Can tell complicated stories of past events. Easily uses complex and compound sentences. Reads simple stories with ease and can write simple compositions.

SOURCE: Child Development Institute. 2004. http://www.childdevelopmentinfo.com.

(Table by GGS Information Services.)

- encourage the child to ask questions
- read books about familiar things, with pictures, rhymes, repetitive lines, and few words
- read favorite books repeatedly, allowing the child to join in with familiar words
- encourage the child to pretend to read
- not interrupt children when they are speaking

Parents of four to six-year-olds should:

- not speak until the child is fully attentive
- pause after speaking to give the child a chance to respond
- acknowledge, encourage, and praise speech
- introduce new words
- talk about spatial relationships and opposites
- introduce limericks, songs, and poems
- talk about the television programs that they watch
- encourage the child to give directions
- give their full attention when the child initiates a conversation

Parents of six to 12-year-olds should talk to the children, not at them, encourage conversation by asking questions that require more than a yes-or-no answer, and listen attentively as the child recounts the day's activities.

Additional recommendations for parents and caregivers, by the American Academy of Pediatrics and others, include:

- talking at eye level with a child and supplementing words with body language, gestures, and facial expressions to enhance language comprehension
- talking in ways that catch a child's attention
- using language to comfort a child
- using correct pronunciations
- using expressive language to discuss objects, actions, and emotions
- playing with sounds and words
- labeling objects and actions with words
- providing objects and experiences to talk about
- choosing activities that promote language
- listening carefully to children and responding in ways that let them know that they have been understood, as well as encouraging further communication
- using complete sentences and adding detail to expand on what a child has said
- knowing when to remain silent
- reading to a child by six months of age at the latest

KEY TERMS

Apraxia—Impairment of the ability to make purposeful movements, but not paralysis or loss of sensation.

Expressive aphasia—A developmental disorder in which a child has lower-than-normal proficiency in vocabulary, production of complex sentences, and word recall, although language comprehension is normal.

Expressive language—Communicating with language.

Expressive language development—A style of language development in which a child's babble mimics the cadence and rhythm of adult speech.

Receptive aphasia—A developmental disorder in which a child has difficulty comprehending spoken and written language.

Receptive language—The comprehension of language.

Referential language development—A style of language development in which a child first speaks single words and then joins words together into two- and three-word sentences.

- encouraging children to ask questions and seek new information
- encouraging children to listen to and ask questions of each other

Television viewing does not promote language development.

When to call the doctor

Parents should call the pediatrician immediately if they suspect that their child may have a language delay or a hearing problem. Warning signs of language delay in toddlers include:

- avoiding eye contact
- neither understanding nor speaking words by 18 months of age
- difficulty learning nursery rhymes or simple songs
- not recognizing or labeling common objects
- inability to pay attention to a book or movie
- poor articulation, such that a parent cannot understand the child more than 50 percent of the time

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Margaret Alic, PhD

Language disorders

Definition

A language disorder is a deficit or problem with any function of language and communication.

Description

Speech and language disorders are extremely common. They can range from slow acquisition of language to sound substitution or **stuttering** to the inability to understand or produce and language at all. The federal Agency for Healthcare Research and Quality estimated in 2002 that communication disorders cost the United States between $30 and $154 billion annually in lost productivity and money spent on medical care, **special education**, and remediation.

Language disorders and the brain

Speech and language pathologists and neurologists (doctors who specialize in the brain and nervous system) have known for about 100 years that certain areas in the left hemisphere of the brain—Broca's area in the posterior frontal lobe and Wernicke's area in the temporal lobe—are centrally involved in language functions. Damage to Broca's area results in problems with language fluency: shortened sentences, impaired flow of speech, poor control of rhythm and intonation, and a telegraphic style with missing inflections. Damage to Wernicke's area produces speech that is fluent and often rapid, but with relatively senseless content, many invented words, and word substitutions.

With the invention of new technologies, including **computed tomography** (CT) scans and **magnetic resonance imaging** (MRI), several studies have looked at the **language development** in very young children with lesions in the traditional language areas of the brain. There is surprising agreement among the studies in their results: all find initial delays in language development followed by remarkably similar progress after about age two to three years. Lasting deficits have not been noticed in these children. Surprisingly, there are also no dramatic effects of laterality; lesions to either side of the brain seem to produce virtually the same effects. However, most of the data comes from conversational analysis or relatively unstructured testing, and these children have not been followed until school age. Nevertheless, the findings suggest remarkable plasticity and robustness of language in spite of brain lesions that would devastate an adult's language abilities.

Language disorders and hearing loss

Children with a hearing loss, either from birth or acquired during the first year or two of life, generally have a serious delay in spoken language development. The hearing loss occurs despite very early diagnosis and fitting with appropriate hearing aids. However, in the unusual case that sign language is the medium of communication in the **family** rather than speech, the child shows no delay in learning to use that language. Hearing development is always one of the first things checked if a pediatrician or parent suspects a **language delay**. The deaf child exposed only to speech will usually begin to babble ("baba, gaga") at a slightly later point than the hearing child. Recent work suggests that the babbling is neither as varied nor as sustained as in hearing children. However, there is often a long delay until the first words are spoken, sometimes not until age two years or older.

Depending on the severity of the hearing loss, the stages of early language development are also quite delayed. It is not unusual for the profoundly deaf child at age four or five years to only have two-word spoken sentences. It is only on entering specialized training programs for oral language development that the profoundly deaf child begins to acquire more spoken language. Often, such children do not make the usual **preschool** language gains until they reach grade school. Many deaf children learning English have pronounced difficulties in articulation and speech quality, especially if they are profoundly deaf, since they get no feedback in how they sound. A child who has hearing for the first few years of life has an enormous advantage in speech quality and oral language learning over a child who is deaf from birth or within his or her first year.

Apart from speech difficulties, deaf children learning English often show considerable difficulty with the

inflection and syntax of the language, which marks their writing as well as their speech. The ramifications of this delayed language are also significant for learning to read, and reading proficiently. The average deaf high school student often only reads at fourth grade level.

Language disorders and mental retardation

Mental retardation can also affect the age at which children learn to talk. A mentally retarded child is defined as one who falls in the lower end of the range of **intelligence**, usually with an IQ (intelligence quotient) below 80 on some standardized IQ tests. There are many causes of mental retardation, including identified genetic syndromes such as **Down syndrome**, **Williams syndrome**, or **fragile X syndrome**.

Retardation can also be caused by damage to the fetus during pregnancy due to alcohol, drug abuse or toxicity, and disorders of the developing nervous system such as **hydrocephalus**. Finally, there are environmental causes following birth such as **lead poisoning**, anoxia, or **meningitis**.

Any of these situations is likely to slow down the child's rate of development in general, and thus to have effects on language development. However, most children with very low IQs develop some language, suggesting it is a relatively "buffered" system that can survive a good deal of insult to the developing brain. In cases of hydrocephalus, for example, it has been noted that children who are otherwise quite impaired intellectually can have impressive conversational language skills. Sometimes called the "chatterbox syndrome," this linguistic sophistication belies their poor ability to deal with the world. In an extreme case, a young man with a tested IQ in the retarded range has an apparent gift for acquiring foreign languages, and could learn a new one with very little exposure. For example, he could do fair translations at a rapid pace from written languages as diverse as Danish, Dutch, Hindi, Polish, French, Spanish, and Greek. He is, in fact, a savant in the area of language, and delights in comparing linguistic systems, although he does not have the mental capacity to live independently.

Adults should not consider retarded children to be a uniform class; different patterns can arise with different syndromes. For example, in hydrocephalic children and Williams syndrome, language skills may be preserved to a degree greater than their general intellectual level. In other groups, including Down syndrome, there may be more delay in language than in other mental abilities.

Most retarded children babble during the first year and develop their first words within a normal time span, but are then slow to develop sentences or a varied vocabulary. Vocabulary size is one of the primary components of standardized tests of verbal intelligence, and it grows slowly in retarded children. Nevertheless, the process of vocabulary development seems quite similar: retarded children also learn words from context and by incidental learning, not just by direct instruction.

Grammatical development, though slow, comes in the same way, and in the same order, as it does for normal IQ children. The child's conversation, however, may contain more repetition. The Down syndrome adolescent with an IQ of around 50 points does not seem to progress beyond the grammatical level of the normally intelligent child at three years, with short sentences that are restricted in variety and complexity. Children with Down syndrome are also particularly delayed in speech development. This is due in part to the facial abnormalities that characterize this syndrome, including a relatively large tongue. It is also linked to the higher risk they appear to suffer from ear infections and hearing loss.

Specific language impairment

Specific language impairment describes a condition of markedly delayed language development in the absence of any apparent handicapping conditions such as deafness, **autism**, or mental retardation. Specific language impairment (SLI) is also sometimes called childhood dysphasia, or developmental language disorder.

Children with SLI usually begin to talk at approximately the same age as normal children, but are markedly slower in their progress. They seem to have particular problems with inflection and word forms, such as leaving off endings when forming verb tenses (for example, the -ed ending when forming the past tense). This problem can persist much longer than early childhood, often into grade school and beyond, where these children encounter difficulties in reading and writing. The child with SLI often has difficulties learning language "incidentally," (picking up the meaning of a new word from context or generalizing a new syntactic form). This is in contrast to the normal child's development, where incidental learning and generalization are the hallmarks of language acquisition. Children with SLI are not cognitively impaired and are not withdrawn or socially aloof like the autistic child.

Very little is known about the cause or origin of specific language impairment, although evidence is growing that the underlying condition may be a form of brain abnormality. However, any such brain abnormality is not readily apparent with existing diagnostic technologies. When compared to other children, SLI children do not

Speech therapists help children overcome their speech and language disorders. *(© Bob Rowan; Progressive Image/Corbis.)*

have clear brain lesions or marked anatomical differences in either brain hemisphere.

Demographics

About one in six people, or 42 million individuals in the United States, have some type of communication disorder. About 28 million have speech, voice, or language problems associated with hearing loss, and about 14 million have similar problems not associated with impaired hearing. More than one million children in special education classes are categorized as having a speech or language disability.

Causes and symptoms

Language disorders can arise at many points in the language production process such as:

- from damage to the part of the brain that produces language
- from damage to the part of the brain that understands language
- from hearing loss
- from damage to the muscles and tissues of the mouth and throat needed for speech (e.g. **cleft palate**)
- from neurological disorders that interrupt the transmission of information necessary to receive and produce language
- from unknown (idiopathic) causes

Symptoms of language disorders vary widely, but include:

- slow acquisition of speech and language
- inability to make the physical sounds associated with language production (mutism)
- failure to make sense of spoken or written words
- inability to speak under certain social circumstances (selective mutism)

KEY TERMS

Speech pathologist—An individual certified by the American Speech-Language-Hearing Association (ASHA) to treat speech disorders.

- transformations of words or sounds when speaking
- inability to recall known words

When to call the doctor

Parents should talk to their pediatrician immediately if their child appears to have **hearing impairment**. They should also consult with their doctor if the child does not babble or begin to use single words within the normal time frame. Parents of older children may need a referral to a speech and language specialist if their child stutters, lisps, has difficulty forming words or producing coherent speech, or exhibits certain learning disabilities.

Diagnosis

Speech and language disorders are usually diagnosed by a speech and language pathologist, often with the help of a pediatrician, audiologist (hearing specialist), and neurologist. Many **assessment** tests are designed specifically for use in children, including the Clinical Evaluation of Language Fundamentals (also available in Spanish); the Preschool Language Scale (also available in Spanish); the Test of Language Development, Primary; and the Test of Language, Intermediate. There are assessments designed to evaluate speech production, such as the Goldman-Fristoe test of Articulation.

Treatment

Treatment varies, depending on the type and cause of the language disorder. However, in all language disorders and delays, early intervention is key to improvement. Many educators of the deaf now urge early compensatory programs in signed languages, because the deaf child shows no handicap in learning a visually based language. Deaf children born to signing parents begin to ''babble'' in sign at the same point in infancy that hearing infants babble speech, and proceed from there to learn a fully expressive language. However, only 10 percent of deaf children are born to deaf parents, so hearing parents must show a commitment and willingness to learn sign language. Furthermore, command of at least written English is still a necessity for such children to be able to function in the larger community.

Speech therapy can be a considerable aid to many children with language disorders For example, it can help to make a Down syndrome child's speech more intelligible. Despite the delay, children with Down syndrome are often quite sociable and interested in language for conversation.

Surgery, followed by speech therapy, can correct physical deformities, such as cleft palate, that interfere with speech production.

Psychotherapy can help older children whose language disorders are psychologically based.

Prognosis

Prognosis varies on an individual basis, depending on the cause, type, and severity of the language disorder. Those children who receive early intervention therapies are more likely to have a better outcome than those for whom services are delayed.

Prevention

Many language disorders are not preventable. However, those that arise from damage to the fetus due to the mother's use of drugs or alcohol during pregnancy can be prevented by avoiding these substances.

Parental concerns

Language is such a critical part of our society that parents are justly concerned when their child has a language disorder. The parents' approach to the disorder can greatly influence the child's self-image, **self-esteem**, and ultimately his or her success in reaching the fullest language potential.

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Laxatives

Definition

Laxatives are products that promote bowel movements.

Description

Laxatives may be grouped by mechanism of action.

Saline cathartics include dibasic sodium phosphate (Phospo-Soda), magnesium citrate, magnesium hydroxide (milk of magnesia), magnesium sulfate (Epsom salts), sodium biphosphate, and others. They act by attracting and holding water in the intestinal lumen, and may produce a watery stool. Magnesium sulfate is the most potent of the laxatives in this group.

Stimulant and irritant laxatives increase the peristaltic movement of the intestine. Examples include cascara and bisadocyl (Dulcolax). Castor oil works in a similar fashion.

Bulk producing laxatives increase the volume of the stool, and will both soften the stool and stimulate intestinal motility. Psyillium (Metamucil, Konsil) and methylcellulose (Citrucel) are examples of this type. The overall effect is similar to that of eating high-fiber foods, and this class of laxative is most suitable for regular use.

Docusate (Colace) is the only representative example of the stool softener class. It holds water within the fecal mass, providing a larger, softer stool. Docusate has no effect on acute **constipation**, since it must be present before the fecal mass forms to have any effect, but may be useful for prevention of constipation in patients with recurrent problems, or those who are about to take a constipating drug, such as narcotic **analgesics**.

Mineral oil is an emollient laxative. It acts by retarding intestinal absorption of fecal water, thereby softening the stool.

The hyperosmotic laxatives are glycerin and lactulose (Chronulac, Duphalac), both of which act by holding water within the intestine. Lactulose may also increase peristaltic action of the intestine.

General use

Laxatives are used to treat constipation—the passage of small amounts of hard, dry stools, usually fewer than three times a week. Constipation may be caused by several conditions, some of which are potentially serious, and require medical attention:

- Neurologic—caused by failure of nerves to stimulate movement of the muscles of the intestines.
- Obstructive—failure of the muscles in the intestine to open, or presence of a mass that's blocking passage of the feces through the intestine.
- Endocrine/metabolic—caused by some diseases including hypothyroidism.
- Medicinal—caused by some drugs, including narcotic analgesics, iron, and some drugs used in **cancer** treatment.

Chronic constipation occurs in 1–4% of children between the ages of four and 10. If constipation continues, it should be treated by a physician.

A reasonable first step is to assure that there is enough fiber in the diet. This may be done by switching to a high fiber breakfast cereal. If this doesn't lead to improvement, then medical attention is necessary.

Precautions

Short term use of laxatives is generally safe except in **appendicitis**, fecal impaction, or intestinal obstruction. Lactulose is composed of two sugar molecules; galactose and fructose, and should not be administered to patients who require a low galactose diet.

Chronic use of laxatives may result in fluid and electrolyte imbalances, steatorrhea, osteomalacia, **diarrhea**, cathartic colon, and liver disease. Excessive intake of mineral oil may cause impaired absorption of oil soluble

KEY TERMS

Carbohydrates—Compounds, such as cellulose, sugar, and starch, that contain only carbon, hydrogen, and oxygen, and are a major part of the diets of people and other animals.

Cathartic colon—A poorly functioning colon, resulting from the chronic abuse of stimulant cathartics.

Colon—The part of the large intestine that extends from the cecum to the rectum. The sigmoid colon is the area of the intestine just above the rectum; linking the descending colon with the rectum. It is shaped like the letter S.

Diverticulitis—Inflammation of the diverticula (small outpouchings) along the wall of the colon, the large intestine.

Fiber—Carbohydrate material in food that cannot be digested.

Hyperosmotic—Hypertonic, containing a higher concentration of salts or other dissolved materials than normal tissues.

Osteomalacia—A bone disease that occurs in adults due to a prolonged period of vitamin D deficiency. It is characterized by softening of the bone and is sometimes referred to as adult rickets.

Steatorrhea—An excessive amount of fat in the feces due to poor fat absorption in the gastrointestinal tract.

Stool—The solid waste that is left after food is digested. Stool forms in the intestines and passes out of the body through the anus.

vitamins, particularly A and D. Excessive use of magnesium salts may cause hypermanesemia.

Side effects

Excessive use of laxatives may result in dependency on these products. This may cause a condition known as cathartic colon.

Excessive use of laxatives that contain sodium or magnesium may result in dangerously high blood levels of these elements.

Interactions

Mineral oil and docusate should not be used in combination. Docusate is an emulsifying agent which will increase the absorption of mineral oil.

Bisacodyl tablets are enteric coated, and so should not be used in combination with antacids. The antacids will cause premature rupture of the enteric coating.

Preventing side effects

Used properly, laxatives are very safe. Do not overuse or give in doses larger than those labeled. If constipation persists, obtain medical help.

Parental concerns

Laxatives should not be used too frequently. Bulk laxatives such as psyllium should be the normal first choice.

Mineral oil should not be given to infants or other children while laying down. This may result in the oil going into the lungs.

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Samuel Uretsky, PharmD

Lazy eye *see* **Amblyopia**

Lead poisoning

Definition

Lead **poisoning** occurs when a person swallows, absorbs, or inhales lead in any form. The result can be damaging to the brain, nerves, and many other parts of the body. Acute lead poisoning, which is somewhat rare, occurs when a relatively large amount of lead is taken into the body over a short period of time. Chronic lead poisoning is a common problem in children that occurs when small amounts of lead are ingested over a longer period. The Centers for Disease Control and Prevention (CDC) defines childhood lead poisoning as a whole-blood lead concentration equal to or greater than 10 micrograms/dL.

Description

Lead can damage almost every system in the human body, and it can also cause high blood pressure (**hypertension**). It is particularly harmful to the developing brain of fetuses and young children. The higher the level of lead in a child's blood and the longer this elevated level lasts, the greater the chance of ill effects. Over the long term, lead poisoning in a child can lead to learning disabilities, behavioral problems, and even **mental retardation**. At very high levels, lead poisoning can cause seizures, coma, and even death.

Many children with elevated blood levels are exposed to lead through peeling paint in older homes. Others are exposed through dust or soil that has been contaminated by old paint or past emissions of leaded gasoline. Since children between the ages of 12 and 36 months are apt to put things in their mouths, they are more likely than older children to take in lead. Pregnant women who come into contact with lead can pass it along to the fetus.

Over 80 percent of American homes built before 1978 have lead-based paint in them, according to the Centers for Disease Control and Prevention (CDC). The older the home, the more likely it is to contain lead paint, and the higher the concentration of lead in the paint is apt to be. Some homes also have lead in the water pipes or plumbing. Without knowing it, people may have lead in the paint, dust, or soil around their homes or in their drinking water, since lead cannot be seen, smelled, or tasted. Because lead does not break down naturally, it can continue to cause problems until it is removed.

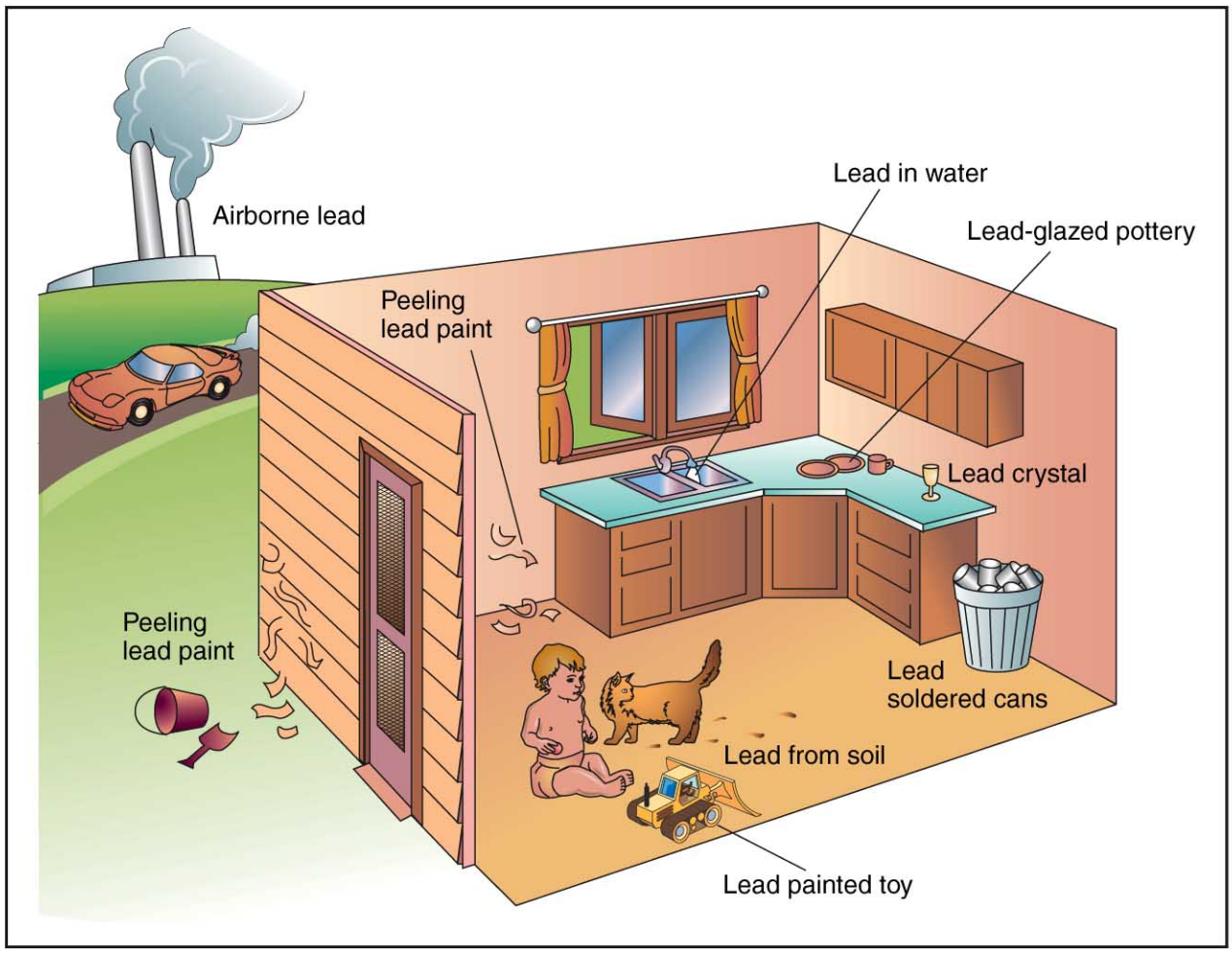
Demographics

According to the Agency for Toxic Substances and Disease Registry, approximately one out of every six children in the United States has a high level of lead in the blood. According to the National Center for Environmental Health, there were about 200 deaths from lead poisoning in the United States between 1979 and 1998. Most of the deaths were among males (74%), African Americans (67%), adults over the age of 45 (76%), and Southerners (70%).

Causes and symptoms

Before scientists knew how harmful it could be, lead was widely used in paint, gasoline, water pipes, and many other products. In the early 2000s house paint is almost lead-free, gasoline is unleaded, and household plumbing is no longer made with lead materials. Still, remnants of the old hazards remain. Following are some sources of lead exposure:

- Lead-based paint: This is the most common source of exposure to large amounts of lead among preschoolers. Children may eat paint chips from older homes that have fallen into disrepair. They may also chew on painted surfaces such as windowsills. In addition, paint may be disturbed during remodeling.
- Dust and soil: These can be contaminated with lead from old paint or past emissions of leaded gasoline. In addition, pollution from operating or abandoned industrial sites and smelters can find its way into the soil, resulting in soil contamination.
- Drinking water: Exposure may come from lead water pipes, found in many homes built before 1930. Even newer copper pipes may have lead solder. Also, some new homes have brass faucets and fittings that can leach lead.
- Jobs and hobbies: A number of activities can expose participants to lead. These include making pottery or stained glass, refinishing furniture, doing home repairs,



Continuous exposure to lead can damage nearly every system in the human body and is particularly harmful to the developing brain of fetuses and young children. Common sources of lead exposure include lead-based paint, dust and soil, drinking water, food from cans, and eating utensils, such as plates and drinking glasses, that are lead-based. *(Illustration by Electronic Illustrators Group.)*

and using indoor firing ranges. When adults take part in such activities, they may inadvertently expose children to lead residue that is on their clothing or on scrap materials.

- Food: Imported food cans often have lead solder. Lead may also be found in leaded crystal glassware and some imported ceramic or old ceramic dishes (e.g., ceramic dishes from Mexico). A 2003 study of cases of lead poisoning in pregnant women found that 70 percent of affected people were Hispanics, most of whom had absorbed the lead from their pottery. In addition, food may be contaminated by lead in the water or soil.
- Folk medicines: Certain folk medicines (for example, alarcon, alkohl, azarcon, bali goli, coral, ghasard, greta, liga, pay-loo-ah, and rueda) and traditional cosmetics (kohl, for example) contain high concentrations of lead.
- Moonshine whiskey: Lead poisoning from drinking illegally distilled liquor is still a cause of death among adults in the southern United States.
- Gunshot **wounds**: Toxic amounts of lead can be absorbed from bullets or bullet fragments that remain in the body after emergency surgery.

Evidence as of 2004 suggested that lead may be harmful to children even at low levels that were once thought to be safe, and the risk of damage rises as blood levels of lead increase. The symptoms of chronic lead poisoning take time to develop, however. Children can appear healthy despite having high levels of lead in their blood. Over time, though, problems such as the following may arise:

- learning disabilities
- hyperactivity

- mental retardation
- slowed growth
- hearing loss
- headaches

It is also known that certain genetic factors increase the harmful effects of lead poisoning in susceptible children; however, these factors are not completely understood.

Lead poisoning is also harmful to adults, in whom it can cause high blood pressure, digestive problems, nerve disorders, memory loss, and muscle and joint **pain**. In addition, it can lead to difficulties during pregnancy, as well as cause reproductive problems in both men and women.

In the early 2000s, chronic exposure to lead in the environment has been found to speed up the progression of kidney disorders in people without diabetes.

Acute lead poisoning

Acute lead poisoning, while less common, shows up more quickly and can be fatal. Symptoms such as the following may occur:

- severe abdominal pain
- diarrhea
- nausea and vomiting
- weakness of the limbs
- seizures
- coma

When to call the doctor

The CDC recommends testing all children at 12 months of age and, if possible, again at 24 months. Testing should start at six months for children at risk for lead poisoning.

Diagnosis

A high level of lead in the blood can be detected with a simple blood test. In fact, testing is the only way to know for sure if children without symptoms have been exposed to lead, since they can appear healthy even as long-term damage occurs. Based on test results and a child's risk factors, the doctor will then decide whether further testing is needed and how often. In some states, more frequent testing is required by law.

Children at risk

Children with an increased risk of lead poisoning include those for whom the following is true:

- They live in or regularly visit a house built before 1978 in which chipped or peeling paint is present.
- They live in or regularly visit a house that was built before 1978 where remodeling is planned or underway.
- They have a brother or sister, housemate, or playmate who has been diagnosed with lead poisoning.
- They have the habit of eating dirt or have been diagnosed with pica.
- They live with an adult whose job or hobby involves exposure to lead.
- They live near an active lead smelter, battery-recycling plant, or other industry that can create lead pollution.

Adults at risk

Testing is also important for adults whose job or hobby puts them at risk for lead poisoning. This need applies to people who take part in the following activities:

- glazed pottery or stained glass production
- furniture refinishing
- home renovation
- target shooting at indoor firing ranges
- battery reclamation
- precious metal refining
- radiator repair
- art restoration

Treatment

The first step in treating lead poisoning is to avoid further contact with lead. For adults, this usually means making changes at work or in hobbies. For children, it means that parents and guardians need to find and remove sources of lead in the home. In most states, the public health department can help assess the home and identify lead sources.

If the problem is lead paint, a professional with special training should remove it. Removal of lead paint is not a do-it-yourself project. Scraping or sanding lead paint creates large amounts of dust that can poison people in the home. This dust can stay around long after the work is completed. In addition, heating lead paint can release lead into the air. For these reasons, lead paint should only be removed by someone who knows how to do the job safely

KEY TERMS

Chelation therapy—A treatment using chelating agents, compounds that surround and bind to target substances allowing them to be excreted from the body.

Dimercaprol—A chemical agent used to remove excess lead from the body.

Edetate calcium disodium—A chemical chelating agent used to remove excess lead from the body.

Penicillamine (Cuprimine, Depen)—A drug used to treat medical problems (such as excess copper in the body and rheumatoid arthritis) and to prevent kidney stones. It is also sometimes prescribed to remove excess lead from the body.

Pica—A desire that sometimes arises in pregnancy to eat nonfood substances, such as dirt or clay.

Succimer—A chelating agent that is used to remove excess lead from the body. Sold under the trade name Chemet.

and has the equipment to clean up thoroughly. Occupants, especially children and pregnant women, should leave the home until the cleanup is finished.

Medical professionals should take all necessary steps to remove bullets or bullet fragments from people with gunshot injuries.

If blood levels of lead are high enough, the doctor may also prescribe chelation therapy. This refers to treatment with chemicals that bind to the lead and help the body pass it in urine at a faster rate. There are four chemical agents that may be used for this purpose, either alone or in combination. Edetate calcium disodium (EDTA calcium) and dimercaprol (BAL) are given through an intravenous line or in shots, while succimer (Chemet) and penicillamine (Cuprimine, Depen) are taken by mouth. (Although many doctors prescribe penicillamine for lead poisoning, this use of the drug has not been approved by the Food and Drug Administration.)

Changes in diet are no substitute for medical treatment. However, getting enough calcium, zinc, and protein may help reduce the amount of lead the body absorbs. Iron is also important, since people who are deficient in this nutrient absorb more lead. Garlic and thiamine, a B-complex vitamin, have been used to treat lead poisoning in animals. However, their usefulness in humans for this purpose has not as of 2004 been demonstrated. Nutritional, botanical, and homeopathic medicines can be administered once the source is removed to help correct any imbalances brought on by lead toxicity.

Prognosis

If acute lead poisoning reaches the stage of seizures and coma, there is a high risk of death. Even if the person survives, there is a good chance of permanent brain damage. The long-term effects of lower levels of lead can also be permanent and severe. However, if chronic lead poisoning is caught early, these negative effects can be limited by reducing future exposure to lead and getting proper medical treatment.

Prevention

Many cases of lead poisoning can be prevented. The following steps can help:

- Keep the areas where children **play** as clean and dust-free as possible.
- Wash pacifiers and bottles when they fall to the floor and wash stuffed animals and **toys** often.
- Make sure children wash their hands before meals and at bedtime.
- Mop floors and wipe windowsills and other chewable surfaces, such as cribs, twice a week with a solution of powdered dishwasher detergent in warm water.
- Plant bushes next to an older home with painted exterior walls to keep children at a distance.
- Plant grass or another ground cover in soil that is likely to be contaminated, such as soil around a home built before 1960 or located near a major highway.
- Have household tap water tested to find out if it contains lead.
- Use only water from the cold-water tap for drinking, cooking, and making baby formula, since hot water is likely to contain higher levels of lead.
- If the cold water has not been used for six hours or more, run it for several seconds, until it becomes as cold as it will get, before using it for drinking or cooking. The more time water has been sitting in the pipes, the more lead it may contain.
- Do not store food in open cans, especially imported cans.
- Do not store or serve food in pottery meant for decorative use.
- People who work with lead in a job or hobby should change their clothes before they go home.

Nutritional concerns

Avoid preparing or serving food in containers that have lead in their glazing. Do not consume homemade liquor that has been distilled.

Parental concerns

Lead tastes sweet. Parents living in homes built prior to 1978 should be vigilant regarding removing all flaking or peeling paint. Simply re-painting such surfaces will not resolve the problem. Parents must monitor the environments in which their children play and the objects that go into their children's mouths. Cleanliness is a must if old paint is in a child's environment. Removal (stripping paint to bare metal or bare wood) of lead is the best way to prevent lead exposure in children.

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American Academy of Family Physicians. 11400 Tomahawk Creek Parkway, Leawood, KS 66211–2672. Web site: <www.aafp.org/>.

American Academy of Pediatrics. 141 Northwest Point Boulevard, Elk Grove Village, IL 60007–1098. Web site: <www.aap.org/default.htm>.

American Association of Poison Control Centers. 3201 New Mexico Avenue NW, Washington, DC 20016. Web site: <www.aapcc.org/>.

American College of Occupational and Environmental Medicine. 55 West Seegers Road, Arlington Heights, IL 60005. Web site: <www.acoem.org/>.

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Learning disorders

Definition

Learning disorders are academic difficulties experienced by children and adults of average to above-average **intelligence**. People with learning disorders have difficulty with reading, writing, mathematics, or a combina-

tion of the three. These difficulties significantly interfere with academic achievement or daily living.

Description

Children with learning disorders, or disabilities, have specific impairments in acquiring, retaining, and processing information. Standardized tests place them well below their IQ range in their area of difficulty. The five main types of learning disorders are reading disorders, mathematics disorders, disorders of written expression, disorders of **fine motor skills**, and information processing disorders.

Reading disorders

Reading disorders are the most common type of learning disorder. Children with reading disorders have difficulty recognizing and interpreting letters and words (**dyslexia**). They are unable to recognize and decode the sounds and syllables (phonetic structure) behind written words and language in general. This condition lowers accuracy and comprehension in reading.

Mathematics disorders

Children with mathematics disorders (dyscalculia) have problems recognizing and counting numbers correctly. They have difficulty using numbers in everyday settings. Mathematics disorders are typically diagnosed in the first few years of elementary school when formal teaching of numbers and basic math concepts begins. Children with mathematics disorders usually have a coexisting reading disorder, a disorder of written expression, or both.

Disorders of written expression

Disorders of written expression typically occur in combination with reading disorders or mathematics disorders or both. The condition is characterized by difficulty with written compositions (dysgraphia). Children with this type of learning disorder have problems with spelling, punctuation, grammar, and organizing their thoughts in writing.

Disorders of fine motor skills

Children with motor skill disorders (dyspraxia) have coordination problems and may have difficulty with handwriting tasks and speech patterns. Dyspraxia tends to affect boys more than girls.

Information processing disorders

Information processing disorders often occur along with other types of learning disorders. Children with this problem have difficulty processing the sensory input they receive, specifically sight and sound information. They can see and hear adequately, but they have difficulty distinguishing between different visual cues and auditory signals, and may have problems understanding spatial relationships and sequencing the sights and sounds they observe.

Demographics

Learning disorders affect approximately two million children between the ages of six and 17 (5 percent of public school children), although some experts think the figure may be as high as 15 percent. The male to female ratio for learning disorders is about five to one.

Causes and symptoms

Learning disorders are thought to be caused by neurological abnormalities or differences that trigger impairments in the regions of the brain that control visual and language processing and attention and planning. These traits may be genetically linked. Children from families with a history of learning disorders are more likely to develop disorders themselves. In 2003, a team of Finnish researchers reported finding a candidate gene for developmental dyslexia on human chromosome 15q21.

Learning difficulties may also be caused by such medical conditions as a traumatic brain injury or brain infections such as **encephalitis** or **meningitis**.

The defining symptom of a learning disorder is academic performance that is markedly below a child's age and grade capabilities and measured IQ. Children with a reading disorder may confuse or transpose words or letters and omit or add syllables to words. The written homework of children with disorders of written expression is filled with grammatical, spelling, punctuation, and organizational errors. The child's handwriting is often extremely poor. Children with mathematical disorders are often unable to count in the correct sequence, to name numbers, and to understand numerical concepts.

When to call the doctor

A child thought to have a learning disorder should undergo a complete medical examination to rule out an organic cause of the problem. This may include an eye exam by an ophthalmologist, a psychological exam by a psychologist, and an exam by an otolaryngologist (an ear, nose, and throat doctor, or ENT).

Diagnosis

Problems with vision or hearing, mental disorders (depression, **attention-deficit/hyperactivity disorder**), **mental retardation**, cultural and language differences, and inadequate teaching may be mistaken for learning disorders or may complicate a diagnosis. A comprehensive medical, psychological, and educational **assessment** is critical to making a correct diagnosis.

A psychoeducational assessment should be performed by a psychologist, psychiatrist, neurologist, neuropsychologist, or learning specialist. A complete medical, **family**, social, and educational history is compiled from existing medical and school records and from interviews with the child and the child's parents and teachers. A series of written and verbal tests are then given to the child to evaluate his or her cognitive and intellectual functioning. Commonly used tests include the Wechsler Intelligence Scale for Children (WISC-III), the Woodcock-Johnson Psychoeducational Battery, the Peabody Individual Achievement Test-Revised (PIAT-R) and the California Verbal Learning Test (CVLT). Federal legislation mandates that this testing is free of charge within the public school system.

Treatment

Once a learning disorder has been diagnosed, an individual education plan (IEP) is developed for the child in question. IEPs are based on psychoeducational test findings. They provide for annual retesting to measure a child's progress. Learning-disordered students may receive special instruction within a regular general education class or they may be taught in a **special education** or learning center for a portion of the day.

Common strategies for the treatment of reading disorders focus first on improving a child's recognition of the sounds of letters and language through phonics training. Later strategies focus on comprehension, retention, and study skills. Students with disorders of written expression are often encouraged to keep journals and to write with a computer keyboard instead of a pencil. Instruction for students with mathematical disorders emphasizes real-world uses of arithmetic, such as balancing a checkbook or comparing prices.

Prognosis

The high school dropout rate for children with learning disabilities is almost 40 percent. Children with learning disabilities that go undiagnosed or are improperly treated may never achieve functional literacy. They often develop serious behavior problems as a result of their frustration with school. In addition, their learning problems are often stressful for other family members and may strain family relationships. The key to helping these students reach their fullest potential is early detection and the implementation of an appropriate individualized education plan (IEP). The prognosis is good for a large percentage of children with reading disorders that are identified and treated early. Learning disorders typically persist into adulthood, but with proper educational and vocational training, an individual can complete college and pursue a challenging career. Studies of the occupational choices of adults with dyslexia indicate that they do particularly well in people-oriented professions and occupations, such as nursing or sales.

Common strategies for the treatment of reading disorders focus first on improving a child's recognition of the sounds of letters and language through phonics training. *(© Robert Maass/Corbis.)*

Prevention

Some studies have indicated that one-on-one tutoring of children at risk for developing learning disorders

KEY TERMS

Dyslexia—A type of reading disorder often characterized by reversal of letters or words.

IEP—Individualized Education Plan. Under federal law governing special education, every child in public schools who is determined through assessment to have special mental disability needs has an IEP. An IEP is typically developed by a team of professionals that may include special education teachers, physical, occupational and speech therapists, psychologists, parents or guardians, and others who may be called on to provide expertise. The team meets at least once a year to set goals for the next school year and to assess progress on already established goals. Parents who are not satisfied with school-based assessments have the right to ask for independent assessments that must be paid for by the school system.

IQ—Intelligence quotient, a measure of intellectual functioning determined by performance on standardized intelligence tests. It is usually calculated by dividing an individual's mental age (determined by testing) by his/her chronological age and multiplying that result by 100.

Phonics—A system to teach reading by teaching the speech sounds associated with single letters, letter combinations, and syllables.

may be effective in preventing later reading and writing problems.

Parental concerns

Parents of children with learning disorders should stay in close contact with educators and school administrators to ensure that their child's IEP undergoes a regular review and continues to provide the maximum educational benefit for their child.

Resources

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ORGANIZATIONS

Learning Disabilities Association of America. 4156 Library Road, Pittsburg, PA 15234. (412) 341–1515. Web site: <www.ldanatl.org>.

National Center for Learning Disabilities (NCLD). 381 Park Avenue South, Suite 1401, New York, NY 10016. (410) 296–0232. Web site: <www.ncld.org>.

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Left-handedness *see* **Handedness**

Legg-Calé-Perthes disease *see* **Osteochondroses**

Lesbian issues *see* **Homosexuality and bisexuality**

Leukemias, acute

Definition

Leukemia is a **cancer** that starts in the organs that make blood, namely the bone marrow and the lymph system. Depending on their characteristics, leukemias can be divided into two broad types. Acute leukemias are the rapidly progressing leukemias, while the **chronic leukemias** progress more slowly. The vast majority of the childhood leukemias are of the acute form.

Description

The cells that make up blood are produced in the bone marrow and the lymph system. The bone marrow is the spongy tissue found in the large bones of the body. The lymph system includes the spleen (an organ in the upper abdomen), the thymus (a small organ beneath the breastbone), and the tonsils (an organ in the throat). In addition, the lymph vessels (tiny tubes that branch like blood vessels into all parts of the body) and lymph nodes (pea-shaped organs that are found along the network of lymph vessels) are also part of the lymph system. Lymph is a milky fluid that contains cells. Clusters of lymph nodes are found in the neck, underarm, pelvis, abdomen, and chest.

Blood is made up of red blood cells (RBCs), which carry oxygen and other materials to all tissues of the body; white blood cells (WBCs), which fight infection; and platelets, which play a part in the clotting of the blood. The white blood cells can be further subdivided into three main types: granulocytes, monocytes, and lymphocytes.

The granulocytes, as their name suggests, contain particles (granules). These granules contain special proteins (enzymes) and several other substances that can break down chemicals and destroy microorganisms, such as bacteria. Monocytes are the second type of white blood cell. They are also important in defending the body against pathogens.

The lymphocytes form the third type of white blood cell. There are two main types of lymphocytes: T lymphocytes and B lymphocytes. They have different functions within the immune system. The B cells protect the body by making antibodies, which are proteins that can attach to the surfaces of bacteria and viruses. This attachment sends signals to many other cell types to come and destroy the antibody-coated organism. The T cells protect the body against viruses. When a virus enters a cell, it produces certain proteins that are projected onto the surface of the infected cell. The T cells recognize these proteins and make certain chemicals that are capable of destroying the virus-infected cells. In addition, the T cells can destroy some types of cancer cells.

The bone marrow makes stem cells, which are the precursors of the different blood cells. These stem cells mature through stages into RBCs, WBCs, or platelets. In acute leukemias, the maturation process of the white blood cells is interrupted. The immature cells (blasts) proliferate rapidly and begin to accumulate in various organs and tissues, thereby affecting their normal function. This uncontrolled proliferation of the immature cells in the bone marrow affects the production of the normal red blood cells and platelets as well.

Acute leukemias are of two types: acute lymphocytic leukemia and acute myelogenous leukemia. Different types of white blood cells are involved in the two leukemias. In acute lymphocytic leukemia (ALL), it is the T or the B lymphocytes that become cancerous. The B cell leukemias are more common than T cell leukemias. Acute myelogenous leukemia, also known as acute nonlymphocytic leukemia (ANLL), is a cancer of the monocytes and/or granulocytes.

Demographics

Leukemias account for 2 percent of all cancers. Because leukemia is the most common form of childhood cancer, it is often regarded as a disease of childhood. However, leukemias affect nine times as many adults as children. Half of the cases occur in people who are 60 years of age or older. The incidence of acute and chronic leukemias is about the same.

Leukemia strikes both sexes and all ages. The human T-cell leukemia virus (HTLV-I) is believed to be the causative agent for some kinds of leukemias. However, as of 2004, the cause of most leukemias is not known. Acute lymphoid leukemia (ALL) is more common among Caucasians than among African-Americans, while acute myeloid leukemia (AML) affects both races equally. The incidence of acute leukemia is slightly higher among men than women. People with Jewish ancestry have a higher likelihood of getting leukemia. A higher incidence of leukemia has also been observed among persons with **Down syndrome** and some other genetic abnormalities.

Causes and symptoms

Exposure to ionizing radiation and to certain organic chemicals, such as benzene, is believed to increase the risk of developing leukemia. Having a history of diseases that damage the bone marrow, such as aplastic anemia, or a history of cancers of the lymphatic system puts people at a high risk for developing acute leukemias. Similarly, the use of anticancer medications, immunosuppressants, and the antibiotic chloramphenicol are also considered risk factors for developing acute leukemias.

The symptoms of leukemia are generally vague and non-specific. A patient may experience all or some of the following symptoms:

- weakness or chronic fatigue
- fever of unknown origin

- weight loss that is not due to dieting or exercise
- frequent bacterial or viral infections
- headaches
- skin rash
- non-specific bone pain
- easy bruising
- bleeding from gums or nose
- blood in urine or stools
- enlarged lymph nodes and/or spleen
- abdominal fullness

Diagnosis

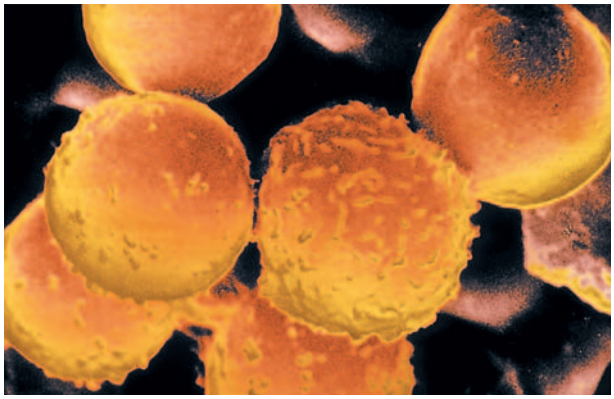
Like all cancers, acute leukemias are most successfully treated when found early. There were as of 2004 no screening tests available.

If the doctor has reason to suspect leukemia, he or she will conduct a very thorough physical examination to look for enlarged lymph nodes in the neck, underarm, and pelvic region. Swollen gums, enlarged liver or spleen, **bruises**, or pinpoint red **rashes** all over the body are some of the signs of leukemia. Urine and blood tests may be ordered to check for microscopic amounts of blood in the urine and to obtain a complete differential blood count. This count gives the numbers and percentages of the different cells found in the blood. An abnormal blood test might suggest leukemia; however, the diagnosis has to be confirmed by more specific tests.

The doctor may perform a bone marrow biopsy to confirm the diagnosis of leukemia. During the biopsy, a cylindrical piece of bone and marrow is removed. The tissue is generally taken out of the hipbone. These samples are sent to the laboratory for examination. In addition to diagnosis, the biopsy is also repeated during the treatment phase of the disease to see if the leukemia is responding to therapy.

A spinal tap (lumbar puncture) is another procedure that the doctor may order to diagnose leukemia. In this procedure, a small needle is inserted into the spinal cavity in the lower back to withdraw some cerebrospinal fluid and to look for leukemic cells.

Standard imaging tests, such as x rays, **computed tomography** scans (CT scans), and **magnetic resonance imaging** (MRI) may be used to check whether the leukemic cells have invaded other areas of the body, such as the bones, chest, kidneys, abdomen, or brain. A gallium scan or bone scan is a test in which a radioactive chemical is injected into the body. This chemical accumulates in the areas of cancer or infection, allowing them to be viewed with a special camera.



An enhanced transmission electron microscopy (TEM) image of white blood cells from a patient with acute myelogenous leukemia cells. *(© Professor Aaron Polliack, Science Source/Photo Researchers, Inc.)*

Treatment

There are two phases of treatment for leukemia. The first phase is called induction therapy. As the name suggests, during this phase, the main aim of the treatment is to reduce the number of leukemic cells as far as possible and induce a remission in the patient. Once the patient shows no obvious signs of leukemia (no leukemic cells are detected in blood tests and bone marrow biopsies), the patient is said to be in remission. The second phase of treatment is then initiated. This is called continuation or maintenance therapy, and the aim in this case is to kill any remaining cells and to maintain the remission for as long as possible.

Chemotherapy is the use of drugs to kill cancer cells. It is usually the treatment of choice and is used to relieve symptoms and achieve long-term remission of the disease. Generally, combination chemotherapy, in which multiple drugs are used, is more efficient than using a single drug for the treatment. Some drugs may be administered intravenously (through a vein) in the arm; others may be given by mouth in the form of pills. If the cancer cells have invaded the brain, then chemotherapeutic drugs may be put into the fluid that surrounds the brain through a needle in the brain or back. This is known as intrathecal chemotherapy.

Because leukemia cells can spread to all the organs via the blood stream and the lymph vessels, surgery is not considered an option for treating leukemias.

KEY TERMS

Antibody—A special protein made by the body's immune system as a defense against foreign material (bacteria, viruses, etc.) that enters the body. It is uniquely designed to attack and neutralize the specific antigen that triggered the immune response.

Biopsy—The surgical removal and microscopic examination of living tissue for diagnostic purposes or to follow the course of a disease. Most commonly the term refers to the collection and analysis of tissue from a suspected tumor to establish malignancy.

Chemotherapy—Any treatment of an illness with chemical agents. The term is usually used to describe the treatment of cancer with drugs that inhibit cancer growth or destroy cancer cells.

Computed tomography (CT)—An imaging technique in which cross-sectional x rays of the body are compiled to create a three-dimensional image of the body's internal structures; also called computed axial tomography.

Cytokines— Chemicals made by the cells that act on other cells to stimulate or inhibit their function. They are important controllers of immune functions.

Immunotherapy—A mode of cancer treatment in which the immune system is stimulated to fight the cancer.

Lumbar puncture—A procedure in which the doctor inserts a small needle into the spinal cavity in the lower back to withdraw spinal fluid for testing. Also known as a spinal tap.

Magnetic resonance imaging (MRI)—An imaging technique that uses a large circular magnet and radio waves to generate signals from atoms in the body. These signals are used to construct detailed images of internal body structures and organs, including the brain.

Maturation—The process by which stem cells transform from immature cells without a specific function into a particular type of blood cell with defined functions.

Radiation therapy—A cancer treatment that uses high-energy rays or particles to kill or weaken cancer cells. Radiation may be delivered externally or internally via surgically implanted pellets. Also called radiotherapy.

Remission—A disappearance of a disease and its symptoms. Complete remission means that all disease is gone. Partial remission means that the disease is significantly improved, but residual traces of the disease are still present. A remission may be due to treatment or may be spontaneous.

Radiation therapy, which involves the use of x-rays or other high-energy rays to kill cancer cells and shrink tumors, may be used in some cases. For acute leukemias, the source of radiation is usually outside the body (external radiation therapy). If the leukemic cells have spread to the brain, radiation therapy can be given to the brain.

Bone marrow transplantation is a process in which the patient's diseased bone marrow is replaced with healthy marrow. There are two ways of doing a bone marrow transplant. In an allogeneic bone marrow transplant, healthy marrow is taken from a donor whose tissue is either the same as or very closely resembles the patient's tissues. The donor may be a twin, a sibling, or a person who is not related at all. First, the patient's bone marrow is destroyed with very high doses of chemotherapy and radiation therapy. Healthy marrow from the donor is then given to the patient through a needle in a vein to replace the destroyed marrow.

In the second type of bone marrow transplant, called an autologous bone marrow transplant, some of the patient's own marrow is taken out and treated with a combination of anticancer drugs to kill all the abnormal cells. This marrow is then frozen and saved. The marrow remaining in the patient's body is destroyed with high-dose chemotherapy and radiation therapy. The marrow that was frozen is then thawed and given back to the patient through a needle in a vein. This mode of bone marrow transplant is in the early 2000s being investigated in clinical trials.

Biological therapy or immunotherapy is a mode of treatment in which the body's own immune system is harnessed to fight the cancer. Substances that are routinely made by the immune system (such as growth factors, hormones, and disease-fighting proteins) are either synthetically made in a laboratory or their effectiveness is boosted and they are then put back into the patient's body. This treatment mode is also being investigated in the early 2000s in clinical trials all over the United States at major cancer centers.

Prognosis

Like all cancers, the prognosis for leukemia depends on the patient's age and general health. According to sta-

tistics, more than 60 percent of the patients with leukemia survive for at least a year after diagnosis. Acute myelocytic leukemia (AML) has a poorer prognosis rate than acute lymphocytic leukemias (ALL) and the chronic leukemias. Between 1985 and 2004, the five-year survival rate for patients with ALL increased from 38 to 57 percent.

Interestingly enough, since most childhood leukemias are of the ALL type, chemotherapy has been highly successful in their treatment. This is because chemotherapeutic drugs are most effective against actively growing cells. Due to the new combinations of anticancer drugs being used, the survival rates among children with ALL have improved dramatically. Eighty percent of the children diagnosed with ALL as of 2004 survive for five years or more, as compared to 50 percent in the late 1970s.

Prevention

Most cancers can be prevented by changes in lifestyle or diet, which will reduce the risk factors. However, in leukemias, there are as of 2004 no such known risk factors. Therefore, as of 2004, no way is known to prevent leukemias from developing. People who are at an increased risk for developing leukemia because of proven exposure to ionizing radiation or exposure to the toxic liquid benzene, and people with Down syndrome, should undergo periodic medical checkups.

Parental concerns

Parents of a child with leukemia must balance their own fears for their child's health with the child's fears and worries. Also, given the large financial burden leukemia treatment entails, parents will want to make sure they are aware of what and what is not covered by their insurance. Parents can find a variety of sources, written and online, that will help them deal with the new circumstances of themselves and their **family**.

Resources

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Tubergen, David G., and Archie Bleyer. "The Leukemias." In *Nelson Textbook of Pediatrics*. Edited by Richard E. Behrman et al. Philadelphia: Saunders, 2004.

ORGANIZATIONS

American Cancer Society. 1599 Clifton Rd., NE, Atlanta, GA 30329–4251. Web site: <www.cancer.org>.

Cancer Research Institute. 681 Fifth Ave., New York, NY 10022. Web site: <www.cancerresearch.org>.

Leukemia Society of America Inc. 600 Third Ave., New York, NY 10016. Web site: <www.leukemia.org>.

National Cancer Institute. Building 31, Room 10A31, 31 Center Drive, MSC 2580, Bethesda, MD 20892–2580. <www.nci.nih.gov>.

Lata Cherath, PhD
Rosalyn Carson-DeWitt, MD

Leukemias, chronic

Definition

Chronic leukemia is a disease in which abnormal, cancerous white blood cells are made in the bone marrow. Depending on the type of white blood cell that is involved, chronic leukemia can be classified as chronic lymphocytic leukemia or chronic myeloid leukemia.

Description

Chronic leukemia is a **cancer** that starts in the blood cells made in the bone marrow. The bone marrow is the spongy tissue found in the large bones of the body. The bone marrow makes precursor cells called blasts or stem cells, which mature into different types of blood cells. Unlike **acute leukemias**, in which the process of maturation of the blast cells is interrupted, in chronic leukemias, the cells do mature and only a few remain as immature cells. However, even though the cells appear normal, they do not function as normal cells.

Bone marrow produces different types of cells: red blood cells (RBCs), which carry oxygen and other materials to all tissues of the body; white blood cells (WBCs), which fight infection; and platelets, which play a part in the clotting of the blood. The white blood cells can be further subdivided into three main types: the granulocytes, monocytes, and the lymphocytes.

The granulocytes, as their name suggests, contain granules (particles). These granules contain special proteins (enzymes) and several other substances that can break down chemicals and destroy microorganisms such as bacteria.

Monocytes are the second type of white blood cell. They are also important in defending the body against pathogens.

The lymphocytes form the third type of white blood cell. There are two main types of lymphocytes: T lymphocytes and B lymphocytes. They have different functions within the immune system. The B cells protect the body by making antibodies, which are proteins that can attach to the surfaces of bacteria and viruses. This attachment sends signals to many other cell types to come and destroy the antibody-coated organism. The T cell protects the body against viruses. When a virus enters a cell, it produces certain proteins that are projected onto the surface of the infected cell. The T cells can recognize these proteins and produce certain chemicals (cytokines) that are capable of destroying the virus-infected cells. In addition, the T cells can destroy some types of cancer cells.

Chronic leukemias develop very gradually. The abnormal lymphocytes multiply slowly, but in a poorly regulated manner. They live much longer and thus their numbers build up in the body. The two types of chronic leukemias can be easily distinguished under the microscope. Chronic lymphocytic leukemia (CLL) involves the T or B lymphocytes. B cell abnormalities are more common than T cell abnormalities. T cells are affected in only 5 percent of the patients. The T and B lymphocytes can be differentiated from the other types of white blood cells based on their size and by the absence of granules inside them. In chronic myelogenous leukemia (CML), the cells that are affected are the granulocytes.

Chronic lymphocytic leukemia (CLL) often has no symptoms at first and may remain undetected for a long time. Chronic myelogenous leukemia (CML), by contrast, may progress to a more acute form.

Demographics

Chronic leukemias account for 1.2 percent of all cancers. Because leukemia is the most common form of childhood cancer, it is often regarded as a disease of childhood. However, leukemias affect nine times as many adults as children. In chronic lymphoid leukemia, 90 percent of the cases are seen in people who are 50 years or older, with the average age at diagnosis being 65. The incidence of the disease increases with age. It is almost never seen in children. Chronic myeloid leukemias are generally seen in people in their mid-40s. In addition, it accounts for about 4 percent of childhood leukemia cases.

Causes and symptoms

Leukemia strikes both sexes and all ages. Although the cause is unknown, chronic leukemia is linked to genetic abnormalities and environmental factors. For example, exposure to ionizing radiation and to certain organic chemicals, such as benzene, is believed to increase the risks for getting leukemia. Chronic leukemia occurs in some people who are infected with two human retroviruses (HTLV-I and HTLV-II). An abnormal chromosome known as the Philadelphia chromosome is seen in 90 percent of those with CML. The incidence of chronic leukemia is slightly higher among men than women.

The symptoms of chronic leukemia are generally vague and non-specific. In chronic lymphoid leukemia (CLL), a patient may experience all or some of the following symptoms:

- swollen lymph nodes
- an enlarged spleen, which could make the patient complain of abdominal fullness
- chronic fatigue
- a general feeling of ill-health
- fever of unknown origin
- night sweats
- weight loss that is not due to dieting or exercise
- frequent bacterial or viral infections

In the early stages of chronic myeloid leukemia (CML), the symptoms are more or less similar to CLL. In the later stages of the disease, the patient may experience the following symptoms:

- non-specific bone **pain**
- bleeding problems
- mucus membrane irritation
- frequent infections
- a pale color due to a low red blood cell count (anemia)
- swollen lymph glands
- fever
- night sweats

Diagnosis

There are no screening tests available for chronic leukemias. The detection of these diseases may occur by chance during a routine physical examination.

If the doctor has reason to suspect leukemia, he or she will conduct a very thorough physical examination

to look for enlarged lymph nodes in the neck, underarm, and pelvic region. Swollen gums, an enlarged liver or spleen, **bruises**, or pinpoint red **rashes** all over the body are some of the signs of leukemia. Urine and blood tests may be ordered to check for microscopic amounts of blood in the urine and to obtain a complete differential blood count. This count will give the numbers and percentages of the different cells found in the blood. An abnormal blood test might suggest leukemia; however, the diagnosis has to be confirmed by more specific tests.

The doctor may perform a bone marrow biopsy to confirm the diagnosis of leukemia. During the bone marrow biopsy, a cylindrical piece of bone and marrow is removed. The tissue is generally taken out of the hipbone. These samples are sent to the laboratory for examination. In addition to diagnosis, bone marrow biopsy is also done during the treatment phase of the disease to see if the leukemia is responding to therapy.

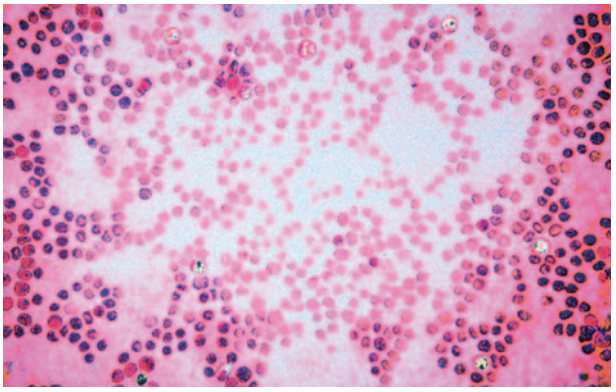
Standard imaging tests such as x-rays, **computed tomography** scans (CT scans), and **magnetic resonance imaging** (MRI) may be used to check whether the leukemic cells have invaded other organs of the body, such as the bones, chest, kidneys, abdomen, or brain.

Treatment

The treatment depends on the specific type of chronic leukemia and its stage. In general, **chemotherapy** is the standard approach to both CLL and CML. Radiation therapy is occasionally used. Because leukemia cells can spread to all the organs via the blood stream and the lymph vessels, surgery is not considered an option for treating leukemias.

Bone marrow transplantation (BMT) is in the early 2000s becoming the treatment of choice for CML because it has the possibility of curing the illness. BMT is generally not considered an option in treating CLL because CLL primarily affects older people, who are not considered to be good candidates for the procedure.

In BMT, the patient's diseased bone marrow is replaced with healthy marrow. There are two ways of doing a bone marrow transplant. In an allogeneic bone marrow transplant, healthy marrow is taken from another person (donor) whose tissue is either the same or very closely resembles the patient's tissues. The donor may be a twin, a sibling, or a person who is not related at all. First, the patient's bone marrow is destroyed with very high doses of chemotherapy and radiation therapy. To replace the destroyed marrow, healthy marrow from the donor is given to the patient through a needle in the vein.



Chronic lyphocytic leukemia cells, colorized and magnified 400 times. *(© 1999 Custom Medical Stock Photo, Inc.)*

In the second type of bone marrow transplant, called an autologous bone marrow transplant, some of the patient's own marrow is taken out and treated with a combination of anticancer drugs to kill all the abnormal cells. This marrow is then frozen to save it. The marrow remaining in the patient's body is then destroyed with high dose chemotherapy and radiation therapy. Following that, the patient's own marrow that was frozen is thawed and given back to the patient through a needle in the vein. This mode of bone marrow transplant is as of the early 2000s being investigated in clinical trials.

In chronic lymphoid leukemia (CLL), chemotherapy is generally the treatment of choice. Depending on the stage of the disease, single or multiple drugs may be given. Drugs commonly prescribed are steroids, chlorambucil, fludarabine, and cladribine. Low dose radiation therapy may be given to the whole body, or it may be used to alleviate the symptoms and discomfort due to an enlarged spleen and lymph nodes. The spleen may be removed in a procedure called a splenectomy.

In chronic myeloid leukemia (CML), the treatment of choice is bone marrow transplantation. During the slow progress (chronic phase) of the disease, chemotherapy may be given to try to improve the cell counts. Radiation therapy, which involves the use of x rays or other high-energy rays to kill cancer cells and shrink tumors, may be used in some cases to reduce the discomfort and pain due to an enlarged spleen. For chronic leukemias, the source of radiation is usually outside the body (external radiation therapy). If the leukemic cells have spread to the brain, radiation therapy can be directed at the brain. As the disease progresses, the spleen may be removed in an attempt to try to control the pain and to improve the blood counts.

KEY TERMS

Antibody—A special protein made by the body's immune system as a defense against foreign material (bacteria, viruses, etc.) that enters the body. It is uniquely designed to attack and neutralize the specific antigen that triggered the immune response.

Biopsy—The surgical removal and microscopic examination of living tissue for diagnostic purposes or to follow the course of a disease. Most commonly the term refers to the collection and analysis of tissue from a suspected tumor to establish malignancy.

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Lumbar puncture—A procedure in which the doctor inserts a small needle into the spinal cavity in the lower back to withdraw spinal fluid for testing. Also known as a spinal tap.

Magnetic resonance imaging (MRI)—An imaging technique that uses a large circular magnet and radio waves to generate signals from atoms in the body. These signals are used to construct detailed images of internal body structures and organs, including the brain.

Maturation—The process by which stem cells transform from immature cells without a specific function into a particular type of blood cell with defined functions.

Radiation therapy—A cancer treatment that uses high-energy rays or particles to kill or weaken cancer cells. Radiation may be delivered externally or internally via surgically implanted pellets. Also called radiotherapy.

Remission—A disappearance of a disease and its symptoms. Complete remission means that all disease is gone. Partial remission means that the disease is significantly improved, but residual traces of the disease are still present. A remission may be due to treatment or may be spontaneous.

In the acute phase of CML, aggressive chemotherapy is given. Combination chemotherapy, in which multiple drugs are used, is more efficient than using a single drug for the treatment. The drugs may either be administered intravenously (through a vein) in the arm or by mouth in the form of pills. If the cancer cells have invaded the central nervous system (CNS), chemotherapeutic drugs may be put into the fluid that surrounds the brain through a needle in the brain or back. This is known as intrathecal chemotherapy.

Biological therapy or immunotherapy is a mode of treatment in which the body's own immune system is harnessed to fight the cancer. Substances that are routinely made by the immune system (such as growth factors, hormones, and disease-fighting proteins) are either synthetically made in a laboratory, or their effectiveness is boosted and they are then put back into the patient's body. This treatment mode in the early 2000s is also being investigated in clinical trials all over the United States at major cancer centers.

Prognosis

The prognosis for leukemia depends on the patient's age and general health. According to statistics, in chronic lymphoid leukemia, the overall survival for all stages of the disease is nine years. Most of the deaths in people with CLL are due to infections or other illnesses that occur as a result of the leukemia.

In CML, if bone marrow transplantation is performed within one to three years of diagnosis, 50 to 60 percent of the patients survive three years or more. If the disease progresses to the acute phase, the prognosis is poor. Less than 20 percent of these patients go into remission.

Prevention

Most cancers can be prevented by changes in lifestyle or diet, which will reduce the risk factors. However, in leukemias, there were as of 2004 no known risk factors. Therefore, as of 2004, there was no way known to prevent the leukemias from developing. People who are at an increased risk for developing leukemia because of

proven exposure to ionizing radiation, the organic liquid benzene, or people who have a history of other cancers of the lymphoid system (Hodgkin's lymphoma) should undergo periodic medical checkups.

Parental concerns

Parents of a child with leukemia must balance their own fears for their child's health with the child's fears and worries. Also, given the large financial burden leukemia treatment entails, parents will want to make sure they are aware of what and what is not covered by their insurance. Parents can find a variety of sources, written and online, that will help them deal with the new circumstances of themselves and their **family**.

Resources

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Tubergen, David G., and Archie Bleyer. "The Leukemias." In *Nelson Textbook of Pediatrics*. Edited by Richard E. Behrman et al. Philadelphia: Saunders, 2004.

ORGANIZATIONS

American Cancer Society. 1599 Clifton Rd., NE, Atlanta, GA 30329–4251. Web site: <www.cancer.org>.

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Lice infestation

Definition

A lice infestation, or pediculosis, is caused by parasites living on human skin. Lice are tiny, wingless insects with sucking mouthparts that feed on human blood and lay eggs on body hair or in clothing. Lice **bites** can cause intense **itching**.

Description

There are three related species of human lice:

- head lice, *Pediculus humanus capitis*
- body lice, *Pediculosis humanus corpus*
- pubic lice, *Phthirus pubis*, commonly called crab lice

Pediculosis capitis is an infestation of head lice. A body lice infestation is called pediculosis corporis. Pediculosis palpebrarum or phthiriasis palpebrarum, caused by crab lice, is an infestation of the pubic hair.

Head lice live and crawl on the scalp, sucking blood every three to six hours. Their claws are adapted for clinging to hair or clothing. Adult head lice can be silvery-white to reddish-brown. They are about the size of a sesame seed. Female lice lay their eggs in sacs called nits that are about 0.04 in (1 mm) long and are glued to shafts of hair close to the scalp. During her one-month lifespan a female louse may lay more than 100 eggs. The nymphs hatch in three to 14 days and must feed on blood within one day. Nymphs are smaller and lighter in color than adults and become sexually mature after nine to 12 days.

Body lice lay their nits in clothing or bedding. Occasionally the nits are attached to body hair. Body lice nits are oval and yellow to white in color. They may not hatch for up to 30 days. Nymphs mature in about seven days.

Pubic lice have large front legs and look like tiny crabs. Females are larger than males. Nits hatch in about one week and the nymphs mature in about seven days.

Transmission

Lice are endemic in human populations, spreading through personal contact or contact with infested clothing or other personal items. They can be transmitted when unaffected clothing is stored with infested items. Among children head lice are commonly transmitted by the sharing of hats, combs, brushes, hair accessories, headphones, pillows, and stuffed **toys**. Pubic lice are sexually transmitted, although occasionally they can be transmitted through infested bedding, towels, or clothing.

Lice do not jump, hop, or fly and they do not live on pets. Head lice cannot survive without a human host for more than a few days at most. Body lice can live without

human contact for up to 10 days. Pubic lice can survive for one to two weeks without human contact.

Demographics

Head lice infestations are extremely common among children in schools, childcare facilities, camps, and playgrounds. They are the second most common communicable health problem in children, after the **common cold**, and appear to be on the increase. Some 6 to 12 million American children get head lice every year. In developing countries more than 50 percent of the general population may be infested. Although anyone can get head lice, children aged three to ten and their families are most affected. Girls and women are more susceptible than boys and men. Although American black children are much less likely to have head lice than white or Hispanic children, the incidence is increasing, particularly in black children with thick hair, hair extensions, or wraps. In Africa head lice have adapted their claws to the curly, elliptical hair shafts of blacks. Neither frequent brushing or shampooing nor hair length affects the likelihood of a head lice infestation.

In general body lice infestations occur in crowded, unsanitary facilities, such as prisons and military or refugee camps. They usually are associated with poor personal hygiene, as may occur during war or natural disasters or in cold climates. They are common among the homeless.

Causes and symptoms

Lice infestations are characterized by intense itching caused by an allergic reaction to a toxin in lice saliva. The itching can interfere with **sleep** and concentration. Repeated bites can lead to generalized skin eruptions or inflammation. Swelling or inflammation of the neck glands are common complications of head lice.

Body lice bites first appear as small red pimples or puncture marks and may cause a generalized skin rash. Intense itching can result in deep scratches around the shoulders, flanks, or neck. If the infestation is not treated, complications may develop, including **headache**, **fever**, and skin infection with scarring.

When to call the doctor

A doctor may need to distinguish between body lice and **scabies** (a disease caused by skin mites) and between pubic lice and eczema (a skin condition). A doctor should be consulted if complications develop from a lice infestation or if a child contracts a bacterial infection from scratching the bites.

Diagnosis

Lice usually are diagnosed by the itching; however, itching may not occur until several weeks after infestation, if at all. The tickling caused by moving lice may be noticeable. Definite diagnosis requires identification of lice or their nits.

Head lice may cause irritability in children and scalp irritations or sores may be present. Head lice in children are usually confined to the scalp. An adult louse may be visible as movement on the scalp, especially around the ears, nape of the neck, and centerline of the crown, the warmest parts of the head. Since less than 20 mature lice may be present at a given time during infestation, the nits often are easier to spot. Nits vary in color from grayish-white to yellow, brown, or black. They are visible at the base or on the shaft of individual hairs. Applying about 10 oz (280 gm) of isopropyl (rubbing) alcohol to the hair and rubbing with a white towel for about 30 seconds releases lice onto the towel for identification.

Body lice appear similar to head lice; however, they burrow into the skin and are rarely seen except on clothing where they lay their nits in seams. Over time body lice infestations can lead to a thickening and discoloring of the skin around the waist, groin, and upper thighs.

Pubic lice usually appear first on genital hair, although they may spread to other body hair. In young children pubic lice usually are seen on the eyebrows or eyelashes. Pubic lice appear as brown or gray moving dots on the skin. There are usually only a few live lice present and they move very quickly away from light. Their white nits can be seen on hair shafts close to the skin. Although pubic lice sometimes produce small, bluish spots called maculae ceruleae on the trunk or thighs, usually it is easier to spot scratching marks. Small, dark-brown specks of lice excretion may be visible on underwear.

Since pediculicides (medications for treating lice) are usually strong insecticides with potential side effects, it is important to rule out other causes of scratching and skin inflammation. Oval-shaped head lice nits can be distinguished from dandruff because they are glued at an angle to the hair shaft, whereas flat, irregularly shaped flakes of dandruff shake off easily.

Treatment

Most authorities believe that head lice should be treated immediately upon discovery. Before beginning any treatment, parents should test a small scalp section for allergic reactions to the medication, use a vinegar

rinse to help loosen nits, and wash hair with regular shampoo.

Infested eyebrows should be treated with petroleum jelly for several days and the nits should be plucked off with tweezers or fingernails. Infested eyelashes are treated with a thick coating of prescription petroleum ointment, applied twice daily for ten days.

The treatment for body lice is a thorough washing of the entire body and replacement of infected clothing. Clothing and bedding should be washed at 140°F (60°C) and dried at high temperature or dry-cleaned.

Pediculicides

Head and pubic lice infestations usually are treated with insecticidal lotions, shampoos, or cream rinses. These pediculicides should not be used on children under two; near broken skin, eyes, or mucous membranes; in the bathtub or shower; by those with **allergies**, **asthma**, epilepsy, or certain other medical conditions. Itching may not subside for several days following treatment.

All U.S. Food and Drug Administration (FDA)-approved non-prescription pediculicides contain relatively safe and effective pyrethroids. Insecticidal pyrethrins (0.33%) (RID, A-200) are extracts from chrysanthemum flowers. Permethrin (1%)(Nix) is a more stable synthetic pyrethrin. Pyrethroid pediculicides also usually contain 4 percent piperonyl butoxide.

Pyrethroids are applied for a specified length of time (usually ten minutes) and then thoroughly rinsed out. The hair should not be washed for one or two days after treatment. Cream rinse, conditioner, hair spray, mousse, gel, mayonnaise, or vinegar should not be used before treatment or within one week after treatment since these products can reduce pediculicide effectiveness.

Prescription insecticides are used when other lice treatments fail or cannot be used. The following are prescription insecticides, which carry certain risks:

- Malathion (0.5% in Ovide), a neurotoxic organophosphate, was withdrawn from the U.S. market due to an increase in malathion-resistant lice. It was reintroduced in 1999. It is foul-smelling and flammable. Sometimes infested clothing is treated with a 1 percent malathion powder.
- Lindane (1% or higher) (Kwell), an organochloride neurotoxin, can induce seizures and death in susceptible people, even when used according to the directions. In 2003 the FDA required new labeling and a reduction in bottle size.
- Ivermectin (Stromectol), an oral treatment for intestinal parasites, is effective against head lice but as of 2004 had not been approved for that use by the FDA.

Experts disagree about the effectiveness and/or safety of pediculicides. Pediculicides do not kill nits, so nit removal and a second application in seven to 10 days are required. During the 1990s, as schools began requiring children to be lice- and nit-free, the use of pyrethroids rose significantly and the FDA began receiving reports of their ineffectiveness. Pediculicides can be poisonous if used improperly or too frequently and overuse can lead to the proliferation of chemically resistant lice. Pediculicide residue may remain on the hair for several weeks and can cause skin or eye irritations.

Alternative treatment

Olive oil or petroleum ointment may be used to smother head lice. After applying to the hair and scalp, the child's head is covered with a shower cap for four to six hours. The treatment is repeated daily for three or four days. Cutting the hair or shaving the head also may be effective. Other treatments for head lice include:

- olive oil (three parts) and essential oil of lavender (one part)
- herbal shampoos or pomades
- RID Pure Alternative, a nontoxic, hypoallergenic, dye- and fragrance-free product
- a spray containing phenethyl propionate, cedar oil, peppermint oil, and sodium lauryl sulfate (LiceFree)
- cocamide DEA (a lathering agent), triethanolamine (a local irritant), and disodium EDTA (a chelator) (SafeTek) are both a nontoxic pediculicide and a conditioner for combing out lice and nits.

A common herbal treatment for pubic lice consists of pennyroyal oil (25%), garlic oil (25%), distilled water (50%). The mixture is applied to the pubic hair once a day for three days.

Nit removal

Treatment does not kill all lice nits. Hair and pubic lice nits must be removed manually to prevent re-infestation as the eggs hatch. Manual removal alone may treat a lice infestation effectively.

Before removing head lice nits, one of the following procedures may be used:

- a 50 percent vinegar rinse to loosen the nits
- wiping individual locks of hair from base to tip with a cloth soaked in vinegar

- catching live lice with a comb, tweezers, fingernails, or by sticking them with double-sided tape
- an enzymatic lice-egg remover

In addition, hair should be clean, damp, and untangled; clothing should be removed and a towel placed between the hair and shoulders; the hair should be divided into square-inch sections with the use of clips or elastics to divide long hair.

Head lice nits are removed by combing through each hair section from scalp to tip. Between each passing, the comb should be dipped in water and wiped with a paper towel to remove lice and nits. The comb should be held up to the light to be sure it is clean. If necessary the comb may be cleaned with a toothbrush, fingernail brush, or dental floss. Good light and possibly a magnifying glass are required. Long, thick hair may take an hour to comb out thoroughly. Towels and clothing should be washed after combing. The process should be repeated at least twice a week for at least two weeks.

Nits also can be manually removed with any fine-toothed comb, including pet flea combs, a specialized nit comb (LiceMeister, LiceOut), a battery-powered vibrating or anti-static comb, tweezers, baby safety scissors, and fingernails.

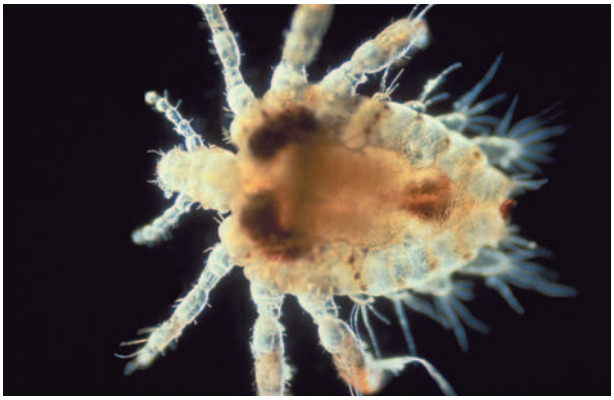
Re-infestation

Re-infestation occurs often with all types of lice due to the following:

- ineffective or incomplete treatment
- chemical-resistant lice
- failure to remove live nits
- failure to treat all infected household members and playmates
- failure to remove nits from clothing, bedding, towels, or other items
- re-infestation from another source

Head lice re-infestation can be prevented by the following:

- repeating lice checks and nit removal daily until no more are found
- notifying school, camp, or daycare, and parents of playmates
- checking and treating household members, playmates, schoolmates, school or daycare staff, and others in close contact with the child
- treating combs and brushes with rubbing alcohol, Lysol, or soapy water above 130°F (54°C)



A close-up view of a body louse. *(Custom Medical Stock Photo Inc.)*

- washing all bedding, clothing, headgear, scarves, and coats with soapy water at 130°F (54°C) and drying with high heat for at least 20 minutes
- washing or vacuuming stuffed animals and other toys
- vacuuming all helmets, carpets, rugs, mattresses, pillows, upholstery, and car seats (Permethrin sprays for treating mattresses, furniture, and other items are not recommended.)
- removing the vacuum-cleaner bag after use, sealing in a plastic bag, and placing in the outside garbage
- dry-cleaning non-washable items or sealing them in a plastic bag for up to four weeks
- repeating treatment if necessary

Re-infestation with body or pubic lice can be prevented by washing underclothes, sleepwear, bedding, and towels in hot, soapy water and drying with high heat for at least 20 minutes. Clothing infected with body lice should be ironed under high heat.

Prognosis

Lice infestations are not usually dangerous. Despite the presence of chemically-resistant lice and the thoroughness required to prevent re-infestation, all lice infestations are eradicated eventually.

Prevention

Prevention of lice infestation depends on adequate personal hygiene and consistently not sharing combs, brushes, hair accessories, hats, towels, or bedding. Hair should be checked weekly for lice and nits.

KEY TERMS

Crabs—An informal or slang term for pubic lice.

Endemic—Natural to or characteristic of a particular place, population, or climate.

Insecticide—Any substance used to kill insects.

Lindane—A benzene compound that is used to kill body and pubic lice. Lindane is absorbed into the louse's central nervous system, causing seizures and death.

Malathion—An insecticide that can be used in 1% powdered form to disinfect the clothes of patients with body lice.

Neurotoxin—A poison that acts directly on the central nervous system.

Nits—The eggs produced by head or pubic lice, usually grayish white in color and visible at the base of hair shafts.

Pediculicide—Any substance that kills lice.

Pediculosis—A lice infestation.

Permethrin—A medication used to rid the scalp of head lice. Permethrin works by paralyzing the lice, so that they cannot feed within the 24 hours after hatching required for survival.

Petroleum jelly or ointment—Petrolatum, a gelatinous substance obtained from oil that is used as a protective dressing.

Piperonyl butoxide—A liquid organic compound that enhances the activity of insecticides.

Pyrethrin, pyrethroid—A naturally occurring insecticide extracted from chrysanthemum flowers. It paralyzes lice so that they cannot feed.

Regular lice checks in schools and "no nit" reentry policies have not been shown to be effective. The American Academy of Pediatrics, the Harvard School of Public Health, and the National Association of School Nurses recommend their elimination, although many healthcare professionals disagree.

Parental concerns

Usually children are not allowed to return to daycare or school until they are lice- and nit-free. The discovery of head lice may cause distress for children and their families. Parents should stay calm and explain to the child:

- what head lice are and how they are transmitted
- that lice are very common and that many—if not most—children have them at some point
- that it is nothing to be embarrassed or ashamed about
- that the infestation is not the child's fault

Scratching or scraping at lice bites may cause **hives** or abrasions that can lead to bacterial skin infections. In developing countries head lice infestations are a significant cause of contagious bacterial infections. Body lice can carry and transmit disease-causing organisms. Although pubic lice do not carry diseases, they often are found in association with other **sexually transmitted diseases**. Crab lice in children may be an indication of sexual activity or abuse.

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American Academy of Pediatrics (AAP). 141 Northwest Point Boulevard, Elk Grove Village, IL 60007-1098. Web site: <www.aap.org>.

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Lipidoses

Definition

Lipidoses are genetic disorders, passed from parents to their children, characterized by defects of the digestive system that impair the way the body uses dietary fat. When the body is unable to properly digest fats, lipids accumulate in body tissues in abnormal amounts.

Description

The digestion, storage, and use of fats (lipids) from foods are complex processes that involve hundreds of chemical reactions in the body. In most people, the body is already programmed by its genetic code to produce all of the enzymes and chemicals necessary to carry out these functions. These genetic instructions are passed from parents to their offspring.

People with lipidoses are born without the genetic codes needed to tell their bodies how to complete a particular part of the fat digestion and utilization process. In most of these disorders, the body does not produce a certain enzyme, or specialized chemical. Over 30 different disorders of fat metabolism are related to genetic defects. Some people can carry the gene for these defects, but be free of symptoms; although the defects are passed from parents to children, the parents often do not have the disorders themselves.

There is great variance in the symptoms, available treatments, and long-term consequences of these conditions. Some of the conditions become apparent shortly after the infant is born. In other lipid disorders, symptoms may not develop until adulthood. For most of the lipidoses, diagnosis is suspected based on symptoms and **family** history. Tests of blood, urine, and tissue can be used to confirm the diagnosis. Genetic testing can be used, in some cases, to identify the defective gene. Some of these disorders can be controlled with changes in the diet, medications, or enzyme supplements. However, for many of these diseases, no treatment is available. Some may cause death in childhood or contribute to a shortened life expectancy. This section focuses on some of the most common or most serious lipidoses.

Demographics

Lipidoses are very rare. The number of people affected depends on the disease, but for many diseases incidence is as little as one in 40,000 people. Some diseases have a higher prevalence in specific populations.

Fabry's disease

Causes and symptoms

Approximately one in every 40,000 males is born with Fabry's disease. This condition has an X-linked, recessive pattern of inheritance, meaning that the defective gene is carried on the X chromosome. A female who carries a defective recessive gene on one of her two X chromosomes will not have the disease because she also has one good X chromosome. However, she has a 50 percent chance of passing the defective gene to her sons. The sons inheriting one defective gene will develop the disorder because a male has only one X chromosome, which he receives from his mother and one Y chromosome from his father. The mother also has a 50 percent chance of passing the defective recessive gene to her daughters who will be carriers of the disorder (like their mother), but will not show symptoms of the disease. Some female carriers of Fabry's disease show mild signs of the disorder, especially cloudiness of the cornea.

The gene that is defective in Fabry's disease causes a deficiency of the enzyme alpha-galactosidase A. Without this enzyme, fatty compounds start to line the blood vessels. The collection of fatty deposits eventually affects blood vessels in the skin, heart, kidneys, and nervous system. The first symptoms in childhood are **pain** and discomfort in the hands and feet brought on by **exercise**, **fever**, stress, or changes in the weather. A raised rash of dark red-purple spots is common, especially on skin between the waist and the knees. Other symptoms include a decreased ability to sweat and changes in the cornea or outer layer of the eye. Although the disease begins in childhood, it progresses very slowly. Kidney and heart problems develop in adulthood.

Diagnosis

A diagnosis of Fabry's disease can be confirmed by a blood test to measure for alpha-galactosidase A. Women who are carriers of the defective gene can also be identified by a blood test.

Treatment

Treatment focuses on prevention of symptoms and long-term complications. Daily doses of diphenylhydantoin (Dilantin) or carbamazapine (Tegretol) can prevent or reduce the severity of pain in the hands and feet associated with this condition. A diet low in sodium and protein may be beneficial to those individuals who have some kidney complications. If kidney problems progress, kidney dialysis or kidney transplantation may be required. Enzyme replacement therapy is being explored.

Prognosis

Although patients with Fabry's disease usually survive to adulthood, they are at increased risk for **stroke**, heart attack, and kidney damage.

Gaucher disease

Causes and symptoms

Gaucher (pronounced go-shay) disease is the most common of the lipid storage disorders. It is found in populations all over the world (20,000–40,000 people have a type of the disease), and it occurs with equal frequency in males and females. Gaucher disease has a recessive pattern of inheritance, meaning that a person must inherit a copy of the defective gene from both parents in order to have symptoms of the disease. The genetic defect causes a deficiency of the enzyme glucocerebrosidase that is responsible for breaking down a certain type of fat and releasing it from fat cells. These fat cells begin to crowd out healthy cells in the liver, spleen, bones, and nervous system. Symptoms of Gaucher disease can start in infancy, childhood, or adulthood.

Three types of Gaucher disease have been identified, but there are many variations in how symptoms develop. Type 1 is the most common and affects both children and adults. It occurs much more often in people of Eastern European and Russian Jewish (Ashkenazi) ancestry, affecting one out of every 450 live births in this population. The first signs of the disease include an enlarged liver and spleen, causing the abdomen to swell. Children with this condition may be shorter than normal. Other symptoms include tiredness, pain, bone deterioration, broken bones, anemia, and increased bruising. Type 2 Gaucher disease is more serious, beginning within the first few months after birth. Symptoms, which are similar to those in type 1, progress rapidly, but also include nervous system damage. Symptoms of type 3 Gaucher disease begin during early childhood with symptoms like type 1. Unlike type 2, the progress of the disease is slower, although it also includes nervous system damage.

Diagnosis

Gaucher disease may be suspected, based on symptoms, and is confirmed with a blood test for levels of the deficient enzyme. Samples of tissue from an affected area may also confirm a diagnosis.

Treatment

The symptoms of Gaucher disease can be stopped and even reversed by treatment with enzyme replacement injections. Two enzyme drugs available are alglucerase (Ceredase) and imiglucerase (Cerezyme). Other treatments address specific symptoms such as anemia, broken bones, or pain.

Prognosis

The pain and deformities associated with symptoms can make coping with this illness very challenging for individuals and families. With treatment and control of symptoms, people with type 1 Gaucher disease may lead fairly long and normal lives. Most infants with type 2 die before age two. Children with type 3 Gaucher disease may survive to **adolescence** and early adulthood.

Krabbe's disease

Causes and symptoms

Krabbe's disease is caused by a deficiency of the enzyme galactoside beta- galactosidase. It has a recessive pattern of inheritance and is believed to occur in one out of 40,000 births in the United States. This condition, which is also called globoid cell leukodystrophy or Krabbe leukodystrophy, is characterized by acute nervous system degeneration. It develops in early infancy with initial symptoms of irritability, **vomiting**, and episodes of partial unconsciousness. Symptoms progress rapidly to seizures, difficulty swallowing, blindness, deafness, **mental retardation**, and paralysis.

Treatment

No treatment is available.

Prognosis

Children born with Krabbe's disease die in infancy.

Niemann-pick disease

Causes and symptoms

At least five different forms of Niemann-Pick disease (NPD) have been identified. The different types seem to be related to the activity level of the enzyme sphingomyelinase. In patients with types A and B NPD, there is a build-up of sphingomyelin in cells of the brain, liver, spleen, kidney, and lung. Type A is the most common form of NPD and the most serious, with death usually occurring by the age of 18 months. Symptoms develop within the first few months of life and include poor appetite, failure to grow, enlarged liver and spleen, and the appearance of cherry red spots in the retina of the eye. Type B develops in infancy or childhood with symptoms of mild liver or spleen enlargement and lung problems. Types C and D NPD are related to cholesterol transfer out of cells. Children with types C or D grow normally in early childhood, but eventually develop difficulty in walking and loss of muscle coordination. Ultimately, the nervous system becomes severely damaged and these patients die. Type C occurs in any population, while type D has been identified only in individuals from Nova Scotia, Canada.

Diagnosis

Diagnosis is confirmed by analyzing a sample of tissue. Prenatal diagnosis of types A and B NPD can be done with **amniocentesis** or chorionic villi sampling.

Treatment

Treatment consists of supportive care to deal with symptoms and the development of complications. Bone marrow transplantation is being investigated as a possible treatment. Low-cholesterol diets may be helpful for patients with types C and D.

Prognosis

Patients with type A NPD usually die within the first 18 months of life. Type B patients generally live to adulthood but suffer from significant liver and lung problems. With types C and D NPD, there is significant nervous system damage leading to severe **muscle spasms**, seizures, and eventually coma and death. Some patients with types C and D die in childhood, while less severely affected individuals may survive to adulthood.

Refsum's disease

Causes and symptoms

Refsum's disease has a recessive pattern of inheritance and affects populations from Northern Europe, particularly Scandinavians. It is due to a deficiency of phytanic acid hydroxylase, an enzyme that breaks down a fatty acid called phytanic acid. This condition affects the nervous system, eyes, bones, and skin. Symptoms, which usually appear by age 20, include vision problems (retinitis pigmentosa and rhythmic eye movements, or **nystagmus**), loss of muscle coordination, loss of sense of smell (anosmia), pain, **numbness**, and elevated protein in the cerebrospinal fluid.

Treatment

A diet free of phytanic acid (found in dairy products, tuna, cod, haddock, lamb, stewed beef, white bread, white rice, boiled potatoes, and egg yolk) can reduce some of the symptoms. Plasmapheresis—a process where whole blood is removed from the body, processed through a filtering system, and then returned to the body—may be used to filter phytanic acid from the blood.

Tay-Sachs disease

Causes and symptoms

Tay-Sachs disease (TSD) is a fatal condition caused by a deficiency of the enzyme hexosaminidase A (Hex-A). The defective gene that causes this disorder is found in roughly one in 250 people in the general population. However, certain populations have significantly higher rates of TSD. French-Canadians living near the St. Lawr-

ence River and in the Cajun regions of Louisiana are at higher risk of having a child with TSD. The highest risk seems to be in people of Eastern European and Russian Jewish (Ashkenazi) descent. Tay-Sachs disease has a recessive pattern of inheritance, and approximately one in every 27 people of Jewish ancestry in the United States carries the TSD gene. Symptoms develop in infancy and are due to the accumulation of a fatty acid compound in the nervous system. Early symptoms include loss of vision and physical coordination, seizures, and mental retardation. Eventually, the child develops problems with breathing and swallowing. Blindness, paralysis, and death follow.

Diagnosis

Carriers of the Tay-Sachs related gene can be identified with a blood test. Amniocentesis or chorionic villi sampling can be used to determine if the fetus has Tay-Sachs disease.

Treatment

There is no treatment for Tay-Sachs disease. Parents who are identified as carriers may want to seek genetic counseling. If a fetus is identified as having TSD, parents can then consider their options.

Prognosis

Children born with Tay-Sachs disease become increasingly debilitated; most die by about age four.

Wolman's disease

Causes and symptoms

Wolman's disease is caused by a genetic defect with a recessive pattern of inheritance that results in a deficiency of an enzyme that breaks down cholesterol. This causes large amounts of fat to accumulate in body tissues. Symptoms begin in the first few weeks of life and include an enlarged liver and spleen, adrenal calcification (hardening of adrenal tissue due to deposits of calcium salts), and fatty stools.

Treatment

No treatment is available.

Prognosis

Death generally occurs before six months of age.

KEY TERMS

Amniocentesis—A procedure performed at 16-18 weeks of pregnancy in which a needle is inserted through a woman's abdomen into her uterus to draw out a small sample of the amniotic fluid from around the baby for analysis. Either the fluid itself or cells from the fluid can be used for a variety of tests to obtain information about genetic disorders and other medical conditions in the fetus.

Chorionic villus sampling—A procedure performed at 10 to 12 weeks of pregnancy in which a needle is inserted either through the mother's vagina or abdominal wall into the placenta to withdraw a small amount of chorionic membrane from around the early embryo. The amniotic fluid can be examined for signs of chromosome abnormalities or other genetic diseases.

Lipids—Organic compounds not soluble in water, but soluble in fat solvents such as alcohol. Lipids are stored in the body as energy reserves and are also important components of cell membranes. Commonly known as fats.

Recessive—Refers to an inherited trait that is outwardly obvious only when two copies of the gene for that trait are present. An individual displaying a recessive trait must have inherited one copy of the defective gene from each parent.

X-linked—A gene carried on the X chromosome, one of the two sex chromosomes.

Prevention

There is no known way to prevent lipidoses. Couples who have family histories of genetic defects can undergo genetic testing and counseling to see if they are at risk for having a child with one of the lipidoses disorders. During pregnancy, cell samples can be collected from the fetus using amniocentesis or chorionic villi sampling. The results of these tests can indicate if the developing fetus has a lipidosis disorder.

Parental concerns

Lipidoses have a variety of different symptoms and progressions. There are treatments for some—but not all—lipidoses. Treating the symptoms is always an integral part of lessening the impact of the disease. If parents have one child with a lipidoses disorder and are consider-

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National Organization for Rare Diseases. 55 Kenosia Avenue, PO Box 1968, Danbury, CT 06813-1968. (203) 744-0100 or 1-800- 999-6673. Fax: (203) 798-2291. Web site: <www.rarediseases.org>.

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Lisping

Definition

A lisp is a functional speech disorder that involves the inability to correctly pronounce one or more sibilant consonant sounds, usually *s* or *z*.

Description

Lisping is a speech disorder characterized by the inability to correctly pronounce the sounds of *s* or *z*, known as the sibilant consonants. Usually *th* sounds are substituted for the sibilants. The word "lisp," for example, would be pronounced "lithp" by someone with this speech disorder.

Many children lisp at certain stages of speech development, especially when they lose their front primary teeth. Lisping is, therefore, sometimes called a developmental phonetic disorder. Frontal or interdental lisp is produced when the tongue protrudes through the front teeth when teeth are missing and is the most familiar type of lisp. Sibilant production may be interfered with in a number of other ways as well. These are all classified as lisping and include excessive pressure by the tongue against the teeth, the tongue held too far back along the midline of the palate, and a "substitute hiss" produced in the throat or larynx.

Sometimes children with functional **speech disorders** have problems making other sounds, such as *sh*, *l*, *r*, and *ch*. When a child cannot produce these sounds correctly, the condition is usually not considered a true lisp, but is a functional speech disorder.

Children can have a functional speech disorder as well as a developmental phonological disorder. The latter is not a matter of being able to physically make a specific sound but is a language disorder. These children have trouble organizing the sounds of speech in common patterns and may consistently replace one sound with another. For example, a child may say "wun" instead of "sun" or "doe" instead of "so."

There are four main types of lisps.

- Interdental lisp—occurs when the tongue protrudes between the front teeth and the *s* or *z* is pronounced like *th*.
- Dentalized lisp or dentalized production—occurs when the tongue pushes against the front teeth.
- Lateral lisp—sounds wet because the air flows around the tongue, which is in the normal position to produce the *l* sound.
- Palatal lisp—the middle of the tongue touches the soft palate, or roof of the mouth, when trying to produce the *s* sound.

Demographics

According to the National Institute on Deafness and Other Communication Disorders, about one in six people in the United States (42 million adults and children) has a communication disorder. Of them, 14 million have a speech, voice, or language disorder that is not linked to hearing loss. Functional speech disorders with no known cause, such as lisping, affect 10 percent of the population; 8–9 percent serious enough to require treatment. Nearly 5 percent of first graders have functional speech disorders, and 50–70 percent of all children with functional speech disorders struggle academically throughout elementary school and high school.

Causes and symptoms

As a functional speech disorder, lisping has no clear known cause. It is often referred to as a speech delay of unknown origin. Structural irregularities of the tongue, palate, or teeth (including abnormalities in the number or position of the teeth) may be implicated in lisping, but they generally are not the main causes. Mild hearing loss involving high frequencies may also impair a child's ability to hear language correctly and be able to repeat phonetic sounds. In some cases, a child with no physical abnormality will develop a lisp. It has been thought that some of these children may be imitating another child or an adult who lisps.

Lisping is also associated with immature development. Some children will adopt a lisp as a means of gaining attention. Other children will begin to lisp after they have experienced unusual stress or trauma. This behavior is part of a regression into a more secure period and can include other types of regressive behaviors such as bed wetting or wanting to **sleep** with the light on in the bedroom.

One theory of the cause of lisping is the result of tongue thrusting, a physiological behavior that causes the tongue to flatten and thrust forward during swallowing and speaking. It is suggested that **thumb sucking**, overuse of pacifiers, bottle feeding, and recurrent upper respiratory illnesses cause tongue thrusting. Thumbs (or fingers), artificial nipples, and pacifiers keep the tongue flat and do not allow the muscles of the tongue to develop in a normal fashion. When the child speaks, the tongue shoots forward, creating a lisp.

Frequent upper respiratory illnesses often stuff the nose, forcing these children to breathe through their mouths. The sounds that they make when they speak may be thick and garbled, and may encourage lisping. Closing the mouth and teeth to make *s* or *z* sounds cuts off the breath, so children compensate by trying to speak without closing their mouths completely. Thus, a lisp develops.

When to call the doctor

The interdental lisp and the dentalized lisp are common in normal speech development. However, if they persist well past four-and-one-half years and garble the speech so that the child is not understood, he or she should be evaluated. The evaluation will determine if there is a physiological basis for the lisp and identify the type of lisp. In some cases, the child will be evaluated and observed for several months or longer to see if the condition can be outgrown. Lateral and palatal lisps are not found in typical speech development and should be evaluated by a speech-language pathologist. If untreated, lisping can persist into adulthood.

For some children, everything they say seems to be interdental. In these cases, there may be an obstruction of the nose because of infection, allergy, enlarged adenoids, or other facial problems. Excessive interdental speech can also be related to mouth breathing and sucking habits. These children should be seen by a physician to treat the health problems and then referred to a speech-language pathologist to correct the lisp.

Diagnosis

A physician can determine whether there are structural irregularities within the mouth or problems with the child's hearing, and can treat related **allergies** and nasal problems. However, true assessment of a child's ability to make speech sounds must be done by a speech-language pathologist. The child's medical history will be taken and the speech-language pathologist will examine the anatomy of the child's mouth and the movements it can make. Next, the child's speech and reading aloud is often recorded for later analysis. This speech sample will also yield information about the quality of the child's voice, how fluent speech occurs, and the child's semantic and physical sound- making skill.

Treatment

Typical treatment is called articulation therapy. The speech-language pathologist finds out whether the child can hear proper speech sounds, and proceeds to read a list of words with specific sounds that the child is having trouble articulating. Lists of contrast words are also read so that the child can hear the subtle differences in word sounds. Therapy then moves to working on the position in the word where the sound occurs; that is, at the beginning, in the middle, or at the end. Specific word exercises follow, beginning with single sounds, then syllables, and moving on to words, phrases, and sentences. Finally, the child participates in controlled conversations such as talking casually during a meal.

Prognosis

Most lisps are developmental and resolve themselves in children by the time they are about five to eight years old. If they last longer or are of a specific type, speech therapy is recommended. The outcome of speech therapy is usually quite good. Depending on the specifics of the therapy and the nature of the lisp, treatment can be relatively short term, lasting only a few months. Some cases may take a year or more.

Prevention

Parents can reduce the risk of a lisp developing because of tongue thrusting by restricting **pacifier use** or choosing to breastfeed their babies. They can also speak clearly in complete sentences around their children and not use baby talk. They should treat allergies and respiratory illnesses immediately to keep the nose open and breathing free. The child's hearing and teeth should be checked periodically to make sure he or she can hear speech clearly and form words correctly. Parents can also encourage the musculature of the mouth by showing children how to drink from straws and how to blow bubbles. In addition, playing word and naming games encourages good speech development and stimulates learning.

Parental concerns

In many families, a child's lisp goes unnoticed, especially if it does not interfere with understanding what the child is saying. These children may grow up content to keep a lisp, feeling that it is a specific part of what makes them who they are, just as some people keep a gap between their front teeth and see it as distinct characteristic. (Lauren Hutton kept the gap between her teeth even as she became a top model, and Boris Karloff had one of the most famous lisps in the world.)

Outside of the home, some children may be teased by other children or feel embarrassed to speak up in the classroom. They may have trouble spelling or even reading because they cannot make some of the sounds necessary to read and write well. These children may have serious **self-esteem** issues related to their lisps. In these cases, seeing a speech-language pathologist early in their lives and correcting their lisps could bolster self-confidence and ability to learn.

Still other families may think that a child's lisp is endearing and cute. They may even encourage the child to continue lisping because he or she receives positive regard whenever the lisping sounds are made. Sometimes, these same families suddenly decide that lisping is no longer cute and want their children to drop their lisps and grow up. These children still can benefit from speech therapy, but they may become resistant to treatment because they are confused about the abrupt change in the family's behavior. In this case, counseling is recommended in addition to sessions with a speech-language pathologist.

A lisp can be a source of distress for adolescent boys and young men who may be told that they are gay because they lisp. A functional speech disorder has no connection with a person's sexual orientation. Many young men, although they **fear** it may be too late, seek out speech-language pathologists as adults to correct their lisps because of this teasing.

KEY TERMS

Palate—The roof of the mouth.

Tongue thrusting—A physiological behavior that causes the tongue to flatten and thrust forward during swallowing and speaking.

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Council for Exceptional Children. Division for Children with Communication Disorders. 1920 Association Drive Reston, VA 22091. (703) 620-3660.

National Institute on Deafness and Other Communication Disorders National Institutes of Health. 31 Center Drive, MSC 2320 Bethesda, MD 20892-2320.

Janie Franz

Listeriosis

Definition

Listeriosis is an illness caused by the bacterium *Listeria monocytogenes* that is acquired by eating contaminated food. The organism can spread to the blood stream and central nervous system. In women who contract listeriosis while pregnant, the disease often causes miscarriage or stillbirth.

Description

Listeriosis is caused by an infection with the bacterium *Listeria monocytogenes*. These bacteria can be carried by many animals and birds, and they have been found in soil, water, sewage, and animal feed. Five out of every 100 people carry *Listeria monocytogenes* in their intestines. Listeriosis is considered a food-borne illness because most people are probably infected after eating food contaminated with *Listeria monocytogenes*. However, a woman can pass the bacteria to her baby during pregnancy. In addition, there have been a few cases where workers have developed *Listeria* skin infections by touching infected calves or poultry.

In the 1980s, the United States government began taking measures to decrease the occurrence of listeriosis. Subsequently, processed meats and dairy products were tested for the presence of *Listeria monocytogenes*. The Food and Drug Administration (FDA) and the Food Safety and Inspection Service (FSIS) can legally prevent food from being shipped, or order food recalls, if they detect any *Listeria* bacteria. These inspections, in combination with the public education regarding the proper handling of uncooked foods, appear to be helping. Nonetheless, as of 2004, about 2,500 individuals become seriously ill from *Listeria* annually, with about 500 deaths.

Demographics

Persons at particular risk for listeriosis include the elderly, pregnant women, newborns, and those with a weakened immune system (called immunocompromised). Risk is increased when a person suffers from diseases such as **AIDS**, **cancer**, kidney disease, **diabetes mellitus**, or by the use of certain medications. Infection is most common in babies younger than one month old and adults over 60 years of age. Pregnant women account for 27 percent of the cases, and immunocompromised persons account for almost 70 percent. Persons with AIDS are 280 times more likely to get listeriosis than others.

Causes and symptoms

As noted, persons become infected with *Listeria monocytogenes* by eating contaminated food. *Listeria* has been found on raw vegetables, fish, poultry, raw (unpasteurized) milk, fresh meat, processed meat (such as deli meat, hot dogs, and canned meat), and certain soft cheeses. Listeriosis outbreaks in the United States since the 1980s have been linked to cole slaw, milk, Mexican-style cheese, undercooked hot dogs, undercooked chicken, and delicatessen foods. Unlike most other bacteria, *Listeria monocytogenes* does not stop growing when food is in the refrigerator; its growth is merely slowed. Fortunately, typical cooking temperatures and the pasteurization process do kill this bacteria.

Listeria bacteria can pass through the wall of the intestines, and from there they can get into the blood stream. Once in the blood stream, they can be transported anywhere in the body but are commonly found the central nervous system (brain and spinal cord); and in pregnant women they are often found in the placenta (the organ which connects the baby's umbilical cord to the uterus). *Listeria monocytogenes* live inside specific white blood cells called macrophages. Inside macrophages, the bacteria can hide from immune responses and become inaccessible to certain **antibiotics**. *Listeria* bacteria are capable of multiplying within macrophages and then may spread to other macrophages.

After people consume food contaminated with this bacteria, they may see symptoms of infection 11 to 70 days later. Most people do not get any noticeable symptoms. Scientists suspect that *Listeria monocytogenes* can cause upset stomach and intestinal problems just like other food-borne illnesses. Persons with listeriosis may develop flu-like symptoms such as **fever**, **headache**, **nausea and vomiting**, tiredness, and **diarrhea**.

Pregnant women experience a mild, flu-like illness with fever, muscle aches, upset stomach, and intestinal problems. They recover, but the infection can cause miscarriage, premature labor, early rupture of the birth sac, and stillbirth. Half of the newborns infected with *Listeria* die from the illness.

There are two types of listeriosis in the newborn baby: early-onset disease and late-onset disease. Early-onset disease refers to a serious illness that is present at birth and usually causes the baby to be born prematurely. Babies infected during the pregnancy usually have a blood infection (sepsis) and may have a serious, whole body infection called granulomatosis infantisepticum. When a full-term baby becomes infected with *Listeria* during **childbirth**, that situation is called late-onset disease. Commonly, symptoms of late-onset listeriosis

appear about two weeks after birth. Babies with late-term disease typically have **meningitis** (inflammation of the brain and spinal tissues); yet they have a better chance of surviving than those with early-onset disease.

Immunocompromised adults are at risk for a serious infection of the blood stream and central nervous system (brain and spinal cord). Meningitis occurs in about half of the cases of adult listeriosis. Symptoms of listerial meningitis occur about four days after the flu-like symptoms and include fever, personality change, uncoordinated muscle movement, tremors, muscle contractions, seizures, and slipping in and out of consciousness.

Listeria monocytogenes causes endocarditis in about 7.5 percent of the cases. Endocarditis is an inflammation of heart tissue due to the bacterial infection. Listerial endocarditis causes death in about half of the patients. Diseases which have been caused by *Listeria monocytogenes* include brain abscess, eye infection, hepatitis (liver disease), peritonitis (abdominal infection), lung infection, joint infection, arthritis, heart disease, bone infection, and gallbladder infection.

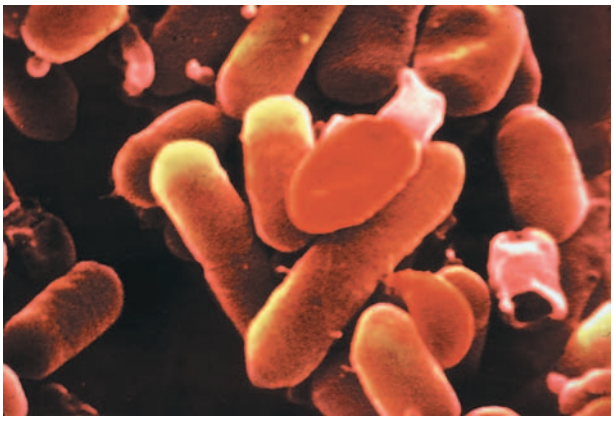
Diagnosis

Listeriosis may be diagnosed and treated by infectious disease specialists and internal medicine specialists. The diagnosis and treatment of this infection should be covered by most insurance providers.

The only way to diagnose listeriosis is to isolate *Listeria monocytogenes* from blood, cerebrospinal fluid, or stool. A sample of cerebrospinal fluid is removed from the spinal cord using a needle and syringe. This procedure is commonly called a spinal tap. The amniotic fluid (the fluid which bathes the unborn baby) may be tested in pregnant women with listeriosis. This sample is obtained by inserting a needle through the abdomen into the uterus and withdrawing fluid. *Listeria* grows well in laboratory media, and test results can be available within a few days.

Treatment

Listeriosis is treated with the antibiotics ampicillin (Omnipen) or sulfamethoxazole-trimethoprim (Bactrim, Septra). Because the bacteria live within macrophage cells, treatment may be difficult, and the treatment periods may vary. Usually, pregnant women are treated for two weeks; newborns, two to three weeks; adults with mild disease, two to four weeks; persons with meningitis, three weeks; persons with brain abscesses, six weeks; and persons with endocarditis, four to six weeks.



Scanning electron micrograph (SEM) scan of lysteria monocytogenes. *(© CNRI/Phototake.)*

Patients are often hospitalized for treatment and monitoring. Other drugs may be provided to relieve **pain** and fever and to treat other reactions to the infection.

Prognosis

The overall death rate for listeriosis is 26 percent. This high death rate is due to the serious illness suffered by newborns, the elderly, and immunocompromised persons. Healthy adults and older children have a low death rate. Complications of *Listeria* infection include: meningitis, sepsis, miscarriage, stillbirth, **pneumonia**, shock endocarditis, abscess (localized infection) formation, and eye inflammation.

Prevention

As of the early 2000s the United States government has done much to prevent listeriosis. Persons at extremely high risk (pregnant women, immunocompromised persons, etc.) must use extra caution. High risk persons should avoid soft cheeses, such as Mexican cheese, feta, Brie, Camembert, and blue cheese (cottage cheese is safe); thoroughly cook leftovers and ready-to-eat foods (such as hot-dogs); and avoid foods from the deli.

For all people, the risk of listeriosis can be reduced by taking these precautions:

- Completely cook all meats and eggs.
- Carefully wash raw vegetables before eating.
- Keep raw meat away from raw vegetables and prepared foods. After cutting raw meat, wash the cutting board with detergent before using it for vegetables.

KEY TERMS

Abscess—A localized collection of pus in the skin or other body tissue caused by infection.

Immunocompromised—A state in which the immune system is suppressed or not functioning properly.

Macrophage—A large white blood cell that engulfs and digests foreign invaders, such as bacteria and viruses, in an attempt to stop them from causing disease within the body.

Meningitis—An infection or inflammation of the membranes that cover the brain and spinal cord. It is usually caused by bacteria or a virus.

Sepsis—A severe systemic infection in which bacteria have entered the bloodstream or body tissues.

- Avoid drinking unpasteurized milk or foods made from such milk.
- Wash hands thoroughly after handling raw meat.
- Follow the instructions on food labels. Observe food expiration dates and storage conditions.

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Liver function test *see* **Bilirubin test**

Lockjaw *see* **Tetanus**

Louis-Bar syndrome *see* **Ataxia telangiectasia/chromosome breakage disorders**

Low blood sugar *see* **Hypoglycemia**

Lumbar puncture *see* **Cerebrospinal fluid (CSF) analysis**

Lung function test *see* **Pulmonary function test**

Lying

Definition

A lie is any deliberate deviation from the truth; it is a falsehood communicated with the intention to mislead or deceive.

Description

Lies differ in type, incidence, magnitude and consequence, with many gradations of severity, from harmless exaggeration and embellishment of stories, to intentional and habitual deceit. Behavioral scientist Wendy Gamble identified four basic types of lies for a University of Arizona study in 2000:

- Prosocial: Lying to protect someone, to benefit or help others.
- Self-enhancement: Lying to save face, to avoid embarrassment, disapproval or punishment.
- Selfish: Lying to protect the self at the expense of another, and/or to conceal a misdeed.
- Antisocial: Lying to hurt someone else intentionally.

Lying is considered by most child development specialists to be a natural developmental occurrence in childhood. Though there is no empirical data about how children learn to lie, parental honesty is recognized as a primary influence on the development of truthfulness in children.

Preschool

Making up stories is part of a normal fantasy life for young children. It is a positive sign of developing **intelligence** and of an active and healthy imagination. Preschool children who are beginning to express themselves through language are not yet able to make a clear

distinction between reality and make-believe. Storytelling at this age is seldom an intentional effort to deceive. When preschool children do engage in intentional deceit, it is usually to avoid reprimand. They are concerned with pleasing the parent, and may **fear** the punishment for admitting a mistake or misdeed.

Many children are socialized by their parents at a very early age to tell "white"; lies to avoid hurting another's feelings. "White lies" or "fibs" are commonplace in many households and social settings and are observed and imitated by children. The incidence of prosocial or "white lies," tends to increase in children as they grow older.

Dr. Kang Lee of the Department of Psychology at Queens University in Kingston, Ontario, Canada, observed young children telling so-called "white lies" to avoid disappointing the researcher. Such prosocial lying behavior occurred in children as young as age three. Dr. Lee's research found that over 60 percent of the 400 boys and girls he studied would pretend to be pleased when asked how they liked a used bar of soap, given as a prize after playing a game with researchers. When parents instructed the children to "be polite" when the researcher asked if they liked the soap, as many as 80 percent of these children, ages three to 11 years of age were dishonest.

Dr. Michael Lewis of Robert Wood Johnson Medical School, has found that as many as 65 percent of the children he studied had learned to lie by age two and one half. This research also reveals a correlation between higher IQ and the incidence of lying in children.

School-age children

Children from age five or six have learned the difference between lies and truth. The motives for lying in this age group are more complex. Prosocial lying may increase, particularly among peers, to avoid hurting another's feelings. In addition, if a parent's expectations for the child's performance are too high, the child may engage in self-enhancing lies out of fear of censure. School-age children also experiment with selfish lies to avoid punishment, or to gain advantage. They are testing the limits as they try to understand how the rules work and what the consequences may be for stepping out of bounds.

By age seven children have developed the ability to convincingly sustain a lie. This capacity has serious implications with regard to children's competency to testify in a court of law. The veracity of child witnesses and their understanding of the concept of an oath are important research issues. Children at this age recognize the difference between what they are thinking and how they can manipulate the thinking of another to serve their own ends.

The type and frequency of lies and the reasons why a child may be dishonest are also related to their stage of **moral development**.

Children progress sequentially through several stages of moral development, according to psychologist Lawrence Kohlberg:

- avoiding punishment
- doing right for self-serving reasons
- fitting in with and pleasing others
- doing one's duty
- following agreed upon rules
- acting on principles

Adolescents are developmentally involved in becoming independent persons. They are working hard to establish their own identity, one that is separate from that of their parents. Peer approval is more important than parental approval during **adolescence**. Conflicts during these years between parental control versus personal autonomy may lead to increased lying to preserve a sense of separation and power from parents, teachers, and other authority figures. Adolescents may also lie to cover up serious behavior problems. A discerning parent will attempt to discover the motive behind the lie.

Common problems

Childhood lying has many causes, including the need to maintain parental approval, to gain attention, to avoid disappointing others, to evade the consequences of misbehavior, or to avoid responsibility. Older children may lie as a means of breaking away from parental control. Issues of **self-esteem**, fear of consequences, the desire to have one's own way, the need to gain attention, or to protect oneself from harm, are also a factor. Difficult circumstances in the home and social environment of the child may increase the likelihood of problem lying.

Early intervention in the case of compulsive lying may reduce the risk of the child developing a life-time habit of deceit. Children who are chronic liars are often found to engage in other antisocial behaviors. If a child's lying is accompanied by fighting, cheating, **stealing**, cruelty, and other impulse control problems, appropriate intervention is required. Lying that is consistently self-serving with no prosocial motive is a serious issue. Lying with malice and without any sign of remorse may indicate that the child has not yet developed a moral con-

science, and may need help to move toward a higher stage of moral development, one that includes a concern for the impact of one's actions upon others.

Children become more adept liars with practice. As they grow older it may become increasingly difficult for a parent, teacher or caregiver to detect dishonesty. Close observation and familiarity with the child, as well as an understanding of their developmental stage, are critical to the diagnosis of problem lying.

Most children with the benefit of a loving **family** environment, one where honesty is valued and modeled and dishonesty is appropriately challenged, will more often than not come to recognize that lying is not an acceptable behavior. Early and appropriate intervention when problem lying persists will increase the possibility that the child will choose honesty in subsequent interactions.

Children may observe much routine dishonesty in the home, school and surrounding culture. Parental examples of honesty in interpersonal relationships are critical if a child is to develop an ethic of truthfulness. Children commonly experiment with lying in the natural course of development. They need help recognizing and understanding the distinction between prosocial and antisocial lying.

Exaggeration and embellishment when relating incidents or telling stories, and the so-called "white lies," told to avoid disappointing or hurting others feelings, do not have the negative, antisocial consequences of serious lying. Parents should intervene when the lying is of a serious nature and explain the impact of dishonesty on another's feelings. This will help the child to develop a moral sense of right and wrong and to value honesty in interpersonal relationships.

Parental concerns

Repetitive lying can develop into a serious habit leading to adjustment problems later in life. Lying that persists and worsens year after year is cause for concern. Chronic lying is often accompanied by other antisocial behaviors. Adolescents may lie to cover up illicit drug or alcohol abuse. Early parental intervention in situations of serious lying may interrupt the formation of a habit of lying in young children. Parents who model truth telling and praise honesty will encourage trust in the parent-child relationship.

When to call the doctor

Serious and repetitive lying may require the professional intervention of a school psychologist or a community mental health agency. Counseling may help to uncover any underlying conditions such as attention-deficit/hyperactivity disorder (AD/HD), **bipolar disorder**, or learning disabilities. Pathological lying often accompanies serious psychiatric problems such as **conduct disorder** or **antisocial personality disorder**, which normally have their onset during adolescence. Children who use lying as a primary means of avoiding personal responsibility, particularly in adolescence, may be attempting to cover up more serious problems with substance abuse.

KEY TERMS

Antisocial personality disorder—A disorder characterized by a behavior pattern that disregards for the rights of others. People with this disorder often deceive and manipulate, or their behavior might include aggression to people or animals or property destruction, for example. This disorder has also been called sociopathy or psychopathy.

Conduct disorder—A behavioral and emotional disorder of childhood and adolescence. Children with a conduct disorder act inappropriately, infringe on the rights of others, and violate societal norms.

Prosocial behaviors—Social behavior characterized by positive, cooperative, and reciprocal social exchanges.

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Lyme disease

Definition

Lyme disease is an inflammatory disease transmitted through the bite of a deer tick carrying the spiral-shaped bacterium *Borrelia burgdorferi*. Symptoms can include skin rash, joint inflammation, **fever**, **headache**, fatigue, and muscle **pain**. Lyme disease is also called Lyme borreliosis.

Description

Lyme disease is an inflammatory, systemic disease, meaning that it affects multiple body systems. Although clinical signs of Lyme disease have been reported for more than 100 years, the disease was not recognized as a distinct illness until 1975, when a cluster of unusual arthritis cases in Lyme, Connecticut, led physicians to discover that town residents living near heavily wooded areas were most affected by arthritis and other symptoms. Tick **bites** were then linked to the cause of the arthritis cases. *Borrelia burgdorferi*, the spiral-shaped bacterium called a spirochete, that causes Lyme disease, was not discovered until 1981 by Willy Burgdorfer.

Although Lyme disease is easily treated, it is not easily diagnosed, since symptoms are often attributed to other conditions. If not treated early and properly with **antibiotics**, Lyme disease can have long-term and disabling effects. In its early stages, Lyme disease affects the skin and produces flu-like symptoms; the disease spreads to the joints and nervous system in its later stages.

Transmission

Lyme disease is a vector-borne disease, meaning that it is transmitted from one host to another by a carrier—called a vector—that transmits but does not become infected with the disease. In the United States, the deer tick in the genus *Ixodes* is the vector for *Borrelia burgdorferi* and Lyme disease transmission. Lyme dis-

ease is transmitted when a tick carrying the *Borrelia burgdorferi* bacterium bites a human to feed on blood. The bacterium is transferred from the intestines of the tick through the mouthparts and into the bloodstream while the tick is feeding. Ticks are most likely to transmit *Borrelia burgdorferi* after remaining attached and feeding for two or more days. In most areas, ticks are most active from April to October, but in milder climates, ticks may bite year-round.

During their two-year life cycle and three life stages (larva, nymph, and adult), deer ticks feed on a number of mammals that may carry the *Borrelia burgdorferi* bacterium in their blood, but the white-footed mouse is the most common source of infection. In the summer, the larval ticks hatch from eggs laid in the ground and feed by attaching themselves to small animals and birds. At this stage, they are not a problem for humans. It is the next stage—the nymph—that causes most cases of Lyme disease. Nymphs are very active from spring through early summer, at the height of outdoor activity for most people. Because they are still quite small (less than 2 mm), they are difficult to spot, giving them ample opportunity to transmit *Borrelia burgdorferi* while feeding. Although far more adult ticks than nymphs carry *Borrelia burgdorferi*, the adult ticks are much larger, more easily noticed, and more likely to be removed before they have fed long enough to transmit *Borrelia burgdorferi*. Neither *Borrelia burgdorferi* nor Lyme disease can be transmitted directly from one person to another or from pets to humans.

Demographics

Lyme disease is the most common vector-borne disease in the United States. In 2002 alone, 23,763 cases were reported to the Centers for Disease Control and Prevention (CDC), a 40-percent increase over the number reported in 2001. According to the CDC, the actual number of Lyme cases may exceed 200,000 due to underreporting and limitations in disease surveillance methods. CDC statistics indicate that the largest proportion of Lyme disease cases occurs in children aged five to 14 years, and more than 50 percent of Lyme disease cases involve children under age 12. Although cases of Lyme disease have been reported in 49 of the 50 states, more than 95 percent of reported cases occur in just twelve states: Connecticut, Rhode Island, New York, Pennsylvania, Delaware, New Jersey, Maryland, Maine, New Hampshire, Minnesota, Massachusetts, and Wisconsin. In the United States, the Great Lakes region and the Pacific Northwest also have a higher incidence of Lyme disease. The disease is also found in Scandinavia, continental Europe, the countries of the former Soviet Union, Japan, China, and Australia.

Causes and symptoms

Lyme disease is caused by the *Borrelia burgdorferi* bacterium. Once *Borrelia burgdorferi* gains entry to the body through a tick bite, it can move through the bloodstream quickly. Only 12 hours after entering the bloodstream, *Borrelia burgdorferi* can be found in cerebrospinal fluid (which means it can affect the nervous system). Treating Lyme disease early and thoroughly is important because Lyme disease can hide for long periods within the body in a clinically latent state. That ability explains why symptoms can recur in cycles and can flare up after months, years, or decades.

Lyme disease is usually described in terms of length of infection (time since the person was bitten by a tick infected with Lyme disease) and whether *Borrelia burgdorferi* is localized or disseminated (spread through the body by fluids and cells carrying *Borrelia burgdorferi*). Furthermore, when and how symptoms of Lyme disease appear can vary widely from patient to patient. People who experience recurrent bouts of symptoms over time are said to have chronic Lyme disease.

Early localized Lyme disease

The most recognizable indicator of Lyme disease is a rash around the site of the tick bite. Often, the tick exposure has not been recognized. The eruption might be warm or itch. The rash—erythema migrans (EM)—generally develops within three to 30 days and usually begins as a round, red patch that expands outward from the tick bite. About 80 percent of patients with Lyme disease develop EM. Clearing may take place from the center out, leaving a bull's-eye effect; in some cases, the center gets redder instead of clearing. On children with dark skin, the rash may look like a bruise. Of those who develop Lyme disease, about 50 percent notice flu-like symptoms, including fatigue, headache, chills and fever, muscle and joint pain, and lymph node swelling. Many children with Lyme disease can develop neurologic symptoms within a few weeks following a tick bite. Neurologic symptoms in children with early Lyme disease include **dizziness**, stiff neck, unilateral or bilateral facial palsy, inflammation of brain membranes (a form of **meningitis**), knee and/or wrist arthralgia, tingling/numbness, **sleep** disturbance, and difficulties with memory, concentration, and learning.

Late disseminated disease and chronic Lyme disease

Weeks, months, or even years after an untreated tick bite, symptoms can appear in several forms, including the following:

- fatigue, forgetfulness, confusion, mood swings, irritability, numbness
- neurologic problems, such as pain (unexplained and not triggered by an injury), **Bell's palsy** (facial paralysis, usually one-sided but possibly on both sides), a mimicking of the inflammation of brain membranes known as meningitis fever, and severe headache
- arthritis (short episodes of pain and swelling in joints) and other musculoskeletal complaints (Arthritis eventually develops in about 60 percent of patients with untreated Lyme disease.)

In adults, less common effects of Lyme disease are heart abnormalities (such as irregular rhythm or cardiac block) and eye abnormalities (such as swelling of the cornea, tissue, or eye muscles and nerves). However, children with Lyme disease frequently complain of chest pain and have papilledema (swelling of the optic nerve). In addition, children with late-stage Lyme disease are more likely than adults to have fever and joint swelling and pain.

When to call the doctor

A child should see a doctor if an attached tick is found that is engorged with blood (usually indicating attachment for more than six hours). Parents should remove the tick gently with tweezers. Medical laboratories can test the tick for *Borrelia burgdorferi* if the tick is alive; parents should place the tick in a tightly sealed plastic bag or small bottle with a moistened cotton ball and take it to the doctor. Most doctors will not prescribe antibiotics immediately following a tick bite but will ask parents to monitor their child for symptoms of early Lyme disease.

Less than 50 percent of children realize that they have been bitten by a tick. And, according to pediatricians specializing in Lyme disease, many children already have chronic Lyme disease when they are first diagnosed because children have difficulties effectively verbalizing their symptoms and their symptoms may be misdiagnosed. Any child that develops a round, bull's-eye skin rash, joint pain, flu-like symptoms, and/or neurologic symptoms as described above should see a doctor. Because many children do not develop a rash or the rash may not be readily visible (e.g., on the scalp under hair), children living in or visiting areas with a high incidence of Lyme disease and those participating in frequent outdoor activities during active tick months who develop joint pain and neurologic symptoms should see a doctor.

Diagnosis

In children, symptoms of Lyme disease can mimic those of other common childhood conditions, and children may not realize they have been bitten by a tick; therefore, diagnosis of Lyme disease in children can be difficult. Therefore, diagnosis of Lyme disease relies on information the patient and parents provide and the doctor's clinical judgment, particularly through elimination of other possible causes of the symptoms. Differential diagnosis (distinguishing Lyme disease from other diseases) is based on clinical evaluation with laboratory tests used for clarification when necessary. A two-test approach is common to confirm the results. Because of the potential for misleading results (false-positive and false-negative), laboratory tests alone cannot establish the diagnosis.

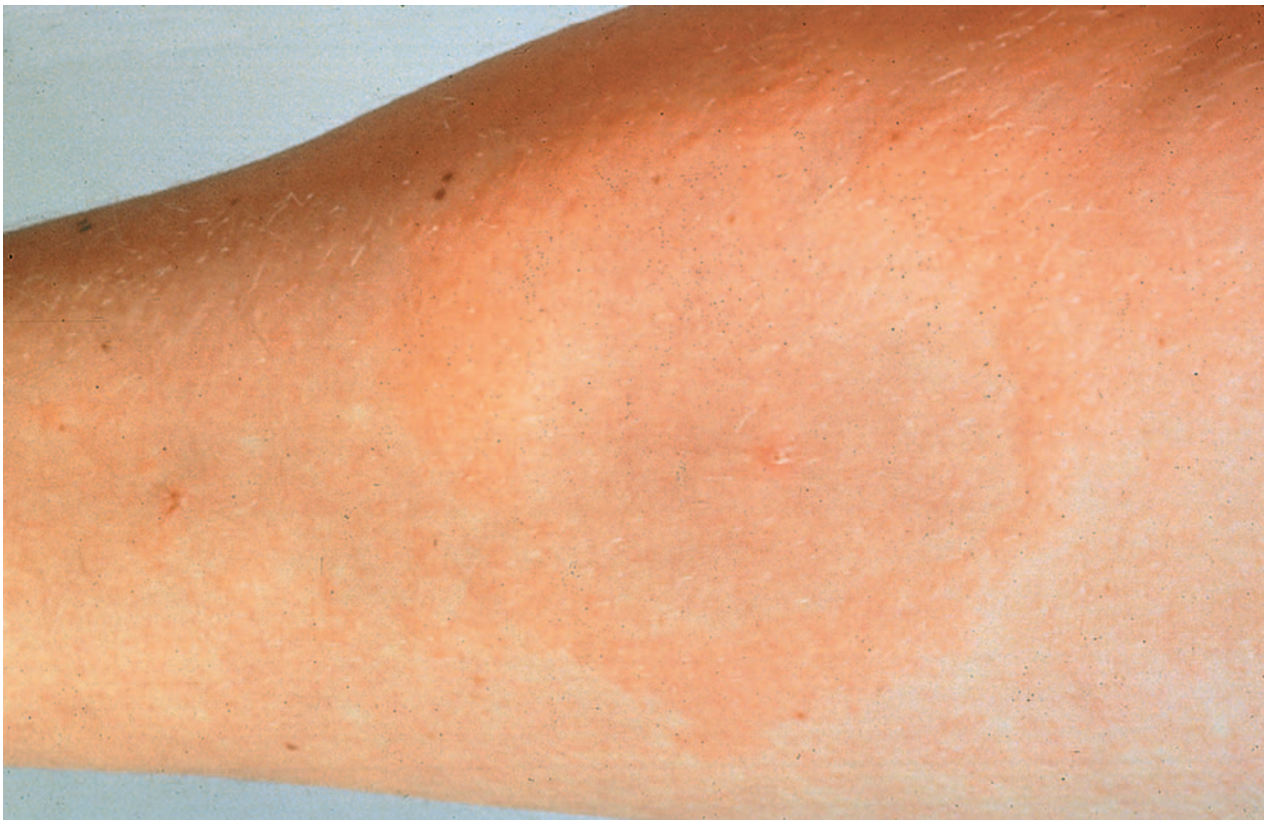
In February 1999 the Food and Drug Administration (FDA) approved a new blood test for Lyme disease called PreVue. The test, which searches for antigens (substances that stimulate the production of antibodies) produced by *Borrelia burgdorferi*, gives results within one hour in the doctor's office. A positive result from the PreVue test is confirmed by a second blood test known as the Western blot, which must be done in a laboratory.

Doctors generally know which disease-causing organisms are common in their geographic area. The most helpful piece of information is whether a tick bite or rash was noticed and whether it happened locally or while traveling. Doctors may not consider Lyme disease if it is rare locally but will take it into account if a patient mentions vacationing in an area where the disease is commonly found.

Treatment

The treatment for Lyme disease is antibiotic therapy. If a child has strong indications of Lyme disease (symptoms and medical history), the doctor will probably begin treatment on the presumption of this disease. The American College of Physicians recommends treatment for a patient with a rash resembling EM or who has arthritis, a history of an EM-type rash, and a previous tick bite.

The benefits of early treatment must be weighed against the risks of overtreatment. The longer a patient is ill with Lyme disease before treatment, the longer the course of therapy must be, and the more aggressive the



The first sign of lyme disease is usually an itchy bull's-eye rash around the site of the tick bite. *(© 1993 Science Photo Library. Custom Medical Stock Photo, Inc.)*

treatment. The development of opportunistic organisms may produce other symptoms. For example, after long-term antibiotic therapy, patients can become more susceptible to yeast infections. Treatment may also be associated with adverse drug reactions.

For most children, oral antibiotics (amoxicillin) are prescribed for 21 days. When symptoms indicate nervous system involvement or a severe episode of Lyme disease, an intravenous antibiotic (ceftriaxone, cefotaxime, ampicillin) may be given for four to six weeks or longer. Some physicians consider intravenous ceftriaxone the best therapy for any late manifestation of disease, but treatments for late Lyme disease are still controversial as of 2004. Corticosteroids (oral) may be prescribed if eye abnormalities occur, but they should not be used without first consulting an eye doctor. Nonsteroidal anti-inflammatory medications (ibuprofen) may be prescribed for joint pain and inflammation.

The doctor may have to adjust the treatment regimen or change medications based on the patient's response. Treatment can be difficult because *Borrelia burgdorferi* comes in several strains (some may react to different antibiotics than others) and may even have the ability to switch forms during the course of infection. Also, *Borrelia burgdorferi* can shut itself up in cell niches, allowing it to hide from antibiotics. Finally, antibiotics can kill *Borrelia burgdorferi* only while it is active rather than dormant.

Alternative treatment

Supportive therapies may minimize symptoms of Lyme disease or improve the immune response. These include vitamin and nutritional supplements, mostly for chronic fatigue and increased susceptibility to infection. For example, yogurt and *Lactobacillus acidophilus* preparations help fight yeast infections, which are common in patients on long-term antibiotic therapy. In addition, botanical medicine and homeopathy can be considered to help bring the body's systems back to a state of health and well-being. A Western herb, spilanthes (*Spilanthes* spp.), may be effective in treating diseases such as Lyme disease that are caused by spirochetes (spiral-shaped bacteria). Therapy using a low-current electrical field or magnetic pulses is also as of 2004 under research to treat bacterial infections. It is important to note that no

alternative treatments have been proven to cure Lyme disease.

Prognosis

If aggressive antibiotic therapy is given early and the patient cooperates fully and sticks to the medication schedule, recovery should be complete. Only a small percentage of Lyme disease patients fail to respond or relapse (have recurring episodes). Most long-term effects of the disease result when diagnosis and treatment is delayed or missed. Co-infection with other infectious organisms spread by ticks in the same areas as *Borrelia burgdorferi* (babesiosis and ehrlichiosis, for instance) may be responsible for treatment failures or more severe symptoms. Most fatalities reported with Lyme disease involved patients coinfected with babesiosis.

Prevention

Lyme disease can be prevented by taking the following measures to reduce exposure to tick bites:

- Avoid areas likely to be infested with ticks, especially during spring and summer, when tick nymphs are most likely to feed. Areas most likely to be infested with ticks include moist and shady areas, wooded and brushy areas, overgrown grassy areas, and areas with a high rodent and deer population.
- When outdoors, wear light-colored clothing, long-sleeved shirts, and long pants tucked into socks or boots.
- Use insect repellents according to **safety** guidelines for children.
- Perform a full-body "tick check" after outdoor activities and use tweezers to gently remove and dispose of ticks.
- Do not try to remove the tick by using petroleum jelly, alcohol, or a lit match.
- Place the tick in a closed container (for species identification later, should symptoms develop) or dispose of it by flushing or by placing the tick between scotch tape.
- Check pets frequently for ticks, since ticks can migrate to children from pets.

Update on vaccination

A vaccine for Lyme disease known as LYMErix was available from 1998 to 2002, when it was removed from the United States market. The decision was influenced by reports that LYMErix may be responsible for neurologic complications in vaccinated patients. As of late 2004, the best prevention strategy was minimizing risk of exposure to ticks and using personal protection precautions.

KEY TERMS

Babesiosis—A infection transmitted by the bite of a tick and characterized by fever, headache, nausea, and muscle pain.

Bell's palsy—Facial paralysis or weakness with a sudden onset, caused by swelling or inflammation of the seventh cranial nerve, which controls the facial muscles. Disseminated Lyme disease sometimes causes Bell's palsy.

Blood-brain barrier—An arrangement of cells within the blood vessels of the brain that prevents the passage of toxic substances, including infectious agents, from the blood and into the brain. It also makes it difficult for certain medications to pass into brain tissue.

Cerebrospinal fluid—The clear, normally colorless fluid that fills the brain cavities (ventricles), the subarachnoid space around the brain, and the spinal cord and acts as a shock absorber.

Disseminated—Spread to other tissues.

Erythema migrans—A red skin rash that is one of the first signs of Lyme disease in about 75% of patients.

Lyme borreliosis—Another name for Lyme disease.

Spirochete—A type of bacterium with a long, slender, coiled shape. Syphilis and Lyme disease are caused by spirochetes.

Vector—A carrier organism (such as a fly or mosquito) which serves to deliver a virus (or other agent of infection) to a host. Also refers to a retrovirus that had been modified and is used to introduce specific genes into the genome of an organism.

Parental concerns

Because most children do not realize they have been in tick-infested areas or been bitten by a tick and because deer ticks can be the size of a poppy seed or smaller, parents should be diligent about checking children for ticks, especially if the **family** lives in or visits an area with a high incidence of Lyme disease or an area near tick habitats. Also, because Lyme disease is difficult to diagnose in children, parents who suspect Lyme disease in their children should inform their doctor about the possibility

of the disease and be proactive in requesting further medical evaluation and treatment.

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Lyme Disease Network of New Jersey Inc. 43 Winton Road, East Brunswick, NJ 08816. Web site: <www.lymenet.org>.

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Lymphadenitis

Definition

Lymphadenitis is the inflammation of lymph nodes. It is often a complication of bacterial infections, although it can also be caused by viruses or other disease agents. Lymphadenitis may be either generalized, involving a number of lymph nodes, or limited to a few nodes in the area of a localized infection. Lymphadenitis is sometimes accompanied by lymphangitis, which is the inflammation of the lymphatic vessels that connect the lymph nodes.

Description

The lymphatic system is a network of vessels (channels), nodes (glands), and organs. It is part of the immune system, which protects against and fights infections, inflammation, and cancers. The lymphatic system also participates in the transport of fluids, fats, proteins, and other substances throughout the body. The lymph nodes are small structures that filter the lymph fluid and contain many white blood cells to fight infections. Lymphadenitis is marked by swollen lymph nodes that develop when the glands are overwhelmed by bacteria, virus, fungi, or other organisms. The nodes may be tender and hard or soft and "rubbery" if an abscess has formed. The skin over an inflamed node may be red and hot. The location of the affected nodes is usually associated with the site of an underlying infection, inflammation, or tumor. In most cases, the infectious organisms are *Streptococci* or *Staphylococci*. If the lymphatic vessels are also infected, in a condition referred to as lymphangitis, there will be red streaks extending from the wound in the direction of the lymph nodes, throbbing **pain**, and high **fever** and/or chills. The child will generally feel ill, with loss of appetite, **headache**, and muscle aches.

The extensive network of lymphatic vessels throughout the body and their relation to the lymph

nodes helps to explain why bacterial infection of the nodes can spread rapidly to or from other parts of the body. Lymphadenitis in children often occurs in the neck area because these lymph nodes are close to the ears and throat, which are frequent locations of bacterial infections in children.

Lymphadenitis is also referred to as lymph node infection, lymph gland infection, or localized lymphadenopathy.

Demographics

Lymphadenitis and lymphangitis are common complications of bacterial infections.

Causes and symptoms

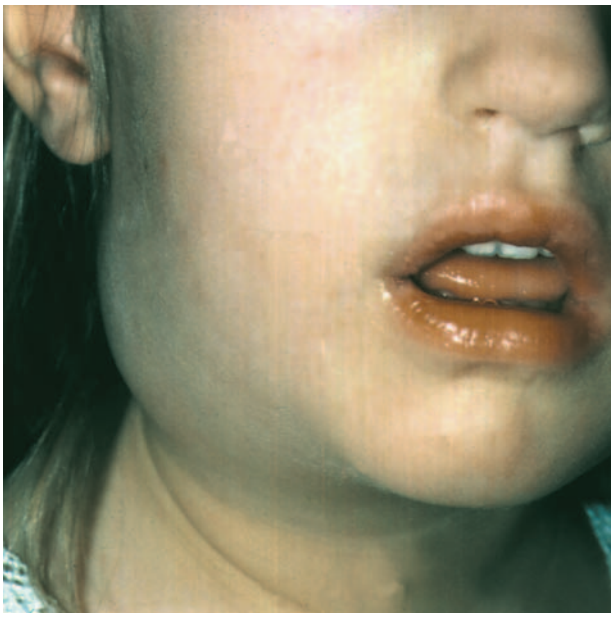
Streptococcal and staphylococcal bacteria are the most common causes of lymphadenitis, although viruses, protozoa, rickettsiae, fungi, and the **tuberculosis** bacillus can also infect the lymph nodes. Diseases or disorders that involve lymph nodes in specific areas of the body include rabbit fever (tularemia), **cat-scratch disease**, lymphogranuloma venereum, chancroid, genital herpes, infected **acne**, dental abscesses, and bubonic plague. Lymphadenitis can also occur in conjunction with cellulitis, which is a deep, widespread tissue infection that develops from a cut or sore. In children, **tonsillitis** or bacterial sore throats are the most common causes of lymphadenitis in the neck area. Diseases that involve lymph nodes throughout the body include mononucleosis, **cytomegalovirus infection**, **toxoplasmosis**, and brucellosis.

The early symptoms of lymphadenitis are swelling of the nodes caused by a build-up of tissue fluid and an increased number of white blood cells resulting from the body's response to the infection. Further developments include fever with chills, loss of appetite, heavy perspiration, a rapid pulse, and general weakness.

Diagnosis

Physical examination

The diagnosis of lymphadenitis is usually based on a combination of the child's medical history, external symptoms, and laboratory cultures. The doctor will press (palpate) the affected lymph nodes to see if they are sore or tender, and search for an entry point for the infection, like a scratch or bite. Swollen nodes without soreness are sometimes caused by cat-scratch disease, which is an uncommon illness. In children, if the lymphadenitis is severe or persistent, the doctor may need to rule out



Swollen lymph node glands in a young girl's neck. *(Custom Medical Stock Photo Inc.)*

mumps, HIV, tumors in the neck region, and congenital cysts that resemble swollen lymph nodes.

Although lymphadenitis is usually diagnosed in lymph nodes in the neck, arms, or legs, it can also occur in lymph nodes in the chest or abdomen. If the child has acutely swollen lymph nodes in the groin, the doctor will need to rule out a **hernia** in the groin that has failed to reduce (incarcerated inguinal hernia). Hernias occur in 1 percent of the general population; 85 percent of children with hernias are male.

Laboratory tests

The most significant tests are a white blood cell count (WBC) and a blood culture to identify the organism. A high proportion of immature white blood cells indicates a bacterial infection. Blood cultures may be positive, most often for a species of staphylococcus or streptococcus. In some cases, the doctor may order a biopsy of the lymph node to look for unusual infection or lymphoma.

When to call the doctor

If a child develops symptoms of lymphadenitis, he or she should be taken to the doctor or emergency room.

KEY TERMS

Hernia—A rupture in the wall of a body cavity, through which an organ may protrude.

Lymph nodes—Small, bean-shaped collections of tissue located throughout the lymphatic system. They produce cells and proteins that fight infection and filter lymph. Nodes are sometimes called lymph glands.

Lymphangitis—Inflammation of the lymphatic vessels. It often occurs together with lymphadenitis (inflammation of the lymph nodes).

Septicemia—A systemic infection due to the presence of bacteria and their toxins in the bloodstream. Septicemia is sometimes called blood poisoning.

Streptococcus—Plural, *streptococci*. Any of several species of spherical bacteria that form pairs or chains. They cause a wide variety of infections including scarlet fever, tonsillitis, and pneumonia.

Treatment

Medications

The medications given for lymphadenitis vary according to the bacterium or virus that causes it. For bacterial infections, the child will be treated with **antibiotics**, usually a penicillin, clindamycin, a cephalosporin, or erythromycin.

Supportive care

Supportive care of lymphadenitis includes resting the affected area and applying hot moist compresses to reduce inflammation and pain.

Surgery

Cellulitis associated with lymphadenitis should not be treated surgically because of the risk of spreading the infection. Pus is drained only if there is an abscess and usually after the child has begun antibiotic treatment. In some cases, biopsy of an inflamed lymph node is necessary if no diagnosis has been made and no response to treatment has occurred.

Inflammation of lymph nodes due to other diseases requires treatment of the underlying causes.

Prognosis

The prognosis for recovery is good if the child is treated promptly with antibiotics. In most cases, the infection can be brought under control in three or four days. However, in some cases it may take weeks or months for swelling to disappear; the length of recovery depends on the underlying cause of the infection. Children with untreated lymphadenitis may develop abscesses, cellulitis, or blood poisoning (septicemia), which is sometimes fatal.

Prevention

Prevention of lymphadenitis depends on prompt treatment of bacterial and viral infections.

Parental concerns

Parents may be concerned that enlarged lymph nodes in their child are malignant. They should seek immediate medical attention for the child so concerns can be addressed in a timely manner.

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M

Macrocephaly

Definition

Macrocephaly is a condition in which the head is larger than normal.

Description

Also called macrocephalia and megalocephaly, macrocephaly is diagnosed when the circumference of the head is more than two standard deviations above average for the child's age, sex, race, and period of gestation. The fontanelle (soft spot) of the newborn is wide, but facial features are usually normal. Macrocephaly is distinguished from **hydrocephalus** in that there is no increase in pressure within the head; however, hydrocephalus can result in macrocephaly in some children. The disorder can result from a defect in formation during the embryonic stage, as a result of certain degenerative diseases, as a part of various genetic syndromes, or as an inherited **family** trait. Mental deficiency, seizures, and **movement disorders** are common in macrocephalic children.

Demographics

Because of the many conditions that cause macrocephaly, a true assessment of its incidence is difficult. It is a relatively rare condition that does not appear to affect children of any particular race, gender, or nationality with more frequency.

Causes and symptoms

Macrocephaly may be caused by many conditions. The most common causes for an enlarged head are megalencephaly, or an enlarged brain, and hydrocephalus, or excessive cerebrospinal fluid (**CSF**) in the brain.

When macrocephaly is a result of megalencephaly, it is often impossible to determine the cause. However, megalencephaly is often associated with metabolic diseases such as Canavan's disease or Alexander's disease or with syndromes such as **gigantism**, achondroplasia (**dwarfism** or small stature), **osteogenesis imperfecta**, **neurofibromatosis**, and some chromosomal anomalies. In each of these disorders, there is an enlargement of brain tissues.

In hydrocephalus, excess CSF collects in the large sections of the brain called the ventricles. This may occur for many reasons, including **Chiari malformation**, abnormal cysts within the brain, and infections such as **meningitis**.

In some cases, a child may have benign macrocephaly. In these children, the only abnormality is an enlarged head. Usually there are other family members with large heads, and the condition is considered a family trait. These children do not have an underlying condition and usually do not have any additional complications.

The major symptom of macrocephaly is an enlarged head circumference. Other symptoms can include, delay in reaching developmental milestones, **mental retardation**, rapid head growth, and slowed growth of the rest of the body.

Diagnosis

Macrocephaly is usually diagnosed by the pediatrician during a physical examination. In some cases this may be the only diagnosis necessary. Some children will require additional diagnostic imaging procedures, such as **computed tomography** scan (CAT scan), x ray, and **magnetic resonance imaging** (MRI), to determine the cause of the macrocephaly and the appropriate treatment.

KEY TERMS

Achondroplasia—A congenital disturbance of growth plate development in long bones that results in a person having shortened limbs and a normal trunk.

Alexander's disease—A progressive, degenerative disorder of the central nervous system.

Canavan disease—A serious genetic disease more common in the Eastern European Jewish population that causes mental retardation and early death. Canavan disease is caused by the lack of an enzyme called aspartoacylase.

Chiari II anomaly—A structural abnormality of the lower portion of the brain (cerebellum and brainstem) associated with spina bifida. The lower structures of the brain are crowded and may be forced into the foramen magnum, the opening through which the brain and spinal cord are connected.

Computed tomography (CT)—An imaging technique in which cross-sectional x rays of the body are compiled to create a three-dimensional image of the body's internal structures; also called computed axial tomography.

Fontanelle—One of several "soft spots" on the skull where the developing bones of the skull have yet to fuse.

Gigantism—Excessive growth, especially in height, resulting from overproduction of growth hormone during childhood or adolescence by a pituitary tumor. Untreated, the tumor eventually destroys the pituitary gland, resulting in death during early adulthood. If the tumor develops after growth has stopped, the result is acromegaly, not gigantism.

Magnetic resonance imaging (MRI)—An imaging technique that uses a large circular magnet and radio waves to generate signals from atoms in the body. These signals are used to construct detailed images of internal body structures and organs, including the brain.

Megalencephaly—A condition in which the brain is abnormally large.

Megalocephaly—An abnormally large head.

Neurofibromatosis—A progressive genetic condition often including multiple café-au-lait spots, multiple raised nodules on the skin (neurofibromas), developmental delays, slifhtly larger head size, and freckles in the armpits, groin, and iris. Also known as von Recklinghausen's disease.

Osteogensis imperfecta—An inherited disorder of the connective tissue which involves multiple symptoms, including weakened bones that break easily.

Standard deviation—A measure of the distribution of scores around the average (mean). In a normal distribution, two standard deviations above and below the mean includes about 95% of all samples.

Treatment

There is no specific treatment for macrocephaly. Medical care for children with macrocephaly focuses on management of specific symptoms such as developmental delays and mental retardation and treatment of the primary diagnosis responsible for the macrocephaly.

Prognosis

For children with benign familial macrocephaly, the prognosis is excellent. These children usually do not have any complications and have normal **intelligence**. For other children with macrocephaly, the prognosis is dependent upon the cause. In children with hydrocephalus, the prognosis can be excellent depending on what type of hydrocephalus they have. Unfortunately, many children with macrocephaly experience delayed development, slow growth, seizure disorders, and limited intelligence. All of these are related to the underlying condition that caused the macrocephaly.

Prevention

Macrocephaly is often present at birth or is a result of conditions that are present at birth. As of 2004 there was no known prevention.

Parental concerns

When mental deficiency and the attendant diseases or disorders are severe, the child may require a life-support system. When the mental deficiency is less severe, the child may be diagnosed with minimal brain dysfunction or as neurologically handicapped. Minimal brain dysfunction can include any or all of the following: memory and language problems, neuromotor functioning problems, and behavior and social problems. The degree of dysfunction is a key factor in parents' deciding whether the child can continue to live at home and what type of schooling is appropriate. Parents and teachers need to be cognizant of the nature of the child's dysfunction. What was once seen as laziness and lack of motivation on the child's part has begun in the early 2000s to be

recognized as a medical condition that can be corrected or modified through psychotherapy. Sometimes, though, a child may suffer several years of frustrating failure and abnormal development or behavior before the problem is recognized and he or she is properly diagnosed.

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Magnetic resonance imaging

Definition

Magnetic resonance imaging (MRI) is a diagnostic imaging procedure that uses radio waves, a magnetic field, and a computer to generate images of the anatomy.

Purpose

MRI is used to visualize the body to assist doctors in their efforts to diagnose certain diseases or conditions and to evaluate injuries. For pediatric imaging, MRI is used for a variety of purposes, including the following:

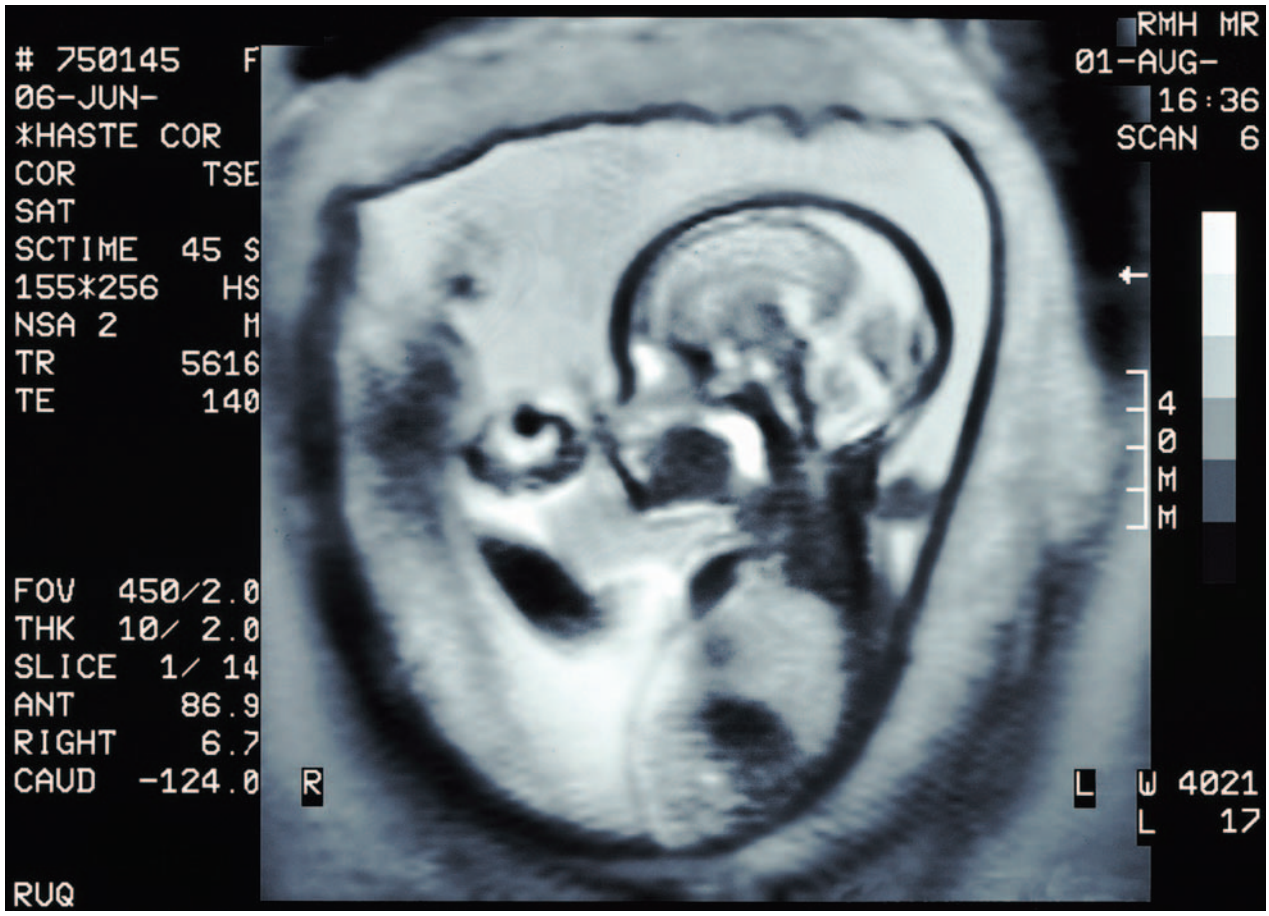
- diagnosing diseases of the central nervous system, including the brain and spine
- detecting musculoskeletal disorders and injuries
- identifying complications of infectious diseases, such as those associated with **Lyme disease** or acquired **immunodeficiency** syndrome (AIDS)
- imaging the cardiovascular system
- detecting congenital heart defects in neonates
- determining the stage of certain types of **cancer**
- evaluating bone marrow disease
- assessing blood vessels in the brain for **stroke** and other abnormalities
- assisting in the planning of surgery and cancer treatment
- evaluating the urinary tract

MRI provides images with excellent contrast that allow clinicians to clearly see details of soft tissue, bone, joints, and ligaments. MRI angiography is an imaging technique used to evaluate the blood vessels, for example, to detect aneurysms or cardiovascular problems. Because MRI does not use ionizing radiation to produce images, like x ray and CT, it is often the examination of choice for pediatric imaging and for imaging the male and female reproductive systems, pelvis and hips, and urinary tract and bladder.

MRI can also be used to evaluate brain function for assessing language, senses, neurologic disorders, and **pain**. This technique, called functional MRI, involves rapid imaging to display changes in the brain's blood flow in response to tasks or visual and auditory stimuli. Functional MRI is being researched to image neurologic disorders, such as attention deficit hyperactivity disorder (ADHD), delayed **cognitive development**, and epilepsy.

MRI spectroscopy is another emerging imaging technique for evaluating pediatric brain disorders. In MRI spectroscopy, chemicals in the brain are measured and brain tissue is imaged. This technique is being investigated to evaluate traumatic brain injury, speech delay, creatine deficiency syndromes, and **mood disorders** in young children.

Interventional and intraoperative MRI is another developing field that involves performing interventional procedures, primarily brain surgeries, using a specially designed MRI unit in an operating room.



Magnetic resonance imaging (MRI) of a fetus. *(© Lester Lefkowitz/Corbis.)*

Description

MRI is performed using a specialized scanner, a patient table, systems that generate radio waves and magnetic fields, and a computer workstation. The scanner, which is usually shaped like a large rectangle with a hole in the center, contains the systems that generate the magnetic field. A motorized and computer-controlled patient table moves into the scanner's center hole during the scan. A technologist operates the MRI scanner from an adjacent control room that contains a computer system and an intercom system for communicating with the patient during the scan.

In most MRI scanners, the patient opening is like a long tube, and some patients may become claustrophobic. To be more patient-friendly, different types of MRI scanners have been developed. Newer MRI scanners have shorter patient openings that allows the patient's head to remain outside the machine during body scans. Open MRI scanners are available with columns and open sides to alleviate claustrophobia.

Depending on the body area being scanned, special body coils may be used to enhance the images. These coils are foam and plastic braces or wraparound pads that are placed on the body part being imaged. For head imaging, the coil may be shaped like a head or neck rest.

Children undergoing an MRI scan are appropriately positioned on the patient table by the technologist. For some scans, an injected contrast material may be used and is administered using an intravenous catheter. Once the patient is positioned, the technologist goes to an adjacent control room to operate the scanner. The technologist uses an intercom system to instruct the child to hold their breath or remain still at certain times during the scan. Scans range from 30 minutes to 90 minutes, depending on the type of scan. When the MRI machine is scanning, the child hears loud clanging and whirring noises. To alleviate **fear** or stress related to hearing this noise and being in the small scanning tube, the child may be offered earplugs or specially designed head phones for listening to music. Centers that specialize in pediatric imaging often also have special video goggles so that the child can watch a cartoon or movie during the scan. For infants, neonatal noise guards—special padded ear shields—are available.

MRI scans are performed in a hospital radiology department for inpatients and emergency cases. For scans requested by a physician, the MRI examination can be performed in the hospital radiology department on an outpatient basis or in an imaging center. Hospitals that do not have their own MRI systems may schedule MRI scans by contracting with a company that brings an MRI scanner in a specially designed mobile trailer. Mobile MRI services are frequently used in rural areas. For some conditions, such as orthopedic disorders or injuries, an MRI may be performed in a physician's office using a small MRI unit called an extremity MRI scanner. These scanners are designed to image only the joints or the head. During this type of scan, only the body part to be scanned is placed in the smaller scanner while the patient lies on a couch or sits in a chair.

The images from an MRI examination are called slices, because they are acquired in very small (millimeter-size) sections of the body. The image slices are displayed on a computer monitor for viewing or printed as a film. A specialist called a radiologist interprets the images produced during the MRI examination. For emergency scans, images are interpreted immediately so that the child can be treated quickly. For non-urgent outpatient MRI scans, the radiologist interprets the images and sends a report to the referring physician within a few days.

Precautions

MRI is a safe procedure that does not involve radiation. However, the magnetic field generated during an MRI examination is so strong that metal objects or objects with metal in them, such as jewelry, **eyeglasses**, oxygen canisters, and even wheelchairs, will be pulled toward the machine. Therefore, MRI staff must take special precautions to ensure that no metallic objects enter the MRI suite. MRI technologists inspect patient clothing and accessories to make sure there are no metals on them during the scan.

Preparation

Prior to any MRI scan, patients are required to remove all metal objects and remove any clothing with metal on them (zippers, snaps). In most cases, parents have to complete a survey regarding their child's past surgical procedures and medical history to indicate whether the child has any metallic implants. Metallic implants include artificial joints, pacemakers, aneurysm clips, metal plates, pins or screws, and surgical staples. Children with metallic implants are likely to undergo a **computed tomography** (CT) examination instead of an MRI.

Unlike CT, no fasting or **laxatives** are required prior to an MRI scan. Only one type of MRI scan, called a magnetic resonance cholangiopancreatography (MRCP), which scans the bile ducts, requires that the child not eat or drink anything for two to three hours prior to the scan.

During the examination, the child must lie still. The MRI scanner does make loud noises throughout the examination, which can be frightening for some children. Before the examination, the procedure should be explained to the child, and it should be emphasized that the examination is painless. Most facilities have specially designed music systems so that patients can wear headsets and listen to music during the scan; some facilities even have special video goggles so children can watch a cartoon or movie during the scan.

Aftercare

No special aftercare is required following MRI scans, unless sedation or general anesthesia was used during the scan. Then children are required to remain in a supervised recovery area for an hour or more following the procedure to monitor for reactions to anesthesia. If injected contrast material is used, some minor first aid (small bandage, pain relief) for the injection site may be necessary.

KEY TERMS

Anaphylaxis—Also called anaphylactic shock; a severe allergic reaction characterized by airway constriction, tissue swelling, and lowered blood pressure.

Cholangiopancreatography—An examination of the bile ducts and pancreas.

Claustrophobia—Fear of small, enclosed spaces.

Computed tomography (CT)—An imaging technique in which cross-sectional x rays of the body are compiled to create a three-dimensional image of the body's internal structures; also called computed axial tomography.

Intravenous—Into a vein; a needle is inserted into a vein in the back of the hand, inside the elbow, or some other location on the body. Fluids, nutrients, and drugs can be injected. Commonly called IV.

Radiography—Examination of any part of the body through the use of x rays. The process produces an image of shadows and contrasts on film.

Radiologist—A medical doctor specially trained in radiology, the branch of medicine concerned with radioactive substances and their use for the diagnosis and treatment of disease.

Risks

MRIs present no radiation exposure. Magnetic fields used in MRI have no side effects for the patient. The contrast material used in MRI contains a material called gadolinium, that is much less likely to cause severe anaphylactic (allergic) reactions than the iodinated material used for CT scans.

Because the MRI examination is long and the patient opening in the machine is small, some children and adolescents may feel claustrophobic. Light sedation or relaxants may be administered, or an MRI scanner with a more open design may be used. For younger infants and children that require sedation or anesthesia to complete the examination, reactions to the anesthesia are possible, including headaches and **vomiting**.

Parental concerns

Younger children may be frightened of the MRI scanner, and a parent or other **family** member may be required to be present in the scanning room. To help alleviate fear, taking the child into the MRI room to see the equipment prior to the procedure may be helpful. Anyone remaining in the scanning room during the MRI examination must remove any metal objects, including jewelry and eyeglasses.

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Jennifer Sisk, MA

Malnutrition

Definition

Malnutrition is a condition that develops when the body does not get the proper amount of protein, energy (calories), **vitamins**, and other nutrients it needs to maintain healthy tissues and organ function.

Description

Poor eating habits or lack of available food may lead to malnutrition. Malnutrition occurs in children who are either undernourished or overnourished. Children who are overnourished may become overweight or obese, which may lead to long-term health problems and social stress.

Undernutrition

Undernutrition is a consequence of consuming little energy and other essential nutrients, or using or excreting them more rapidly than they can be replaced. This state of malnutrition is often characterized by infections and disease. Malnutrition intensifies the effect of every disease. Severe malnutrition is most often found in developing countries. Rarely in the United States do children suffer from severe malnutrition that is not related to severe chronic illness. Deficiency in one nutrient occurs less often than deficiency in several nutrients. A child suffering from malnutrition is usually deficient in a variety of nutrients.

The leading cause of death in children in developing countries is **protein-energy malnutrition**. This type of malnutrition is the result of inadequate intake of protein and energy. Children who are already undernourished can suffer from protein-energy malnutrition when rapid growth, infection, or disease increases the need for protein and essential nutrients.

Overnutrition

In the United States, nutritional deficiencies have generally been replaced by dietary imbalances or excesses

associated with many of the leading causes of death and disability. Overnutrition results from eating too much, eating too many of the wrong foods, not exercising enough, or taking too many vitamins or other dietary replacements.

Risk of overnutrition is also increased by being more than 20 percent overweight, consuming a diet high in fat and salt, and taking high doses of:

- nicotinic acid (niacin) to lower elevated cholesterol levels
- vitamin B_6 to relieve premenstrual syndrome
- vitamin A to clear up skin problems
- iron or other trace **minerals** not prescribed by a doctor

Nutritional disorders can affect any system in the body and the senses of sight, taste, and smell. Malnutrition begins with changes in nutrient levels in blood and tissues. Alterations in enzyme levels, tissue abnormalities, and organ malfunction may be followed by illness and death.

Complications

Poorly nourished children often have weakened immune systems, thus increasing their chances of illness. Underweight, malnourished teenagers (such as those with an eating disorder) have an increased risk of osteoporosis and may not have menstrual periods. They may have heart and other organ problems with severe malnutrition. Malnutrition, if left untreated, can lead to physical or mental disability, or even death.

Children who are overweight have an increased risk for long-term conditions and diseases, including cardiovascular disease, **high cholesterol**, high blood pressure, type 2 diabetes, **asthma**, **sleep** apnea, and certain cancers. Health consequences range from a higher risk of premature death to chronic conditions that reduce a person's quality of life.

Demographics

Malnutrition is a major cause of illness and death throughout the world. Throughout the developing world, malnutrition affects almost 800 million people, or 20 percent of the population. Approximately half of the 10.4 million children who die each year are malnourished. It often causes disease and disability in the children who survive. Diarrheal diseases are also a major world health problem, and may be a cause of malnutrition. Nearly all of these deaths occur in impoverished parts of Africa and Asia, where they often result from contamination of the water supply by animal and human feces.

Worldwide, the most common form of malnutrition is iron deficiency, affecting up to 80 percent of the population, as many as four or five billion people.

In contrast, children in many parts of the world are becoming increasingly overweight. What was thought of as a problem for industrialized nations only until recently, is now affecting children in developing countries. Approximately 25–30 percent of school-age children in the United States are overweight.

Causes and symptoms

Worldwide, poverty and lack of food are the primary reasons why malnutrition occurs. Families of low-income households do not always have enough healthy food to eat. When there is a household food shortage, children are the most vulnerable to malnutrition because of their high energy needs.

There is an increased risk of malnutrition associated with chronic diseases, especially disease of the intestinal tract, kidneys, and liver. Children with chronic diseases like **cancer**, **cystic fibrosis**, **AIDS**, **celiac disease**, and intestinal disorders may lose weight rapidly and become susceptible to malnutrition because they cannot absorb valuable vitamins, iron, and other necessary nutrients. Children who are lactose intolerant have difficulty digesting milk and milk products, and may be at risk for malnutrition, particularly a calcium deficiency.

Symptoms of malnutrition vary, depending on what nutrients are deficient in the body. Unintentionally losing weight may be a sign of malnutrition. Children who are malnourished may be skinny or bloated and may be short for their age (stunted). Their skin is pale, thick, dry, and easily bruised. **Rashes** and changes in pigmentation are common.

Hair is thin, tightly curled, and easily pulled out. Joints ache and bones are soft and tender. The gums bleed. The tongue may be swollen, or shriveled and cracked. Visual disturbances include night blindness and increased sensitivity to light and glare.

Other symptoms of malnutrition include:

- fatigue
- dizziness
- anemia
- **diarrhea**
- disorientation
- goiter (enlarged thyroid gland)
- loss of reflexes and lack of coordination
- muscle twitches
- decreased immune response
- scaling and cracking of the lips and mouth

Children who are overnourished are visibly overweight or obese, and consume more food than their bodies need (or expend too little energy through physical activity).

When to call the doctor

Parents who worry about malnutrition can discuss their concerns with a doctor, registered dietitian, or other health care provider. Though not an exhaustive list, treatment should be sought for a child if:

- there is a change in bodily functions (impairment)
- the child is not growing
- the child faints
- the child rapidly loses hair
- a girl at **puberty** stops menstruating or is underweight and fails to start menstruating

Diagnosis

Overall appearance, behavior, body-fat distribution, and organ function can alert a **family** physician, internist, or **nutrition** specialist to the presence of malnutrition. Parents may be asked to record what a child eats during a specific period. **X rays** or a CT scan can determine bone density and reveal gastrointestinal disturbances, as well as heart and lung damage.

Blood and urine tests are used to measure levels of vitamins, minerals, and waste products. Nutritional status can also be determined by:

- comparing a child's weight to standardized charts
- calculating body mass index (BMI) according to a formula that divides height into weight
- measuring skin-fold thickness or the circumference of the upper arm

Treatment

Normalizing nutritional status starts with a nutritional **assessment**. This process enables a registered dietitian or nutritionist to confirm the presence of malnutrition, assess the effects of the disorder, and formulate a diet that will restore adequate nutrition. For children suffering malnutrition due to an illness or underlying disorder, the condition should be treated concurrently.

Nutritional concerns

Children who cannot or will not eat, or who are unable to absorb nutrients taken by mouth, may be fed intravenously (parenteral nutrition) or through a tube inserted into the gastrointestinal tract (enteral nutrition).

Child suffering from the severe effects of malnutrition. *(Photograph by Bruce Brander. National Audubon Society Collection/Photo Researchers, Inc.)*

Tube feeding is often used to provide nutrients to children who have **burns**, inflammatory bowel disease, or other long-term conditions that cause chronic malnutrition or malabsorption (e.g. cystic fibrosis or AIDS), and interfere with the ability to take in enough calories. This procedure involves inserting a thin tube through the nose and carefully guiding it along the throat until it reaches the stomach or small intestine. If long-term tube feeding is necessary, the tube may be placed directly into the stomach or small intestine through an incision in the abdomen.

Tube feeding cannot always deliver adequate nutrients to children who:

- are severely malnourished
- require surgery
- are undergoing **chemotherapy** or radiation treatments
- have been seriously burned
- have persistent diarrhea or vomiting
- have a gastrointestinal tract that is not functional

Intravenous feeding can also supply some or all of the nutrients these children need.

Doctors or registered dietitians can help parents can monitor overweight or obese children. These professionals may suggest a weight loss program if the child is more than 40 percent overweight. Keeping weight gain under control can be accomplished by changing eating habits, lowering fat intake, and increasing physical activity.

Prognosis

Some children with protein-energy malnutrition recover completely. Others have many health problems throughout life, including mental disabilities and the inability to absorb nutrients through the intestinal tract. Prognosis is dependent on age and the length and severity of the malnutrition, with young children having the highest rate of long-term complications and death. Death usually results from heart failure, electrolyte imbalance, or low body temperature. Children with semiconsciousness, persistent diarrhea, **jaundice**, or low blood sodium levels have a poorer prognosis.

A good prognosis exists for overweight children who make lifestyle changes and adhere to a diet and **exercise** program.

Prevention

Every child admitted to the hospital for poor weight gain or malnutrition should be screened for the presence of illnesses and conditions that could lead to protein-energy malnutrition. Children with higher-than-average risk for malnutrition should be more closely assessed, and evaluated often.

Nutritional concerns

Proper nutrition is required to ensure optimal health. Consumption of a wide variety of foods, with adequate vitamin and mineral intake, is the basis of a healthy diet. Researchers state that no single nutrient is the key to good health, but that optimum nutrition is derived from eating a diverse diet, including a variety of fruits and vegetables. Because foods such as fruits and vegetables provide many more nutrients than vitamin supplements, food is the best source for acquiring needed vitamins and minerals.

Breastfeeding a baby for at least six months is considered the best way to prevent early-childhood malnutrition. The United States Department of Agriculture and Health and Human Services recommends that all Americans over the age of two:

- consume plenty of fruits, grains, and vegetables
- eat a variety of foods that are low in fats and cholesterol, and contain only moderate amounts of salt, sugars, and sodium
- engage in moderate physical activity for at least 30 minutes, at least several times a week
- achieve or maintain their ideal weight
- use alcohol sparingly or avoid it altogether

Iron deficiency can be prevented by consuming red meat, egg yolks, and fortified breads, flour, and cereals.

KEY TERMS

Anemia— A condition in which there is an abnormally low number of red blood cells in the bloodstream. It may be due to loss of blood, an increase in red blood cell destruction, or a decrease in red blood cell production. Major symptoms are paleness, shortness of breath, unusually fast or strong heart beats, and tiredness.

Electrolytes—Salts and minerals that produce electrically charged particles (ions) in body fluids. Common human electrolytes are sodium chloride, potassium, calcium, and sodium bicarbonate. Electrolytes control the fluid balance of the body and are important in muscle contraction, energy generation, and almost all major biochemical reactions in the body.

Minerals—Inorganic chemical elements that are found in plants and animals and are essential for life. There are two types of minerals: major minerals, which the body requires in large amounts, and trace elements, which the body needs only in minute amounts.

Nutrient—Substances in food that supply the body with the elements needed for metabolism. Examples of nutrients are vitamins, minerals, carbohydrates, fats, and proteins.

Vitamins—Small compounds required for metabolism that must be supplied by diet, microorganisms in the gut (vitamin K) or sunlight (UV light converts pre-vitamin D to vitamin D).

Parental concerns

Infants, young children, and teenagers need additional nutrients to provide for growth requirements. This is also true for women who are pregnant or breastfeeding; a mother's nutritional status affects her baby. Nutrient loss can be accelerated by diarrhea, excessive sweating, heavy bleeding (hemorrhage), or kidney failure. Nutrient intake can be restricted by age-related illnesses and conditions, excessive dieting, severe injury,

serious illness, a lengthy **hospitalization**, or substance abuse.

Children usually eat as much or as little as they need in order to feel satisfied. Children should be allowed to select what they want to eat among healthy food choices; they should be allowed to stop eating when they feel full. An underweight, overweight, or normal weight child should be allowed to decide how much to eat or whether to eat at all, within reason.

Parents must proactively prevent childhood **obesity** by recognizing weight imbalances when they begin. They can help an overweight child to lose weight (if medically necessary) by being supportive, rather than scolding. Parents should offer their children nutritious food choices and encourage physical activity. With proper intervention, an overweight child is not destined to become an overweight adult, but weight loss goals should be realistic.

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American College of Emergency Physicians. 1125 Executive Circle, Irving, TX 75038-2522. (800) 798-1822.

American College of Nutrition. 300 S. Duncan Ave. Ste. 225, Clearwater, FL 33755. (727) 446-6086.

American Dietetic Association. 120 South Riverside Plaza, Suite 2000, Chicago, IL 60606-6995. (800) 877-1600.

Food and Nutrition Information Center. Agricultural Research Service, USDA, National Agricultural Library, Room 105, 10301 Baltimore Boulevard, Beltsville, MD 20705-2351. Web site: <www.nal.usda.gov/fnic/fniccomments.html>.

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Malocclusion

Definition

Malocclusion is the misalignment of the upper and lower teeth when biting or chewing.

Description

The word malocclusion literally means "bad bite." The condition may also be referred to as an irregular bite, crossbite, or overbite. Malocclusion may be seen as crooked, crowded, or protruding teeth. It may affect a child's appearance, speech, and/or ability to eat.

Demographics

Most children have some degree of malocclusion. Malocclusion usually does not require treatment except for cosmetic reasons. It is more likely to occur if the parents have malocclusion, the child sucks his or her thumb or a pacifier, or if a tooth is lost prematurely.

Causes and symptoms

Malocclusions are most often inherited, but may be acquired. Inherited conditions include too many or too few teeth, too much or too little space between teeth, irregular mouth and jaw size and shape, and atypical formations of the jaws and face, such as a **cleft palate**. Malocclusions may be acquired from habits like finger or **thumb sucking**, tongue thrusting, premature loss of teeth from an accident or dental disease, and possibly from medical conditions such as enlarged tonsils and adenoids that lead to mouth breathing.

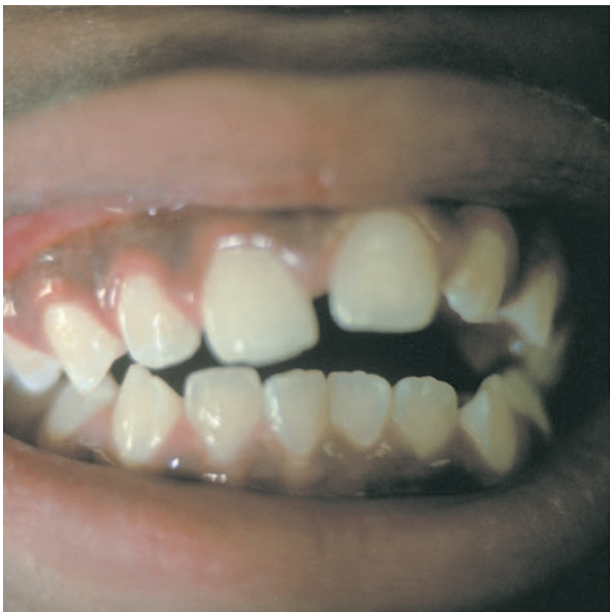
Malocclusions may cause no symptoms, or they may produce **pain** from the increased stress on oral structures. Teeth may show abnormal signs of wear on the chewing surfaces or decay in areas of tight overlap. Chewing may be difficult.

When to call the doctor

A dentist or orthodontist should be consulted if a child's teeth seem to be particularly misaligned or if a child complains of dental or jaw pain.

Diagnosis

Malocclusion is most often found during a routine dental examination. A dentist will check a patient's occlusion by watching how the teeth make contact when the child bites down normally. The dentist may ask the



This patient's teeth are misarranged because of excessive thumb sucking. *(Photograph by K.L. Boyd. Custom Medical Stock Photo, Inc.)*

KEY TERMS

Braces—An orthodontic appliance consisting of brackets cemented to the surface of each tooth and wires of stainless steel or nickel titanium alloy. Braces are used to treat malocclusion by changing the position of the teeth.

Impression—In dentistry, an imprint of the upper or lower teeth made in a pliable material that sets. When this material has hardened, it may be filled with plaster, plastic, or artificial stone to make an exact model of the teeth.

Occlusion—The way upper and lower teeth fit together during biting and chewing. Also refers to the blockage of some area or channel of the body.

Retainer—An orthodontic appliance that is worn to stabilize teeth in a new position.

Space maintainer—An orthodontic appliance that is worn to prevent adjacent teeth from moving into the space left by an unerupted or prematurely lost tooth.

child to bite down with a piece of coated paper between the upper and lower teeth. This paper will leave colored marks at the points of contact. When malocclusion is suspected, photographs and **x rays** of the face and mouth may be taken for further study. To confirm the presence and extent of malocclusion, the dentist makes plaster or plastic models of the patient's teeth from impressions. These models duplicate the fit of the teeth and are very useful in planning treatment.

Treatment

Malocclusion may be remedied by orthodontic treatment. **Orthodontics** is a specialty of dentistry that manages the growth and correction of dental and facial structures. Braces are the most commonly used orthodontic appliances in the treatment of malocclusion. At any given time, approximately four million people in the United States are wearing braces, most of whom are children and teenagers.

Braces apply constant gentle force to slowly change the position of the teeth, straightening and properly aligning them with the opposing teeth. Braces consist of brackets cemented to the surface of each tooth and wires of stainless steel or nickel titanium alloy. When the wires are threaded through the brackets, they exert pressure against the teeth, causing them to gradually move.

Braces are not removable for daily tooth brushing. To prevent **tooth decay**, the child must be especially diligent about keeping the mouth clean and removing food particles that become easily trapped. Crunchy foods should be avoided to minimize the risk of breaking the appliance. Hard fruits, vegetables, and breads must be cut into bite-sized pieces before eating. Foods that are sticky, including chewing gum, should be avoided because they may pull off the brackets or weaken the cement. Carbonated beverages may also weaken the cement, as well as contribute to tooth decay. Teeth should be brushed immediately after eating. Special floss threaders are available to make flossing easier.

If overcrowding is creating malocclusion, one or more teeth may be extracted (surgically removed), giving the others room to move. If a tooth has not yet erupted or is prematurely lost, the orthodontist may insert an appliance, called a space maintainer, to keep the other teeth from moving out of their natural position. In severe cases of malocclusion, surgery may be necessary and the patient is referred to another specialist, an oral or maxillofacial surgeon.

Once the teeth have been moved into their new position, the braces are removed, and a retainer is worn until the teeth stabilize in that position. Retainers do not move teeth, they only hold them in place. Often a retainer is initially worn all the time; its use is gradually tapered until it is only worn at night, and eventually not at all.

Orthodontic treatment is the only effective treatment for malocclusion not requiring surgery. However,

depending on the cause and severity of the condition, an orthodontist may be able to suggest other appliances as alternatives to braces. If the malocclusion is thought to be caused by the child sucking on fingers or a pacifier and the child is stopped early enough, the malocclusion may resolve spontaneously without treatment.

Alternative treatment

There are some techniques of craniosacral therapy that can alter structure. This therapy may allow correction of some cases of malocclusion. If surgery is required, pre- and post-surgical care with homeopathic remedies, as well as vitamin and mineral supplements, can enhance recovery. Night guards are sometimes recommended to ease the strain on the jaw and to limit teeth grinding.

Prognosis

Depending on the cause and severity of the malocclusion and the appliance used in treatment, a patient may expect correction of the condition to take two or more years. Patients typically wear braces 18–24 months, and a retainer for another year. Treatment is faster and more successful in children and teens whose teeth and bones are still developing. The time needed for treatment is also affected by how well the patient follows orthodontic instructions.

Prevention

In general, malocclusion is not preventable. It may be minimized by controlling habits such as thumb sucking. An initial consultation with an orthodontist before a child is seven years of age may lead to appropriate management of the growth and development of the child's dental and facial structures, circumventing many of the factors contributing to malocclusion.

Parental concerns

Most of the time, malocclusion is treated for cosmetic reasons. Children, however, may not want treatment because they will have to wear braces. It is usually possible to schedule the beginning of treatment for a time that is convenient for the child and the parent. Talking with children or teenagers and obtaining their input about treatment may be beneficial in increasing compliance. Full compliance with the orthodontist's instructions helps to ensure that the treatment is successful.

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ORGANIZATIONS

American Association of Orthodontists. 401 North Lindberg Boulevard, St. Louis, MO 63141-7816. (800) STRAIGHT Fax: (3314)-997-1745. Web site: </www.braces.org>.

American Dental Association. 211 East Chicago Avenue, Chicago IL, 60611-2678. (312) 440-2500. Web site: <http://www.ada.org>.

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Manic depression *see* **Bipolar disorder**

Marfan syndrome

Definition

Marfan syndrome is an inherited disorder of the connective tissue that causes abnormalities of a child's eyes, cardiovascular system, and musculoskeletal system. It is named for the French pediatrician, Antoine Marfan (1858-1942), who first described it in 1896.

Description

Marfan syndrome affects three major organ systems of the body: the heart and circulatory system, the bones and muscles, and the eyes. The genetic mutation responsible for Marfan was discovered in 1991. It affects the body's production of fibrillin, which is a protein that is an important part of connective tissue. Fibrillin is the primary component of the microfibrils that allow tissues to stretch repeatedly without weakening. Because the child's fibrillin is abnormal, his or her connective tissues are looser than usual, which weakens or damages the support structures of the entire body.

The most common external signs associated with Marfan syndrome include excessively long arms and legs, with the child's arm span being greater than his or her height. The fingers and toes may be long and slender, with loose joints that can be bent beyond their normal

limits. This unusual flexibility is called hypermobility. The child's face may also be long and narrow, and he or she may have a noticeable curvature of the spine. It is important to note, however, that children with Marfan vary widely in the external signs of their disorder and in their severity; even two children from the same **family** may look quite different. Most of the external features of Marfan syndrome become more pronounced as the child gets older, so that diagnosis of the disorder is often easier in adults than in children. In many cases, the child may have few or very minor outward signs of the disorder, and the diagnosis may be missed until the child develops vision problems or cardiac symptoms.

Marfan syndrome by itself does not affect a child's **intelligence** or ability to learn. There is, however, some clinical evidence that children with Marfan have a slightly higher rate of hyperactivity and attention-deficit disorder (ADD) than the general population. In addition, a child with undiagnosed nearsightedness related to Marfan may have difficulty seeing the blackboard or reading printed materials, and thus do poorly in school.

Marfan syndrome affects males and females equally, and appears to be distributed equally among all races and ethnic groups. The rate of mutation of the fibrillin gene, however, appears to be related to the age of the child's father; older fathers are more likely to have new mutations appear in chromosome 15.

Marfan syndrome is sometimes called arachnodactyly, which means "spider-like fingers" in Greek, since one of the characteristic signs of the disease is disproportionately long fingers and toes.

Demographics

It is estimated that one person in every 3000-5000 has Marfan syndrome, or about 50,000 people in the United States. Marfan syndrome is one of the more common inheritable disorders.

Causes and symptoms

Marfan syndrome is caused by a single gene for fibrillin on chromosome 15, which is inherited in most cases from an affected parent. Between 15 and 25 percent of cases result from spontaneous mutations. Mutations of the fibrillin gene (FBNI) are unique to each family affected by Marfan, which makes rapid genetic diagnosis impossible, given present technology. The syndrome is an autosomal dominant disorder, which means that someone who has it has a 50 percent chance of passing it on to any offspring.

Another important genetic characteristic of Marfan syndrome is variable expression. This term means that the mutated fibrillin gene can produce a variety of symptoms of very different degrees of severity, even in members of the same family.

Cardiac and circulatory abnormalities

The most important complications of Marfan are those affecting the heart and major blood vessels; some are potentially life-threatening. About 90 percent of children with Marfan will develop cardiac complications, including:

- Aortic enlargement. This is the most serious potential complication of Marfan syndrome. Because of the abnormalities of the child's fibrillin, the walls of the aorta (the large blood vessel that carries blood away from the heart) are weaker than normal and tend to stretch and bulge out of shape. This stretching increases the likelihood of an aortic dissection, which is a tear or separation between the layers of tissue that make up the aorta. An aortic dissection usually causes severe **pain** in the abdomen, back, or chest, depending on the section of the aorta that is affected. Rupture of the aorta is a medical emergency requiring immediate surgery and medication.
- Aortic regurgitation. A weakened and enlarged aorta may allow some blood to leak back into the heart during each heartbeat; this condition is called aortic regurgitation. Aortic regurgitation occasionally causes shortness of breath during normal activity. In serious cases, it causes the left ventricle of the heart to enlarge and may eventually lead to heart failure.
- Mitral valve prolapse. Between 75 and 85% of children with Marfan have loose or "floppy" mitral valves, which are the valves that separate the chambers of the heart. When these valves do not cover the opening between the chambers completely, the condition is called mitral valve prolapse. Complications of mitral valve prolapse include **heart murmurs** and arrhythmias. In rare cases, mitral valve prolapse can cause sudden death.
- Infective endocarditis. Infective endocarditis is an infection of the endothelium, the tissue that lines the heart. In children with Marfan, it is the abnormal mitral valve that is most likely to become infected.
- Other complications. Some children with Marfan develop cystic disease of the lungs or recurrent spontaneous pneumothorax, which is a condition in which air accumulates in the space around the lungs. Many will also eventually develop emphysema.

Musculoskeletal abnormalities

Marfan syndrome causes an increase in the length of the child's bones, with decreased support from the ligaments that hold the bones together. As a result, the child

may develop various deformities of the skeleton or disorders related to the relative looseness of the ligaments.

Disorders of the spine

Children with Marfan syndrome also can experience spinal disorders, including:

- **Scoliosis**. Scoliosis, or curvature of the spine, is a disorder in which the vertebrae that make up the spine twist out of line from side to side into an S-shape or a spiral. It is caused by a combination of the rapid growth of children with Marfan, and the looseness of the ligaments that help the spine to keep its shape.
- Kyphosis. Kyphosis is an abnormal outward curvature of the spine at the back, sometimes called hunch back when it occurs in the upper back. Children with Marfan may develop kyphosis either in the upper (thoracic) spine or the lower (lumbar) spine.
- Spondylolisthesis. Spondylolisthesis is the medical term for a forward slippage of one vertebra on the one below it. It produces an ache or stiffness in the lower back.
- Dural ectasia. The dura is the tough, fibrous outermost membrane covering the brain and the spinal cord. The weak dura in a child with Marfan swells or bulges under the pressure of the spinal fluid. This swelling is called ectasia. In most cases, dural ectasia occurs in the lower spine, producing low back ache, a burning feeling, or **numbness** or weakness in the legs.

Disorders of the chest and lower body

Disorders of the chest and lower body of children with Marfan include:

- Pectus excavatum. Pectus excavatum is a malformation of the chest in which the child's breastbone, or sternum, is sunken inward. It can cause difficulties in breathing, especially if the heart, spine, and lung have been affected by Marfan. It also usually causes concerns about appearance.
- Pectus carinatum. In other children with Marfan, the sternum is pushed outward and narrowed. Although pectus carinatum does not cause breathing difficulties, it can cause embarassment about appearance. A few children with Marfan may have a pectus excavatum on one side of their chest and a pectus carinatum on the other.
- Foot disorders. Children with Marfan are more likely to develop pes planus (flat feet) or so-called "claw" or "hammer" toes than people in the general population. They are also more likely to suffer from chronic pain in their feet.
- Protrusio acetabulae. The acetabulum is the socket of the hip joint. In a child with Marfan, the acetabulum becomes deeper than normal during growth, for reasons that are not yet understood. Although protrusio acetabulae does not cause problems during childhood and **adolescence**, it can lead to a painful form of arthritis in adult life.

Disorders of the eyes and face

Although the visual problems that are related to Marfan syndrome are rarely life-threatening, they are important in that they may be the child's first indication of the disorder. Eye disorders related to the syndrome include the following:

- Myopia (nearsightedness). Most children with Marfan develop nearsightedness, usually in childhood.
- Ectopia lentis. Ectopia lentis is the medical term for dislocation of the lens of the eye. Between 65 and 75 percent of children with Marfan have dislocated lenses. This condition is an important indication for diagnosis of the syndrome because there are relatively few other disorders that produce it.
- Glaucoma. This condition is much more prevalent in children with Marfan syndrome than in the general population.
- Cataracts. Children with Marfan are more likely to develop cataracts, and to develop them much earlier in life, sometimes as early as 40 years of age.
- Retinal detachment. Children with Marfan are more vulnerable to this disorder because of the weakness of their connective tissues. Untreated retinal detachment can cause blindness. The danger of retinal detachment is an important reason for children to avoid contact **sports** or other activities that could cause a blow on the head or being knocked to the ground.
- Other facial problems. Children with Marfan sometimes develop dental problems related to crowding of the teeth caused by a high-arched palate and a narrow jaw.

Other disorders

Other disorders associated with Marfan syndrome include:

- Striae. Striae are stretch marks in the skin caused by rapid weight gain or growth; they frequently occur in pregnant women, for example. Children with Marfan often develop striae over the shoulders, hips, and lower back at an early age because of rapid bone growth. Although the child may be self-conscious about the striae, they are not a danger to health.
- Obstructive **sleep** apnea. Obstructive sleep apnea refers to partial obstruction of the airway during sleep, causing irregular breathing and sometimes snoring. In children with Marfan, obstructive sleep apnea is

caused by the unusual flexibility of the tissues lining the child's airway. This disturbed breathing pattern increases the risk of aortic dissection.

When to call the doctor

Prospective parents with a family history of Marfan syndrome should check with their doctor concerning genetic counseling. Also a doctor should be called if a child has symptoms suggestive of Marfan syndrome.

Diagnosis

Presently, there is no objective diagnostic test for Marfan syndrome, in part because the disorder does not produce any measurable biochemical changes in the child's blood or body fluids, or cellular changes that can be detected from a tissue sample. Although researchers in molecular biology are currently investigating the FBNI gene through a process called mutational analysis, it is presently not useful as a diagnostic test because there is evidence that there can be mutations in the fibrillin gene that do not produce Marfan. Similarly, there is no reliable prenatal test, although some physicians have used ultrasound to try to determine the length of fetal limbs in at-risk pregnancies.

The diagnosis is made by taking a family history and a thorough examination of the child's eyes, heart, and bone structure. The examination should include an echocardiogram taken by a cardiologist, a slit-lamp eye examination by an ophthalmologist, and a work-up of the child's spinal column by an orthopedic specialist. In terms of the cardiac examination, a standard electrocardiogram (EKG) is not sufficient for diagnosis; only the echocardiogram can detect possible enlargement of the aorta. The importance of the slit-lamp examination is that it allows the doctor to detect a dislocated lens, which is a significant indication of the syndrome.

The symptoms of Marfan syndrome in some children resemble the symptoms of homocystinuria, which is an inherited disorder marked by extremely high levels of homocystine in the child's blood and urine. This possibility can be excluded by a urine test.

In other cases, the diagnosis remains uncertain because of the mildness of the child's symptoms, the absence of a family history of the syndrome, and other variables. These borderline conditions are sometimes referred to as marfanoid syndromes.

Treatment

The treatment and management of Marfan is tailored to the specific symptoms of each child. Some children find that the syndrome has little impact on their overall lifestyle; others have found their lives centered on the disorder.

Cardiovascular system

After a child has been diagnosed with Marfan, he or she should be monitored with an echocardiogram every six months until it is clear that the aorta is not growing larger. After that, he or she should have an echocardiogram once a year. If the echocardiogram does not allow the physician to visualize all portions of the aorta, CT (**computed tomography**) or MRI (**magnetic resonance imaging**) may be used. In cases involving a possible aortic dissection, the child may be given a TEE (transesophageal echocardiogram).

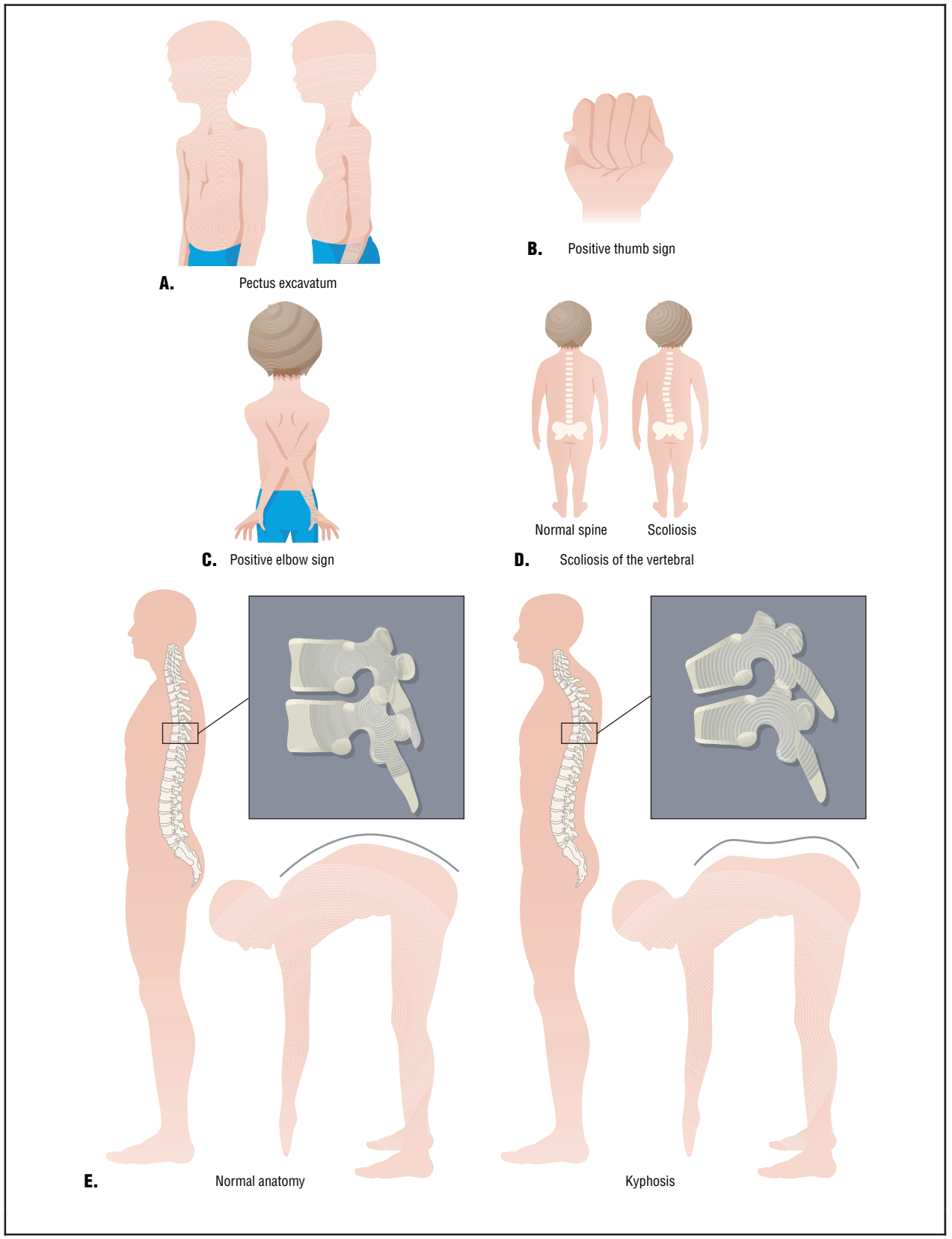
Medications. A child with Marfan may be given drugs called beta-blockers to slow down the rate of aortic enlargement and decrease the risk of dissection by lowering the blood pressure and decreasing the forcefulness of the heartbeat. The most commonly used beta-blockers in chidren with Marfan are propranolol (Inderal) and atenolol (Tenormin). Children who are allergic to beta-blockers may be given a calcium blocker such as verapamil.

Because children with Marfan are at increased risk for infective endocarditis, they must take a prophylactic dose of an antibiotic before having dental work or minor surgery, as these procedures may allow bacteria to enter the bloodstream. Penicillin and amoxicillin are the **antibiotics** most often used.

Surgical treatment. Surgery may be necessary if the width of the child's aorta increases rapidly or reaches a critical size (about 2 inches [5 cm]). The most common surgical treatment involves replacing the child's aortic valve and several inches of the aorta itself with a composite graft, which is a prosthetic heart valve sewn into one end of a Dacron tube. This surgery has been performed widely since about 1985; most children who have had a composite graft have not needed additional surgery. Children who have had a valve replaced must take an anticoagulant medication, usually warfarin (Coumadin), in order to minimize the possibility of a clot forming on the prosthetic valve.

Musculoskeletal system

Children diagnosed with Marfan should be checked for scoliosis by their pediatricians at each annual physical examination. The doctor simply asks the child to bend forward while the back is examined for changes in the curvature. In addition, the child's spine should be x rayed in order to measure the extent of scoliosis or kyphosis. The curve is measured in degrees by the angle between the vertebrae as seen on the x ray. Curves of 20 degrees or less are not likely to become worse. Curves between 20 and 40



Five clinical signs of Marfan syndrome: (left to right) pectus excavatum, positive thumb sign, positive elbow sign, normal spine compared with scoliosis, normal anatomy compared with kyphosis. *(Illustration by Argosy, Inc.)*

degrees are likely to increase in children or adolescents. Curves of 40 degrees or more are highly likely to worsen, even in an adult, because the spine is so badly imbalanced that the force of gravity will increase the curvature.

Scoliosis between 20 and 40 degrees in children is usually treated with a back brace. The child must wear this appliance about 23 hours a day until growth is complete. If the spinal curvature increases to 40 or 50 degrees, the child may require surgery in order to prevent lung problems, back pain, and further deformity. Surgical treatment of scoliosis involves straightening the spine with metal rods and fusing the vertebrae in the straightened position.

Spondylolisthesis is treated with a brace in mild cases. If the slippage is more than 30 degrees, the slipped vertebra may require surgical realignment.

Dural ectasia can be distinguished from other causes of back pain on an MRI. Mild cases are usually not treated. Medication or spinal shunting to remove some of the spinal fluid are used to treat severe cases.

Pectus excavatum and pectus carinatum can be treated by surgery. In pectus excavatum, the deformed breastbone and ribs are raised and straightened by a metal bar. After four to six months, the bar is removed in an outpatient procedure.

Protrusio acetabulae may require artificial hip joint surgery in adult life, if the arthritic pains are severe.

Pain in the feet or limbs is usually treated with a mild analgesic such as **acetaminophen**. Children with Marfan should consider wearing shoes with low heels, special cushions, or orthotic inserts. Foot surgery is rarely necessary.

Visual and dental concerns

Children with Marfan should have a thorough eye examination, including a slit-lamp examination, to test for dislocation of the lens as well as nearsightedness. Dislocation can be treated by a combination of special glasses and daily use of one percent atropine sulfate ophthalmic drops, or by surgery.

Because children with Marfan are at increased risk of glaucoma, they should have the fluid pressure inside the eye measured every year as part of an eye examination. Glaucoma can be treated with medications or with surgery.

Cataracts are treated with increasing success by implant surgery. It is important, however, to seek treatment at medical centers with eye surgeons familiar with the possible complications of cataract surgery in children with Marfan syndrome.

All children with Marfan should be taught to recognize the signs of retinal detachment (sudden blurring of vision in one eye becoming progressively worse without pain or redness) and ask their parents to seek professional help immediately.

Children with Marfan should be evaluated by their dentist at each checkup for crowding of the teeth and possible misalignment and referred to an orthodontist if necessary.

Athletic activities and occupational choice

Children with Marfan should avoid sports or occupations that require heavy weight lifting, rough physical contact, or rapid changes in atmospheric pressure (e.g., scuba diving). Weight lifting increases blood pressure, which in turn may enlarge the aorta. Rough physical contact may cause retinal detachment. Sudden changes in air pressure may produce pneumothorax. Regular noncompetitive physical **exercise**, however, is beneficial for children with Marfan. Good choices include brisk walking, shooting baskets, and slow-paced tennis.

Social and lifestyle issues

Smoking is particularly harmful for children and adolescents with Marfan because it increases their risk of emphysema.

KEY TERMS

Arachnodactyly—A condition characterized by abnormally long and slender fingers and toes.

Ectopia lentis—Dislocation of the lens of the eye. It is one of the most important single indicators in diagnosing Marfan syndrome.

Fibrillin—A protein that is an important part of the structure of the body's connective tissue. In Marfan's syndrome, the gene responsible for fibrillin has mutated, causing the body to produce a defective protein.

Hypermobility—Unusual flexibility of the joints, allowing them to be bent or moved beyond their normal range of motion.

Kyphosis—An extreme, abnormal outward curvature of the spine, with a hump at the upper back.

Pectus carinatum—An abnormality of the chest in which the sternum (breastbone) is pushed outward. It is sometimes called "pigeon breast."

Pectus excavatum—An abnormality of the chest in which the sternum (breastbone) sinks inward; sometimes called "funnel chest."

Scoliosis—An abnormal, side-to-side curvature of the spine.

Children and adolescents with Marfan may benefit from supportive counseling regarding appearance, particularly if their symptoms are severe enough to cause them to withdraw from social activities.

Prognosis

The prognosis for children with Marfan has improved markedly in recent years. By 1995, the life expectancy of people with the syndrome increased to 72 years, up from 48 years in 1972. This dramatic improvement is attributed to new surgical techniques, improved diagnosis, and new techniques of medical treatment.

The most important single factor in improving the child's prognosis is early diagnosis. The earlier that a child can benefit from the new techniques and lifestyle modifications, the more likely he or she is to have a longer life expectancy.

Prevention

Marfan syndrome that occurs because of spontaneous new mutations (15% to 25% of the cases) cannot be prevented. However, for prospective parents with a family history of Marfan syndrome, genetic counseling is recommended. Also, older fathers are more likely to have new mutations appear in chromosome 15.

Parental concerns

Families may wish to seek counseling regarding the effects of the syndrome on relationships within the family. Many people respond with guilt, **fear**, or blame when a genetic disorder is diagnosed in the family, or they may overprotect the affected member. Support groups are often good sources of information about Marfan; they can offer helpful suggestions about living with it as well as emotional support.

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Martin-Bell syndrome *see* **Fragile X syndrome**

Massage therapy

Definition

Massage therapy is the scientific manipulation of the soft tissues of the body, consisting primarily of manual (hands-on) techniques such as applying fixed or movable pressure, holding, and moving muscles and body tissues.

Purpose

Generally, massage is delivered to improve the flow of blood and lymph (fluid in lymph glands, part of immune system), to reduce muscular tension or flaccidity, to affect the nervous system through stimulation or sedation, and to enhance tissue healing. Therapeutic massage may be recommended for children and adults to deliver benefits such as the following:

- reducing muscle tension and stiffness
- relieving muscle spasms
- increasing joint and limb flexibility and range of motion
- increasing ease and efficiency of movement
- relieving points of tension and overall stress; inducing relaxation
- promoting deeper and easier breathing

- improving blood circulation and movement of lymph
- relieving tension-related headaches and eyestrain
- promoting faster healing of soft tissue injuries, such as pulled muscles and sprained ligaments
- reducing **pain** and swelling related to injuries
- reducing the formation of scar tissue following soft tissue injuries
- enhancing health and nourishment of skin
- improving posture by changing tension patterns that affect posture
- reducing emotional or physical stress and reducing **anxiety**
- promoting feelings of well-being
- increasing awareness of the mind-body connection and improving mental awareness and alertness generally

Massage therapy may also be recommended for its documented clinical benefits such as improving pulmonary function in young **asthma** patients, reducing psychoemotional distress in individuals who suffer from chronic inflammatory bowel disease, helping with weight gain, improving motor development in premature infants, and enhancing immune system functioning.

Description

Massage therapy is one of the oldest healthcare practices known. References to massage are found in ancient Chinese medical texts written more than 4,000 years ago. Massage has been advocated in Western healthcare practices since the time of Hippocrates, the "father of medicine."

Massage therapy is the scientific manipulation of the soft tissues of the body for the purpose of normalizing those tissues and consists of a group of manual techniques that include applying fixed or movable pressure, holding, and/or causing movement to parts of the body. While massage therapy is applied primarily with the hands, sometimes the forearms or elbows are used. These techniques affect the muscular, skeletal, circulatory, lymphatic, nervous, and other systems of the body. The basic philosophy of massage therapy embraces the concept of *vis Medicatrix naturae*, which means "aiding the ability of the body to heal itself."

Touch is the fundamental medium of massage therapy. While massage can be described in terms of the type of techniques performed, touch is not used solely in a mechanistic way in massage therapy. Because massage usually involves applying touch with some degree of pressure and movement, the massage therapist must use touch with sensitivity in order to determine the optimal amount of pressure to use for each person. For example, using too much pressure may cause the body to tense up, while using too little may not have enough effect. Touch used with sensitivity also allows the massage therapist to receive useful information via his or her hands about the individual's body, such as locating areas of muscle tension and other soft tissue problems. Because touch is also a form of communication, sensitive touch can convey a sense of caring to the person receiving massage, enhancing the individual's sense of self and well being.

In practice, many massage therapists use more than one technique or method in their work and sometimes combine several. Effective massage therapists ascertain each person's needs and then use the techniques that will best meet those needs.

Swedish massage is the most commonly used form of massage. It uses a system of long gliding strokes, kneading, and friction techniques on the more superficial layers of muscles, generally in the direction of blood flow toward the heart, and sometimes combined with active and passive movements of the joints. It is used to promote general relaxation, improve circulation and range of motion, and relieve muscle tension.

Deep tissue massage is used to release chronic patterns of muscular tension using slow strokes, direct pressure, or friction directed across the grain of the muscles. It is applied with greater pressure and to deeper layers of muscle than Swedish, which is why it is called deep tissue and is effective for chronic muscular tension.

Sports massage uses techniques that are similar to Swedish and deep tissue but are specially adapted to deal with the effects of athletic performance on the body and the needs of athletes regarding training, performing, and recovery from injury.

Neuromuscular massage is a form of deep massage that is applied to individual muscles. It is used primarily to release trigger points (intense knots of muscle tension that refer pain to other parts of the body) and also to increase blood flow. It is often used to reduce pain. Trigger point massage and myotherapy are similar forms.

Acupressure applies finger or thumb pressure to specific points located on the energy pathways or "meridians" in order to release blocked energy along these meridians that may be causing physical discomfort. The rebalance of energy flow releases tension and restores function of organs and muscles in the body. Shiatsu is a Japanese form of acupressure that applies these principles.

Massage therapy sessions can be at home or in a professional office. Most sessions are one hour. Frequency of massage sessions can vary widely as needed based on

A young boy receives a massage. *(© Owen Franken/Corbis.)*

the condition being treated. The cost of massage therapy varies according to geographic location, experience of the massage therapist, and length of the massage. In the United States, as of 2004, the average range is from $35 to $60 for a one-hour session.

The first appointment generally begins with information gathering, such as the reason for getting massage therapy, physical condition and medical history, and other areas. The client is asked to remove clothing to one's level of comfort. Undressing takes place in private, and a sheet or towel is provided for draping. The massage therapist will undrape only the part of the body being massaged. The individual's modesty is respected at all times. The massage therapist may use an oil or cream, which is quickly absorbed into the skin.

Insurance coverage for massage therapy varies widely. There tends to be greater coverage in states that license massage therapy. In most cases, a physician's prescription for massage therapy is needed. Once massage therapy is prescribed, authorization from the insurer may be needed if coverage is not clearly spelled out in one's policy or plan.

Massage therapy may be recommended for children to help relieve conditions such as **allergies**, anxiety and stress, arthritis, asthma and **bronchitis**, joint or limb injuries, post-surgical muscle rehabilitation, chronic and temporary pain, circulatory problems, depression, digestive disorders, tension headaches, **sleep** problems or insomnia, myofascial pain, **sports injuries**, and eating problems associated with temporomandibular joint dysfunction.

Precautions

Massage is comparatively safe; however, it should not be used if the child has one of the following conditions.

- advanced heart disease
- hypertension (high blood pressure)
- phlebitis
- thrombosis
- embolism
- kidney failure

If the child has **cancer**, massage is not advisable if the cancer is the kind that can spread to other organs (metastatic cancer) or if it involves tissue damage due to

chemotherapy or other treatment. Massage may also not be advisable if the child has any of the following conditions.

- a cold
- an infectious disease
- a contagious skin conditions
- an acute inflammation
- an infected injuries
- an unhealed fractures
- dislocations
- is postoperative with a condition in which pain and muscular splinting are increased
- has frostbite
- has large hernias
- has torn ligaments
- has any condition prone to hemorrhage
- has a psychosis
- has any other psychological state that may impair communication or perception

Massage should not be used locally on affected areas (i.e., avoid using massage on the specific areas of the body that are affected by the condition) for the following conditions: eczema, goiter (thyroid dysfunction), and open skin lesions. Massage may be used on the areas of the body that are not affected by these conditions. The decision to use massage must be based on whether it may cause harm. A physician's recommendation is appropriate before a child with any health condition receives massage therapy.

Preparation

Going for a massage requires little in the way of preparation. Generally, one should be clean and should not eat just before a massage. Massage therapists generally work by appointment and usually provide information about how to prepare for an appointment. To receive the most benefit from a massage, parents should give the therapist accurate health information about the child and report discomfort of any kind (whether it is from the massage itself or due to the room temperature or any other distractions). The child can be encouraged to be as receptive to the process as possible.

Aftercare

There are no special recommendations for after a massage. A period of quiet activity or rest following the massage helps maintain full benefits from the procedure.

KEY TERMS

Lymph—Clear, slightly yellow fluid carried by a network of thin tubes to every part of the body. Cells that fight infection are carried in the lymph.

Manipulation—Moving muscles or connective tissue to enhance function, ease tension, and reduce pain in those tissues as well as other beneficial effects.

Mind-body connection—Rather than relying on an understanding of the term "psychosomatic," mind-body medicine acknowledges the influence of thinking and the cognitive process on the behavior of chemicals in the body, involving the mind in both creating the conditions for disease and helping to heal the effects of disease.

Psychoses—Mental illness that interferes with an individual's ability to manage life's challenges and everyday activities. The impairment of cognitive ability that distorts reality.

Risks

Massage therapy does not have notable side effects. Rather than feeling too relaxed or too mentally unfocused after a massage, a child may be both more relaxed and more alert.

Parental concerns

Parents who may not have experienced therapeutic massage themselves or who have doubts about its effectiveness may be interested in the results of research studies, particularly those conducted on groups of children. Well designed studies have documented the benefits of massage therapy for the treatment of acute and chronic pain, acute and chronic inflammation, chronic lymphedema, **nausea**, muscle spasm, various soft tissue dysfunctions, anxiety, depression, insomnia, and psychoemotional stress, which may aggravate mental illness.

Premature infants treated with daily massage therapy gain more weight and have shorter hospital stays than infants who are not massaged. A study of 40 low-birth-weight babies found that the 20 massaged babies had a 47 percent greater weight gain per day and stayed in the hospital an average of six fewer days than 20 infants who did not receive massage, resulting in a cost savings of approximately $3,000 per infant. Cocaine-exposed, preterm infants given massage three times daily for a 10-day period showed significant improvement. Results indicated that massaged infants had fewer postnatal complications and exhibited fewer stress behaviors

during the 10-day period, had a 28 percent greater daily weight gain, and demonstrated more mature motor behaviors.

A study comparing 52 hospitalized depressed and adjustment disorder children and adolescents with a control group that viewed relaxation videotapes, found massage therapy subjects were less depressed and anxious and had lower saliva cortisol levels (an indicator of less depression).

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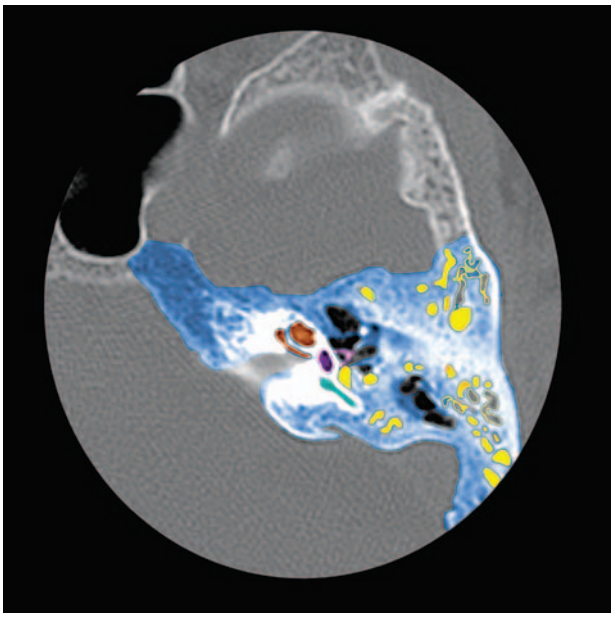
Mastoiditis

Definition

Mastoiditis is a bacterial infection of the air cells in the mastoid bone of the skull. Mastoiditis most commonly affects children. Before the use of **antibiotics**, mastoiditis was one of the leading causes of death in children. As of the early 2000s, it is a relatively uncommon and much less dangerous disorder.

Description

Mastoiditis is usually a consequence of a middle ear infection called acute **otitis media** (AOM). The infection may spread from the ear to the mastoid bone of the skull, which is the bony bump off the base of the skull, located just behind the ears slightly above the level of the earlobe. The mastoid bone is composed of air cells that are in communication with the middle ear. If the air cells fill with infected materials, the mastoid honeycomb-like structure may deteriorate. Mastoiditis has been classified into two types, acute and subacute. Acute or classic mastoiditis refers to acute disease following AOM and involves the development of an abscess behind the ear. Subacute mastoiditis refers to a more chronic disease, often following partial treatment of AOM with antibiotics.



Computed tomography scan (CT scan) showing inflammation and fluid within the air spaces (represented in yellow) of the mastoid. *(© Neil Borden/Photo Researchers, Inc.)*

Demographics

In the United States and first world countries, the incidence of mastoiditis is 0.004 percent. Developing countries have a higher incidence of mastoiditis, presumably resulting from untreated otitis media. The highest incidence occurs in infants aged six to 13 months. As of 2004 reports indicated that acute mastoiditis is on the increase.

Causes and symptoms

The bacteria that cause mastoiditis are those most commonly associated with AOM. They include the following:

KEY TERMS

Abscess—A localized collection of pus in the skin or other body tissue caused by infection.

Audiogram—A chart or graph of the results of a hearing test conducted with audiographic equipment. The chart reflects the softest (lowest volume) sounds that can be heard at various frequencies or pitches.

Computed tomography (CT)—An imaging technique in which cross-sectional x rays of the body are compiled to create a three-dimensional image of the body's internal structures; also called computed axial tomography.

Gram-negative—Refers tof bacteria that have a cell wall composed of a thin layer of peptidoglycan surrounded by an outer membrane made of polysaccharides and proteins. They take on the red color of the counterstain used in the Gram stain procedure.

Mastoid bone—The prominent bone behind the ear that projects from the temporal bone of the skull.

Mastoiditis—An inflammation of the bone behind the ear (the mastoid bone) caused by an infection spreading from the middle ear to the cavity in the mastoid bone.

Myringotomy—A surgical procedure in which an incision is made in the ear drum to allow fluid or pus to escape from the middle ear.

Otitis—Inflammation of the ear, which may be marked by pain, fever, abnormalities of hearing, hearing loss, noise in the ears, and dizzy spells.

Otoscope—A hand-held instrument with a tiny light and a funnel-shaped attachment called an ear speculum, which is used to examine the ear canal and eardrum.

- *Streptococcus pneumoniae*
- *Haemophilus influenzae*
- *Moraxella catarrhalis*
- *Staphylococcus aureus*
- *Pseuodomonas aeruginosa*
- *Klebsiella*
- *Escherichia coli*
- *Proteus*
- *Prevotella*
- *Fusobacterium*
- *Porphyromonas*
- *Bacteroides*

Gram-negative organisms are found more frequently in chronic mastoiditis, and in young infants, which may be due to prolonged antibiotic therapy.

The main symptoms of mastoiditis are increasing earache, **fever**, and the development of redness and swelling behind the ear. The eardrum is inflamed with swelling of the ear canal wall. Mastoiditis typically develops over the few days following an ear infection. This interval is sometimes more prolonged particularly if the initial infection was treated with antibiotics but not completely eliminated (subacute mastoiditis).

When to call the doctor

Children will usually complain of severe **pain** in the affected ear, which may become swollen. Parents should contact their healthcare provider if a child's symptoms indicate mastoiditis. Appointments with the healthcare provider should also be made if a known ear infection does not respond to treatment or is followed by new symptoms.

Diagnosis

In addition to a complete medical history and physical examination, the physician inspects using an otoscope the outer ears and eardrums of the child. Diagnosis is established by clinical tests showing bacterial growth in cultures of ear drainage. Pus taken from the ear or sucked out of the abscess with a needle is sent to a laboratory so that the infecting bacteria can be identified. Imaging studies are used to confirm diagnosis. X rays are considered unreliable but can show clouding of the mastoid air cells. A head CT scan or CT of the ear may show a fluid-filled middle ear and an abnormality in the mastoid bone. Audiograms can also be performed to assess hearing loss.

Treatment

Antibiotics are typically the first course of action in treating mastoiditis. If these do not work, a tube can be inserted to drain off pus or fluid. An incision can be made with the same end purpose. Surgery may also be a recourse, to remove the affected portion of the mastoid bone, to remove a cyst should one be present, and do any further repair required.

Prognosis

Mastoiditis is curable with treatment but may be hard to treat and may recur. Acute mastoiditis usually recovers completely after treatment with no long term damage to hearing and no increased risk of further ear trouble later in life if complications have not occurred.

Prevention

Rapid and complete treatment of ear infections significantly lowers the risk of developing mastoiditis.

Parental concerns

Ear pain is a common complaint from children, but parents should suspect serious ear infection if the ear area is red and swollen. Mastoiditis often causes the ear to be sticking out at an angle. Parents should be aware that ear infections are very common in children, especially those younger than two years of age.

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American Hearing Research Foundation. 55 E. Washington St., Suite 2022, Chicago, IL 60602. Web site: <www.american-hearing.org>.

Better Hearing Institute. 515 King Street, Suite 420, Alexandria, VA 22314. Web site: <www.betterhearing.org>.

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Monique Laberge, Ph.D.

Masturbation

Definition

Masturbation is the erotic stimulation of one's own genitals for pleasure.

Description

Masturbation is the self-stimulation of the sex organs, most often to the point of orgasm. Sixty to ninety percent of adolescent boys and 40 percent of girls masturbate. Although people's attitudes about masturbation differ widely, there is no evidence that masturbation is in any way physically, psychologically, or emotionally harmful. For many young people, masturbation is an opportunity for private sexual exploration before deciding to engage is sexual activity with another person. It is also considered the safest form of sex in the prevention of **sexually transmitted diseases**, including human **immunodeficiency** virus (HIV).

Masturbation allows a healthy way to express and explore one's sexuality and to release sexual tension without the associated risks of sexual intercourse, according to many healthcare providers. They also agree that masturbation is a natural, normal, and healthy way of self-exploration and sexual expression.

It is increasingly recognized among mental health professionals that masturbation can relieve depression and lead to a higher sense of self worth. Masturbation can also be particularly useful in relationships in which one partner wants more sexual activity than the other, in which case masturbation provides a balancing effect.

Many conservative religious groups teach that masturbation is a sinful practice. The Catechism of the Catholic Church, paragraph 2352, lists masturbation as

one of the "Offenses against Chastity" and calls it "an intrinsically and gravely disordered action" because "use of the sexual faculty, for whatever reason, outside of marriage is essentially contrary to its purpose." It goes on to caution that extenuating factors could exist, such as immaturity, habitual, or psychological problems.

The discussion of masturbation has been controversial for hundreds of years and still is to some extent in the early 2000s, more so in the United States than Europe and other Western nations. Children caught by their parents masturbating are often punished and told it is a sin. In fact, there is no mention of the word "masturbation" or "self-pleasure" in the Bible. Children are also often told it is wrong or unhealthy, myths that are not supported by medical research.

In the early 2000s, masturbation has become more accepted for both males and females yet there is still a stigma about discussing it openly. College courses on human sexuality include materials and discussion of masturbation, and many parenting manuals deal with ways to affirm a child's self-pleasing habits rather than degrading or punishing the child. Many sex therapists believe that to have better sexual experiences with a partner, an individual needs to learn to masturbate first since it is the best way to learn what one likes and does not like in his or her sex acts.

Most people think of masturbation as a very personal and private act involving using only the hands to manipulate the genitals. Ways of masturbating common to both males and females include pressing or rubbing the genital area against an object, inserting a finger or other object into the anus, and stimulating the penis or clitoris with electric vibrators, which can also be inserted into the anus or vagina. Some males and females enjoy touching, rubbing, or pinching their nipples while masturbating, and both sexes also sometimes use lubricants, such as hand lotion, to improve the sensation.

Masturbation in males

The most common form of masturbation, especially in circumcised males, is to wrap one or both hands or several fingers and thumb around the erect penis and stroke it up and down until ejaculation. This action results in no direct stimulation of the head of the penis and ejaculation is achieved almost entirely from stimulation of the penis shaft and its contact against the underside of the head of the penis only. In uncircumcised males, it is most common to grip the skin of the penis and move it up and down, resulting in repeated sliding of the foreskin back and forth over the head of the penis until orgasm is reached.

Another common method is to rub the erect penis against a smooth surface, such as a mattress or pillow until ejaculation is reached. Less common masturbation techniques include use of an artificial vagina or other "sex toy."

In 2003, an Australian research team led by Graham Giles of the **Cancer** Council published a medical study that concluded frequent masturbation by males may help prevent the development of prostate cancer and that it would be more helpful than ejaculation through sexual intercourse because intercourse can transmit diseases which can increase the risk of cancer instead.

Masturbation in females

Females most commonly masturbate by stroking or rubbing the vulva, especially the clitoris, with hands and fingers until orgasm is reached. Females also may use running water to stimulate the vulva or insert fingers or a hard object into the vagina. Many women are only able to achieve orgasm through masturbation. Some women can experience sexual stimulation simply by crossing their legs tightly.

One enduring myth is that female masturbation can lead to decreased sensitivity of the clitoris resulting in a decrease in the frequency and intensity of female orgasm. However, the evidence points the other way and suggests that women who have engaged in masturbation have a better understanding of their own genital anatomy and can guide their sexual partners in appreciating the specific sexual acts that contribute to female orgasm.

Infancy and toddlerhood

Some and probably all children are capable of what appear to be sexual responses even in earliest years. Most infants probably explore and fondle their own genitals, but not in a goal directed way. Masturbation by infants is also referred to as gratification disorder or infantile masturbation. It is sometimes mistakenly identified by physicians for epilepsy. A study published in the March 2004 issue of *Archives of Disease in Childhood* reported the median age at first symptoms was ten and one-half months, with an age range of three months to five years and five months. The median frequency was seven times a week and the median length was two and one-half minutes. Masturbation in infants is difficult to recognize because it often does not involve manual stimulation of the genitals at all, the study reported.

Preschool

Occasional masturbation is a normal behavior in preschool-age children and most commonly occurs

"when a child is sleepy, bored, watching television, or under stress," according to a 2002 advisory in the annual journal *Clinical Reference Systems*. The advisory states that up to one third of preschool-age children discover masturbation while exploring their bodies. They often continue to masturbate simply because it feels good. Some children masturbate frequently because they are unhappy or under stress or are reacting to punishment or pressure to stop masturbation completely. Once a child discovers masturbation, he or she seldom stops doing it completely, according to the advisory. It is not abnormal or excessive unless it is deliberately done in public places after age five or six, when most children learn discretion and masturbate only in private.

"It is impossible to eliminate masturbation in a child. Accept the fact you're your child has learned about it and enjoys it," the advisory states. "The only thing you can control is where he or she does it. A reasonable goal is to permit it in the bedroom and bathroom only. . . . If you completely ignore the masturbation, no matter where it's done, your child will think he or she can do it freely in any setting."

School age

As a child grows, masturbation to orgasm becomes more and more likely. Researchers and experts disagree on how many children masturbate before **adolescence**. Most children seem to have the biological capacity to derive pleasure from self-stimulation. Masturbation becomes almost universal at **puberty** in response to normal surges in sex hormones and sexual drive. Most studies suggest that approximately 94 percent of teenage males and about 70 percent of teenage girls admit they masturbate. The actual number of youngsters who masturbate is believed to be higher, since the use of the word "admit" in surveys can imply wrong-doing.

Most males learn to masturbate during adolescence; fewer females do. Some sex therapists believe that girls who do not masturbate miss an important step in their sexual development, since masturbation provides an opportunity to learn how one's body responds to erotic stimulation. Because boys usually masturbate and girls often do not, boys are more likely to learn a sexuality that is genitally focused. Boys learn their sexuality in a context with other boys who bestow a sense of esteem on them. Boys often masturbate with another boy or group of boys. This in itself does not imply **homosexuality** or **bisexuality**. Girls who masturbate almost always discover it alone. Girls generally talk among themselves about masturbation but do not perform with other girls or in front of others. There is no peer support for sexual exploration or reward for teaching orgasm. Boys emerge from adolescence both sexually advantaged and disadvantaged. They are practiced at having orgasms and comfortable with the physical aspects of sex. They are less adept at handling emotional relationships with girls.

Common problems

There is no credible scientific or medical evidence that manual masturbation is damaging to either one's physical or mental health. The exception to this includes some cases of Peyronie's disease in which aggressive manipulation, such as inversion during adolescence, and bending or twisting of the penis, results in a localized benign tumor, distorting the erectile appearance.

Contrary to popular myth, masturbation does not make the palms hairy or cause blindness or genital shrinkage. It has also been alleged that masturbation can reduce sensitivity in the male penis. This statement is also false. The only side-effects recorded are that repeated masturbation may result in tiredness or soreness, which tend to make repeated masturbation self-limiting in any case and that the volume of ejaculate is temporarily reduced in men after multiple ejaculations until normal semen volume is regained in a day or so. Also, people from a socially conservative or religious background may experience feelings of guilt during or after masturbation.

Parental concerns

Studies show that kids who feel they can talk with their parents about masturbation and other sexual issues—because their moms and dads speak openly and listen carefully to them—are less likely to engage in high-risk behavior as teens than kids who do not feel they can talk with their parents about the subject. Parents should explore their own feelings about sex and masturbation. Parents who are uncomfortable with the subject should read books or articles on masturbation and discuss their feelings with a trusted friend, relative, physician, or clergy member. The more parents examine the subject, the more confident they will feel discussing it. If a child has not started asking questions about masturbation, parents should look for a good opportunity to mention it.

While children need to know the biological facts about masturbation, they also need to understand that sexual relationships involve caring, concern, and responsibility. If parents discuss with their children the emotional aspect of a sexual relationships, the children will be better informed to make decisions later on and to resist **peer pressure**.

KEY TERMS

Circumcision—A surgical procedure, usually with religious or cultural significance, where the prepuce or skin covering the tip of the penis on a boy, or the clitoris on a girl, is cut away.

Clitoris—The most sensitive area of the external genitals. Stimulation of the clitoris causes most women to reach orgasm.

Ejaculation—The process by which semen (made up in part of prostatic fluid) is ejected by the erect penis.

Genital—Refers to the sexual or reproductive organs that are visible outside the body.

Infantile masturbation—The masturbation by infants, also called gratification disorder.

Orgasm—Another word for sexual climax. In the male, orgasm is usually accompanied by ejaculation but may be experienced as distinct from ejaculation.

Peyronie's disease—A disease of unknown origin which causes a hardening of the corpora cavernosa, the erectile tissue of the penis. The penis may become misshapen and/or curved as a result and erections are painful.

Vulva—The external genital organs of a woman, including the outer and inner lips, clitoris, and opening of the vagina.

When to call the doctor

In the vast majority of cases masturbation is considered to be a normal activity but the following scenarios may suggest that a problem exists:

- If a child masturbates frequently and appears to be relating to adults in a sexually precocious manner.
- If masturbation becomes a compulsive activity and the person is driven to do it at certain times each day and it almost becomes a ritualistic activity, at the exclusion of almost all else.
- If masturbation takes place in a public place.

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Ken R. Wells

Maxillofacial trauma

Definition

Maxillofacial trauma refers to any injury to the face or jaw caused by physical force, the presence of **foreign objects**, animal or human **bites**, or **burns**.

Description

Maxillofacial trauma includes injuries to any of the bony or fleshy structures of the face. Any part of the face may be affected. Teeth may be knocked out or loosened.

The eyes and their muscles, nerves, and blood vessels may be injured as well as the eye socket (orbit), which can be fractured by a forceful blow. The lower jaw (mandible) may be dislocated by force. Although anchored by strong muscles for chewing, the jaw is unstable in comparison with other bones and is easily dislocated from the temporomandibular joints that attach it to the skull. A fractured nose or jaw may affect the ability to breathe or eat. Any maxillofacial injury may also prevent the passage of air or be severe enough to cause a **concussion** or more serious brain damage.

Athletes are particularly at risk of maxillofacial injuries; one researcher estimates that 10.4 percent of all **fractures** of the facial bones are related to **sports**. Boxers suffer repeated blows to the face and occasional knockouts (traumatic brain injury). Football, basketball, hockey, and soccer players, and many other athletes are at risk for milder forms of brain injury called concussions. Burns to the face are also categorized as maxillofacial trauma.

Demographics

About 3 million injuries to the face and jaw occur in the United States each year. Falls account for 78 percent of facial injuries in preschoolers and 47 percent of such injuries in children between the ages of six and 15. In older adolescents and adults, violent crime or other personal assaults account for almost 50 percent of facial injuries, with automobile accidents accounting for 29 percent and sports-related accidents for another 11 percent. One researcher estimates that about 2 percent of all children or adolescents who participate in sports eventually suffer a facial injury severe enough to require medical attention. Patients between the ages of 17 and 30 are more likely to suffer facial injuries from gunshot **wounds**, while older adults are more likely to be injured by attacks with blunt objects. About 10 percent of facial injuries in young children are caused by parental abuse.

Children who grow up on farms are at significant risk for injury by animals. Of one group of 96 children who required inpatient treatment for head or facial injuries, 39 had been kicked or bitten by horses or other farm animals. Another 37 children had been injured by farm machinery, most commonly a tractor.

Causes and symptoms

Causes

Automobile accidents are a major cause of maxillofacial trauma, as well as participation in sports, fights, and other violent acts. Athletes may sustain facial injuries from colliding with other players (as in football or rugby), from direct contact with equipment (baseball bats, hockey sticks, goal posts, parallel bars, etc.), or from contact with other objects related to the sport (baseballs, hockey pucks, lacrosse balls, skis, etc.) People most at risk are athletes, anyone who drives a vehicle or rides in one, and those who live on farms, do dangerous work, or engage in aggressive types of behavior.

Animals are a common cause of maxillofacial trauma. Horses and other large farm animals can cause severe injury to the face and jaw from kicks or bites. In addition, some large pet dogs can bite hard enough to fracture a small child's facial bones.

Domestic violence and abuse is also a common cause of facial injuries in children and adolescents.

Symptoms

The major symptoms of most facial injuries are **pain**, swelling, bleeding, and bruising, although a fractured jaw also prevents the person from working his jaw properly. Symptoms of a fractured nose include black eyes and possible blockage of the airway due to swelling and bleeding.

Symptoms of eye injury or orbital fracture can include blurred or double vision, decreased mobility of the eye, and **numbness** in the area of the eye. In severe injuries there can be temporary or permanent loss of vision.

Burn symptoms include pain, redness, and possibly blisters, **fever**, and **headache**. Extensive burns can cause the victim to go into shock. In that situation, the person will have low blood pressure and a rapid pulse.

Symptoms of traumatic brain injury include problems with thinking, memory, and judgment as well as mood swings and difficulty with coordination and balance. These symptoms may linger for weeks or months and in severe cases can be permanent. Double vision for months after the injury is not uncommon.

When to call the doctor

Parents should call 911 or take their child to an emergency room at once in the event of a facial injury. The following describe emergency situations that require immediate medical care:

- The child or adolescent is bleeding profusely.
- The patient is having difficulty breathing normally.
- The child or adolescent has lost consciousness or is comatose.
- The patient is nauseated and **vomiting**.

- There are penetrating injuries of the skin, ear, or eye.
- The injury involves a gun, blunt instrument, or animal or human bites.
- The child is seeing double, has other visual disturbances, or staggers when trying to walk.
- Blood or watery fluid is leaking from the child's nose or ears.

Diagnosis

Maxillofacial trauma is often diagnosed and treated by specialists in emergency medicine. About 50 percent of patients with facial injuries have suffered trauma to other organ systems or other parts of the body, however, and may need care from specialists in ophthalmology, plastic surgery, otolaryngology, trauma surgery, oral surgery, and psychiatry as well as from doctors with specialized training in emergency medicine. Injuries to the face and jaw area require special attention because they involve the senses of sight, hearing, taste, and smell as well as such vital functions as breathing. From a psychological perspective, maxillofacial trauma can be additionally upsetting if the patient's appearance is permanently affected.

The doctor will begin by taking a history, either from the patient if he is able to talk or from the parents or other witnesses. In the case of a known accident, sports injury, or assault, the doctor begin with the ABCs, which means that he or she will check the child or adolescent's *airway*, *breathing*, and *circulation*. The doctor will usually have the patient sit upright or lie on one side and will remove blood clots, broken teeth, vomitus, or other foreign bodies from the nose or throat. He or she will then carry out a systematic examination of the patient's face and head. The most common pattern of examination moves from the inside of the nose and mouth to the outside of the face, and from the bottom of the face to the top.

The doctor looks for signs of bruising and tissue swelling as well as bleeding and gently palpates, or touches, the various facial bones for movement and stability. If the doctor suspects that the nose is fractured, he or she will listen for crepitus (a crackling sound) when the nose is gently moved and will look for evidence of a dislocated septum or a septal hematoma, which appears as a bluish bulging mass within the nasal septum. The child's teeth will be examined for looseness, and the muscles and nerves of the face will be evaluated. If there is a discharge from the nose, the doctor will look at it to see whether it contains cerebrospinal fluid, which would indicate damage to the bones of the skull as well as the nasal bones. Lastly, the doctor examines the patient's eyes to make sure that the pupils are responding normally to light and that the patient is not seeing double or having other visual problems that might indicate nerve damage or damage to the eye itself.

In cases involving animal kicks, bites, or other tearing or crushing injuries to the skin and external tissues of the face and jaw, the doctor carefully cleanses the broken skin with soap and water or benzalkonium chloride and checks for fractured facial bones.

Emergency room doctors are required by law to report to local law enforcement authorities cases of suspected **family** abuse.

Treatment

Treatment of maxillofacial trauma varies according to the type and extent of the injury.

Jaw

Dislocation of the jaw can be treated by a primary care physician by exerting pressure in the proper manner. If muscle spasm prevents the jaw from moving back into alignment, a sedative is administered intravenously (IV) to relax the muscles. Afterward, the child must avoid opening the jaw wide to minimize the risk of another dislocation.

A jaw fracture may be minor enough to heal with simple limitation of movement and time. More serious fractures require complicated multistep treatment. The jaw must be surgically immobilized by a qualified oral or maxillofacial surgeon or an otolaryngologist. The jaw is properly aligned and secured with metal pins and wires. Proper alignment is necessary to ensure that the bite is correct. If the bite is off, the patient may develop a painful disorder called temporomandibular joint syndrome.

During the weeks of healing the patient is limited to a liquid diet sipped through a straw and must be careful not to choke or vomit since he cannot open his mouth to expel the vomitus. The surgeon will prescribe pain relievers and perhaps muscle relaxants. The recovery time varies according to the patient's overall health but takes at least several weeks.

Nose

Another common maxillofacial fracture is a broken nose. The bones that form the bridge of the nose may be fractured, but cartilage may also be damaged, particularly the nasal septum that separates the two nostrils. If the child's nose is hit from the side, the bones and cartilage are displaced to the side, but if hit from the front,

they are splayed out. Severe swelling can inhibit diagnosis and treatment. Mild trauma to the nose can sometimes heal without the person being aware of the fracture unless there is an obvious deformity. The nose will be tender for at least three weeks.

Either before the swelling begins or after it subsides, some ten days after the injury, the doctor can assess the extent of the damage. Physical examination of the inside using a speculum and the outside, in addition to a detailed history of how the injury occurred, determines appropriate treatment. The doctor should be informed of any previous nasal fractures, nasal surgery, or such chronic diseases as diabetes or bleeding disorders. Sometimes an x-ray is useful for diagnosis, but it is not always required.

A primary care physician may treat a nasal fracture himself, but if there is extensive damage or the air passage is blocked, he will refer the patient to an otolaryngologist or a plastic surgeon for treatment. Initially the nose may be packed to control bleeding and hold the shape. It is reset under anesthesia. A protective shield or bandage may be placed over it while the fracture heals.

Eyes

In the case of orbital fractures, there is great danger of permanent damage to vision. Double vision and decreased mobility of the eye are common complications of facial trauma. Surgical reconstruction may be required if the fracture changes the position of the eye or there is other facial deformity. Proper treatment of these injuries requires a maxillofacial surgeon.

When the eyes have been exposed to chemicals, they must be washed out for 15 minutes with clear water. **Contact lenses** may be removed only after rinsing the eyes. The eyes should then be kept covered until the person can be evaluated by a primary care physician or ophthalmologist.

When a foreign object is lodged in the eye, the person should not rub the eye or put pressure on it which would further injure the eyeball. The eye should be covered to protect it until medical attention can be obtained.

Mouth and teeth

Several kinds of traumatic injuries can occur to the mouth. A person can suffer a laceration (cut) to the lips or tongue or loosening of teeth or have teeth knocked out. Such injuries often accompany a jaw fracture or other facial injury. Wounds to the soft tissues of the mouth bleed freely, but the plentiful blood supply that leads to this heavy bleeding also helps healing. It is important to clean mouth wounds thoroughly with salt water or a hydrogen peroxide rinse to prevent infection. Large cuts may require sutures and should be done by a maxillofacial surgeon for a good cosmetic result, particularly when the laceration is on the edge of the lip line (vermilion). The doctor will prescribe an antibiotic because there is normally a large amount of bacteria present in the mouth.

Any injury to the teeth should be evaluated by a dentist for treatment and prevention of infection. Implantation of a tooth is sometimes possible if it has been handled carefully and protected. The tooth should be held by the crown, not the root, and kept in milk, saline, or contact lens fluid. The child's dentist can refer him to a specialist in this field.

Facial burns

For first-degree burns, the child's parent can put a cold-water compress on the area or run cold water on it and cover it with a clean bandage for protection. Second- and third-degree burn victims must be taken to the hospital for treatment.

In the hospital, the child will be given replacement fluids through an IV. This treatment is vital since a patient in shock will die unless those lost fluids are replaced quickly. **Antibiotics** are given to combat infection since the burns make the body vulnerable to infection.

Head injuries

Treatment for a **head injury** requires examination by a primary care physician unless the child's symptoms point to a more serious injury. In that case, the victim must seek emergency care. A concussion is treated with rest and avoidance of contact sports. Very often athletes who have suffered a concussion are allowed to **play** again too soon, perhaps in the mistaken impression that the injury is not so bad if the player did not lose consciousness. Anyone who has had one concussion is at increased risk of another one.

Danger signs that a head injury is more serious include worsening headaches, vomiting, weakness, numbness, unsteadiness, change in the appearance of the eyes, seizures, slurred speech, confusion, agitation, or a change in mental status. These signs require immediate transport to the hospital. A neurologist will evaluate the situation, usually with a CT scan. A stay in a rehabilitation facility may be necessary.

In the case of animal bites on the face or head, the child may be given passive or active immunization against **rabies** if there is a chance that the animal is rabid. This precaution is particularly important, as the

incubation period of the rabies virus is much shorter for bites on the head and neck than for bites elsewhere on the body.

Alternative treatment

Fractures, burns, and deep lacerations require treatment by a doctor but alternative treatments can help the body withstand injury and assist the healing process. Calcium, **minerals**, **vitamins**, all part of a balanced and nutrient-rich diet, as well as regular **exercise**, build strong bones that can withstand force well. After an injury, craniosacral therapy may help healing and ease the headaches that follow a concussion or other head trauma. A physical therapist can offer ultrasound treatment, which raises skin temperature to ease pain, or biofeedback, a technique in which the patient learns how to tense and relax muscles to relieve pain. Hydrotherapy may ease the emotional stress of recovering from trauma. Traditional Chinese medicine seeks to reconnect the chi (energy flow) along the body's meridians and thus aid healing. Homeopathic physicians may prescribe such remedies as *Arnica* or *Symphytum* to enhance healing.

Prognosis

When appropriate treatment is obtained quickly after a facial injury, the prognosis can be excellent. If the child or adolescent has a weakened immune system or a debilitating chronic disease, healing is more problematic. Healing also depends upon the extent of the injury. An automobile accident or a gunshot wound, for example, can cause severe facial trauma that may require multiple surgical procedures and a considerable amount of time to heal. Burns and lacerations cause scarring that might be improved by plastic surgery.

Prevention

Safety equipment is vital for preventing maxillofacial trauma from automobile accidents and sports. Here is a partial list of equipment people should always use:

- seatbelts
- automobile air bags
- approved child safety seats
- helmets for riding motorcycles or bicycles, skateboarding, snowboarding, and other sports
- safety glasses for yard work and sports
- such other approved safety equipment for sports as mouthguards, masks, and goggles

KEY TERMS

Corneal abrasion—A scratch on the surface of the cornea.

Crepitus—A crackling sound.

Hematoma—A localized collection of blood, often clotted, in body tissue or an organ, usually due to a break or tear in the wall of blood vessel.

Mandible—The lower jaw, a U-shaped bone attached to the skull at the temporomandibular joints.

Maxilla—The bone of the upper jaw which serves as a foundation of the face and supports the orbits.

Nasal septum—The partition that separates the nostrils.

Orbit—The eye socket which contains the eyeball, muscles, nerves, and blood vessels that serve the eye.

Otolaryngologist—A doctor who is trained to treat injuries, defects, diseases, or conditions of the ear, nose, and throat. Also sometimes known as an otorhinolaryngologist.

Shock—A medical emergency in which the organs and tissues of the body are not receiving an adequate flow of blood. This deprives the organs and tissues of oxygen and allows the build-up of waste products. Shock can be caused by certain diseases, serious injury, or blood loss.

Temporomandibular joint disorder—Inflammation, irritation, and pain of the jaw caused by improper opening and closing of the temporomandibular joint. Other symptoms include clicking of the jaw and a limited range of motion. Also called temporomandibular joint syndrome.

Temporomandibular joint (TMJ)—One of a pair of joints that attaches the mandible of the jaw to the temporal bone of the skull. It is a combination of a hinge and a gliding joint.

Vermilion border—The line between the lip and the skin.

Parental concerns

Parental concerns regarding maxillofacial trauma depend on the cause and severity of the injury. Minor **bruises** and uncomplicated fractures caused by accidents generally heal without problems and are quickly absorbed into the family's routine. Complex fractures or

other injuries requiring a second operation may require explanation or discussion with the child. Facial injuries, particularly repeated injuries related to the neighborhood or farm setting, lifestyle choices, or family violence, however, suggest the importance of professional counseling and changes in the family's structure, geographical location, or level of functioning. Children or adolescents who are severely disfigured by facial injuries may require extra reassurance from family members as well as professional counseling in order to cope with their changed appearance.

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American Academy of Otolaryngology—Head and Neck Surgery. One Prince Street, Alexandria, VA 22314–3357. Web site: <www.entnet.org>.

American Association of Oral & Maxillofacial Surgeons. 9700 W. Bryn Mawr Ave., Rosemont, IL 60018. Web site: <www.acoms.org>.

American College of Sports Medicine (ACSM). 401 West Michigan Street, Indianapolis, IN 46202–3233. Web site: <www.acsm.org>.

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Measles

Definition

Measles is an infection caused by a virus, which causes an illness displaying a characteristic skin rash known as an exanthem. Measles is also sometimes called rubeola, five-day measles, or hard measles.

Description

Measles is a very contagious disease primarily characterized by **cough**, runny nose, red eyes (**conjunctivitis**), and a characteristic rash on the skin and inside of the cheeks. The most common complications are ear infection and **diarrhea**, although more serious complications can include **pneumonia**, **meningitis**, or **encephalitis**. Measles is fatal (due to complications) in about two out of every 1,000 cases.

Demographics

Measles infections appear all over the world. Prior the effective immunization program used in the early

2000s, large-scale measles outbreaks occurred on a two to three-year cycle, usually in the winter and spring. Smaller outbreaks occurred during the off years. Babies up to about eight months of age are usually protected from contracting measles, due to immune cells they receive from their mothers in the uterus. Once someone has had measles infection, he or she can never get it again.

Causes and symptoms

Measles is caused by a type of virus called a paramyxovirus. It is an extremely contagious infection, spread through the tiny droplets that may spray into the air when an individual carrying the virus sneezes or coughs. About 85 percent of those people exposed to the virus will become infected with it. About 95 percent of those people infected with the virus will develop the illness called measles. Once someone is infected with the virus, it takes about seven to 18 days before he or she actually becomes ill. The most contagious time period is the three to five days before symptoms begin through about four days after the characteristic measles rash has begun to appear.

The first signs of measles infection are **fever**; extremely runny nose; red, runny eyes; and a cough. A few days later, a rash appears in the mouth, particularly on the mucous membrane that lines the cheeks. This rash consists of tiny white dots (like grains of salt or sand) on a reddish bump. These are called Koplik's spots and are unique to measles infection. The throat becomes red, swollen, and sore.

A couple of days after the appearance of the Koplik's spots, the measles rash begins. It appears in a characteristic progression, from the head, face, and neck, to the trunk, then abdomen, and next out along the arms and legs. The rash starts out as flat, red patches but eventually develops some bumps. The rash may be somewhat itchy. When the rash begins to appear, the fever usually climbs higher, sometimes reaching as high as 105°F (40.5°C). There may be **nausea**, **vomiting**, diarrhea, and multiple swollen lymph nodes. The cough is usually more problematic at this point, and the patient feels awful. The rash usually lasts about five days. As it fades, it turns a brownish color and eventually the affected skin becomes dry and flaky.

Many patients (about 5–15%) develop other complications. Bacterial infections, such as ear infections, sinus infections, and pneumonia are common, especially in children. Other viral infections may also strike the patient, including **croup**, **bronchitis**, laryngitis, or viral pneumonia. Inflammation of the liver, appendix, intestine, or lymph nodes within the abdomen may cause other complications. Rarely, inflammations of the heart or kidneys, a drop in **platelet count** (causing episodes of difficult-to-control bleeding), or reactivation of an old **tuberculosis** infection can occur.

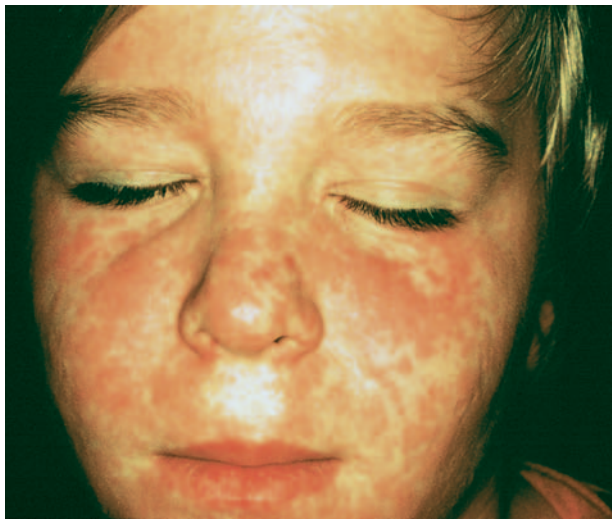
An extremely serious complication of measles infection is swelling of the brain. Called encephalitis, this condition can occur up to several weeks after the basic measles symptoms have resolved. About one out of every thousand patients develops this complication, and about 10 to 15 percent of these patients die. Symptoms include fever, **headache**, sleepiness, seizures, and coma. Long-term problems following recovery from measles encephalitis may include seizures and **mental retardation**.

A very rare complication of measles can occur up to ten years following the initial infection. Called subacute sclerosing panencephalitis, this is a slowly progressing, smoldering swelling and destruction of the entire brain. It is most common among people who had measles infection prior to the age of two years. Symptoms include changes in personality, decreased **intelligence** with accompanying school problems, decreased coordination, involuntary jerks and movements of the body. The disease progresses so that the individual becomes increasingly dependent, ultimately becoming bedridden and unaware of his or her surroundings. Blindness may develop, and the temperature may spike (rise rapidly) and fall unpredictably as the brain structures responsible for temperature regulation are affected. Death is inevitable.

Measles during pregnancy is a serious disease, leading to increased risk of a miscarriage or stillbirth. In addition, the mother's illness may progress to pneumonia.

Diagnosis

Measles infection is almost always diagnosed based on its characteristic symptoms, including Koplik's spots, and a rash which spreads from central body structures out towards the arms and legs. If there is any doubt as to the diagnosis, then a specimen of body fluids (mucus, urine) can be collected and combined with fluorescent-tagged measles virus antibodies. Antibodies are produced by the body's immune cells that can recognize and bind to markers (antigens) on the outside of specific organisms, in this case the measles virus. Once the fluorescent antibodies have attached themselves to the measles antigens in the specimen, the specimen can be viewed under a special microscope to verify the presence of measles virus.



Measles rash on a child's face. *(© CNRI/Photo Researchers, Inc.)*

Treatment

As of 2004 there are no treatments available to stop measles infection. Treatment is primarily aimed at helping the patient to be as comfortable as possible and watching carefully so that **antibiotics** can be started promptly if a bacterial infection develops. Fever and discomfort can be treated with **acetaminophen**. Children with measles should never be given aspirin, as aspirin is correlated with the fatal disease **Reye's syndrome**. A cool-mist vaporizer may help decrease the cough. Patients should be given a lot of liquids to drink, in order to avoid **dehydration** from the fever.

Some studies have shown that children with measles encephalitis benefit from relatively large doses of vitamin A.

Prognosis

The prognosis for an otherwise healthy, well-nourished child who contracts measles is usually quite good. In developing countries, however, death rates may reach 15 to 25 percent. Adolescents and adults usually have a more difficult course. Women who contract the disease while pregnant may give birth to a baby with **hearing impairment**. Although only one in 1,000 patients with measles will develop encephalitis, 10 to 15 percent of those who do will die, and about another 25 percent will be left with permanent brain damage.

KEY TERMS

Antibody—A special protein made by the body's immune system as a defense against foreign material (bacteria, viruses, etc.) that enters the body. It is uniquely designed to attack and neutralize the specific antigen that triggered the immune response.

Antigen—A substance (usually a protein) identified as foreign by the body's immune system, triggering the release of antibodies as part of the body's immune response.

Encephalitis—Inflammation of the brain, usually caused by a virus. The inflammation may interfere with normal brain function and may cause seizures, sleepiness, confusion, personality changes, weakness in one or more parts of the body, and even coma.

Exanthem—A skin eruption regarded as a characteristic sign of such diseases as measles, German measles, and scarlet fever.

Koplik's spots—Tiny spots occurring inside the mouth, especially on the inside of the cheek. These spots consist of minuscule white dots (like grains of salt or sand) set onto a reddened bump and are characteristic of measles.

Prevention

Measles is a highly preventable infection. A very effective vaccine exists, made of live measles viruses that have been treated so that they cannot cause actual infection. The important markers on the viruses are intact, however, which causes an individual's immune system to react. Immune cells called antibodies are produced, which in the event of a future infection with measles virus quickly recognize the organism and kill it off. Measles vaccines are usually given to children at about 15 months of age; prior to that age, the baby's immune system is not mature enough to initiate a reaction strong enough to insure long-term protection from the virus. A repeat injection should be given at about ten or 11 years of age. Outbreaks on college campuses have occurred among students who were not immunized or who were incorrectly immunized.

Measles vaccine should not be given to pregnant women, however, in spite of the seriousness of gestational measles. The reason for not giving this particular vaccine during pregnancy is the risk of transmitting measles to the unborn child.

Parental concerns

New cases of measles began being reported in some countries—including Great Britain—in 2001 because of parents' fears about vaccine safety. The combined vaccine for measles, **mumps**, and **rubella** (MMR) was claimed to cause **autism** or bowel disorders in some children. However, the World Health Organization (WHO) position is there is no scientific merit to these claims. The United Nations expressed concern that unwarranted **fear** of the vaccine would begin spreading the disease in developing countries and ultimately in developed countries as well. Parents in Britain began demanding the measles vaccine as a separate dose, and scientists were exploring that option as an alternative to the combined **MMR vaccine**. Unfortunately, several children died during an outbreak of measles in Dublin because they had not received the vaccine. Child mortality due to measles is considered largely preventable, and making the MMR vaccine widely available in developing countries is part of WHO strategy to reduce child mortality by two-thirds by the year 2015.

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Centers for Disease Control and Prevention. 1600 Clifton Rd., NE, Atlanta, GA 30333. Web site: <www.cdc.gov>.

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Measles, mumps, rubella vaccine *see* **MMR vaccine**

Meditation *see* **Yoga**

Mediterranean fever *see* **Familial Mediterranean fever**

Meningitis

Definition

Meningitis is a serious inflammation of the meninges, the membranes (lining) that surround the brain and spinal cord. It can be of bacterial, viral, or fungal origin.

Description

Meningitis is usually the result of a viral or bacterial infection. Viral meningitis, also called aseptic meningitis, is generally less severe and often disappears without specific treatment, while bacterial meningitis can be quite serious and may result in brain damage, hearing loss, or learning disabilities in children. The infection may even cause death.

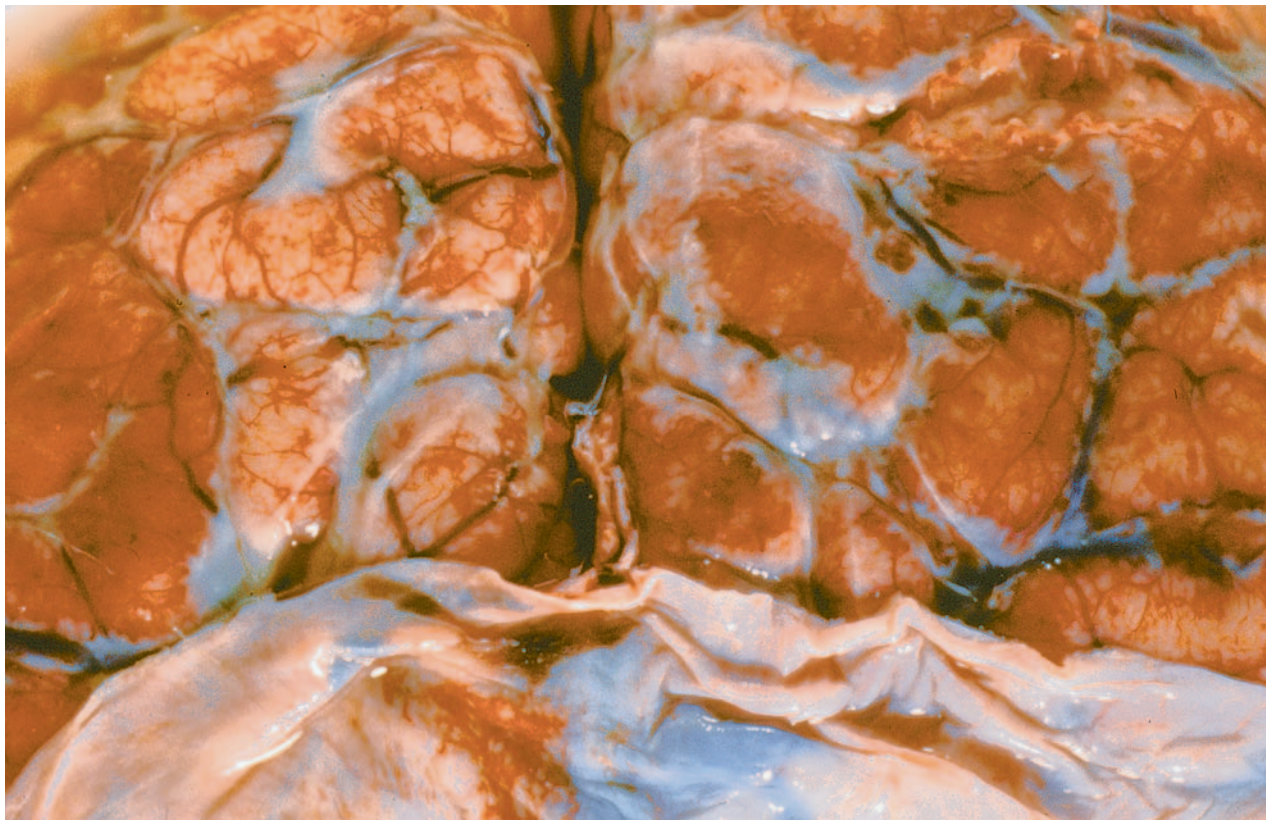
Bacterial meningitis is either monococcal or pneumococcal, depending on the type of bacteria responsible for the infection. Meningitis caused by *Haemophilus influenzae* and related strains (A, B C, Y, and W135) is also called meningococcal meningitis. Similarly, meningitis due to *Streptococcus pneumoniae* is also called pneumococcal meningitis.

Transmission

Most types of meningitis are contagious. A person may be exposed to meningitis bacteria when someone with meningitis coughs or sneezes. The bacteria can also spread through kissing or sharing eating utensils or a toothbrush.

Demographics

According to the National Institute of Neurological Disorders and Stroke (NINDS), some 6,000 cases of pneumococcal meningitis are reported in the United States each year. Meningococcal meningitis is common in minors ages two to 18. Each year about 2,600 people get this highly contagious disease. High-risk groups include infants under the age of one year, people with suppressed immune systems, travelers to foreign countries where the disease is endemic, and college students and Army recruits who reside in dormitories and other close quarters. Between 10 and 15 percent of cases are fatal, with another 10 to 15 percent involving brain damage and other serious side effects.



Brain tissue infected with acute meningitis. *(© Custom Medical Stock Photo, Inc.)*

Causes and symptoms

The bacteria which cause bacterial meningitis live in the back of the nose and throat region and are carried by 10 to 25 percent of the population. They cause meningitis when they get into the bloodstream and travel to the meninges.

At least 50 kinds of bacteria can cause bacterial meningitis. According to the Centers for Disease Control (CDC), before the 1990s, *Haemophilus influenzae* type b (Hib) was the leading cause of bacterial meningitis, but subsequent vaccines given to all children as part of their routine immunizations have reduced the occurrence of the disease due to *H. influenzae*. As of 2004, *Streptococcus pneumoniae* and *Neisseria meningitidis* were the leading causes of bacterial meningitis.

In newborns, the most common agents of meningitis are those that are contracted from the newborn's mother, including Group B streptococci (becoming an increasingly common infecting organism in the newborn period), *Escherichia coli*, and *Listeria monocytogenes*. The highest incidence of meningitis occurs in babies less than a month old, with an increased risk of meningitis continuing through about two years of age.

Older children are more frequently infected by the bacteria *Haemophilus influenzae*, *Neisseria meningitidis*, and *Streptococci pneumoniae*.

Most cases of viral meningitis are caused by enteroviruses (viruses that typically cause stomach flu). However, many other types of viruses, such as the **herpes simplex** virus, the **mumps** and **measles** viruses (against which most children are protected due to mass immunization programs), the virus that causes **chickenpox**, the **rabies** virus, and a number of viruses that are acquired through the **bites** of infected mosquitoes.

Meningitis symptoms include high **fever**, **headache**, and stiff neck in children over the age of two years. These symptoms can develop over several hours, or they may take one to two days. Other symptoms may include **nausea**, **vomiting**, discomfort looking into bright lights, confusion, and sleepiness. In some cases, a rash may be present. In newborns and small infants, these symptoms may be absent or difficult to detect, and the infant may only appear slow or inactive, or be irritable, have vomiting, or be feeding poorly. As the disease progresses, patients of any age may also have seizures.

When to call the doctor

Parents should call a doctor if a child has a temperature above 101°F (38.5°C). If any meningitis symptoms occur, the child should see a doctor immediately, as early diagnosis and treatment are very important for a successful outcome.

Diagnosis

Viral meningitis often remains undiagnosed because its symptoms are similar to those of the common flu. As for bacterial meningitis, the diagnosis is established by growing bacteria from a sample of spinal fluid. The spinal fluid is obtained by performing a lumbar puncture (also called a spinal tap), in which a needle is inserted into an area in the lower back where fluid in the spinal canal is readily accessible.

Treatment

Bacterial meningitis treatment usually involves intravenous administered **antibiotics**, for a minimum of four days. The type of meningitis contracted will determine the specific antibiotic used. It is imperative that treatment start as early as possible, in order to avoid brain damage and death.

Viral meningitis cases usually resolve without complications, but typically, antibiotics are ineffective in treating it, so none are prescribed. The child will be told to get as much rest as he or she can. If the child has **pain** related to the disease such as headaches or other body pains, medication can be used to treat it.

Prognosis

The long-term outlook for children who develop bacterial meningitis varies significantly. The outcome depends on the child's age, the bacteria causing the infection, complications, and the treatment the child receives. The complications of bacterial meningitis can be severe and include neurological problems such as hearing loss, visual impairment, seizures, and learning disabilities. The heart, kidneys, and adrenal glands may also be affected. Although some children develop long-lasting problems, most children who receive prompt diagnosis and treatment recover fully.

The majority of cases of viral meningitis resolve with no complications.

KEY TERMS

Analgesics—A class of pain-relieving medicines, including aspirin and Tylenol.

Antibody—A special protein made by the body's immune system as a defense against foreign material (bacteria, viruses, etc.) that enters the body. It is uniquely designed to attack and neutralize the specific antigen that triggered the immune response.

Antigen—A substance (usually a protein) identified as foreign by the body's immune system, triggering the release of antibodies as part of the body's immune response.

Bacterial meningitis—Meningitis caused by bacteria. Depending on the type of bacteria responsible for the infection, bacterial meningitis is either classified as monococcal or pneumococcal.

Immunization—A process or procedure that protects the body against an infectious disease by stimulating the production of antibodies. A vaccination is a type of immunization.

Meninges—The three-layer membranous covering of the brain and spinal cord, composed of the dura mater, arachnoid, and pia mater. It provides protection for the brain and spinal cord, as well as housing many blood vessels and participating in the appropriate flow of cerebrospinal fluid.

Viral meningitis—Meningitis caused by a virus. Also called aseptic meningitis.

Prevention

Many children as of 2004 routinely receive vaccines against meningitis, starting at about two months of age. Immunizations are recommended by the American Academy of Pediatrics and many other organizations. If a child has not been vaccinated, parents should talk to their doctor about the Hib and pneumococcal (Prevnar 7) vaccines.

Vaccines are available for both meningococcal and pneumococcal meningitis. Specifically, there are vaccines against Hib and against some strains of *N. meningitidis* and many types of *Streptococcus pneumoniae*. The vaccines against Hib are very safe and highly effective. There is a vaccine that protects against four strains of *N. meningitidis*, but it is not routinely used in the United States. There are also vaccines to prevent meningitis due

to *S. pneumoniae*, which can also prevent other forms of infection due to *S. pneumoniae*.

Parents should teach children to wash their hands often, especially before they eat and after using the bathroom, or after petting animals. They should be taught how to wash their hands vigorously, covering both the front and back of each hand with soap and rinsing thoroughly under running water.

Parental concerns

Some forms of bacterial meningitis are contagious. The bacteria are spread through coughing, kissing, and sneezing. Fortunately, the bacteria that cause meningitis are not as contagious as the **common cold** or the flu, and they are not spread by casual contact or by simply breathing the air where a person with meningitis has been. However, people in the same household or daycare center, or anyone with direct contact with a patient's oral secretions is considered at increased risk of acquiring the infection.

Awareness of the symptoms and signs of meningitis, especially the rash which may accompany meningococcal meningitis is very important.

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ORGANIZATIONS

Meningitis Foundation of America. 6610 North Shadeland Avenue, Suite 200, Indianapolis, IN 46220–4393. Web site: <www.musa.org>.

National Institute of Allergy and Infectious Diseases (NIAID). National Institutes of Health, 6610 Rockledge Drive, MSC 6612, Bethesda, MD 20892–6612. Web site: <www.meningitis.org>.

National Meningitis Foundation. 22910 Chestnut Road, Lexington Park, MD 20653. Web site: <www.nmaus.org>.

Meningitis Research Foundation. Midland Way, Thornbury, Bristol BS35 2BS, UK. Web site: <www.meningitis.org>.

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Monique Laberge, Ph.D.

Meningocele *see* **Spina bifida**

Meningococcal meningitis vaccine

Definition

The meningococcal **meningitis** vaccine is given by injection (shots) to provide immunization against meningococcal disease and meningitis caused by the bacterium *Neisseria meningitides*.

Description

Meningococcal disease, or **meningococcemia**, is a leading cause of meningitis in children, and then disease can also lead to infections of the blood. People who acquire the disease can become very ill, especially the young children. Meningococcal disease is treated with **antibiotics**, and the vaccine is not routinely recommended for most people in the United States. Particularly, it is not for children under age two, except under special circumstances.

Meningococcal meningitis is different from the meningitis in infants for which **vaccination** is routinely given. Before the 1990s, *Haemophilus influenzae* type b (Hib) was the leading cause of bacterial meningitis. However, vaccines given to all children as part of their routine immunization have reduced the frequency of the invasive diseases caused by *H. influenzae* and *Streptococcus pneumonia,* leaving neisseria meningitis as one of the leading causes of bacterial meningitis.

The meningococcal vaccine contains inactivated bacteria and cannot cause the disease. It is effective against four of the five subtypes of meningococcal meningitis. It is a one-time injection (except for the very young), and the effects last for four to five years. Adverse reactions are uncommon with this vaccine. Localized redness at the injection site lasting one or two days may occur. Less likely is an allergic reaction to the vaccine.

General use

Meningococcal vaccine is recommended for children and young adults as follows:

- children two years old and older in a population where an outbreak has occurred
- college students who live in close quarters (dormitories), who consume alcohol, smoke, or are regularly around smokers
- those with certain chronic conditions, including those with spleen damage or immune disease
- anyone traveling to or living in a part of the world where meningococcal disease is common, such as West Africa
- household or institutional members who have contact with anyone with meningococcal disease (The same individuals should also receive prophylaxis antibiotic therapy.)
- medical and laboratory personnel at risk of exposure to meningococcal disease

Precautions

Children who are mildly ill at the time the shot is due can still get meningococcal vaccine. Children with moderately severe illnesses should wait until they recover. Children two years old and over receive one dose, while children three months to two years old need two doses, three months apart. Immunizations should be deferred during any acute illness. Pregnant women should not receive the vaccine because it may affect the fetus.

KEY TERMS

Acute—Refers to a disease or symptom that has a sudden onset and lasts a relatively short period of time.

Antibiotics—Drugs that are designed to kill or inhibit the growth of the bacteria that cause infections.

Antibody—A special protein made by the body's immune system as a defense against foreign material (bacteria, viruses, etc.) that enters the body. It is uniquely designed to attack and neutralize the specific antigen that triggered the immune response.

Antigen—A substance (usually a protein) identified as foreign by the body's immune system, triggering the release of antibodies as part of the body's immune response.

Efficacy—The effectiveness of a drug in treating a disease or condition.

Immunity—Ability to resist the effects of agents, such as bacteria and viruses, that cause disease.

Side effects

Children who get the meningitis vaccine may have mild side effects, such as tenderness, redness, or a painful lump on the skin at the injection site; symptoms usually last one to two days. A small percentage of the patients who receive the vaccine develop a slight **fever**. The meningitis vaccine, like any other injection, may in rare cases lead to a serious allergic reaction. Symptoms of allergic reaction include swelling in the mouth or throat, trouble breathing, weakness, hoarseness or wheezing, a fast heart beat, **hives**, **dizziness**, paleness, and a high fever. If a serious allergic reaction occurs the symptoms will start within a few minutes to a few hours after the shot. The child should be seen immediately by a doctor. The doctor will need to know the date of the vaccination and when exactly the symptoms started. A health-care provider should file a report using the vaccine adverse events reporting system (VAERS) form.

Interactions

If the vaccine is given to children receiving immunosuppressive therapy, as in **chemotherapy** for **cancer** or HIV/AIDs, the immune response may not take place. Moreover, the meningitis vaccine should not be given to

individuals known to be sensitive to thimerosal (mercury derivative) or other ingredients of the vaccine.

Parental concerns

Meningitis passes from person to person, mainly by coughing and sneezing. The risk of contracting the disease increases if the child spends time in close contact with the local population at schools, crowded markets, or public buildings. In addition, young adults living in close quarters on college campuses are at risk for contracting the disease. Vaccines are available at student health services on campuses.

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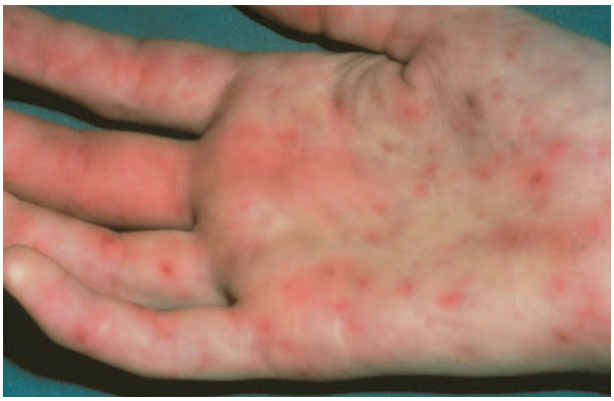
Meningococcemia

Definition

Meningococcemia is the presence of meningococcus in the bloodstream. Meningococcus, a bacteria formally called *Neisseria meningitidis*, can be one of the most dramatic and rapidly fatal of all infectious diseases.

Causes and symptoms

Meningococcemia, a relatively uncommon infection, occurs most commonly in children and young adults. In susceptible people, it may cause a very severe illness that can produce death within hours. The bacteria, which can spread from person to person, usually first causes a colonization in the upper airway, but without symptoms. From there, it can penetrate into the bloodstream to the central nervous system and cause **meningitis** or develop into a full-blown bloodstream infection (meningococcemia). Fortunately in most colonized people, this does not happen, and the result of this colonization is long-lasting immunity against the particular strain.



A close-up image of a person's hand with meningococcemia, caused by *Neisseria meningitidis*. The organism can cause multiple illnesses and can damage small blood vessels. *(Custom Medical Stock Photo Inc.)*

After colonization is established, symptoms can develop within one day to one to two weeks. After a short period of time (one hour up to one to two days) when the patient complains of **fever** and muscle aches, more severe symptoms can develop. Unfortunately during this early stage, a doctor cannot tell this illness from any other illness, such as a viral infection like **influenza**. Unless the case is occurring in a person known to have been exposed to or in the midst of an epidemic of meningococcal disease, there may be no specific symptoms or signs found that help the doctor diagnose the problem. Rarely, a low-grade bloodstream infection called chronic meningococcemia can occur.

After this initial period, the patient will often complain of continued fever, shaking chills, overwhelming weakness, and even a feeling of impending doom. The organism is multiplying in the bloodstream, unchecked by the immune system. The severity of the illness and its dire complications are caused by the damage the organism does to the small blood vessel walls. This damage is called a vasculitis, an inflammation of a blood vessel. Damage to the small vessels causes them to become leaky. The first signs of the infection's severity are small bleeding spots seen on the skin (petechiae). A doctor

should always suspect meningococcemia when he or she finds an acutely ill patient with fever, chills, and petechiae.

Quickly (within hours), the blood vessel damage increases, and large bleeding areas on the skin (purpura) are seen. The same changes are taking place in the affected person's internal organs. The blood pressure is often low, and there may be signs of bleeding from other organs (like coughing up blood, nose bleeds, blood in the urine). The organism not only damages the blood vessels by causing them to leak, but also causes clotting inside the vessels. If this clotting occurs in the larger arteries, it results in major tissue damage. Essentially, large areas of skin, muscle, and internal organs die from lack of blood and oxygen. Even if the disease is quickly diagnosed and treated, the patient has a high risk of dying.

Diagnosis

The diagnosis of meningococcemia can be made by the growth of the organism from blood cultures. Treatment should begin when the diagnosis is suspected and should not be delayed by the doctor's waiting for positive cultures. Obtaining fluid from a petechial spot and staining it in the laboratory can assist in quickly seeing the organism.

Treatment

Immediate treatment of a suspected case of meningococcemia begins with **antibiotics** that work against the organism. Possible choices include penicillin G, ceftriaxone (Rocephin), cefotaxime (Claforan), or trimethoprim/sulfamethoxazole (Bactrim, Septra). If the patient is diagnosed in a doctor's office, antibiotics should be given immediately if possible, even before transfer to the hospital and even if cultures cannot be obtained before treatment. It is most likely that the speed of initial treatment will affect the ultimate outcome.

Prognosis

As many as 15 to 20 percent of patients with meningococcemia will die as a result of the acute infection. A significant percentage of the survivors have tissue damage that requires surgical treatment. This treatment may consist of skin grafts, or even partial or full amputations of an arm or leg. Certain people with immune system defects (particularly those with defects in the complement system) may have recurrent episodes of meningococcemia. These patients, however, seem to have a less serious outcome.

KEY TERMS

Blood culture—A procedure where blood is collected from a vein and is placed in a small bottle that contains a special liquid; the liquid will make any organisms that are present in the blood sample grow. These organisms can then be grown and identified in the laboratory so that the proper antibiotic can be given to the patient.

Colonization—The presence of bacteria on a body surface (like on the skin, mouth, intestines or airway) without causing disease in the person.

Complement—One of several proteins in the blood that acts with other proteins to assist in killing bacteria.

Meningitis—An infection or inflammation of the membranes that cover the brain and spinal cord. It is usually caused by bacteria or a virus.

Prevention

Although a vaccine is available for meningococcus, it is still difficult as of 2004 to produce a vaccine for the type B organism, the most common one in the United States. Because of this and the short time that the vaccine seems to offer protection, the product has not been routinely used in the United States. It can be used for travelers going to areas where meningococcal disease is more common or is epidemic. In the early 2000s, the vaccine has been suggested for use in incoming college freshmen, particularly those living in dormitories. These students appear to have a somewhat higher risk of meningococcal infections.

It is, however, recommended that all people take certain antibiotics if they have had contact (like at home or in a daycare) with a person who has meningococcal infection. The most common antibiotics given are rifampin (Rifadin) or ciprofloxacin (Cipro). These medicines are usually taken by mouth twice a day for two days. This treatment will decrease the risk of infection in these people who have been exposed. However, the overall risk to people who have been exposed, even without antibiotic use, is probably no more than 1 to 2 percent.

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Meningomyelocele *see* **Spina bifida**

Menstruation

Definition

Menstruation is the vaginal bleeding that occurs in adolescent girls and women as a result of hormonal changes. It normally happens in a predictable pattern, once a month.

Description

Menstruation is part of the menstrual cycle, which helps a woman's body prepare for the possibility of pregnancy each month. The parts of the body involved in the menstrual cycle include the uterus and cervix, the ovaries, fallopian tubes, the brain and pituitary gland, and the vagina. Certain body chemicals known as hormones rise and fall during the month, causing the menstrual cycle to occur.

In the first half of the menstrual cycle, estrogen levels rise, causing the lining of the uterus to grow and thicken. This lining is called the endometrium. The two small, grape-shaped organs inside the abdomen on either side of the uterus, known as the ovaries, are filled with hundreds of thousands of eggs and are the organs that allow pregnancy to occur. When a girl reaches **puberty**, the ovaries respond to a rise in follicle-stimulating hormone and cause one of the eggs to mature. About half way through the menstrual cycle, a surge of luteinizing hormone takes place, and the egg is released. This mature egg is called an ovum, and its release is called ovulation. When the egg is released it travels through one of the two fallopian tubes and down towards the uterus. If the ovum is fertilized by a sperm at this time, pregnancy occurs. However, if a sperm does not fertilize the egg, the body no longer needs the uterine lining to support the fertilized egg. Estrogen and progesterone levels then drop, triggering the uterine lining to gently fall away from the wall of the uterus, and to be shed through the vagina. The discharge of this lining is the menstrual flow. The entire process is called menstruation.

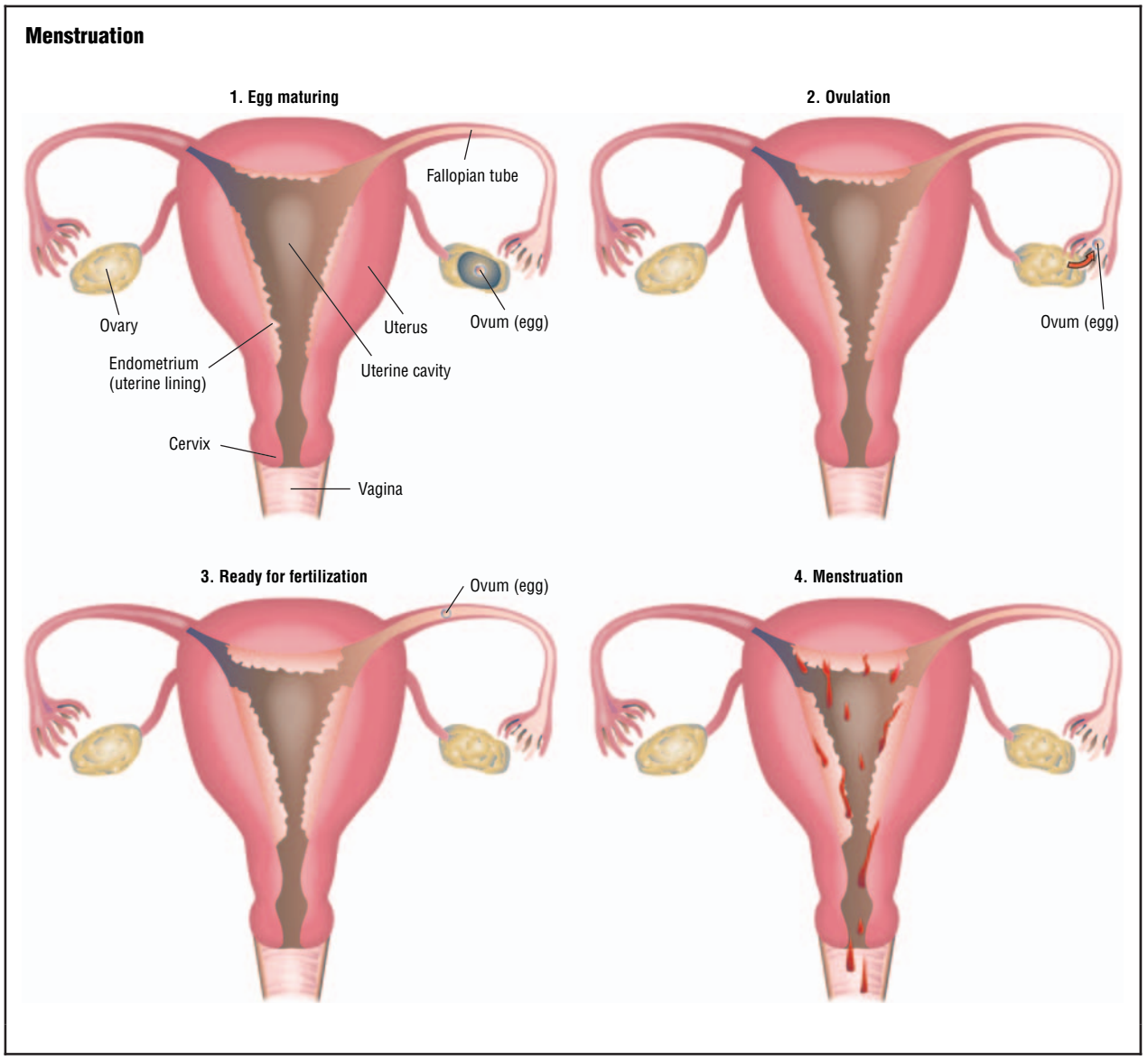
A "normal" menstrual period usually occurs every 28 days, from the first day of a period to the first day of the next. However, this can vary from 22 to 36 days. Each period usually lasts from three to seven days, with the average being five. It may take several years from the start of menstruation for periods to settle into a pattern. Irregular periods are common in early **adolescence**. Even after adolescence, many factors can throw off the timing of menstruation. These include weight changes, starting a new job or school, and relationship problems.

Menstrual hygiene products

Once a girl begins menstruating, she needs to choose from the various menstrual hygiene products which are available. Menstrual hygiene products can be divided into two basic categories: sanitary pads and tampons. Absorbency and a comfortable fit are the main features girls need to look for when purchasing menstrual products. Because a girl's menstrual flow may vary from day to day during the cycle, she may want to use different types of products during her period.

Sanitary pads are worn inside the underwear where they collect the menstrual flow. They come in different sizes, thicknesses, and styles. Some pads have flaps or "wings" that wrap around and attach to the underside of underwear. Others have deodorant and contain perfume. Some girls find that the perfume irritates their skin.

Tampons are the another option for absorbing menstrual flow. Tampons come in various absorbency categories and should be chosen based on the amount of flow experienced. The absorbency of a tampon can be determined by how often it needs to be changed. Girls should use the tampon with the least absorbency necessary to absorb the flow. Tampons should be changed every four to six hours. Tampons also come with a variety of applicators, including plastic and cardboard. Tampons are comfortable to wear and may be a good choice for active girls. They should be inserted carefully to avoid any irritation. A rare, but serious, condition called **toxic shock syndrome** (TSS) can be connected to tampon use. The higher the absorbency of tampons used, the higher the risk for TSS. To decrease the risk of TSS, girls should choose the lowest absorbency necessary.



In this illustration, the menstrual cycle is divided into four stages. First, an egg matures inside the ovary (1), which then releases the egg (2), allowing it to travel through the fallopian tube, where it rests awaiting fertilization (3). If the egg is not fertilized, it is flushed out with the menstrual flow (4). *(Illustration by GGS Information Services.)*

Problems with menstruation

DYSMENORRHEA **Dysmenorrhea** is the medical term for menstrual cramps, the dull or throbbing **pain** in the lower abdomen that many women experience just before and during their menstrual periods. It can be primary or secondary. Primary dysmenorrhea involves no abnormality. Secondary dysmenorrhea involves an underlying physical cause, such as uterine fibroids, pelvic inflammatory disease, or endometriosis. Signs and symptoms of dysmenorrhea, whether it is primary or secondary, may include the following:

- dull, throbbing pain in the lower abdomen
- radiating pain to the lower back and thighs
- nausea, loose stools, sweating, and **dizziness** (though these are much less common)

If menstrual cramps become severe enough to keep a girl from going about her day-to-day routine, she should see a doctor. The doctor will perform a medical history and physical examination, including a pelvic exam, where he or she will look for any abnormalities, signs of infection, and possible causes of secondary dysmenorrheal. In addition, the doctor may request a variety

of diagnostic tests, such as imaging tests, laparoscopy, and hysteroscopy.

Complications can arise from secondary dysmenorrhea. If pelvic inflammatory disease is present, the fallopian tubes may become scarred and possibly cause later infertility or other reproductive problems. Endometriosis can also lead to fertility problems as well.

Many experts believe that prostaglandins, hormone-like substances involved in pain and inflammation and which trigger uterine muscle contractions, are responsible for causing menstrual cramping. Whether the dysmenorrhea is primary or secondary, there are effective ways to treat menstrual pain. **Nonsteroidal anti-inflammatory drugs** (NSAIDS), such as ibuprofen or naproxen, may block the production of prostaglandins and can be very effective in the treatment of menstrual cramps. In the case of severe cramping, doctors may recommend a low-dose oral contraceptive to prevent ovulation, which may reduce the release of prostaglandins and the severity of the cramps.

DYSFUNCTIONAL UTERINE BLEEDING Dysfunctional uterine bleeding (DUB) is prolonged or heavy bleeding that often occurs in a menstrual cycle where ovulation did not occur. Heavy bleeding is defined as more than 15 soaked pads or tampons per period, and prolonged bleeding is that which lasts for more than 8 to 10 days. Although DUB is quite common in the first few years after menstruation starts, it can be frightening and should always be reported to a physician. DUB that is accompanied by dizziness and a low blood pressure should be considered a medical emergency. DUB is usually caused by hormonal imbalances. Other causes of bleeding are sexually transmitted disease, an ectopic pregnancy, ovarian cysts, and uterine fibroids or polyps. Young women within the first menstrual period are not usually treated unless symptoms are exceptionally severe or if anemia develops.

Demographics

Girls may start their menstrual period as early as nine years of age and as late as 16 years old. The average age a girl begins menstruating is 12. Girls who are very active in **sports** or who are quite thin may not develop until a later age. Losing weight while experiencing a growth spurt may also delay menstruation.

In the early 2000s, some people have voiced concern about girls starting their periods at younger and younger ages. However, a study reported in 2003 found that overall, girls in the United States are not beginning menstruation earlier than in the past. Less than 10 percent of girls start their periods before 11 years of age, and 90 percent of all U.S. girls are menstruating by age 14. This age is not significantly different than that reported for girls in 1973. African-American girls on average begin menstruating before Caucasian- and Hispanic-American girls.

Causes and symptoms

The menstrual cycle takes place each month in response to the hormonal changes which occur when pregnancy does not take place. A number of symptoms can occur just before and during a girl's period which may cause discomfort. These include:

- having pelvic area cramps
- feeling bloated or puffy
- breast tenderness or swelling
- headaches and backaches
- acne breakouts
- mild nausea

These symptoms usually stop or lessen a day or two after the period begins.

Diagnosis

There are several reasons why a girl should see her healthcare provider regarding her menstrual cycle. These include:

- if menstruation has not started by the age of 16
- if a menstrual period lasts for more than seven days
- if periods suddenly stop
- if she is experiencing excessive bleeding
- if she feels suddenly ill while using tampons
- if she bleeds more than a few drops between periods
- if she experiences excessive pain during her period

Treatment

No specific medical treatment is necessary for an uncomplicated menstrual cycle, as it is a normal, healthy process in girls and women.

Alternative treatment

Some girls may find relief from menstrual discomfort through meditation, **yoga**, or massage. These stress-relieving activities are unlikely to cause any harm.

KEY TERMS

Dysmenorrhea—Painful menstruation.

Menarche—The first menstrual cycle in a girl's life.

Parental concerns

Though menstruation is no longer the taboo subject it once was, many parents still find that discussing the issue with their daughters can be uncomfortable. This is especially common in families in which the mother is not present. Still, it is important to discuss menstruation with girls when they are preteens, so that they do not experience the potential embarrassment or trauma if they start their first period without knowledge about what is happening to their bodies. Taking the time to prepare may help to make this discussion less awkward.

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American College of Obstetricians and Gynecologists. 409 12th Street, SW, PO Box 96920, Washington, DC 20090–6290. Web site: <www.acog.org>.

The Center for Young Women's Health. 300 Longwood Avenue, Box 310, Boston, MA 02115. Web site: <www.youngwomenshealth.org>.

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Menstruation, infrequent *see* **Oligomenorrhea**

Menstruation, painful *see* **Dysmenorrhea**

Mental development *see* **Cognitive development**

Mental retardation

Definition

Mental retardation is a developmental disability that first appears in children under the age of 18. It is defined as an intellectual functioning level (as measured by standard tests for **intelligence** quotient) that is well below average and significant limitations in daily living skills (adaptive functioning).

Description

According to statistics made available by the Centers for Disease Control and Prevention in the 1990s, mental retardation occurs in 2.5 to 3 percent of the general population. About 6 to 7.5 million mentally retarded individuals live in the United States alone. Mental retardation begins in childhood or **adolescence** before the age of 18. In most cases, it persists throughout adulthood. A diagnosis of mental retardation is made if an individual has an intellectual functioning level well below average and significant limitations in two or more adaptive skill areas. Intellectual functioning level is defined by standardized tests that measure the ability to reason in terms of mental age (intelligence quotient or IQ). Mental retardation is defined as IQ score below 70 to 75. Adaptive skills are the skills needed for daily life. Such skills include the ability to produce and understand language (communication); home-living skills; use of community resources; health, **safety**, leisure, self-care, and social skills; self-direction; functional academic skills (reading, writing, and arithmetic); and work skills.

In general, mentally retarded children reach developmental milestones such as walking and talking much later than the general population. Symptoms of mental retardation may appear at birth or later in childhood. Time of onset depends on the suspected cause of the disability. Some cases of mild mental retardation are not diagnosed before the child enters **preschool**. These children typically have difficulties with social, communication, and functional academic skills. Children who have a neurological disorder or illness such as **encephalitis** or **meningitis** may suddenly show signs of cognitive impairment and adaptive difficulties.

Mental retardation varies in severity. There are four different degrees of mental retardation: mild, moderate, severe, and profound. These categories are based on the functioning level of the individual.

Mild mental retardation

Approximately 85 percent of the mentally retarded population is in the mildly retarded category. Their IQ score ranges from 50 to 75, and they can often acquire academic skills up to the sixth grade level. They can become fairly self-sufficient and in some cases live independently, with community and social support.

Moderate mental retardation

About 10 percent of the mentally retarded population is considered moderately retarded. Moderately retarded individuals have IQ scores ranging from 35 to 55. They can carry out work and self-care tasks with moderate supervision. They typically acquire **communication skills** in childhood and are able to live and function successfully within the community in a supervised environment such as a group home.

Severe mental retardation

About 3 to 4 percent of the mentally retarded population is severely retarded. Severely retarded individuals have IQ scores of 20 to 40. They may master very basic self-care skills and some communication skills. Many severely retarded individuals are able to live in a group home.

Profound mental retardation

Only 1 to 2 percent of the mentally retarded population is classified as profoundly retarded. Profoundly retarded individuals have IQ scores under 20 to 25. They may be able to develop basic self-care and communication skills with appropriate support and training. Their retardation is often caused by an accompanying neurological disorder. The profoundly retarded need a high level of structure and supervision.

The American Association on Mental Retardation (AAMR) has developed another widely accepted diagnostic classification system for mental retardation. The AAMR classification system focuses on the capabilities of the retarded individual rather than on the limitations. The categories describe the level of support required. They are: intermittent support, limited support, extensive support, and pervasive support. Intermittent support, for example, is support needed only occasionally, perhaps during times of stress or crisis. It is the type of support typically required for most mildly retarded individuals. At the other end of the spectrum, pervasive support, or life-long, daily support for most adaptive areas, would be required for profoundly retarded individuals.

Demographics

For children, the mental retardation rate is 11.4 per 1,000 and varies approximately nine fold, ranging from 3.2 in New Jersey to 31.4 in Alabama. For adults, the rate is 6.6 and varies approximately six fold, ranging from 2.5 in Alaska to 15.7 in West Virginia. In 42 states, the rate for children is higher than that for adults; in seven states, the rate for adults is higher, and in two states, both rates are similar. The correlation between state-specific rates for children and for adults is 0.66. Overall, 69 percent of the state-specific variation in prevalence rates for adults is accounted for by median household income, the percentage of total births to teenaged mothers, and the percentage of the population with less than a ninth-grade education. Low educational attainment was the most important correlate of mental retardation rates among adults.

Causes and symptoms

Low IQ scores and limitations in adaptive skills are the hallmarks of mental retardation. Aggression, self-injury, and **mood disorders** are sometimes associated with the disability. The severity of the symptoms and the age at which they first appear depend on the cause. Children who are mentally retarded reach developmental milestones significantly later than expected, if at all. If retardation is caused by chromosomal or other genetic disorders, it is often apparent from infancy. If retardation is caused by childhood illnesses or injuries, learning and adaptive skills that were once easy may suddenly become difficult or impossible to master. In about 35 percent of cases, the cause of mental retardation cannot be found. Biological and environmental factors that can cause mental retardation include genetics, prenatal illnesses and issues, childhood illnesses and injuries, and environmental factors.

Genetics

About 5 percent of mental retardation is caused by hereditary factors. Mental retardation may be caused by an inherited abnormality of the genes, such as **fragile X syndrome**. Fragile X, a defect in the chromosome that determines sex, is the most common inherited cause of mental retardation. Single gene defects such as **phenylketonuria** (PKU) and other inborn errors of metabolism may also cause mental retardation if they are not found and treated early. An accident or mutation in genetic

development may also cause retardation. Examples of such accidents are development of an extra chromosome 18 (trisomy 18) and **Down syndrome**. Down syndrome is caused by an abnormality in the development of chromosome 21. It is the most common genetic cause of mental retardation.

Prenatal illnesses and issues

Fetal alcohol syndrome affects one in 600 children in the United States. It is caused by excessive alcohol intake in the first twelve weeks (trimester) of pregnancy. Some studies have shown that even moderate alcohol use during pregnancy may cause learning disabilities in children. Drug abuse and cigarette **smoking** during pregnancy have also been linked to mental retardation.

Maternal infections and illnesses such as glandular disorders, **rubella**, **toxoplasmosis**, and **cytomegalovirus infection** may cause mental retardation. When the mother has high blood pressure (**hypertension**) or blood poisoning (toxemia), the flow of oxygen to the fetus may be reduced, causing brain damage and mental retardation.

Birth defects that cause physical deformities of the head, brain, and central nervous system frequently cause mental retardation. Neural tube defect, for example, is a birth defect in which the neural tube that forms the spinal cord does not close completely. This defect may cause children to develop an accumulation of cerebrospinal fluid on the brain (**hydrocephalus**). By putting pressure on the brain hydrocephalus can cause learning impairment.

Childhood illnesses and injuries

Hyperthyroidism, **whooping cough**, **chickenpox**, **measles**, and Hib disease (a bacterial infection) may cause mental retardation if they are not treated adequately. An infection of the membrane covering the brain (meningitis) or an inflammation of the brain itself (encephalitis) cause swelling that in turn may cause brain damage and mental retardation. Traumatic brain injury caused by a blow or a violent shake to the head may also cause brain damage and mental retardation in children.

Environmental factors

Ignored or neglected infants who are not provided the mental and physical stimulation required for normal development may suffer irreversible learning impairments. Children who live in poverty and suffer from **malnutrition**, unhealthy living conditions, and improper or inadequate medical care are at a higher risk. Exposure to lead can also cause mental retardation. Many children develop **lead poisoning** by eating the flaking lead-based paint often found in older buildings.

When to call the doctor

If mental retardation is suspected, a comprehensive physical examination and medical history should be done immediately to discover any organic cause of symptoms. Conditions such as hyperthyroidism and PKU are treatable. If these conditions are discovered early, the progression of retardation can be stopped and, in some cases, partially reversed. If a neurological cause such as brain injury is suspected, the child may be referred to a neurologist or neuropsychologist for testing.

The symptoms of mental retardation are usually evident by a child's first or second year. In the case of Down syndrome, which involves distinctive physical characteristics, a diagnosis can usually be made shortly after birth. Mentally retarded children lag behind their peers in developmental milestones such as smiling, sitting up, walking, and talking. They often demonstrate lower than normal levels of interest in their environment and responsiveness to others, and they are slower than other children in reacting to visual or auditory stimulation. By the time a child reaches the age of two or three, retardation can be determined using physical and **psychological tests**. Testing is important at this age if a child shows signs of possible retardation because alternate causes, such as impaired hearing, may be found and treated.

Diagnosis

A complete medical, **family**, social, and educational history is compiled from existing medical and school records (if applicable) and from interviews with parents. Children are given intelligence tests to measure their learning abilities and intellectual functioning. Such tests include the Stanford-Binet Intelligence Scale, the Wechsler Intelligence Scales, the Wechsler Preschool and Primary Scale of Intelligence, and the Kaufmann Assessment Battery for Children. For infants, the **Bayley Scales of Infant Development** may be used to assess motor, language, and problem-solving skills. Interviews with parents or other caregivers are used to assess the child's daily living, muscle control, communication, and social skills. The Woodcock-Johnson Scales of Independent Behavior and the Vineland Adaptive Behavior Scale (VABS) are frequently used to test these skills.

Treatment

Federal legislation entitles mentally retarded children to free testing and appropriate, individualized education and skills training within the school system from ages three to 21. For children under the age of three, many states have established early intervention programs that assess, recommend, and begin treatment programs. Many day schools are available to help train retarded children in basic skills such as bathing and feeding themselves. **Extracurricular activities** and social programs are also important in helping retarded children and adolescents gain **self-esteem**.

Training in independent living and job skills is often begun in early adulthood. The level of training depends on the degree of retardation. Mildly retarded individuals can often acquire the skills needed to live independently and hold an outside job. Moderate to profoundly retarded individuals usually require supervised community living. **Family therapy** can help relatives of the mentally retarded develop coping skills. It can also help parents deal with feelings of guilt or anger. A supportive, warm home environment is essential to help the mentally retarded reach their full potential. However, as of 2004, there is no cure for mental retardation.

A promising but controversial treatment for mental retardation involves stem cell research. In the early 2000s scientists are exploring the potential of adult stem cells in treating mental retardation. They have transplanted bone marrow cells into living embryos in the uteri of animals to approach congenital diseases, birth defects, and mental retardation. Stem cells are primitive cells that are capable of forming diverse types of tissue. Because of this remarkable quality, human stem cells hold huge promise for the development of therapies to regenerate damaged organs and heal people who are suffering from terrible diseases. Embryonic stem cells are derived from human embryos. Their use is controversial because such stem cells cannot be used in research without destroying the living embryo. Other sources of stem cells are available, however, and can be harvested from umbilical cord blood as well as from fat, bone marrow, and other adult tissue without harm to the donor. An enormous amount of research involving adult stem cells is going on as of 2004 in laboratories in the United States.

Prognosis

Individuals with mild to moderate mental retardation are frequently able to achieve some self-sufficiency and to lead happy and fulfilling lives. To reach these goals, they need appropriate and consistent educational, community, social, family, and vocational supports. The outlook is less promising for those with severe to profound retardation. Studies have shown that these individuals have a shortened life expectancy. The diseases that are usually associated with severe retardation may cause the shorter life span. People with Down syndrome develop in later life the brain changes that characterize Alzheimer's disease and may develop the clinical symptoms of this disease as well.

Prevention

Immunization against diseases such as measles and Hib prevents many of the illnesses that can cause mental retardation. In addition, all children should undergo routine developmental screening as part of their pediatric care. Screening is particularly critical for those children who may be neglected or undernourished or may live in disease-producing conditions. Newborn screening and immediate treatment for PKU and hyperthyroidism can usually catch these disorders early enough to prevent retardation. Good prenatal care can also help prevent retardation. Pregnant women should be educated about the risks of drinking and the need to maintain good **nutrition** during pregnancy. Tests such as **amniocentesis** and ultrasonography can determine whether a fetus is developing normally in the womb.

Parental concerns

All states are required by law to offer early intervention programs for mentally retarded children from the time they are born. The sooner the diagnosis of mental retardation is made, the more the child can be helped. With mentally retarded infants, the treatment emphasis is on sensorimotor development, which can be stimulated by exercises and special types of **play**. It is required that **special education** programs be available for retarded children starting at three years of age. These programs concentrate on essential self-care, such as feeding, dressing, and **toilet training**. There is also specialized help available for language and communication difficulties and physical disabilities. As children grow older, training in daily living skills, as well as academic subjects, is offered.

Counseling and therapy are another important type of treatment for the mentally retarded. Retarded children are prone to behavioral problems caused by short attention span, low tolerance for frustration, and poor impulse control. Behavior therapy with a mental health professional can help combat negative behavior patterns and replace them with more functional ones. A counselor or therapist can also help retarded children cope with the

KEY TERMS

Amniocentesis—A procedure performed at 16–18 weeks of pregnancy in which a needle is inserted through a woman's abdomen into her uterus to draw out a small sample of the amniotic fluid from around the baby for analysis. Either the fluid itself or cells from the fluid can be used for a variety of tests to obtain information about genetic disorders and other medical conditions in the fetus.

Developmental delay—The failure of a child to meet certain developmental milestones, such as sitting, walking, and talking, at the average age. Developmental delay may indicate a problem in development of the central nervous system.

Down syndrome—A chromosomal disorder caused by an extra copy or a rearrangement of chromosome 21. Children with Down syndrome have varying degrees of mental retardation and may have heart defects.

Hib disease—An infection caused by *Haemophilus influenza* type b (Hib). This disease mainly affects children under the age of five. In that age group, it is the leading cause of bacterial meningitis, pneumonia, joint and bone infections, and throat inflammations.

Inborn error of metabolism—One of a group of rare conditions characterized by an inherited defect in an enzyme or other protein. Inborn errors of metabolism can cause brain damage and mental retardation if left untreated. Phenylketonuria, Tay-Sachs disease, and galactosemia are inborn errors of metabolism.

Phenylketonuria (PKU)—A rare, inherited, metabolic disorder in which the enzyme necessary to break down and use phenylalanine, an amino acid necessary for normal growth and development, is lacking. As a result, phenylalanine builds up in the body causing mental retardation and other neurological problems.

Trisomy—An abnormal condition where three copies of one chromosome are present in the cells of an individual's body instead of two, the normal number.

Ultrasonography—A medical test in which sound waves are directed against internal structures in the body. As sound waves bounce off the internal structure, they create an image on a video screen. Ultrasonography is often used to diagnose fetal abnormalities, gallstones, heart defects, and tumors. Also called ultrasound imaging.

low self-esteem that often results from the realization that they are different from other children, including siblings. Counseling can also be valuable for the family of a retarded child to help parents cope with painful feelings about the child's condition and with the extra time and patience needed for the care and education of a special-needs child. Siblings may need to talk about the pressures they face, such as accepting the extra time and attention their parents must devote to a retarded brother or sister. Sometimes parents have trouble **bonding** with an infant who is retarded and need professional help and reassurance to establish a close and loving relationship.

Current social and healthcare policies encourage keeping mentally retarded persons in their own homes or in informal group home settings rather than institutions. The variety of social and mental health services available to the mentally retarded, including pre-vocational and vocational training, are geared toward making this possible.

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Methamphetamine *see* **Stimulant drugs**

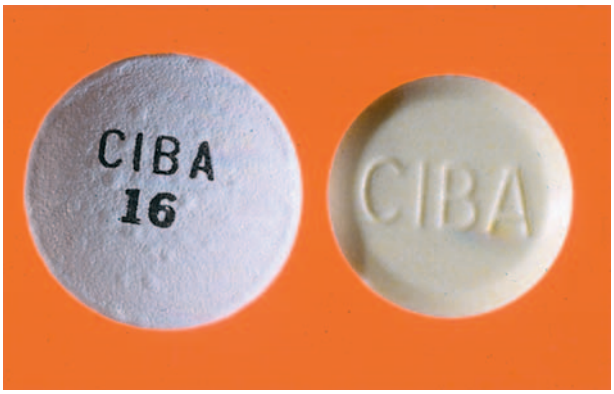
Methylphenidate

Definition

The generic name for the drug Ritalin, the most commonly prescribed medication for treating children with attention-deficit hyperactivity disorder (ADHD).

Description

Methylphenidate is one of a group of drugs called central nervous system (CNS) stimulants. It is used to treat attention-deficit hyperactivity disorder, **narcolepsy** (uncontrollable desire for **sleep** or sudden attacks of deep sleep), and other conditions as determined by a physician or other healthcare provider.



Psychostimulants such as methylphenidate (Ritalin) are commonly prescribed to treat attention-deficit/hyperactivity disorder (AD/HD). *(Photo Researchers, Inc.)*

Methylphenidate contributes to the treatment of ADHD by increasing attention and decreasing restlessness in children and adults who are overactive, cannot concentrate for very long, or are easily distracted, and are impulsive. Methylphenidate is intended to be used as part of a total treatment program that also includes social, educational, and psychological treatment.

A central nervous system stimulant, methylphenidate is also used to control narcolepsy, a condition characterized by an overpowering desire to sleep. Methylphenidate comes in short- and long-acting tablets. The latter should be swallowed whole, never broken into smaller pieces or chewed.

General use

Initially methylphenidate is prescribed in two daily doses of 2.5 mg each, taken at breakfast and lunch time. The dosage is gradually increased until the daily amount reaches 10.0 mg. The dosages should be strictly followed, and since anorexia is an important side effect, the dosages should always be accompanied by a meal or snack. The primary side effect of methylphenidate is growth suppression. Others include irritability, restlessness, agitation, **nausea**, and headaches. Occasionally it causes sleeplessness, in which case the last dosage of the day should be a short-action tablet. Physicians often recommend regular drug-free periods to combat these side effects. In many cases, a child only takes methylphenidate during the school year.

Precautions

Methylphenidate can be addictive and dosage should be tapered off gradually. Signs of physical depen-

KEY TERMS

Attention deficit hyperactivity disorder (ADHD)—A condition in which a person (usually a child) has an unusually high activity level and a short attention span. People with the disorder may act impulsively and may have learning and behavioral problems.

Narcolepsy—A life-long sleep disorder marked by four symptoms: sudden brief sleep attacks, cataplexy (a sudden loss of muscle tone usually lasting up to 30 minutes), temporary paralysis, and hallucinations. The hallucinations are associated with falling asleep or the transition from sleeping to waking.

dency include the need to increase the dosage in order to achieve results, mental depression, unusual behavior, and unusual tiredness or weakness. Some medical professionals believe that methylphenidate is prescribed too often. They call for better diagnostic procedures conducted by trained personnel rather than relying primarily on subjective observations by parents and teachers.

The dosage of methylphenidate is different for different people. It is important to follow the prescribing physician's orders or the instructions that appear on the label of the container. Do not change dosages unless a physician approves such an alteration.

Side effects

Any serious reaction to the drug, such as shortness of breath, irregular heartbeat, or allergic reaction, should be reported to one's doctor. Less severe, more common side effects include blurred vision, insomnia, drowsiness, gastrointestinal distress (nausea or **vomiting**), **dizziness**, headaches, and possible **addiction**.

Interactions

Persons taking methylphenidate should be aware of the possible adverse interactions with the following drugs: amphetamines, appetite suppressants, **caffeine**, chlophedianol, cocaine, **asthma** medication, cold, sinus and hay fever medications, nabilone, pemoline, monoamine oxidase inhibitors, and pimozide. Methylphenidate is also affected by epilepsy, Tourette's syndrome, glaucoma, high blood pressure, psychosis, severe **anxiety**, and **tics**.

Parental concerns

The use of methyphenidate has been subject to controversy over the last several years. Parents concerned about whether the drug is being properly prescribed for their children should seek out the opinion of the most suitable physician for the type of problems their child is having. If a parent is uncomfortable with a physician's response, they should not hesitate to get a second opinion.

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American Medical Association. 515 N. State Street, Chicago, IL 60610. Web site: <www.ama-assn.org/>.

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Midget *see* **Dwarfism**

Mineral deficiency

Definition

Mineral deficiency is a reduced level of any of the **minerals** essential to human health. An abnormally low mineral concentration is usually defined as a level that may impair a function dependent on that mineral.

Description

Minerals are essential nutrients for every living cell in the human body. Defined in the study of human **nutrition** as all the inorganic elements or molecules required for life, minerals assist in body functions such as producing energy, growing, and healing. Minerals are required for fluid balance, blood and bone development, maintaining a healthy nervous system, and regulating muscles, including heart muscles. Minerals, like **vitamins**, function as coenzymes. They participate in all enzyme reactions in the body and help in the assimilation and use of vitamins and other nutrients.

Minerals occur either as bulk minerals (macrominerals) or trace minerals (microminerals). The body needs more bulk minerals than it does trace minerals, although both are essential for health. Minerals are consumed in food from plants and plant-eating animals. These sources of minerals develop in a sequence that takes millions of years, beginning with rock formation, the breakdown of rocks into mineral salts, and the assimilation of these salts into soil that nourishes edible plants.

Recommended daily allowances exist for a number of minerals, such as calcium. However, minimum daily requirements for some minerals such as boron, chromium, and molybdenum, do not exist. The essential bulk minerals include:

- Calcium—essential for strong bones and teeth, healthy gums, and bone growth and mineral density in children. Calcium helps regulate the heart rate and nerve impulses, lower cholesterol, prevent atherosclerosis, develop muscles, and prevent muscle cramping. Calcium is an important component of blood clotting. Calcium and phosphorus are closely related minerals that should be balanced. About 99 percent of calcium and 85 percent of phosphate occur in the skeleton as crystals of calcium phosphate. Both nutrients occur in a variety of foods such as milk, eggs, and green, leafy vegetables. Calcium deficiency due to lack of dietary calcium occurs only rarely and is often due to **vitamin D deficiency**, because vitamin D is required for efficient absorption of dietary calcium. Significant depletion of calcium stores can lead to osteoporosis.
- Magnesium—assists in the utilization of calcium and potassium, and functions in enzyme reactions to produce energy. Magnesium protects the lining of arteries and helps form bones. It helps prevent cardiovascular disease, osteoporosis, and some cancers. By acting with vitamin B_6, magnesium can help prevent or dissolve calcium oxylate kidney stones, the most common kind of stones. Dietary magnesium deficiency is uncommon, but may occur in chronic alcoholics, persons taking diuretic drugs, and as a result of severe, prolonged **diarrhea**.
- Sodium—sodium deficiency (hyponatremia) is a serious deficiency, arising most often after excessive losses of body fluid (**dehydration**) during prolonged and severe diarrhea or **vomiting**. Sodium and potassium are electrolytes that must be balanced in the body. Since most people get more than enough salt in the diet, potassium may be needed to balance it. Together, these minerals control fluid balance through a mechanism called "the sodium/potassium pump." Prolonged imbalances in sodium and potassium can contribute to heart disease.
- Potassium—important for a healthy nervous system and a steady heart rate, helps to prevent **stroke**, and, with sodium, is critical in maintaining fluid balance. Potassium, an electrolyte, must be balanced with sodium. Potassium deficiency is usually associated

with sodium deficiency and both are associated with dehydration stemming from excessive losses of body fluid.

- Phosphorus—helps form bones and teeth, supports cell growth, and regulates heart muscle contraction and kidney function. Phosphorus converts food to energy and supports the utilization of vitamins. Deficiency is rare because phosphate is plentiful in plant and animal foods and is efficiently absorbed from the diet. Phosphorus is closely related to calcium and the two minerals should be in balance with each other and with magnesium. Deficiency in one will affect all and will ultimately have an unwanted effect on body function. Calcium and phosphorus are stored in the bones as crystals of calcium phosphate. Milk, eggs, and green, leafy vegetables are rich in calcium and phosphate.

Trace minerals essential for human health include:

- Boron—required for healthy bones, brain function, alertness, and the metabolism of bulk minerals such as calcium, phosphorus, and magnesium. Deficiencies are rare except in aging, when supplementation may help absorb calcium. A deficiency in boron is associated with vitamin D deficiency. Boron supplements can improve calcium levels as well as vitamin D levels, and can help prevent osteoporosis in postmenopausal women by promoting calcium absorption.
- Chromium—required for maintaining energy levels. Chromium helps metabolize glucose and stabilize glucose levels. It helps the body manufacture and use cholesterol and protein.
- Copper—helps form healthy bones, joints, and nerves as well as hemoglobin and red blood cells. Copper contributes to healing, energy production, taste, and hair and skin color. It is essential in forming collagen for healthy bones and connective tissue, and helps prevent osteoporosis. Except in osteoporosis, copper deficiency is rare, although dramatic changes in copper metabolism occur in two serious genetic diseases, Wilson disease and Menkes' disease.
- Germanium—helps improve the delivery of oxygen to tissues and remove toxins and poisons from the body. Germanium gives garlic its natural antibiotic properties.
- Iodine—helps promote healthy physical and mental development in children. Iodine is required for thyroid gland function and metabolizing fats. Iodine deficiency is a public health problem in parts of the world that have iodine-deficient soils. Iodine is needed to make thyroid hormone, which has a variety of roles in human embryo development. A deficiency during pregnancy can cause serious birth defects. Deficiency in adults can result in an enlarged thyroid gland (goiter) in the neck.
- Iron—critical in the production of hemoglobin, the oxygen-carrying protein in red blood cells, and myoglobin found in muscle tissue. Iron is essential for important enzyme reactions, growth, and maintaining a healthy immune system. In the blood, iron is found in larger amounts than any other mineral. Iron deficiency causes anemia (low hemoglobin and reduced numbers of red blood cells), which results in tiredness and shortness of breath because of poor oxygen delivery.
- Manganese—essential for metabolizing fat and protein, regulating blood glucose, and supporting immune system and nervous system function. Manganese is necessary for normal bone growth and cartilage development. It is involved in reproductive functions and helps produce mother's milk. Along with B vitamins, manganese produces feelings of well-being. Deficiency can lead to convulsions, vision and hearing problems, muscle contractions, tooth-grinding and other problems in children; and atherosclerosis, heart disease, and **hypertension** in older adults.
- Molybdenum—found in bones, kidneys, and liver. Only extremely small amounts are needed to metabolize nitrogen and promote proper cell function. Molybdenum is present in beans, peas, legumes, whole grains, and green leafy vegetables. A diet low in these foods can lead to mouth and gum problems and cancer.
- Selenium—an important antioxidant that works with vitamin E to protect the immune system, heart, and liver, and may help prevent tumor formation. Selenium deficiency occurs in regions of the world where soils are selenium-poor and low-selenium foods are produced. Premature infants are naturally low in selenium with no known serious effects.
- Silicon—helps form bones and connective tissue, nails, skin, and hair. Silicon is important in preventing cardiovascular disease.
- Sulfur—disinfects the blood and helps to rid the body of harmful bacteria and toxic substances.
- Vanadium—vital to cell metabolism, and helps reduce cholesterol and form healthy bones and teeth. Vanadium functions in reproduction. Deficiencies may be associated with heart and kidney disease and reproductive disorders. Vanadium deficiency may be associated with infant mortality.
- Zinc—important in the growth of reproductive organs and regulation of oil glands. Zinc is required for protein synthesis, immune system function, protection of the liver, collagen formation, and wound healing. A component of insulin and major body enzymes, zinc

helps vitamin absorption, particularly vitamins A and E. Deficiency is rare.

Trace and bulk minerals are stored in muscles and bones and delivered to tissue cells through blood circulation. They work together synergistically and must be chemically balanced in the body; if one is deficient or out of balance, it can affect all the others, often resulting in illness. If zinc, for example, is present at high levels, calcium levels will be reduced because the two minerals compete for absorption. Similarly, too much calcium will deplete magnesium, and so on. Deficiency in one nutrient occurs less often than deficiency in several nutrients. A child suffering from **malnutrition** will likely be deficient in a variety of nutrients. Deficiencies in one nutrient do occur, however, such as in populations living in iodine-poor regions, and in iron deficient persons who lose excess iron by abnormal bleeding. All uncorrected mineral deficiencies can affect body functions, produce symptoms, and result in illness.

Demographics

Statistics are not available for most individual mineral deficiencies, most likely because such deficiencies are rare in the United States. Surveys of lower-income families in the United States reveal that about 6 percent of infants are anemic, indicating a possible deficiency of iron in the diet (all **anemias** are not iron-deficiency related).

Diarrheal diseases and related sodium and potassium deficiencies are responsible for about two million infant deaths each year worldwide. Few of these deaths occur in the United States.

Causes and symptoms

Calcium and phosphorus deficiencies

Calcium and phosphorus are plentiful in foods, and dietary deficiencies are rare. Vitamin D deficiency impairs the absorption of dietary calcium and can provoke calcium deficiency (hypocalcemia) even when adequate calcium is consumed. Vitamin D deficiency can be found among young infants and the elderly who may be shielded from sunshine for prolonged periods. As women age, reductions in the hormone estrogen can affect the rate of calcium loss. Significant depletion of calcium stores can lead to osteoporosis. Deficiency of calcium or imbalances with phosphorus and magnesium can produce muscle cramping and digestive problems. Symptoms of calcium deficiency include joint **pain**, brittle nails, eczema, **high cholesterol**, insomnia, high blood pressure, nervousness, and **tooth decay**. Calcium deficiency can also contribute to cognitive problems (confusion, inattention, learning, and memory), convulsions, depression, and hyperactivity. Phosphorus deficiency can produce **anxiety**.

Sodium and potassium deficiencies

Deficiency or imbalance in sodium and potassium does not usually result from a lack of these minerals in the diet, but from imbalances in body fluids. This can be caused by excessive losses of body fluid (dehydration) from severe diarrhea or vomiting; laxative abuse; or during treatment of heart disease or high blood pressure (hypertension) with diuretic drugs, which are used to reduce fluid overload. Sodium and potassium imbalances can cause cardiac arrhythmias and shock (a reduced flow of blood and oxygen to tissues throughout the body). Although diarrheal fluids deplete a number of electrolytes (sodium, potassium, chloride, calcium, phosphorus, and magnesium), the main concern in avoiding shock is replacing sodium and water. Potassium deficiency alone can also affect nerve function.

Magnesium deficiency

Dietary magnesium deficiency is rare because the mineral is found in nearly all foods, but it can occur through poor diet or in malnutrition, or result from excessive losses due to severe diarrhea or vomiting. Symptoms of magnesium deficiency include faulty transmission of nerve and muscle impulses, irritability, nervousness, and **tantrums**. Confusion, poor digestion, rapid or irregular heartbeat (arrhythmia), and seizures can also result. Magnesium deficiency is associated with cardiac arrest, **asthma**, chronic fatigue syndrome, chronic pain, depression, insomnia, **irritable bowel syndrome**, and lung conditions.

Boron deficiency

Boron deficiency is rare, although reduced levels do occur with aging and with reduced levels of vitamin D. Because boron is involved in the absorption of calcium, the only symptom may be reduced levels of calcium or the inability to absorb supplemental calcium.

Chromium deficiency

Many Americans are deficient in dietary chromium, which can be associated with poor regulation of insulin and related imbalances in glucose (either diabetes or **hypoglycemia**). Symptoms include fatigue, anxiety, poor protein metabolism, and glucose intolerance (as in diabetes). In adults, chromium deficiency can be a sign of coronary artery disease.

Copper deficiency

Copper is obtained through a balanced diet and deficiency is rare. Signs of copper deficiency may include anemia, diarrhea, weakness, poor respiratory function, baldness, skin sores, and increased lipid (fat) levels in the blood. Severe alterations in copper metabolism are seen in two rare genetic diseases: Wilson disease and Menkes' disease, which occur in about one in 100,000 births. Both diseases involve mutations in copper transport proteins, special channels that allow copper ions to pass through cell membranes. Menkes' disease, called the "kinky hair disease," results in tangled, grayish, steely, or kinky hair and chubby, rosy cheeks. Untreated Menkes' disease is associated with **mental retardation** and death before three years of age. Wilson disease involves decreases in copper in blood cells, the liver and brain; and increases in copper (copper toxicosis) in the cells of the intestines and kidneys. It results in degenerative changes in the brain, liver disease, and hemolytic anemia. Children older than five years who have any form of liver disease are often evaluated for serum and cellular copper levels to determine if Wilson disease is present.

Germanium deficiency

Germanium deficiency is rare; in fact, there is no established deficiency level.

Iodine deficiency

Iodine deficiency occurs when soil is iodine-poor and foods grown in the soil are correspondingly low in iodine. An iodine intake of 0.10–0.15 mg/day is considered to be nutritionally adequate. Iodine deficiency occurs when intake is below 0.05 mg/day. Goiter, an enlargement of the thyroid gland in the neck, results from iodine deficiency. Although goiter continues to be a problem in other parts of the world, it no longer occurs in the United States because of the fortification of foods with iodine. Iodine deficiency during pregnancy can result in cretinism in newborns, involving mental retardation and a large tongue.

Iron deficiency

Iron deficiency occurs most often because of poor iron intake and poor absorption. In children, iron deficiency is due to periods of dietary deficiency and heavy demands for iron during rapid growth. Human milk and cow's milk both contain low levels of iron; however, the iron in human milk is in a highly absorbable form. Infants are at risk for acquiring iron deficiency because their rapid rate of growth needs a corresponding increased supply of dietary iron, for use in making blood and muscles. Cow's milk formula is fortified with iron. Human milk is a better source of iron than cow's milk, since about half of the iron in human breast milk is absorbed by the infant's digestive tract. In contrast, only 10 percent of the iron in cow's milk is absorbed by the infant. Toddlers who drink excessive whole cow's milk are at risk for iron deficiency. Iron deficiency can also be caused by excess phosphorus in the diet, chronic intestinal bleeding, poor digestion and absorption, prolonged illness, ulcers, and the use of antacids. In women and teenage girls, blood loss through **menstruation** can result in iron deficiency. Symptoms of iron deficiency include anemia and resulting fatigue and weakness, especially during physical exertion. Fragile bones, brittle hair and nails, hair loss, spoon-shaped fingernails or ridges from the base of the nails to the ends, difficulty swallowing, nervousness, paleness, and lagging mental responses are also possible iron deficiency symptoms.

Manganese deficiency

Deficiency of manganese is very rare. Experimental studies of individuals fed a manganese deficient diet have revealed that the deficiency produces a scaly, red rash on the skin of the upper torso.

Selenium deficiency

Selenium deficiency may occur in premature infants who naturally tend to have about one-third the selenium levels of full-term infants. It is not known if these lower levels result in adverse consequences. Selenium deficiency occurs in regions of the world containing low-selenium soils, including parts of China, New Zealand, and Finland. In Keshan Province, China, a condition (Keshan disease) occurs that results in deterioration of regions of the heart and the development of fibers in these areas. Keshan disease, which may be fatal, is thought to result from a combination of selenium deficiency and a virus.

Zinc deficiency

Zinc deficiency can be caused by diarrhea, liver and kidney disease, **alcoholism**, diabetes, malabsorption, and overconsumption of fiber. Symptoms of zinc deficiency include **acne**, recurrent colds and flu, loss of senses of taste and smell, poor night vision, slow growth, lack of sexual maturation, lack of pubic hair, and small stature. Studies have shown that signs of zinc deficiency are detectable after two to five weeks of consuming a zinc-free diet. Signs include a rash on the face, groin, hands and feet, and diarrhea. Administering zinc will correct these symptoms.

When to call the doctor

Mineral deficiencies present with a wide variety of symptoms. Parents should observe children closely and report any unusual symptoms to the pediatrician, such as tiredness, weakness, depression or anxiety, irritability, nervousness, skin irritations, dehydration from vomiting or diarrhea, and slow growth or development of skills. Other than providing regular vitamin supplements and a balanced diet to prevent deficiencies, parents should not attempt to diagnose and treat deficiencies on their own.

Diagnosis

Individual minerals can be measured in blood serum, red blood cells, tissue cells, or urine, to estimate available levels and determine normal or abnormal status. Since each mineral performs strikingly different functions, tests to confirm deficiency are markedly different from each other. Testing can range from simple to extensive. Physicians will consider the possible consequences of each type of deficiency and evaluate the function of organ systems affected by the particular mineral.

In addition to determining serum calcium, phosphorus, and vitamin D levels, the diagnosis of calcium and phosphorus deficiency may involve taking **x rays** of the skeleton.

Diagnosing iron deficiency will require measuring iron levels and investigating anemia by performing blood tests such as a complete blood count (CBC) to determine the number of red blood cells, hemoglobin level, red cell volume, and cell maturity (morphology). A stair-stepping test may be used to evaluate stamina, but a blood test is required to diagnose iron deficiency.

Diagnosing low levels or imbalances of the electrolytes sodium, potassium, calcium, magnesium, or phosphate involves measuring the serum levels of each. Measurement determines the circulating blood level at the time blood was drawn. Laboratory values of sodium and potassium, which are present within cells and in the fluid between cells, can change rapidly depending on the individual's overall condition. They may be measured repeatedly to determine a trend and to monitor correction of the deficiency or imbalance after diagnosis.

Normal serum magnesium levels are 1.2–2.0 mE/l, while levels in deficiency (hypomagnesemia) are below 0.8 mE/l. Because calcium and magnesium must remain balanced, magnesium levels below 0.5 mE/l can provoke a decline in serum calcium levels. Hypomagnesemia can also result in low serum potassium. Symptoms of hypomagnesemia, such as twitching and convulsions, may actually result from the hypocalcemia. Other symptoms, such as cardiac arrhythmias, actually occur because of low potassium. All three minerals will be measured.

Iodine deficiency is diagnosed by measuring the concentration of iodine in urine. A urinary level greater than 0.05 mg iodine per gram of creatinine (another metabolite excreted in urine) indicates adequate iodine status. Levels under 0.025 mg iodine/gram creatinine indicate serious risk. The doctor may also examine the neck with the eyes and hands to see if a goiter is present.

Urinary zinc levels will differ between normal dietary intake (16 mg per day) and low-zinc diets (0.3 mg per day); normal urinary zinc is about 0.45 mg per day while low-zinc urinary levels are about 0.150 mg per day. Plasma zinc levels tend to be maintained during a dietary deficiency in zinc. Plasma and urinary zinc levels can be influenced by a variety of factors, and for this reason cannot provide a clear picture of zinc status.

Selenium can be measured in plasma or red blood cells and compared to normal values. The activity of an enzyme (glutathione peroxidase) in platelets (small blood cells essential in blood clotting) may be evaluated to assess selenium status.

Treatment

Most mineral deficiencies can be successfully treated through diet or supplementation, except when caused by disease, which requires treatment of the disease.

Treating fluid imbalances and related deficiencies in sodium, potassium, calcium, and phosphate usually requires intravenous (IV) infusion of the deficient mineral in fluid over a period of time. Sudden changes in sodium and potassium levels can be just as dangerous as low levels; caution is used to restore balance gradually. Children may be given oral pediatric preparations to gradually restore fluids and minerals.

Iron deficiency requires oral supplementation or injectable iron. Vitamin C helps to assimilate iron.

Iodine deficiency is easily treated and prevented by consuming foods fortified with iodine, such as table salt. Goiter is reversible with treatment but cretinism is not.

A magnesium-rich diet will correct magnesium deficiency. If deficiency is due to prolonged depletion, treatment may include injections of magnesium sulfate; if severe enough to provoke convulsions, intravenous infusions may be given.

Selenium deficiency can be treated by supplementation. Children can be given supplements containing 1.0 mg sodium selenite.

Zinc and copper deficiencies are rare and can be treated with supplementation.

Prognosis

The prognosis for mineral deficiencies depends on the extent of deficiency at diagnosis, the degree of effects or symptoms, and the overall health of the individual. Correction through diet or supplements usually produces good results. Symptoms may sometimes be relieved promptly with supplementation. Some deficiencies produce permanent effects of varying severity.

In iodine deficiency, the prognosis for treating goiter is excellent. Sodium and potassium deficiencies or imbalances can be corrected if diagnosed and treated promptly, but can be life-threatening if untreated. Anemia and other effects of iron deficiency are not usually life-threatening and can be corrected with supplementation. "Silent" or undiagnosed calcium loss may result in osteoporosis, which may produce disability or complications by the time it is diagnosed.

Alternative treatment

Sea vegetables (sea weeds such as dulse, kelp, wakambe, and hijiki) are an excellent source of minerals obtained from the ocean. They can be used to make soup stock, added to stews and casseroles, or served with vegetables. Herbs are a valuable source of minerals as well. For example, calcium is found in alfalfa, burdock root, chamomile, dandelion, flaxseed, paprika, raspberry leaves, rose hips, and other herbs. Iron is found in the same herbs as calcium, and in the Chinese herb *dong quai* (angelica), as well as other herbs.

KEY TERMS

Electrolytes—Salts and minerals that produce electrically charged particles (ions) in body fluids. Common human electrolytes are sodium chloride, potassium, calcium, and sodium bicarbonate. Electrolytes control the fluid balance of the body and are important in muscle contraction, energy generation, and almost all major biochemical reactions in the body.

Nutrient—Substances in food that supply the body with the elements needed for metabolism. Examples of nutrients are vitamins, minerals, carbohydrates, fats, and proteins.

Osteoporosis—Literally meaning "porous bones," this condition occurs when bones lose an excessive amount of their protein and mineral content, particularly calcium. Over time, bone mass and strength are reduced leading to increased risk of fractures.

Recommended Dietary Allowance (RDA)—The Recommended Dietary Allowances (RDAs) are quantities of nutrients in the diet that are required to maintain good health in people. RDAs are established by the Food and Nutrition Board of the National Academy of Sciences, and may be revised every few years. A separate RDA value exists for each nutrient. The RDA values refer to the amount of nutrient expected to maintain good health in people. The actual amounts of each nutrient required to maintain good health in specific individuals differ from person to person.

Prevention

Ensuring an adequate intake of essential nutrients through a balanced diet and supplements is the best way to prevent mineral deficiencies. The Required Dietary Allowances (RDA) guidelines can help ensure that minerals are being obtained. Safe amounts of certain minerals are often included in multivitamins. Because excess mineral levels can also cause health problems, taking excessive amounts of any mineral supplement is not advised unless a deficiency is diagnosed. When mineral deficiency is the result of disease, medical attention, other than preventive measures, is required.

Nutritional concerns

A balanced diet includes fresh vegetables and fruits, legumes, whole grains (cereal, bread, rice, pasta, and other grains), eggs, dairy products, fish, fowl, and lean meat as preferred. A diet high in refined foods, prepared foods, sugars, and fats will not provide sufficient quantities of essential minerals. Water delivers nutrients throughout the body; it is essential to drink enough clean water daily to maintain fluid balance and distribute nutrients.

Essential mineral nutrients are found in a variety of sources, as in these examples:

- Boron is abundant in apples, pears, grapes, leafy greens, carrots, whole grains, and nuts.
- Primary calcium sources are dairy foods, eggs, fish, and green leafy vegetables. Other calcium-rich foods are figs, broccoli, cabbage, oats, almonds and filberts, yogurt, and blackstrap molasses. Spinach and Swiss chard bind calcium in the digestive tract and are not a ready source.

- Chromium is found in brown rice and other whole grains, cheeses, meats, dried beans, corn, eggs, mushrooms, and potatoes.
- Copper is found in nuts, mushrooms, broccoli, garlic, lentils, salmon, and green vegetables. It is also found in plumbing and cooking pots, which may leach into food and water.
- Germanium occurs primarily in garlic, shiitake mushrooms, and onions.
- Iron can be provided by eating green leafy vegetables, raisins, meat, eggs, liver, fish and fowl, nuts, and whole grains. It can also be obtained by cooking in iron skillets.
- Magnesium is found in most foods, primarily animal and fish sources.
- Potassium food sources include dairy foods, fish, fresh and dried fruits, beans and peas, meats, fish and fowl, and whole grains.
- Zinc sources include brewer's yeast, eggs, fish, meats, beans, mushrooms, nuts and seeds, and whole grains.

Parental concerns

Good nutrition is a concern of all parents. In the United States, it is relatively easy to provide a balanced diet with essential nutrients if a wide variety of whole foods are prepared for **family** meals and snacks, while avoiding refined and prepared foods high in fats and sugars. RDA guidelines and public health resources can help assure parents that they can prevent dietary deficiencies in their young children and teens.

Resources

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Mineral toxicity

Definition

The term mineral toxicity refers to a condition in which the concentration in the body of any one of the **minerals** necessary for life is abnormally high, and which has an adverse effect on health.

Description

The mineral nutrients are defined as all the inorganic elements or inorganic molecules that are required for life. As far as human **nutrition** is concerned, the inorganic nutrients include water, sodium, potassium, chloride, calcium, phosphate, sulfate, magnesium, iron, fluorine, copper, zinc, chromium, manganese, iodine, selenium, and molybdenum. The last nine elements in this list are sometimes called trace minerals or micronutrients because humans need only small amounts of them in the diet. In high doses all nine trace minerals can be toxic in humans.

In general, mineral toxicity results when a person accidentally consumes too much of any mineral, as with drinking ocean water (sodium toxicity), or is overexposed to industrial pollutants, household chemicals, or certain drugs. Iron toxicity in children, for example, frequently results from accidental swallowing of dietary supplement tablets.

Mineral toxicity may also refer to toxic conditions resulting from certain diseases or injuries. For example, a disorder known as hemochromatosis leads to iron toxicity, while Wilson's disease results in copper toxicity. Severe trauma can lead to hyperkalemia or potassium toxicity.

Demographics

Iron **poisoning** is the most common form of mineral toxicity in children in the United States and is one of the leading causes of fatal poisoning in children younger than six years of age. About 20,000 children are reported as accidentally swallowing iron tablets each year in the United States, although not all of these cases end in death. In one Indian study of 21 children treated for iron poisoning, four of the patients died.

With regard to diseases leading to mineral toxicity, about one person in ten in the United States has the genetic mutation that can lead to hemochromatosis; however, not everyone with this mutation necessarily develops the disease. It is thought that there are about 1 million persons in the United States with hemachromatosis

as of the early 2000s. About one person in 30,000 has the genetic defect that causes Wilson's disease, while about 1.1 percent of the general population are carriers of the mutant gene. The incidence of Menkes disease, which primarily affects boys, is variously estimated at one in 50,000 to one in 250,000 persons. Wilson's disease and Menkes disease occur at the same rate in all races and ethnic groups.

Causes and symptoms

The causes and symptoms of mineral toxicity depend on the specific mineral in question:

- Sodium: An increase in sodium concentration in the bloodstream can be toxic. The normal concentration of sodium in human blood plasma is 136–145 mM, while levels over 152 mM can result in seizures and death. Increased plasma sodium, which is called hypernatremia, causes the cells in various body tissues, including those of the brain, to shrink. Shrinkage of the brain cells results in confusion, coma, paralysis of the lung muscles, and death. Death has occurred when table salt (sodium chloride) was accidentally used to feed infants instead of sugar. Death due to sodium toxicity has also resulted when baking soda (sodium bicarbonate) was used to treat excessive **diarrhea** or **vomiting**. Although a variety of processed foods contain high levels of sodium chloride, the levels in these items are not enough to result in sodium toxicity.
- Potassium: The normal level of potassium in the bloodstream is in the range of 3.5–5.0 mM, while levels of 6.3–8.0 mM (severe hyperkalemia) result in cardiac arrhythmias or even death due to cardiac arrest. Potassium is potentially quite toxic; however, potassium poisoning is usually prevented because of the vomiting reflex. The consumption of food results in mild increases in the concentration of potassium in the bloodstream, but these levels of potassium do not become toxic because of the uptake of potassium by various cells of the body as well as by the action of the kidneys transferring the potassium ions from the blood to the urine. The body's regulatory mechanisms can easily be overwhelmed, however, when potassium chloride is injected intravenously, as high doses of injected potassium can easily result in death.
- Iodine: Iodine toxicity can result from an intake of 2.0 mg of iodide per day. Toxic levels of iodine inhibit the secretion of thyroid hormone, resulting in lower levels of thyroid hormone in the bloodstream. As a result, the thyroid gland becomes enlarged. This condition is known as goiter or **hyperthyroidism**. Goiter is usually caused by iodine deficiency. In addition to goiter, iodine toxicity produces a brassy taste in the mouth, excessive production of saliva, and ulcers on the skin. This skin condition has been called kelp **acne** because of its association with eating kelp, an ocean plant that contains high levels of iodine. Iodine toxicity occurs fairly frequently in Japan, where people consume large amounts of seaweed.
- Iron: Iron toxicity is not unusual in small children due to the wide distribution of dietary supplements containing iron. A lethal dose of iron is in the range of 200—250 mg iron/kg body weight, meaning that a child who accidentally eats 20 or more iron tablets may die as a result of iron poisoning. Children are unfortunately likely to take large amounts of these pills because they look like candy. Within six hours of ingestion, iron toxicity can result in vomiting, diarrhea, abdominal **pain**, seizures, and possibly coma. In the second period of iron poisoning, the patient's symptoms appear to improve; however, this phase is followed by a terminal phase in which shock, low blood sugar levels, liver damage, convulsions, and death occur 12 to 48 hours after the fatal dose.
- Nitrite: Nitrite poisoning should be considered along with iron toxicity, since nitrite produces its toxic effect by reacting with the iron atom in hemoglobin. Hemoglobin is an iron-containing protein that resides within the red blood cells. This protein is responsible for transporting nearly all of the oxygen acquired from the lungs to various tissues and organs of the body. Hemoglobin accounts for the red color of red blood cells. A very small fraction of hemoglobin spontaneously oxidizes per day, producing a protein of a slightly different structure called methemoglobin. Normally, the amount of methemoglobin constitutes less than 1 percent of the total hemoglobin. Methemoglobin can accumulate in the blood as a result of nitrite poisoning. Infants are especially susceptible to poisoning by nitrite.
- Nitrate: Nitrate is naturally present in green leafy vegetables and in the water supply. It is rapidly converted to nitrite by the bacteria that live in the mouth as well as in the intestines and then absorbed into the bloodstream. The amount of nitrate that is supplied by leafy vegetables and drinking water is generally about 100 to 170 mg/day. The amount of nitrite supplied by a typical diet is much lower, about 0.1 mg nitrite per day. Poisoning by nitrite (or nitrate after its conversion to nitrite) results in the inability of hemoglobin to carry oxygen throughout the body. This condition can be seen by the blue color of the skin. Adverse symptoms occur when over 30 percent of the hemoglobin has been converted to methemoglobin. These symptoms include cardiac arrhythmias, **headache**, **nausea and vomiting**, and in severe cases, seizures.

- Calcium and phosphate: Calcium and phosphate are closely related nutrients. Calcium toxicity is rare, but overconsumption of calcium supplements may lead to deposits of calcium phosphate in the soft tissues of the body. Phosphate toxicity can result from the overuse of **laxatives** or enemas that contain phosphate. Severe phosphate toxicity can result in hypocalcemia and in various symptoms resulting from low plasma calcium levels. Moderate phosphate toxicity occurring over a period of months may result in the deposit of calcium phosphate crystals in various tissues of the body.
- Zinc: Zinc toxicity is rare but is more likely to occur in adults than in children. It is usually related to occupational hazards and has been reported to occur in metal workers exposed to fumes containing zinc. A few instances of zinc toxicity have been reported in people who consumed acidic food or beverages that had been stored in galvanized zinc containers. Taking excessive supplemental zinc can result in **nausea**, vomiting, and diarrhea. The chronic intake of excessive zinc supplements can result in copper deficiency, as zinc inhibits the absorption of copper.
- Copper: Copper toxicity in humans is usually the result of disease. Severe alterations in copper metabolism occur in two genetic diseases, Wilson's disease and Menkes disease. These diseases are rare. They involve mutations in the proteins that transport copper, that is, in special channels that allow the passage of copper ions through cell membranes. Wilson's disease, which is caused by a mutation of the ATP7B gene on chromosome 13, first produces symptoms in teenagers and young adults. Copper accumulates in the liver, kidney, and brain, resulting in damage to the liver and nervous system. In Menkes disease, which is usually first noticed in infancy, impaired transport of copper from the digestive tract results in low levels of copper in the blood, while copper accumulates in the kidney, pancreas, and skeletal muscle. Children with Menkes disease have characteristic kinky hair, seizures, developmental failures, and progressive degeneration of the brain.
- Selenium: Selenium toxicity occurs in a few regions of the world, most notably some parts of China where soils contain high levels of the mineral. A daily intake of 0.75 to 5.0 mg selenium may occur in these regions due to the presence of selenium in foods and water. Early signs of selenium toxicity include nausea, weakness, and diarrhea. Continued intake of selenium results in changes in the fingernails, hair loss, and damage to the nervous system. The person's breath may acquire a characteristic garlic odor as a result of the increased production of dimethylselenide in the body and its release via the lungs.
- Manganese: Manganese toxicity is most likely to affect adults rather than children. It occurs most commonly in workers in manganese mines who must breathe air containing high levels of manganese dust (in a concentration of 5–250 mg/cubic meter). Manganese toxicity in miners has been documented in Chile, India, Japan, Mexico, and elsewhere. Symptoms of manganese poisoning typically occur within several months or years of exposure. These symptoms include a mental disorder resembling **schizophrenia** as well as hyperirritability, violent acts, hallucinations, and difficulty in walking.

When to call the doctor

The most common form of mineral toxicity for children and adolescents in the United States is accidental poisoning from iron supplements. Parents should take a child who is known to have swallowed iron tablets to the doctor or a hospital emergency room for treatment as soon as possible, as an iron overdose is potentially fatal.

Children born into families with a history of Wilson's disease should have a blood test for the disease at some point in their second year of life, before symptoms of the disease develop.

Diagnosis

An initial diagnosis of mineral toxicity requires taking a careful history. The doctor asks the parents of a small child questions intended to identify any unusual aspects of the family's diet or intake of drugs and chemicals. An older teenager in the workforce may be asked about possible occupational exposure. The mineral content of the body may be measured by testing samples of body fluids, most commonly blood plasma, red blood cells from whole blood, and urine. Diagnosis of mineral toxicities also involves measuring the concentration of various metals in the plasma or urine. Concentrations that are above the normal range can confirm the initial suspected diagnosis.

Menkes disease may be diagnosed by the unusual appearance of the hair, skin, and facial features in male infants with the disorder as well as by their developmental problems.

In addition to a deficiency in blood plasma of a protein known as ceruloplasmin, Wilson's disease is characterized by gold or greenish-gold discolorations of the cornea of the eye known as Kayser-Fleischer rings. These rings may be detected by an ophthalmologist during a slit-lamp examination. The doctor may also suspect

Wilson's disease in a child above the age of five with unexplained episodes of hepatitis or such symptoms of copper toxicity in the brain as drooling, loss of coordination, tremor, sudden drop in academic performance, or frank psychotic episodes. The clinical symptoms of Wilson's disease do not appear in young children; however, measurements of serum ceruloplasmin can be taken in children over 12 months of age if a **family** history of Wilson's disease is a risk factor.

Treatment

Iron toxicity is treated by efforts to remove the remaining iron from the stomach by administering a solution of 5 percent sodium bicarbonate. Where plasma iron levels have risen above 0.35 mg/dL, the patient is treated with deferoxamine. Treatment of manganese toxicity involves removal of the patient from the high manganese environment as well as giving him or her lifelong doses of the drug L-dopa. The treatment is only partially successful. Treatment of nitrite or nitrate toxicity involves inhalation of 100 percent oxygen for several hours. If oxygen treatment is not effective, then a solution of 1.0 percent methylene blue may be injected in a dose of 1.0 mg methylene blue/kg body weight.

With regard to disorders of copper metabolism, Wilson's disease can be successfully controlled by lifelong treatment with d-penicillamine, trientine, and zinc acetate. Treatment also involves avoiding foods that are high in copper, such as liver, nuts, chocolate, and mollusks. After an initial period of treatment with penicillamine, Wilson's disease may be treated with zinc (150 mg oral Zn/day). The zinc inhibits the absorption of dietary copper. Patients with this disease must, however, comply with treatment for the rest of their lives, as untreated Wilson's disease is invariably fatal. Patients who develop liver failure as a result of the disease may be candidates for a liver transplant.

Children with Menkes disease are sometimes helped temporarily by intravenous injections of copper supplements. There is, however, no cure for the disease as of the early 2000s, and most children with the disorder live only a few years.

Nutritional concerns

Families consuming a well-balanced diet without overuse of dietary supplements are unlikely to have problems with mineral toxicity. Children or adolescents diagnosed with Wilson's disease must observe the dietary limitations described earlier.

Prognosis

The prognosis for mineral toxicity due to sodium, potassium, calcium, and phosphate is usually excellent. Toxicity due to the deposit of calcium phosphate crystals is not usually reversible. The prognosis for treating iodine toxicity is excellent. For any mineral overdose that causes coma or seizures, the prognosis for recovery is often poor, and death results in a small fraction of patients. For any mineral toxicity that causes nerve damage, the prognosis is often fair to poor. Wilson's disease is fatal, usually before age 30, unless the patient complies with continual lifelong treatment to prevent brain or liver disease. Children diagnosed with Menkes disease rarely live past their third birthday.

Prevention

When mineral toxicity results from the excessive consumption of mineral supplements, toxicity can be prevented by minimizing the use of dietary supplements and keeping iron tablets in particular out of the reach of children. Zinc toxicity may be prevented by not storing food or beverages in zinc containers. In the case of iodine, toxicity can be prevented by avoiding overconsumption of seaweed or kelp. In the case of selenium toxicity resulting from high-selenium soils, toxicity can be prevented by relying on food and water acquired from a low-selenium region.

Such genetic diseases as Wilson's disease and Menkes disease cannot be prevented as of the early 2000s.

Parental concerns

Parental concerns about mineral toxicity in most children should be directed toward preventing accidental consumption of iron and other mineral supplements in young children and in monitoring the adoption of fad diets in teenagers.

In the case of children with hemachromatosis or Wilson's disease, parents will need to make sure that the affected child complies with all aspects of necessary treatment. In the case of a child with Menkes disease, parents should seek genetic counseling, as the grim prognosis of this illness places a heavy emotional as well as economic burden on a family.

See also Heavy metal poisoning; Poisoning.

KEY TERMS

Arrhythmia—Any deviation from a normal heart beat.

Goiter—Chronic enlargement of the thyroid gland.

Hemochromatosis—An inherited blood disorder that causes the body to retain excessive amounts of iron. This iron overload can lead to serious health consequences, including painful joints, diabetes, and liver damage, if the iron concentration is not lowered.

Hyperkalemia—An abnormally high level of potassium in the blood.

Hypernatremia—An abnormally high level of sodium in the blood.

Hypocalcemia—A condition characterized by an abnormally low level of calcium in the blood.

Menkes disease—A genetic disease caused by a mutation on the X chromosome and resulting in impaired transport of copper from the digestive tract. It was first identified in 1962.

Methemoglobin—A compound formed from hemoglobin by oxidation of its iron component. Methemoglobin cannot carry oxygen.

Micronutrient—An organic compound such as vitamins or minerals essential in small amounts and necessary to the growth and health of humans and animals.

Trace element—An element that is required in only minute quantities for the maintenance of good health. Trace elements are also called micronutrients.

Wilson's disease—A rare inherited disease in which excessive amounts of copper accumulate in the liver or brain. It is fatal unless the patient complies with lifelong treatment with penicillamine and zinc oxidase. Wilson's disease is also known as inherited copper toxicosis.

Resources

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American Academy of Emergency Medicine (AAEM). 555 East Wells Street, Suite 1100, Milwaukee, WI 53202. Web site: <www.aaem.org>.

American Academy of Family Physicians (AAFP). 11400 Tomahawk Creek Parkway, Leawood, KS 66211–2672. Web site: <www.aafp.org>.

National Institute of Child Health and Human Development (NICHD). 31 Center Drive, Room 2A32, Bethesda, MD 20892–2425. <www.nichd.nih.gov>.

National Organization for Rare Disorders Inc. (NORD). 55 Kenosia Avenue, Danbury, CT 06813–1968. Web site: <www.rarediseases.org>.

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Tom Brody, PhD

Minerals

Definition

Minerals are inorganic nutrients. That is, they are materials found in foods that are essential for growth and health and do not contain the element carbon. The minerals that are relevant to human **nutrition** are water, sodium, potassium, chloride, calcium, phosphate, sulfate, magnesium, iron, copper, zinc, manganese, iodine, selenium, and molybdenum. Cobalt is a required mineral

for human health, but it is supplied by vitamin B_{12}. There is some evidence that chromium, boron, and other inorganic elements play some part in human nutrition, but their role has not been proven.

Description

Minerals should be provided by a normal, healthy diet. In special cases, additional mineral supplements may be called for. Preterm (low birth weight) infants have special needs for calcium, phosphorus, and sodium, as well as extra needs for vitamin D. Iron supplements may also be recommended.

The amount of each mineral that is needed to support growth during infancy and childhood, to maintain body weight and health, and to facilitate pregnancy and **lactation**, are listed in a table called the Recommended Dietary Allowances (RDA). This table was compiled by the Food and Nutrition Board, a committee that serves the United States government. The values listed in the RDA indicate the daily amounts that are expected to maintain health throughout most of the general population. The actual levels of each inorganic nutrient required by any given individual is likely to be less than that stated by the RDA. The RDAs are all based on studies that provided the exact, minimal requirement of each mineral needed to maintain health. However, the RDA values are actually greater than the minimal requirement, as determined by studies on small groups of healthy human subjects, in order to accommodate the variability expected among the general population.

Because of differences in individual diets and individual needs, the decision regarding any child's need for supplements should be made by the parents after discussion with the pediatrician and, where appropriate, a nutritionist. Children on a well-balanced diet do not require supplements, while those who are picky eaters or who routinely eat a poor diet may benefit from supplementation.

Girls should get their calcium from foods, particularly dairy products, rather than supplements. Dairy products were associated with higher bone mineral density in the spine, while calcium supplements had no such benefit.

General use

The following discussion describes the role of the major minerals in human nutrition.

Iron is essential for the formation of hemoglobin, the chemical in the blood that carries oxygen to the cells. Low levels of iron cause anemia. In severe cases, the children become flabby, and they fail to grow normally. Milder cases of iron deficiency may not produce any physical symptoms, but children may learn at a slower pace than children with a proper amount of iron in their diet. The combination of rice, beans, and meat consumed with fresh citrus fruit provides an excellent source of absorbable iron. Iron supplements are suggested for children who cannot or will not follow a proper diet through the first two years of life.

Calcium is required for proper development of bones and teeth. It is also needed for proper muscle activity and blood clotting. Lack of calcium can cause rickets, a condition in which the bones are soft and develop in abnormal shapes. Calcium must be accompanied by vitamin D in order to have the proper effects. Foods rich in calcium include almonds, swiss cheese, collards, sardines and salmon with bones, spinach, ice cream, kale, beet greens, cheddar cheese, molasses, oysters, milk, and broccoli.

Zinc deficiency has been associated with reduced growth and **mental retardation**. The best foods for zinc are lamb, beef, leafy grains, root vegetables such as potatoes and carrots, shellfish, and organ meats such as liver or kidneys. While a high fiber diet is important for health, too much fiber can reduce the absorption of zinc and lead to a zinc deficiency.

Iodine is needed in the diet for proper thyroid function. The best source of iodine is fish, but table salt normally has iodine added to it, and even modest amounts of salt will meet the daily iodine requirements.

Fluoride is needed for strong teeth. In many areas, drinking water contains fluoride that meets all normal needs, but for children who do not drink water or drink filtered or bottled water, fluoride supplements may be useful. Fluoride supplements may be useful for infants and then may be discontinued as the child gets older and starts drinking water.

Magnesium is found in so many parts of the body that it is almost impossible to describe the effects of low magnesium levels. The most common problems are twitching, and, because of the need for magnesium in the parathyroid gland, soft bones even when calcium and vitamin D are adequate. Because magnesium is found in most foods, deficiency is usually associated with absorption problems and requires medical attention.

Copper is required for blood and nerve fiber development. It is found in liver, nuts, and seafood.

Phosporus is needed for energy production, metabolism, and healthy bone development. The best sources

are milk, cheese, meats, whole grains, eggs, peas, and beans.

Potassium is needed for muscle contractions and nerve function. Good sources of potassium are orange juice, milk, cheese, whole grains, and vegetables.

Selenium is needed for proper thyroid function. It has also been associated with prevention of some types of **cancer** in adults. Selenium supplements are not normally required except in children with **phenylketonuria** receiving a low-protein diet, although it may sometimes be associated with thyroid problems. In these cases, medical care is required.

Precautions

Although the greatest nutritional concern is with inadequate levels of minerals, it is possible to take too much, particularly when people already eating a normally healthy diet take supplements. The daily intake of minerals should be reviewed to prevent adverse effects.

Excess calcium may lead to **constipation** and kidney problems. Too much zinc may lead to **diarrhea**, **vomiting**, and kidney and heart problems. Excess iron may cause problems of the stomach and digestive tract, liver problems, an increased risk of diabetes, and male sexual problems.

Side effects

When minerals are taken properly, they have no side effects.

Interactions

Minerals can interact with drugs and in excess with each other. Iron and calcium are known to bind to drugs of the tetracycline family and inactivate the antibiotic. The compound of calcium and tetracycline may also be absorbed into a child's teeth, causing discoloration.

Too much calcium in the diet may inhibit absorption of iron, magnesium, phosphorus, and zinc. Excess iron may reduce the absorption of zinc.

Parental concerns

Following a proper balanced diet is the best prevention of both **mineral deficiency** and mineral overdose. Since many children and adolescents cannot or will not eat a balanced diet, the possible need for supplements should be discussed with an appropriate professional.

Many children fail to follow a proper diet. This may be because of excess intake of fast foods and snack foods of low nutritional value. It is important for parents to teach children the benefits of proper nutrition and the importance of maintaining a healthful diet.

At the same time, adolescents, particularly those who engage in **sports**, may feel that they will do better with increased levels of nutrients. Because of the risk of toxic reactions to minerals and some **vitamins**, children should be discouraged from taking vitamin supplements unless there is clear evidence of increased need.

KEY TERMS

Inorganic—Pertaining to chemical compounds that are not hydrocarbons or their derivatives.

Parathyroid gland—A pair of glands adjacent to the thyroid gland that primarily regulate blood calcium levels.

Phenylketonuria (PKU)—A rare, inherited, metabolic disorder in which the enzyme necessary to break down and use phenylalanine, an amino acid necessary for normal growth and development, is lacking. As a result, phenylalanine builds up in the body causing mental retardation and other neurological problems.

Rickets—A condition caused by the dietary deficiency of vitamin D, calcium, and usually phosphorus, seen primarily in infancy and childhood, and characterized by abnormal bone formation.

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American Dietetic Association. 120 South Riverside Plaza, Suite 2000, Chicago, IL 60606–6995. Web site: <www.eatright.org>.

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Minnesota Multiphasic Personality Inventory

Definition

The Minnesota Multiphasic Personality Inventory (MMPI-2; MMPI-A) is a written psychological **assessment**, or test, used to diagnose mental disorders.

Purpose

The MMPI is used to screen for personality and psychosocial disorders in adults (i.e., over age 18) and adolescents age 14 to 18. It is also frequently administered as part of a neuropsychological test battery to evaluate cognitive functioning.

Description

The original MMPI was developed at the University of Minnesota and introduced in 1942. The current standardized version for adults 18 and over, the MMPI-2, was released in 1989, with a subsequent revision of certain test elements in early 2001. The MMPI-A, a version of the inventory developed specifically for adolescents age 14 to 18, was published in 1992.

The adolescent inventory is shorter than the standard adult version, was developed at a sixth-grade reading level, and is geared towards adolescent issues and personality "norms." The MMPI-A has 478 true/false items, or questions, (compared to 567 items on the MMPI-2) and takes 45 minutes to an hour to complete (compared to 60 to 90 minutes for the MMPI-2). There is also a short form of the test that is comprised of the first 350 items from the long-form MMPI-A.

The questions asked on the MMPI-A are designed to evaluate the thoughts, emotions, attitudes, and behavioral traits that comprise personality. The results of the test reflect an adolescent's personality strengths and weaknesses, and may identify certain disturbances of personality (psychopathologies) or mental deficits caused by neurological problems.

There are eight validity scales and ten basic clinical or personality scales scored in the MMPI-A, and a number of supplementary scales and subscales that may be used with the test. The validity scales are used to determine whether the test results are actually valid (i.e., if the test taker was truthful, answered cooperatively and not randomly) and to assess the test taker's response style (i.e., cooperative, defensive). Each clinical scale uses a set or subset of MMPI-A questions to evaluate a specific personality trait. Some were designed to assess potential problems that are associated with **adolescence**, such as eating disorders, social problems, **family** conflicts, and alcohol or chemical dependency.

Precautions

The MMPI should be administered, scored, and interpreted by a qualified clinical professional trained in its use, preferably a psychologist or psychiatrist. The MMPI is only one element of psychological assessment, and should never be used as the sole basis for a diagnosis. A detailed history of the test subject and a review of psychological, medical, educational, or other relevant records are required to lay the groundwork for interpreting the results of any psychological measurement.

Cultural and language differences in the test subject may affect test performance and may result in inaccurate MMPI results. The test administrator should be informed before psychological testing begins if the test taker is not fluent in English and/or has a unique cultural background.

Preparation

The administrator should provide the test subject with information on the nature of the test and its intended use, and complete standardized instructions for taking the MMPI (including any time limits, and information on the confidentiality of the results).

The MMPI should be scored and interpreted by a trained professional. When interpreting test results for test subjects, the test administrator will review what the test evaluates, its precision in evaluation and any margins of error involved in scoring, and what the individual scores mean in the context of overall norms for the test and the background of the test subject.

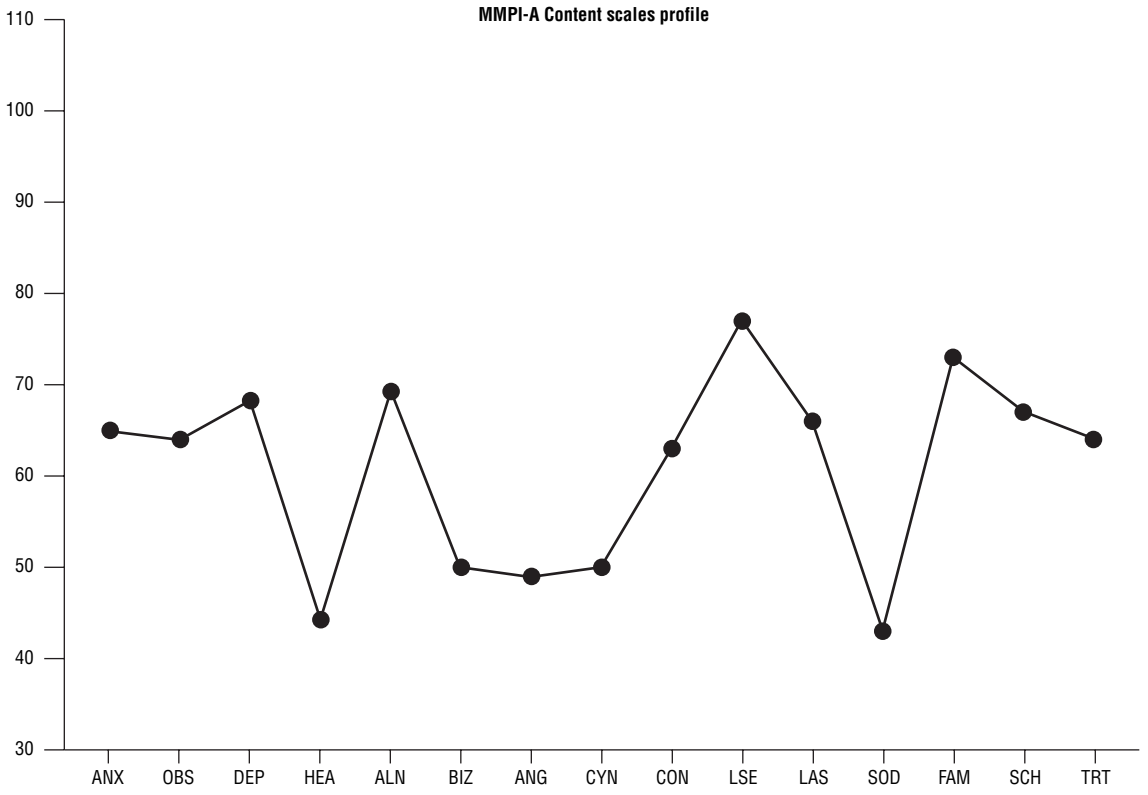
Risks

There are no risks involved in taking the MMPI. However, parents should try to make sure the test is properly administered, and the results evaluated appropriately, to avoid an unnecessary negative label on their child.

Parental concerns

Test anxiety can have an impact on a child's performance, so parents should attempt to take the stress off their child by making sure they understand that the MMPI is not an achievement test and the child's honest answers are all that is required. Parents can also ensure

Minnesota Multiphasic Personality Inventory–Adolescent



	ANX	OBS	DEP	HEA	ALN	BIZ	ANG	CYN	CON	LSE	LAS	SOD	FAM	SCH	TRT
Raw score:	15	12	18	5	12	4	9	14	13	14	10	4	23	11	15
T-score:	65	64	68	44	69	50	49	50	63	77	66	43	73	67	64
Response %:	100	100	100	100	100	100	100	100	100	100	100	100	100	100	100

ANX- Anxiety
OBS-Obsessiveness
DEP-Depression
HEA-Health concerns
ALN-Alienation
BIZ-Bizarre mentation
ANG-Anger
CYN-Cynicism
CON-Conduct problems
LSE-Low self-esteem
LAS-Low aspiration
SOD-Social discomfort
FAM-Family problems
SCH-School problems
TRT-Negative treatment indicators

SOURCE: MMPI-A sample extended score report, Pearson Assessments, 2004.

Sample profile from the Content scales profile of the MMPI-Adolescent. Although the data is interpreted by trained professionals, this adolescent's score shows a low self-esteem and probable family problems. *(Illustration by GGS Information Services.)*

that their children are well-rested on the testing day and have a nutritious meal beforehand.

When interpreting test results for parents, the test administrator will review what the test evaluates, its precision in evaluation and any margins of error involved in scoring, and what the individual scores mean in the context of overall norms for the test and the background of the adolescent.

See also Psychological tests.

Resources

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ORGANIZATIONS

American Psychological Association. Testing and Assessment Office of the Science Directorate. 750 First St., N.E., Washington, DC 20002–4242. (202)336–6000. Web site: <www.apa.org/science/testing.html>.

KEY TERMS

Neuropsychological testing—Tests used to evaluate patients who have experienced a traumatic brain injury, brain damage, or organic neurological problems (e.g., dementia). It may also be used to evaluate the progress of a patient who has undergone treatment or rehabilitation for a neurological injury or illness.

Norms—A fixed or ideal standard; a normative or mean score for a particular age group.

Psychopathology—The study of mental disorders or illnesses, such as schizophrenia, personality disorder, or major depressive disorder.

Standardization—The process of determining established norms and procedures for a test to act as a standard reference point for future test results.

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Paula Ford-Martin

Minority health

Definition

Minority health addresses the special medical and health needs associated with specific ethnic and other minority groups.

Description

The United States, along with many other countries, experiences cultural diversity. This fact poses health issues that are specific to ethnic and other minority groups. Additionally, the propensity for certain diseases or illnesses is of concern in certain minority groups. These specific health issues include **infant mortality** rates, **cancer**, cardiovascular disease, diabetes, **HIV infection**, and immunizations. The primary minority groups in the United States are Hispanics, African Americans, Native Americans and Native Alaskans, Native Hawaiians and other Pacific Islanders, and gays and lesbians.

One of the major health problems in the United States is overweight and **obesity**, which lead to increased risks for a wide variety of conditions, including cardiovascular disease, diabetes, **hypertension**, and cancer. A 2003 study by the Agency for Healthcare Research and Quality shows that African-American and Hispanic children face much higher odds of being overweight than non-Hispanic white or Asian-American and Pacific Islander children. African-American children ages six to 11 are more than twice as likely as non-Hispanic white children to be overweight, and Hispanic children are roughly twice as likely. The odds change dramatically when children become teenagers. For example, as children, Asian Americans and Pacific Islanders have the lowest prevalence of being overweight, but once they reach **adolescence**, the reverse is true. As teens, they have the highest prevalence of being overweight—more than four times that of non-Hispanic white teenagers. African American children have the highest rate of being overweight, but once they reach their teen years, they are no more likely than white children to be overweight. Hispanic teens are one-and-a-half times more likely than white or African American teens to be overweight.

Researchers and policymakers have attributed the poorer health of minority Americans in part to their reduced access to medical care and the lower quality of primary care they receive. Indeed, when asked about the primary care they receive, minority patients—particularly Asian Americans—give the primary care they receive lower marks than white patients do, according to a 2001 report by the Agency for Healthcare Research and Quality. After adjustment for socioeconomic and other factors, Asian Americans gave their primary care significantly lower scores (out of 100 total) than whites for communication (69 versus 79) and comprehensive knowledge of patients (48 versus 56), as well as all other areas of primary care except continuity of care and integration of care. African American and Hispanics reported significantly less financial access to care than whites (60 and 56, respectively, versus 65), and African Americans reported significantly less continuity of care than whites (74 versus 78), but their assessments of other aspects of primary care did not differ significantly from whites. This study agrees with others which show that Asian Americans tend to be the least satisfied with quality of care. However, this study was limited by the small number of Asian and Hispanic patients surveyed, as well as the lack of patient's country of origin and physician's ethnicity, factors that may affect patient evaluations of primary care.

Infant mortality rates

Infant mortality rates (IMRs) in the United States and in all countries worldwide are an accurate indicator of health status. They provide information concerning programs about pregnancy education and counseling, technological advances, and procedures and aftercare. IMRs vary among racial groups. Infant mortality among African Americans in 2000 occurred at a rate of 14.1 deaths per 1,000 live births. This is more than twice the national average of 6.9 deaths per 1,000 live births. The leading causes of infant death include congenital abnormalities, pre-term/low birth weight, **sudden infant death syndrome** (SIDS), problems related to complications of pregnancy, and **respiratory distress syndrome**. SIDS deaths among Native American and Alaska Natives is 2.3 times the rate for non-Hispanic white mothers.

Cancer

Cancer is a serious national, worldwide, and minority health concern. It is the second cause of death in the United States, claiming over 500,000 lives each year. Approximately 50 percent of persons who develop cancer die of the disease. There is great disparity among the cancer rates in minority groups. Across genders, cancer death rates for African Americans are 35 percent higher when compared to statistics for Caucasians. The death rates for prostate cancer (two times more) and lung cancer (27 times more) are disproportionately higher when compared to Caucasians. There are also gender differences among ethnic groups and specific cancers. Lung cancers in African American and Hawaiian men are evaluated compared with Caucasian males. Vietnamese females who live in the United States have five times more new cases of cervical cancer when compared to Caucasian women. Hispanic females also have a greater incidence of cervical cancer than Caucasian females. Additionally, Alaskan native men and women have a greater propensity for cancers in the rectum and colon than do Caucasians.

Cardiovascular disease

Cardiovascular disease is the leading cause of disability and death, about equal to the rate of death from all other diseases combined. Cardiovascular disease can affect the patient's lifestyle and function in addition to having an impact on **family** members. The financial costs are very high. Among ethnic and racial groups cardiovascular disease is the leading cause of death. **Stroke** is the leading cause of cardiovascular-related death, which occurs in higher numbers for Asian-American males when compared to Caucasian men. Mexican-American men and women and African-American males have a higher incidence of hypertension. African American women have higher rates of being overweight, which is a major risk factor of cardiovascular disease. African Americans are 13 percent less likely to undergo coronary angioplasty and one-third less likely to undergo bypass surgery than are Caucasians.

Diabetes

Diabetes, a serious health problem among Americans and ethnic groups, is the seventh leading cause of death in the United States. The prevalence of diabetes in African Americans is about 70 percent higher than Caucasians. The burden of diabetes is much greater for minority populations than the white population. For example, 10.8 percent of non-Hispanic blacks, 10.6 percent of Hispanics, and 9 percent of Native Americans and Native Alaskans have diabetes, compared with 6.2 percent of whites. Certain minorities also have much higher rates of diabetes-related complications and death, in some instances by as much as 50 percent more than the total population. Diabetes-related mortality rates for African Americans, Hispanic Americans, and Native Americans and Native Alaskans are higher than those for white people. Asians and Pacific Islanders have the lowest diabetes-related mortality of any racial/ethnic group in the United States.

HIV and AIDS

HIV infection/AIDS is the most common cause of death for all persons age 25 to 44 years old. Ethnic groups account for 25 percent of the U.S. population and 54 percent of all **AIDS** cases. In addition to sexual transmission there is an increase in HIV among ethnic groups related to intravenous drug usage. African Americans with HIV infection are less likely to be on antiretroviral therapy, less likely to receive prophylaxis for Pneumocystis **pneumonia**, and less likely to be receiving protease inhibitors than other persons with HIV. An HIV infection data coordinating center, under development in 2004, will allow researchers to compare contemporary data on HIV care to determine whether disparities in care among groups are being addressed and to identify any new patterns in treatment that arise. Among children, the disparities are dramatic, with African-American and Hispanic children representing more than 80 percent of pediatric AIDS cases in 2000. Approximately 78 percent of HIV-infected women are minorities and most become infected through heterosexual transmission.

In 2002, African Americans accounted for 50 percent of all new AIDS cases, while Hispanics accounted for 20 percent, according to the Centers for Disease

Control and Prevention (CDC). Although the virus is still most likely to be passed on by gay and bisexual males, as of 2004 more than 25 percent of AIDS cases are women, most of whom are African American or Hispanic. According to the National Center for Health Statistics, black females age 15 and older are 15.5 times more likely to die of AIDS than whites—a figure even more dramatic than the one presented in the vice presidential debates, according to an article in the October 16, 2004 *Los Angeles Times*.

Immunizations

Data show that in 2000 children living below the poverty level have lower immunization coverage rates. Although significant progress has been made in improving childhood immunization rates, some disparities in overall immunization coverage rates among racial and ethnic groups continue. This disparity is of great concern in large urban areas with underserved populations because of the potential for outbreaks of vaccine-preventable diseases.

Demographics

The overall health of the U.S. population improved during the last decades of the twentieth century, but all Americans have not shared equally in these improvements. Among nonelderly adults, for example, 17 percent of Hispanic and 16 percent of black Americans report they are in minimally fair or poor health, compared with 10 percent of white Americans.

Causes and symptoms

Most IMRs are correlated with prenatal care. Women who receive adequate prenatal care tend to have better pregnancy outcomes when compared to those who receive little or no care. Women who receive inadequate prenatal care tend to have increased chances of delivering a very low birth weight (VLBW) infant, which is linked to risk of early death.

Cancer is related to several preventable lifestyle choices. Diet and tobacco and sun exposure can be shaped by lifestyle modifications. Additionally many cancers can occur due to lack of interest in and/or lack of availability for screening and educational programs.

Cardiovascular diseases are higher among persons with high blood cholesterol and high blood pressure. Certain lifestyle choices that may increase the chance for heart disease include lack of **exercise**, overweight, and cigarette use. Cardiovascular disease is responsible for over 50 percent of the deaths in persons with diabetes.

HIV occurs at a higher frequency among gay males (the number of African-American males who have AIDS through sex with men has as of 2004 increased). Additionally unprotected sexual intercourse and sharing used needles for IV drug injection are strongly correlated with infection.

Vaccinations are an effective method of preventing certain disease such as **polio**, **tetanus**, pertussis, **diphtheria**, **influenza**, **hepatitis b**, and pneumococcal infections. Approximately 90 percent of influenza-related mortality is associated with persons aged 65 and older. This is mostly due to neglect of vaccinations. About 45,000 adults each year die of diseases related to hepatitis B, pneumococcal and influenza infections.

When to call the doctor

Parents of minority children should contact their family physician or other healthcare provider when they have any concern about their child's health.

Diagnosis

The diagnosis of VLBW occurs when newborns are weighed. Infants who weigh 52.5 ounces (1,500 grams) are at high risk for death. For cancer, the diagnosis can be made through screening procedures such as mammography (for breast cancer), PAP smear (for cervical cancer). Lifestyle modifications such as avoidance of sun, cessation of cigarette **smoking**, maintaining a balanced diet, and adequate **nutrition**, all positively affect one's health. Other specific screening tests (PSA, prostate surface antigen) are helpful for diagnosing prostate cancer. Cardiovascular diseases can be detected by medical check-up. Blood pressure and cholesterol levels can be measured. Obesity can be diagnosed by assessing a person's weight compared to the person's height. Diabetes and its complications can be detected by blood tests, in-depth eye examinations, and studies that assess the flow of blood through blood vessels in the legs. HIV can be detected through a careful history and physical examination and analysis of blood using a special test called a western blot. Infections caused by lack of immunizations can either be detected by conducting physical examination and culturing the specific microorganism in the laboratory.

Treatment

Treatment should be directed toward the primary causes(s) that minorities have increased chances of developing disease(s). Cancer may require treatment using surgery, radiotherapy, or **chemotherapy**. Cardiovascular diseases may require surgical procedures for

establishing a diagnosis and initiating treatment. Depending on the extent of disease, cardiovascular management can become complicated requiring medications and daily lifestyle modifications. Treatment usually includes medications, dietary modifications, and—if complications arise—specific interventions tailored to alleviating the problem. HIV can be treated with specific medications and more often than not with symptomatic treatment as complications arise. Diseases caused by lack of immunizations are treated based on the primary disease. The best method of treatment is through prevention and generating public awareness through widespread education on the topic.

Alternative treatment

Alternative therapies do exist, but as of 2004 more research is needed to substantiate available data. Most physicians say the diseases that relate to minority health are best treated with nationally accepted standards of care.

Prognosis

Generally the prognosis is related to the diagnosis, patients' state of health, age, and the presence of another disease or complication in addition to the presenting problem. The course for IMRs is related to educational programs and prenatal care, which includes medical and psychological treatments. The prognosis for chronic diseases such as cardiovascular problems, high blood pressure, cancer, and diabetes is variable. As of 2004, these diseases are not cured, and control is achieved by standardized treatment options. Eventually complications, despite treatment, can occur. For HIV the clinical course as of 2004 is death, even though this process may take years. Educational programs with an emphasis on disease prevention can potentially improve outcomes concerning pediatric and geriatric diseases.

Prevention

Prevention is accomplished best through educational programs specific to target populations. IMRs can be prevented by increasing awareness, interest, and accessibility for prenatal care that offers a comprehensive approach for the needs of each patient. Regular physicals and special screening tests can potentially prevent certain cancers in high-risk groups. Educational programs concerning lifestyle modifications, diet, exercise, and testing may prevent the development of cardiovascular disease and diabetes. Educational programs for illicit IV drug abusers and persons who engage in unprotected sexual intercourse may decrease the incidence of HIV infection.

Parental concerns

All children should have regular well-child check ups according to the schedule recommended by their physician or pediatrician. The American Academy of Pediatrics (AAP) advises that children be seen for well-baby check ups at two weeks, two months, four months, six months, nine months, 12 months, 15 months, and 18 months of age. Well-child visits are recommended at ages two, three, four, five, six, eight, ten, and annually thereafter through age 21. Parents can take some precautions to ensure the health of their children. **Childproofing** the home, following a recommended immunization schedule, educating kids on **safety**, learning **cardiopulmonary resuscitation** (CPR), and taking kids for regular well-child check-ups all help to protect against physical harm. In addition, encouraging open communication with children can help them grow both emotionally and socially. Providing a loving and supportive home environment can help to nurture an emotionally healthy child who is independent, self-confident, socially skilled, insightful, and empathetic towards others.

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KEY TERMS

Angioplasty—A medical procedure in which a catheter, or thin tube, is threaded through blood vessels. The catheter is used to place a balloon or stent (a small metal rod) at a narrowed or blocked area and expand it mechanically.

Cardiopulmonary resuscitation (CPR)—An emergency procedure designed to stimulate breathing and blood flow through a combination of chest compressions and rescue breathing. It is used to restore circulation and prevent brain death to a person who has collapsed, is unconscious, is not breathing, and has no pulse.

Cardiovascular—Relating to the heart and blood vessels.

Congenital—Present at birth.

Hypertension—Abnormally high arterial blood pressure, which if left untreated can lead to heart disease and stroke.

Healthcare." *Managed Healthcare Executive* 13 (November 2003): 8.

ORGANIZATIONS

National Center on Minority Health and Health Disparities. National Institutes of Health. 6707 Democracy Blvd., Suite 800, MSC-5465, Bethesda, MD 20892–5465. Web site: <http://ncmhd.nih.gov>.

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Mitochondrial disorders

Definition

Mitochondrial disorders, also called mitochondrial cytopathies, are a diverse group of diseases caused by damage to small structures found in human cells that are essential in converting food to energy. The result is decreased energy production and associated symptoms.

Description

Cells are the building blocks of the human body, microscopic structures that are bound by a membrane and contain numerous components called organelles that are responsible for functions such as cell reproduction, transportation of materials, and protein synthesis. Cellular respiration, a process by which food molecules are converted into high-energy molecules used as a source of energy, takes place in structures called mitochondria. The energy produced by mitochondria is essential for cell functions.

Before the mid-twentieth century, little was known about mitochondrial disorders. The first diagnosis of a mitochondrial disorder occurred in 1959, and the genetic material of microchondria, called mtDNA, was discovered in 1963. In the 1970s and 1980s, as more was learned about the mitochondria and more mitochondrial disorders were discovered, the term "mitochondrial myopathies" (myopathy meaning a disease of muscle tissue) was coined to describe the group of diseases. Further research in the 1990s led to classification of mitochondrial disorders. As it became evident that tissues other than muscle could be affected by mitochondrial defects, the term "mitochondrial cytopathies" (cytopathy meaning cell disorder) was adopted.

Disorders in which skeletal muscle is the primary target of the mitochondrial dysfunction are called mitochondrial **myopathies**. Mitochondrial encephalomyopathies are disorders in which muscle and brain tissue is involved.

Common mitochondrial disorders

As of 2004 there were more than 40 distinct mitochondrial cytopathies. Some of the more common disorders include:

- Kearns-Sayre syndrome (KSS). Onset of KSS usually occurs before the age of 20. Symptoms include progressively constrained eye movements, droopy eye lids, muscle weakness, short stature, hearing loss, loss of coordination, heart problems, cognitive delays, and diabetes.

- Myoclonus epilepsy with ragged-red fibers (MERRF). MERFF is a mitochondrial encephalomyopathy in which a mitochondrial defect as well as a tissue abnormality called "ragged-red fibers" (an accumulation of diseased mitochondria) is found microscopically. The resulting symptoms include seizures, loss of coordination, short stature, build-up of lactic acid in the blood, difficulty speaking, dementia, and muscle weakness.
- Mitochondrial encephalomyopathy with lactic acidosis and stroke-like episodes (MELAS). MELAS is a progressive mitochondrial disease that involves multiple organ systems including the central nervous system, cardiac muscle, skeletal muscle, and gastrointestinal system. Symptoms include muscle weakness, stroke-like events, eye muscle paralysis, and cognitive impairment.
- Leber hereditary optic neuropathy (LHON). LHON causes progressive loss of vision resulting in various degrees of blindness and primarily affects men over the age of 20. Heart abnormalities may also occur.
- Leigh syndrome. This degenerative brain disorder is usually diagnosed at a young age (e.g. before age two). Deterioration is often rapid with symptoms such as seizures, dementia, feeding and speech difficulties, respiratory dysfunction, heart problems, and muscle weakness. Prognosis is poor with death typically occurring within a few years of diagnosis.
- Myoneurogenic gastrointestinal encephalopathy (MNGIE). Key features include symptoms that mimic gastrointestinal obstruction and nervous system abnormalities. Other symptoms may include eye muscle paralysis, muscle weakness, loss of coordination, and brain abnormalities.
- Pearson syndrome. With symptoms usually first appearing in childhood, the characteristics of this rare syndrome include pancreatic dysfunction and anemia (low red blood cells). Difficulty gaining weight, **diarrhea**, and enlarged liver are other signs of Pearson syndrome.
- Neuropathy, ataxia, and retinitis pigmentosa (NARP). The symptoms implied by this disorder's name include nervous system abnormalities, loss of coordination, and progressive loss of vision. Developmental delays, dementia, and muscle weakness may also result. Onset usually occurs in childhood.

Demographics

Approximately 1,000 to 4,000 children are born with mitochondrial disease in the United States each year. Typically, by the age of ten, approximately one in 4,000 American children is diagnosed with mitochondrial disease.

Causes and symptoms

Although mitochondrial disorders may be caused by distinctly different damage to the mitochondrial genetic material, and thus affect any of the hundreds of chemical reactions required to convert food and oxygen into energy, they all share a common feature: the ability of mitochondria to generate energy is damaged. Byproducts of the numerous reactions can begin to accumulate in the cells and interfere with other chemical reactions and over time damage the mitochondria further.

Inheritance of mitochondrial disorders

In many cases, a mitochondrial disorder is passed genetically from parent to child (inheritance). It can often be helpful for the type of inheritance to be determined, as parents can then make an educated decision about the risks of passing the condition on to another child or the risks of another **family** member developing the disease. Genetic defects may be passed through nuclear DNA (nDNA), the genetic material found in each cell that determines the majority of hereditary characteristics, or through mtDNA. Some types of mitochondrial disorder inheritance include:

- Autosomal recessive inheritance. Each individual has two sets of genes, one inherited from each parent. In some genetic diseases, a person needs to have two copies of a defective gene in order to show symptoms of the disease; if only one of the two genes is defective, the person is considered a carrier. In autosomal recessive inheritance, the affected individual has inherited a defective gene from each parent.
- Maternal inheritance. mtDNA is only passed from mother to child because the mitochondria of a sperm is located in the sperm's tail, which is not involved in conception. Some mitochondrial disorders are, therefore, only passed from mother to child.
- X-linked recessive inheritance. The sex of a child is determined through the inheritance of strands of DNA called chromosomes. A female child inherits two X chromosomes, while a male child inherits an X chromosome from one parent and a Y chromosome from the other. If a defective gene encoding for a disease is found on the X chromosome, then a male child cannot have a healthy copy of the gene (since he only has one X chromosome); therefore, he will develop the disorder. Female children are at less risk because they have to have two copies of the defective gene (one on each X chromosome) in order to develop the disease.

- Autosomal dominant inheritance. As opposed to autosomal recessive inheritance, only one defective copy of a gene needs to be inherited in order for an individual to develop the disease. Each successive child, therefore, has a 50 percent chance of developing the disorder.

In some cases, no other family members are affected by the disease and there appears to be no genetic link. These cases are called random or sporadic occurrences and may be caused by a number of environmental factors including certain drugs (e.g. medications used to treat human **immunodeficiency** virus [HIV] have been linked to mitochondrial damage), **anorexia nervosa** (a disease characterized by self-starvation), exposure to certain toxins, prolonged periods of insufficient oxygen, or older parental age (mtDNA mutations may accumulate over time).

Symptoms

Because more than 90 percent of the energy needed by the human body to function is generated by mitochondria, the effects of mitochondrial disorders can be far-reaching. Research has shown that cells of the brain, nerves, skeletal muscles, liver, heart, kidneys, ears, eyes, and pancreas seem to be particularly affected because of their high energy requirements. Some of the more common symptoms of mitochondrial diseases by organ system include the following:

- brain: confusion, memory loss, headaches, seizures, developmental delays, and stroke-like episodes
- nerves: **pain** caused by nerve abnormalities (neuropathic pain), gastrointestinal problems linked to nerve abnormalities, abnormal sweating, and fainting
- skeletal muscles: muscle weakness, muscle cramping, muscle pain, loss of coordination, **exercise** intolerance, and poor growth
- liver: liver failure not due to excessive alcohol use and low blood sugar (hypoglycemia)
- heart: heart muscle weakness and disturbed electrical signals in the heart (called heart block)
- kidneys: abnormalities that cause difficulty with absorbing nutrients and electrolytes back into the body (called Fanconi syndrome)
- ears: hearing loss
- eyes: eye muscle paralysis, progressive loss of vision
- pancreas: diabetes (a group of conditions characterized by excessive urine excretion and persistent thirst) and pancreatic failure

Other symptoms include **failure to thrive** in infants, poor growth, short stature, fatigue, respiratory disorders, swallowing difficulties, and increased risk of infection.

When to call the doctor

The array of symptoms that are displayed by children suffering from mitochondrial disorders are common to many other diseases, and the age of onset can range from early infancy to adulthood. Often, the hallmark sign of a mitochondrial disorder that distinguishes it from other diseases with similar symptoms is additional features (such as the above symptoms) that do not normally appear with the non-mitochondrial disease. Parents should notify their healthcare provider if their child develops symptoms atypical for their previously diagnosed condition or if those symptoms get worse or recur with infection.

Diagnosis

Because of the complex nature of mitochondrial disorders, physicians take a multi-faceted approach to diagnosing such diseases. The process usually starts with a comprehensive physical exam and evaluation of the patient's medical and family history. Often a neurological exam is performed to determine if there are any brain abnormalities. To diagnose a mitochondrial disorder and rule out other diseases, more extensive tests may need to be performed. Some examples are as follows:

- Initial evaluation. The first line of testing usually involves the least invasive methods, such as sending a sample of blood for evaluation. In some cases a diagnosis can be made based on blood tests; in others, blood tests may indicate that further testing is necessary.
- Secondary evaluation. These tests may be more intensive, more invasive, and/or carry more risks. Examples include lumbar puncture (spinal tap), urine collection, **magnetic resonance imaging** (MRI), additional blood tests, or electrocardiogram (ECG).
- Tertiary evaluation. Complex and/or invasive procedures such as skin or muscle biopsy (taking a small sample of tissue for microscopic evaluation) are considered tertiary tests. In some cases such tests are necessary to make a definitive diagnosis.

In some cases, a physician may not be able to diagnose the patient with a specific mitochondrial disorder even after extensive evaluation. Parents should, therefore, be advised that despite the complexity of testing for mitochondrial disorders, diagnosis is not always possible.

Treatment

As of 2004, there are no cures for mitochondrial disorders. Treatment plans focus on delaying progression of the disease or reducing a patient's symptoms. The method of treatment depends on many factors, including the patient's disease, age, affected organs, and health status. Not all patients benefit from treatment; those with less severe disease generally respond better. Treatment may consist of **vitamins**, supplements, physical or occupational therapy, or traditional medications. Examples of these include:

- vitamins, such as B vitamins (thiamine, riboflavin, niacin, folate, biotin, and pantothenic acid), vitamin E, and vitamin C
- coenzyme Q10 (CoQ10), which is involved in cellular respiration in normal mitochondria
- levocarnitine (Carnitor), taken orally or intravenously, to replace a cofactor necessary in cellular respiration
- antioxidant therapy, a treatment under investigation that may help to keep mitochondrial damage under control
- physical or occupational therapy for myopathies

Alternative treatment

For some patients, avoiding physiological stressors such as extreme cold, extreme heat, poor **nutrition**, fasting, and lack of **sleep** may improve their condition. Alcohol, cigarette smoke, and monosodium glutamate (MSG, added to many Asian foods) may also exacerbate a mitochondrial disorder.

Nutritional concerns

In some cases, a properly devised diet is necessary to avoid worsening symptoms. Parents of a child affected with a mitochondrial disorder may be referred to a dietician to help formulate a diet specific to his or her disease. The plan is individualized to the child and may include suggestions such as avoiding long periods of time without eating, eating small but frequent meals, increasing or decreasing the amount of fat consumed, and avoiding or supplementing with certain vitamins or **minerals**.

Prognosis

The prognosis of mitochondrial disease depends on many factors, including the specific disorder, the mode of inheritance, the age of onset, and what organs are affected. Two children suffering from the same mitochondrial disorder may have two distinctly different courses. In some cases, patients may be able to control their symptoms to a great degree with various treatments, or progression of the disease is slow. In other cases, the disease progresses rapidly and inevitably leads to death.

Prevention

Prevention of inherited mitochondrial disorders is not possible unless parents decide against having more children. In the case of mitochondrial cytopathies that are caused by environmental factors such as certain drugs or toxins, avoidance of these substances may minimize the risk of developing mitochondrial disease.

Parental concerns

Because of the potential of passing on inherited mitochondrial disorders to other children, parents may be interested in genetic counseling. Genetic counselors are health professionals who are trained to help families determine the risk or probability of developing or passing on a genetic disorder. Genetic testing, however, cannot determine with certainty if or when a child will develop a mitochondrial disease or what the severity will be.

Resources

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ORGANIZATIONS

United Mitochondrial Disease Foundation. 8085 Saltsburg Rd., Suite 201, Pittsburgh, PA 15239. Web site: <www.umdf.org>.

WEB SITES

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KEY TERMS

Cellular respiration—The process by which food molecules are converted into high-energy molecules used as a source of energy.

Electrocardiagram (ECG, EKG)—A record of the electrical activity of the heart, with each wave being labeled as P, Q, R, S, and T waves. It is often used in the diagnosis of cases of abnormal cardiac rhythm and myocardial damage.

Mitochondrial inheritance—Inheritance associated with the mitochondrial genome which is inherited exclusively from the mother.

Ragged-red fibers—A microscopic accumulation of diseased mitochondria.

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Stephanie Dionne Sherk

MMPI *see* **Minnesota Multiphasic Personality Inventory**

MMR vaccine

Definition

MMR vaccine is a combined vaccine to protect children against **measles**, **mumps**, and **rubella**, which are dangerous and potentially deadly diseases. Alternative names are rubella **vaccination**, mumps vaccination, vaccine-MMR.

Description

The MMR, which does not contain mercury, consists of live viruses that have been weakened (attenuated) so that the vaccine is still capable of inducing a productive immune response but does not cause the disease that the original or "wild-type" viruses can. The MMR vaccine is a mix of three vaccines: attenuvax (measles), mumpsvax (mumps), and meruvax II (rubella).

The three-in-one MMR vaccine protects against measles, mumps, and rubella. Although single antigen (individual) vaccines are available for each part of the MMR, they are only used in specific situations, in which one of the three diseases occurs and public health officials decide to immunize infants six to 15 months of age for that particular disease. (Single antigen vaccines pose less risk to children younger than the recommended age of 15 months for the MMR.)

Measles (rubeola)

Before vaccination, epidemics of measles peaked in the spring every two to four years. Measles is an endemic disease in many undeveloped countries and in countries where measles immunization levels are low. Because the risk of contracting measles in other countries is greater than in the United States, infants and children should be as well protected as possible before traveling.

Measles is caused by a virus that grows in the nose, mouth, throat, and the eyes, and in their secretions. It is highly communicable and may not be recognized early because the symptoms often resemble cold symptoms. The incubation period is 10 to 11 days. Measles begins with slight temperature rise and a runny nose and eyes. About the second or third day, blush-white pinpoint spots with a red rim, known as Koplick's spots, appear in the mouth. Small dark red pimples appear on the head and spread gradually over the body. These pimples grow larger and in groups, giving a blotchy appearance, which is an important difference between measles and **scarlet fever**. In scarlet fever, the skin appears red all over.

The respiratory symptoms grow worse. The child sneezes often, the eyes are sore, and nasal secretion becomes purulent. Light hurts the eyes (photophobia). The child's throat is sore. The rash is greatest about the fourth day, and it may last up to ten days. During the second week, the skin begins to flake off, and it continues to do so for five to ten days.

Treatment is limited to combating the symptoms of measles because **antiviral drugs** as of 2004 are ineffective. The disease has serious possible complications. For example, encephalomyelitis (inflammation of the brain and spinal cord) occurs in one to two cases out of 1000 patients; the disease is fatal at that same rate. Immune globulin injections help prevent or reduce measles infection if given within six days of exposure. Complications can be brought on by measles. **Encephalitis** occurs in one out of 6000 cases; 20 percent of these infections are fatal. Thrombocytopenic purpura (skin hemorrhages because of decreased **platelet count**) occur in one out of 3000 cases.

Mumps (epidemic parotitis)

Mumps, another viral disease, affects the salivary glands, especially the parotid gland. Children under the age of two years old seldom have mumps; adults rarely have this disease. A closer contact is necessary to transmit mumps than other contagious diseases. The incubation period lasts from two to three weeks, averaging about 18 days.

In most cases, the first sign of mumps is a swelling in the parotid glands; occasionally, mumps may begin with a slight fever, **headache**, and malaise before the swelling appears. Sometimes only one of the parotid glands is affected, but both may be inflamed at the same time or one after the other. The glands become swollen and tender and are painful. It hurts for the child to suck while nursing or in older children to open his mouth and eat, but otherwise he may not feel sick at all. After two or three days, the swelling begins to go down, and usually disappears by the tenth day. As a rule, keeping children isolated or out of school for two weeks is long enough to prevent communicating the infection to others. Treatment is entirely palliative; as of 2004, there was no effective antiviral treatment.

Mumps can cause certain complications. The nervous system is affected in 65 percent of patients; 10 percent display symptoms of this, and 2 percent of these cases are fatal. Testicular complications occur in 14 to 35 percent of post-pubertal boys, and complications regarding the ovaries in 7 percent of post-pubertal females. These complications are rare in prepubescent children, however. Deafness in one or both ears occurs in one out of 15,000 cases. More than half of the deaths from mumps occur in those over 19 years of age. Mumps infection during the first trimester of pregnancy increases the risk of spontaneous abortion.

Rubella (German measles)

Rubella is also caused by a virus, but the disease is mild and last only a short time. The symptoms are like measles but are not nearly as severe, and spots never appear on the mucous membranes of the mouth. Sometimes the rash that appears on the face is the first noticeable sign of a rubella infection. The rash spreads quickly and disappears just as rapidly; sometimes it is gone from the face and the neck by the time it reaches the arms and the legs. The rash usually lasts two to four days.

Isolation from other children is brief or not carried out at all; since the infectious stage is so brief, there is little danger of passing on the infection after the rash appears. The greatest risk of German measles is fetal malformations which occur when a mother is infected in the early months of pregnancy.

Children and adults may can have rubella more than once; 3 to 10 percent of those who have had rubella and 14 to 18 percent of those immunized become infected on exposure to the virus. Some reinfections are subclinical (i.e., have no visible symptoms). In fact, some 25 to 50 percent of rubella infections are asymptomatic.

General use

Recommended MMR vaccination schedule

Because the risk of serious disease from infection with either mumps or rubella in infants is low, mumps and rubella vaccines should not be given to infants younger than 12 months old. When the measles vaccine is needed a single-antigen measles vaccine is given. However, parents of an infant less than 12 months of age should be immune to mumps and rubella so they will not expose the infant or become infected if the infant becomes ill.

The first dose of the vaccine is given to children 12 to 15 months old. The second dose of the MMR vaccine should be given at four to six years of age. All children are to be fully immunized before starting school in the United States. Children who have not the second dose as recommended should complete the immunization by 11 or 12 years of age.

MMR traveling recommendations

Before infants and children of 12 months of age or older leave the United States, they should receive two doses of MMR vaccine separated by at least 28 days, with the first dose given on their birthday. Infants under 12 months of age should receive a dose of monovalent (single antigen) measles vaccine before departure. If monovalent vaccine is not available, no specific contraindication exists to giving MMR to infants six to eleven months of age. The risk for serious disease from either mumps or rubella infection among infants is low.

Infants who receive the monovalent measles vaccine or MMR before their first birthday are vulnerable to all three diseases and should be revaccinated with two doses of MMR. The first should be given when the infant is 12 to 15 months of age (12 months if the infant remains in an area where disease risk is high) and the second at least 28 days later.

Parents or adults who travel or live abroad with infants less than 12 months old should have evidence of immunity to rubella and mumps, as well as measles, to

avoid becoming infected if the infants are exposed to the diseases.

An infant less than six months of age is usually protected against measles, mumps, and rubella by maternal antibodies. As a rule, the infant does not need added protection unless the mother is diagnosed with measles.

Maternal immunity to MMR

Most fetuses receive some natural immunity to measles from their mothers in utero. This passive immunity fades over time and is less effective in children of immunized mothers than in children of mothers who had the measles.

The duration of protection is dependent to a great extent on the maternal antibody titer and the antibodies received by the infant during pregnancy. Women who have had the disease have higher measles antibody titers than women who have not had measles but have been vaccinated. Women who have not had measles nor vaccination have no measles antibodies.

Precautions

There are few reasons not to be vaccinated. Some of these are as follows:

- being allergic to gelatin or neomycin or having had an allergic reaction to a previous MMR vaccination
- being moderately or severely ill
- being pregnant
- in males, mumps can cause inflammation of the testes; in female, the ovaries, external genitals, or breasts may be affected

Side effects

Most of the time inactivated vaccines are given intramuscularly (IM), and live virus vaccines are given subcutaneously (SC). Vaccines that are used intramuscularly may cause local reactions (such as irritation, skin discoloration, inflammation, and granuloma formation) if injected into subcutaneous tissue. The vaccine may also be less effective if it is not given by the proper route.

Interactions

There is varying incidence of vaccine reactions. Some of these are as follows:

- fever (one out of six)
- mild rash (one out of 20)
- swollen glands (rare)
- seizure (one out of 3,000)
- **pain** and joint stiffness (one out of 20)
- low platelet count (one out of 30,000)
- serious allergic reaction (less than one out of 1,000,000)

Parental concerns

Parents often express concern about combining three vaccines in one injection. As of 2004 there is no published evidence showing a benefit to separating the

KEY TERMS

Acellular—Without whole cells. An acellular vaccine contains on parts of the cells which can produce immunity in a person receiving the vaccine.

Active immunity—Produced by the body when the immune system is triggered to produce antibodies, either by immunization or a disease.

Adverse effect—A negative side effect of a vaccine.

Anaphylaxis—Also called anaphylactic shock; a severe allergic reaction characterized by airway constriction, tissue swelling, and lowered blood pressure.

Encephalitis—Inflammation of the brain, usually caused by a virus. The inflammation may interfere with normal brain function and may cause seizures, sleepiness, confusion, personality changes, weakness in one or more parts of the body, and even coma.

Incubation period—The time period between exposure to an infectious agent, such as a virus or bacteria, and the appearance of symptoms of illness. Also called the latent period.

Inflammation—Pain, redness, swelling, and heat that develop in response to tissue irritation or injury. It usually is caused by the immune system's response to the body's contact with a foreign substance, such as an allergen or pathogen.

Passive immunity—The body reception of proteins that act as antibodies instead of making the antibodies itself. Immunoglobulins may produce this immunity. All babies have antibodies from their mothers, which give them short-term protection.

combination MMR vaccine into three individual shots. The CDC continues to recommend two doses of the combined MMR vaccine for all children.

Because signs of **autism** may appear around the time children receive the MMR vaccine, some parents worry that the vaccine causes autism. Research has not found a relationship between MMR vaccine and autism.

It is sometimes difficult for parents to adhere to the recommended vaccine schedule, including the spacing between doses. If the intervals between doses is longer than usual, there is no need to restart the series of any vaccine.

MMR vaccinations are appropriate for children with chronic diseases such as diabetes and cardiovascular condition as advised by the pediatrician.

Symptoms of low-grade fever, irritability, and soreness at the injection site following the MMR immunization can be relieved with an analgesic such as acetaminophen as recommended by the pediatrician. Cool compresses to the injection site are also comforting.

Resources

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Aliene Linwood, R.N., DPA, FACHE

Moles

Definition

A mole (nevus) is a pigmented (colored) spot on the outer layer of the skin (epidermis).

Description

Moles can be round, oval, flat, or raised. They can occur singly or in clusters on any part of the body. Most moles are brown, but colors can range from pinkish flesh tones to yellow, dark blue, or black.

A mole usually lasts about 50 years before beginning to fade. Some moles disappear completely, and some never lighten at all. Some moles develop stalks that raise them above the skin's surface; these moles eventually drop off.

Types of moles

Moles that are present at birth are called congenital nevi.

Other types of moles include:

- junctional moles, which are usually brown and may be flat or slightly raised
- compound moles, which are slightly raised, range in color from tan to dark brown, and involve pigment-producing cells (melanocytes) in both the upper and lower layers of the skin (epidermis and dermis)
- dermal moles, which range from flesh-color to brown, are elevated, most common on the upper body, and may contain hairs
- sebaceous moles, which are produced by over-active oil glands and are yellow and rough-textured
- blue moles, which are slightly raised, colored by pigment deep within the skin, and most common on the head, neck, and arms of women

Most moles are benign (not cancerous), but atypical moles (dysplastic nevi) may develop into malignant melanoma, a potentially fatal form of skin **cancer**. Atypical moles are usually hereditary. Most are bigger than a pencil eraser, and the shape and pigmentation are irregular.

Congenital nevi are more apt to become cancerous than moles that develop after birth, especially if they are more than eight inches in diameter. Lentigo maligna (melanotic freckle of Hutchinson), most common on the face and after the age of 50, first appears as a flat spot containing two or more shades of tan. It gradually becomes larger and darker. One in three of these moles develop into a form of skin cancer known as lentigo maligna melanoma.

Demographics

Nearly everyone has at least a few moles. They generally appear by the time a person is 20 and at first resemble freckles. A mole's color and shape do not usually change; however, changes in hormone levels that occur during **puberty** and pregnancy can make moles larger and darker. New moles may also appear during

this period. About 1 to 3 percent of all babies have one or more moles when they are born. Only about one in 1 million moles is cancerous.

Causes and symptoms

The cause of moles is unknown, although atypical moles seem to run in families and result from exposure to sunlight. During the early 2000s, researchers identified two genes known as CDKN2A and CDK4 that govern susceptibility to melanoma in humans. Other susceptibility genes are being sought. Most experts, however, think that these susceptibility genes are not sufficient by themselves to account for moles becoming cancerous but are influenced by a combination of other inherited traits and environmental factors.

When to call the doctor

Only a small percentage of moles require medical attention. A mole that has the following symptoms should be evaluated by a dermatologist (a physician specializing in skin diseases):

- bleeds
- itches
- looks unusual or changes in any way

Diagnosis

A doctor who suspects skin cancer will remove all or part of the mole for microscopic examination. This procedure, which is usually performed in a doctor's office, is simple, relatively painless, and does not take more than a few minutes. It does leave a scar.

The doctor may also use a dermatoscope to examine the mole before removal. The dermatoscope, which can be used to distinguish between benign moles and melanomas, is an instrument that resembles an ophthalmoscope used to look at the eye. An oil is first applied to the mole to make the outer layers of skin transparent.

A combination of high-frequency ultrasound and color Doppler studies has also been shown to have a high degree of accuracy in distinguishing between melanomas and benign moles.

Treatment

If laboratory analysis confirms that a mole is cancerous, the dermatologist will remove the rest of the mole. Patients should realize that slicing off a section of a malignant mole will not cause the cancer to spread.

Removing a mole for cosmetic reasons involves numbing the area and using scissors or a scalpel to remove the elevated portion. The patient is left with a flat mole the same color as the original growth. Cutting out parts of the mole above and beneath the surface of the skin can leave a scar more noticeable than the mole.

Scissors or a razor can be used to temporarily remove hair from a mole. Permanent hair removal, however, requires electrolysis or surgical removal of the mole.

Prognosis

Moles are rarely cancerous and, once removed, unlikely to recur. A dermatologist should be consulted if a mole reappears after being removed.

Prevention

Wearing a sunscreen and limiting sun exposure may prevent some moles. Anyone who has moles should examine them every month and see a dermatologist if changes in size, shape, color, or texture occur or if new moles appear.

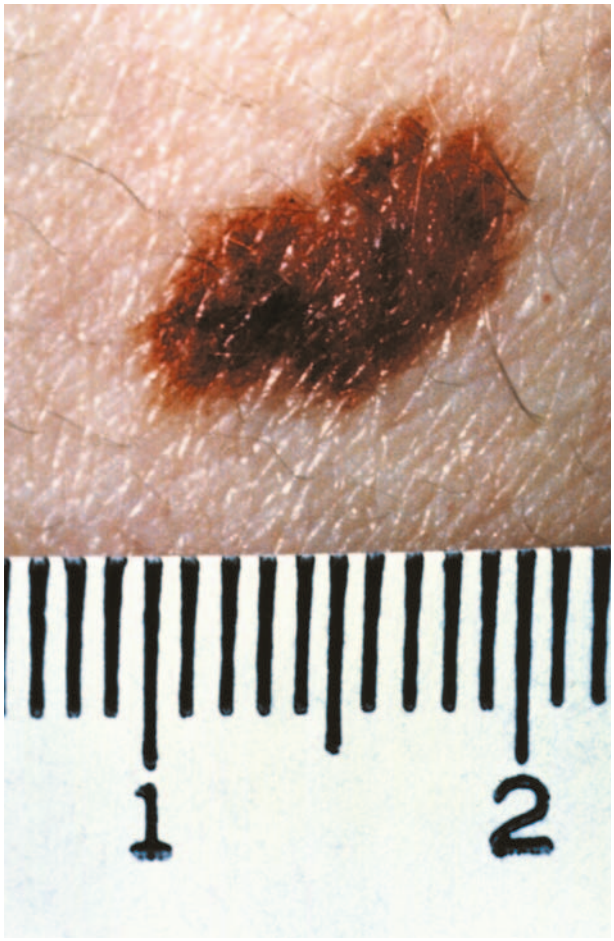
A team of researchers at Duke University reported in 2003 that topical application of a combination of 15 percent vitamin C and 1 percent vitamin E over a four-day period offered significant protection against **sunburn**. The researchers suggest that this combination may protect skin against aging caused by sunlight as well.

Anyone with a **family** history of melanoma should see a dermatologist for an annual skin examination. Everyone should know the ABCDEs of melanoma:

- A: asymmetry, which occurs when the two halves of the mole are not identical
- B: borders that are irregular or indistinct
- C: color that varies in a single mole
- D: diameter, which should be no larger than a pencil eraser (about 6 mm)
- E: elevated above the surrounding tissue

A mole with any of these characteristics should be evaluated by a dermatologist.

Advances in photographic technique had as of 2004 made it easier to track the development of moles with the help of whole-body photographs. A growing number of hospitals are offering these photographs as part of outpatient mole-monitoring services.



Close up of a mole. *(Custom Medical Stock Photo Inc.)*

Parental concerns

Very few moles are cancerous. Moles on the face or other frequently exposed areas of skin may be troubling for children. If a child is particularly troubled by a mole, a dermatologist can be consulted about its possible removal, although this often leaves a scar.

Resources

BOOKS

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Thompson, June. *Spots, Birthmarks, and Rashes: The Complete Guide to Caring for Your Child's Skin*. Westport, CT: Firefly Books, 2003.

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"About Moles, Melanomas, and Lasers: The Dermatologist's Schizophrenic Attitude toward Pigmented Lesions." *Archives of Dermatology* 139 (November 2003): 1405–7.

KEY TERMS

Dermatology—The branch of medicine that studies and treats disorders of the skin.

Malignant melanoma—The most serious of the three types of skin cancer, malignant melanoma arises from the melanocytes, the skin cells that produce the pigment melanin.

Melanin—A pigment that creates hair, skin, and eye color. Melanin also protects the body by absorbing ultraviolet light.

Nevus (plural, nevi)—The medical term for any anomaly of the skin that is present at birth, including moles and birthmarks.

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Chamilin, Sarah L. "Shedding Light on Moles, Melanoma, and the Sun." *Contemporary Pediatrics* 19 (June, 2002): 102–10.

ORGANIZATIONS

American Academy of Dermatologists. PO Box 4014 Schaumburg, IL 60168–4014. Web site: <www.aad.org>.

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Money *see* **Allowance and money management**

Mongolism *see* **Down syndrome**

Mononucleosis *see* **Infectious mononucleosis**

Monosomy X *see* **Turner syndrome**

Mood disorders

Definition

Mood disorders are mental disorders characterized by periods of depression, sometimes alternating with periods of elevated mood.

Description

While many people go through sad or elated moods from time to time, people with mood disorders suffer from severe or prolonged mood states that disrupt their daily functioning. Among the general mood disorders are major depressive disorder, **bipolar disorder**, and dysthymia. In classifying and diagnosing mood disorders, doctors determine if the mood disorder is unipolar or bipolar. When only one extreme in mood (the depressed state) is experienced, this condition is called unipolar. Major depression refers to a single severe period of depression, marked by negative or hopeless thoughts and physical symptoms like fatigue. In major depressive disorder, some patients have isolated episodes of depression. In between these episodes, the patient does not feel depressed or have other symptoms associated with depression. Other patients have more frequent episodes.

Bipolar depression or bipolar disorder (sometimes called manic depression) refers to a condition in which people experience two extremes in mood. They alternate between depression (the low mood) and mania or hypomania (the high mood). These patients go from depression to a frenzied, abnormal elevation in mood. Mania and hypomania are similar, but mania is usually more severe and debilitating to the patient. Dysthymia is a recurrent or lengthy depression that may last a lifetime. It is similar to major depressive disorder, but dysthymia is chronic, long-lasting, persistent, and mild. Patients may have symptoms that are not as severe as major depression, but the symptoms last for many years. It seems that a mild form of the depression is always present. In some cases, people may also experience a major depressive episode on top of their dysthymia, a condition sometimes referred to as double depression.

Psychologists have identified the teenage years as one of the most difficult phases of human life. Although they are often seen as a time for enjoying friendship and engaging in activities that adults would not usually do, the teenage period can be difficult. Many changes in the human mind take place during **puberty**. Apart from the onset of sexual maturity, teenagers must also make key decisions about their future, develop their identities, change schools and meet a new sets of friends, find out about their family's past, and cope with a wide range of other issues. Many young people have difficulty coping with these stresses.

Four out of five teenagers who commit **suicide** are male, but the average female teenager is prone to attempt suicide four more times during her teen years than the average male. White teenage males are more likely to commit suicide than any other ethnic group, but teenage suicide among blacks was as of 2004 increasing. Teenagers who have unsuccessfully tried to commit suicide in the past are more likely to attempt suicide in the future. The odds increase after each failed attempt. There are two groups of teens that are at the highest risk for committing suicide: Native Americans and teens who are gay, lesbian, bisexual, and transgendered.

Gay and bisexual male teens, which represent about 10 percent of the male teen population, are six to seven times more at risk for attempting suicide than their heterosexual peers. Several surveys show gay and lesbian youth account for 30 percent of all suicides among teens, according to the U.S. Department of Health and Human Services. Yet most studies of teen suicide have not been concerned with identifying sexual orientation.

Demographics

As many as 14 percent of children will experience at least one episode of major depression by age 15. Girls are significantly more likely to experience depression than boys after the age of 16. Out of 100,000 adolescents, two to three thousand will have mood disorders out of which 8 to 10 will commit suicide. In the early 2000s, suicide, attempted suicide, and thoughts of committing suicide are growing problems among adolescents in the United States and much of the world. It is the third leading cause of death among 15 to 19 year olds in the United States and the sixth leading cause of death among 10 to 14 year olds. About 2 percent of adolescent girls and 1 percent of adolescent boys attempt suicide each year in the United States. Another 5 to 10 percent of children and teens each year come up with a plan to commit suicide.

Causes and symptoms

Mood disorders tend to run in families. These disorders are associated with imbalances in certain chemicals that carry signals between brain cells (neurotransmitters). These chemicals include serotonin, norepinephrine, and dopamine. Women are more vulnerable to unipolar depression than are men. In adults, major life stressors (like **divorce**, serious financial problems, death of a **family** member, etc.) will often provoke the symptoms of depression in susceptible people. Children's versions of these stressors contribute to their vulnerability to depression.

Major depression is more serious than just feeling sad or "blue." The symptoms of major depression may include the following:

- loss of appetite
- change in the **sleep** pattern, like not sleeping (insomnia) or sleeping too much

Light therapy is a treatment for mood disorders. *(Photograph by Najlah Feanny. © Stock Boston, Inc.)*

- feelings of worthlessness, hopelessness, or inappropriate guilt
- fatigue
- difficulty in concentrating or making decisions
- overwhelming and intense feelings of sadness or grief
- disturbed thinking
- certain physical symptoms such as stomachaches or headaches

Bipolar disorder includes mania or hypomania. Mania is an abnormal elevation in mood. These individuals may be excessively cheerful, have grandiose ideas, and may sleep less. They may talk nonstop for hours, have unending enthusiasm, and demonstrate poor judgment. Sometimes the elevation in mood is marked by irritability and hostility rather than cheerfulness. While the person may at first seem normal with an increase in energy, others who know the person well see a marked difference in behavior. The patient may seem to be in a frenzy and will often make poor, bizarre, or dangerous choices in his or her personal and professional lives. Hypomania is not as severe as mania and does not cause the level of impairment in work and social activities that mania can.

When to call the doctor

There are many methods for helping teenagers deal with mood disorders, both medical and psychological. Most teenagers who have mood disorders believe their problems are too hard or embarrassing to talk about, so it is important for a helper to show they can be trusted and talked to. Seeing a psychologist is widely recommended as well. Psychologists can improve a teenager's vision of life by listening to them and making them feel it will work out for the best.

If a child or teen is so depressed that he or she is talking about suicide, doctors recommend that parents or other helpers do not ask the adolescent what reason they have to think of such a thing to do; rather, one should listen and wait for the child to gain trust enough so that he or she finally can feel comfortable in talking about the problem. Helpers should, however, show understanding of the teenager's situation. Doctors also recommend that helpers do not mention any ''reasons to live'' to the teen-

ager, as that might send the teenager back into depressing thoughts, e.g. "What reason do I have to live?" Many doctors recommend that teenagers be taken to a hospital immediately after they express the desire to commit suicide.

Diagnosis

Doctors diagnose mood disorders based on the patient's description of the symptoms as well as the patient's family history. The length of time the patient has had symptoms is also important. Generally patients are diagnosed with dysthymia if they feel depressed more days than not for at least two years. The depression is mild but long lasting. In major depressive disorder, the patient is depressed almost all day nearly every day of the week for at least two weeks. The depression is severe. Sometimes laboratory tests are performed to rule out other causes for the symptoms (like thyroid disease). The diagnosis may be confirmed when a patient responds well to medication.

Treatment

The most effective treatment for mood disorders is a combination of medication and psychotherapy. The four different classes of drugs used in mood disorders are as follows:

- heterocyclic **antidepressants** (HCAs), such as amitriptyline (Elavil)
- selective serotonin reuptake inhibitors (SSRI inhibitors), such as fluoxetine (Prozac), paroxetine (Paxil), and sertraline (Zoloft)
- monoamine oxidase inhibitors (MAOI inhibitors), such as phenelzine sulfate (Nardil) and tranylcypromine sulfate (Parnate)
- mood stabilizers, such as lithium carbonate (Eskalith) and valproate, often used in people with bipolar mood disorders

A number of psychotherapy approaches are useful as well. Interpersonal psychotherapy helps the patient recognize the interaction between the mood disorder and interpersonal relationships. Cognitive-behavioral therapy explores how the patient's view of the world may be affecting his or her mood and outlook.

When depression fails to respond to treatment or when there is a high risk of suicide, electroconvulsive therapy (ECT) is sometimes used. ECT is believed to affect neurotransmitters like the medications do. Patients are anesthetized and given muscle relaxants to minimize discomfort. Then low-level electric current is passed through the brain to cause a brief convulsion. The most common side effect of ECT is mild, short-term memory loss.

Alternative treatment

There are many alternative therapies that may help in the treatment of mood disorders, including acupuncture, botanical medicine, homeopathy, aromatherapy, constitutional hydrotherapy, and light therapy. The therapy used is an individual choice. Short-term clinical studies have shown that the herb St. John's wort (*Hypericum perforatum*) can effectively treat some types of depression. Though it appears very safe, the herb may have some side effects and its long-term effectiveness has not been proven. It has not been tested in patients with bipolar disorder. St. John's wort and antidepressant drugs should not be taken simultaneously, so patients should tell their doctor if they are taking St. John's wort.

Prognosis

Most cases of mood disorders can be successfully managed if properly diagnosed and treated.

Prevention

People can take steps to improve mild depression and keep it from becoming worse. They can learn stress management (like relaxation training or breathing exercises), **exercise** regularly, and avoid drugs or alcohol.

Parental concerns

Parents who are concerned that their child may have a mood disorder should seek help, such as from a psychiatrist, psychologist, or counselor. Make an appointment with a therapist or counselor who can talk to the child about his or her problems and ways to cope. There are a number of ways parents can help children and teens deal with loneliness, depression, and suicidal feelings. First, parents should let the child do the talking, and they should listen very carefully. They should let the child know they take his or her feelings and thoughts seriously. They should try to find out what is the root of the problem. They may also ask direct questions of the child, such as "Are you thinking of committing suicide?" or "Are you thinking of ending your life?" Third, they should stay with the child. They should not leave the child alone if they say they want to commit suicide. By staying with the child, the parent may be saving the child's life.

KEY TERMS

Cognitive therapy—Psychological treatment aimed at changing a person's way of thinking in order to change his or her behavior and emotional state.

Electroconvulsive therapy (ECT)—A psychological treatment in which a series of controlled electrical impulses are delivered to the brain in order to induce a seizure within the brain. This type of therapy is used to treat major depression and severe mental illness that does not respond to medications.

Neurotransmitter—A chemical messenger that transmits an impulse from one nerve cell to the next.

Serotonin—A widely distributed neurotransmitter that is found in blood platelets, the lining of the digestive tract, and the brain, and that works in combination with norepinephrine. It causes very powerful contractions of smooth muscle and is associated with mood, attention, emotions, and sleep. Low levels of serotonin are associated with depression.

See also Depressive disorders.

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Moral development

Definition

Moral development is the process throught which children develop proper attitudes and behaviors toward other people in society, based on social and cultural norms, rules, and laws.

Description

Moral development is a concern for every parent. Teaching a child to distinguish right from wrong and to behave accordingly is a goal of parenting.

Moral development is a complex issue that—since the beginning of human civilization—has been a topic of discussion among some of the world's most distinguished psychologists, theologians, and culture theorists. It was not studied scientifically until the late 1950s.

Piaget's theory of moral reasoning

Jean Piaget, a Swiss psychologist, explored how children developed moral reasoning. He rejected the idea that children learn and internalize the rules and morals of

society by being given the rules and forced to adhere to them. Through his research on how children formed their judgments about moral behavior, he recognized that children learn morality best by having to deal with others in groups. He reasoned that there was a process by which children conform to society's norms of what is right and wrong, and that the process was active rather than passive.

Piaget found two main differences in how children thought about moral behavior. Very young children's thinking is based on how actions affected them or what the results of an action were. For example, young children will say that when trying to reach a forbidden cookie jar, breaking 10 cups is worse than breaking one. They also recognize the sanctity of rules. For example, they understand that they cannot make up new rules to a game; they have to play by what the rule book says or what is commonly known to be the rules. Piaget called this "moral realism with objective responsibility." It explains why young children are concerned with outcomes rather than intentions.

Older children look at motives behind actions rather than consequences of actions. They are also able to examine rules, determining whether they are fair or not, and apply these rules and their modifications to situations requiring negotiation, assuring that everyone affected by the rules is treated fairly. Piaget felt that the best moral learning came from these cooperative decision-making and problem-solving events. He also believed that children developed moral reasoning quickly and at an early age.

Kohlberg's theory of moral development

Lawrence Kohlberg, an American psychologist, extended Piaget's work in cognitive reasoning into **adolescence** and adulthood. He felt that moral development was a slow process and evolved over time. Still, his six stages of moral development, drafted in 1958, mirrors Piaget's early model. Kohlberg believed that individuals made progress by mastering each stage, one at a time. A person could not skip stages. He also felt that the only way to encourage growth through these stages was by discussion of moral dilemmas and by participation in consensus democracy within small groups. Consensus democracy was rule by agreement of the group, not majority rule. This would stimulate and broaden the thinking of children and adults, allowing them to progress from one stage to another.

PRECONVENTIONAL LEVEL The child at the first and most basic level, the preconventional level, is concerned with avoiding punishment and getting needs met. This level has two stages and applies to children up to 10 years of age.

Stage one is the Punishment-Obedience stage. Children obey rules because they are told to do so by an authority figure (parent or teacher), and they **fear** punishment if they do not follow rules. Children at this stage are not able to see someone else's side.

Stage two is the Individual, Instrumentation, and Exchange stage. Here, the behavior is governed by moral reciprocity. The child will follow rules if there is a known benefit to him or her. Children at this stage also mete out justice in an eye-for-an-eye manner or according to Golden Rule logic. In other words, if one child hits another, the injured child will hit back. This is considered equitable justice. Children in this stage are very concerned with what is fair.

Children will also make deals with each other and even adults. They will agree to behave in a certain way for a payoff. "I'll do this, if you will do that." Sometimes, the payoff is in the knowledge that behaving correctly is in the child's own best interest. They receive approval from authority figures or admiration from peers, avoid blame, or behave in accordance with their concept of self. They are just beginning to understand that others have their own needs and drives.

CONVENTIONAL LEVEL This level broadens the scope of human wants and needs. Children in this level are concerned about being accepted by others and living up to their expectations. This stage begins around age 10 but lasts well into adulthood, and is the stage most adults remain at throughout their lives.

Stage three, Interpersonal Conformity, is often called the "good boy/good girl" stage. Here, children do the right thing because it is good for the **family**, peer group, team, school, or church. They understand the concepts of trust, loyalty, and gratitude. They abide by the Golden Rule as it applies to people around them every day. Morality is acting in accordance to what the social group says is right and moral.

Stage four is the Law and Order, or Social System and Conscience stage. Children and adults at this stage abide by the rules of the society in which they live. These laws and rules become the backbone for all right and wrong actions. Children and adults feel compelled to do their duty and show respect for authority. This is still moral behavior based on authority, but reflects a shift from the social group to society at large.

POST-CONVENTIONAL LEVEL Some teenagers and adults move beyond conventional morality and enter morality based on reason, examining the relative values and opinions of the groups with which they interact. Few adults reach this stage.

Correct behavior is governed by the sixth stage, the Social Contract and Individual Rights stage. Individuals in this stage understand that codes of conduct are relative to their social group. This varies from culture to culture and subgroup to subgroup. With that in mind, the individual enters into a contract with fellow human beings to treat them fairly and kindly and to respect authority when it is equally moral and deserved. They also agree to obey laws and social rules of conduct that promote respect for individuals and value the few universal moral values that they recognize. Moral behavior and moral decisions are based on the greatest good for the greatest number.

Stage six is the Principled Conscience or the Universal/Ethical Principles stage. Here, individuals examine the validity of society's laws and govern themselves by what they consider to be universal moral principles, usually involving equal rights and respect. They obey laws and social rules that fall in line with these universal principles, but not others they deem as aberrant. Adults here are motivated by individual conscience that transcends cultural, religious, or social convention rules. Kohlberg recognized this last stage but found so few people who lived by this concept of moral behavior that he could not study it in detail.

Carol Gilligan and the morality of care

Kohlberg's and Piaget's theories have come under fire. Kohlberg's six stages of moral development, for example, have been criticized for elevating Western, urban, intellectual (upper class) understandings of morality, while discrediting rural, tribal, working class, or Eastern moral understandings. Feminists have pointed out potential sexist elements in moral development theories devised by male researchers using male subjects only (such as Kohlberg's early work). Because women's experiences in the world differ from men's in every culture, it would stand to reason that women's moral development might differ from men's, perhaps in significant ways.

Carol Gilligan deemed Kohlberg's research biased because he only used male subjects to reach his findings. Because of this, his model is based on a concept of morality based on equity and justice, which places most men in stage five or six. Gilligan found that women, who value social interaction more than men, base their moral decisions on a culture of caring for other human beings. This would place them at stage three, making women appear to be inferior morally to men. Men determine immorality based on treating others unfairly, and women base it on turning away someone in need.

Gilligan's work, however, doesn't solve the gender question, because newer research has found that both males and females often base their moral judgments and behaviors on both justice and care. Nevertheless, the morality of care theory opened up explorations of moral reasoning in many groups and cultures.

Bronfenbrenner

Urie Bronfenbrenner studied children and schools in different cultures since many ethnic, religious, and social groups often have their own rules for moral behavior. His research found five moral orientations, regardless of culture, social group, or developmental stage. Movement from the first stage to any of the others was dependent on participation in the family and other social institutions within each culture. Movement to the last stage involved exposure to a different moral system that might be in conflict with one's own. This moral pluralism forces individuals to examine their own moral reasoning and beliefs. This often occurs when people work in other countries or cultures and come face to face with different sets of moral conventions.

Bronfenbrenner also noted that individuals could slide back into a previous moral orientation when they experienced the breakdown of their familiar social order as in war, regime changes, genocide, famine, or large scale natural disasters that destroy social infrastructures. People narrow their attention to their own pressing needs and ignore the welfare of the larger society.

Self-oriented morality coincided with Kohlberg's pre-conventional morality. Behavior is based on self-interest and motivated by who can help children get what they want or who is hindering that process. This stage was found in all children and some adults in all cultures.

Authority-oriented morality again is similar to Kohlberg's Law and Order stage. This applies not only to parents' rules but to teachers, religious leaders, and government officials. This moral orientation was culturally defined. It was very evident in Middle Eastern cultures where religious authority is the law.

Peer-authority morality is moral conformity based on the conventions and rules of a social group. This is evident among teenagers in Western cultures and even among some adults.

Collective-oriented morality is an extension of the peer-authority stage. Here a larger group's rule supercedes individual rights and interests. Duty is the law. This moral orientation was found in Asian cultures.

Objectively-oriented morality is akin to Kohlberg's universal principles stage. Here, however, these rules

transcend individual moral perspectives and become entities in themselves. Like Kohlberg's last stage, this moral orientation was found in relatively few people in any culture.

Other theories

There are several other approaches to the study of moral development, which are categorized in a variety of ways. Briefly, the social learning theory approach claims that humans develop morality by learning the rules of acceptable behavior from their external environment, an essentially behaviorist approach. Psychoanalytic theory proposes instead that morality develops through humans' conflict between their instinctual drives and the demands of society. **Cognitive development** theories view morality as an outgrowth of cognition, or reasoning, whereas personality theories are holistic in their approach, taking into account all the factors that contribute to human development.

The differences between these approaches rest on two questions: How moral are infants at birth? and How is moral maturity defined? The contrasting philosophies at the heart of the answers to these questions determine the essential perspective of each moral development theory. Those who believe infants are born with no moral sense tend toward social learning or behaviorist theories, because all morality must therefore be learned from the external environment. Others who believe humans are innately aggressive and completely self-oriented are more likely to accept psychoanalytic theories where morality is the learned management of socially destructive internal drives. Those who believe it is the reasoning abilities that separate humans from the rest of creation will find cognitive development theories the most attractive. And those who view humans as holistic beings born with a full range of potentialities will most likely be drawn to personality theories.

What constitutes mature morality is a subject of great controversy. Each society develops its own set of norms and standards for acceptable behavior, leading many to say that morality is entirely culturally conditioned. There is debate over whether or not this means that there are no universal truths, and no cross-cultural standards for human behavior. This debate fuels the critiques of many moral development theories.

Definitions of what is or is not moral are in a state of upheaval within individual societies. Controversies rage over the morality of warfare (especially nuclear), ecological conservation, genetic research and manipulation, alternative fertility and childbearing methods, abortion, sexuality, pornography, drug use, euthanasia, racism, sexism, and human rights issues, among others. Determining the limits of moral behavior becomes increasingly difficult as human capabilities, choices, and responsibilities proliferate with advances in technology and scientific knowledge. For example, prenatal testing techniques that determine birth defects in the womb force parents to make new moral choices about whether to give birth to a child.

The rise in crime, drug and alcohol abuse, gang violence, teen parenthood, and **suicide** in Western society has also caused a rise in concern over morality and moral development. Parents and teachers want to know how to raise moral children, and they turn to moral development theorists to find answers. Freudian personality theories became more widely known to the Western public in the 1960s and were understood to imply that repression of a child's natural drives would lead to neuroses. Many parents and teachers were therefore afraid to **discipline** their children, and permissiveness became the rule. Cognitive development theories did little to change things, as they focus on reasoning and disregard behavior. Behaviorist theories, with their complete denial of free will in moral decision-making, are unattractive to many and require precise, dedicated, behavior modification techniques.

Schools are returning to character education programs, popular in the 1920s and 1930s, where certain virtues such as honesty, fairness, and loyalty, are taught to students along with the regular academic subjects. Unfortunately, there is little or no agreement as to which virtues are important and what exactly each virtue entails.

Another approach to moral education that became popular in the 1960s and 1970s is known as values clarification or values modification. The purpose of these programs is to guide students to establish or discern their own system of values on which to base their moral decisions. Students are also taught that others may have different values systems, and that they must be tolerant of those differences. The advantages of this approach are that it promotes self-investigation and awareness and the development of internal moral motivations, which are more reliable than external motivations, and prevents fanaticism, authoritarianism, and moral coercion. The disadvantage is that it encourages moral relativism, the belief that "anything goes." Values clarification is generally seen as a valuable component of moral education, but incomplete on its own.

Lawrence Kohlberg devised a moral education program in the 1960s based on his cognitive development theory. Called the Just Community program, it utilizes age-appropriate or stage-appropriate discussions of moral dilemmas, democratic consensus rule-making, and

the creation of a community context where students and teachers could act on their moral decisions. Just Community programs have been established in schools, prisons, and other institutions with a fair amount of success. Exposure to moral questions and the opportunity to practice moral behavior in a supportive community appear to foster deeper moral reasoning and more constructive behavior.

Overall, democratic family and school systems are much more likely to promote the development of internal self-controls and moral growth than are authoritarian or permissive systems. Permissive systems fail to instill any controls, while authoritarian systems instill only fear of punishment, which is not an effective deterrent unless there is a real chance of being caught or punishment becomes a reward because it brings attention to the offender. True moral behavior involves a number of internal processes that are best developed through warm, caring parenting with clear and consistent expectations, emphasis on the reinforcement of positive behaviors rather than the punishment of negative ones, modeling of moral behavior by adults, and creation of opportunities for the child to practice moral reasoning and actions.

According to personal (social) goal theory, moral behavior is motivated by the desire to satisfy a variety of personal and social goals, some of which are self-oriented (selfish), and some of which are other-oriented (altruistic). The four major internal motivations for moral behavior as presented by personal (social) goal theorists are: 1) empathy; 2) the belief that people are valuable in and of themselves and therefore should be helped; 3) the desire to fulfill moral rules; and 4) self-interest.

In social domain theory, moral reasoning is said to develop within particular social domains: 1) moral (e.g., welfare, justice, rights); 2) social-conventional (social rules for the orderly function of society); and 3) personal (pure self-interest, exempt from social or moral rules).

Most people have more than one moral voice and shift among them depending on the situation. In one context, a person may respond out of empathy and place care for an individual over concern for social rules. In a different context, that same person might instead insist on following social rules for the good of society, even though someone may suffer because of it. People also show a lack of consistent morality by sometimes choosing to act in a way that they know is not moral, while continuing to consider themselves moral people. This discrepancy between moral judgment (perceiving an act as morally right or wrong) and moral choice (deciding whether to act in the morally right way) can be explained in a number of ways, any one of which may be true in a given situation:

- weakness of will (the person is overwhelmed by desire)
- weakness of conscience (guilt feelings are not strong enough to overcome temptation)
- limited/flexible morality (some latitude allowed in moral behavior while still maintaining a "moral" identity)

The Moral Balance model proposes that most humans operate out of a limited or flexible morality. Rather than expecting moral perfection from ourselves or others, people set certain limits beyond which they cannot go. Within those limits, however, there is some flexibility in moral decision-making. Actions such as taking coins left in the change-box of a public telephone may be deemed acceptable (though not perfectly moral), while **stealing** money from an open, unattended cash register is not. Many factors are involved in the determination of moral acceptability from situation to situation, and the limits on moral behavior are often slippery. If given proper encouragement and the opportunity to practice a coherent inner sense of morality, however, most people will develop a balanced morality to guide their day-to-day interactions with their world.

Common problems

Religious development often goes hand in hand with moral development. Children's concepts of divinity, right and wrong, and who is ultimately responsible for the world's woes are shaped by the family and by the religious social group to which each child belongs. Their concepts also mirror cognitive and moral developmental stages.

In general, in the earliest stage (up to age two years), the child knows that religious objects and books are to be respected. The concept of a divine being is vague, but the child enjoys the regularity of the religious rituals such as prayer.

In the next stage (from two to 10 years), children begin to orient religion concepts to themselves as in the catechism litany, "Who made you? God made me." The concept of a divine being is usually described in anthropomorphic ways for children around six years old. In other words, children perceive God to look like a human being only bigger or living in the sky. At this stage, God is physically powerful and often is portrayed as a superhero. God may also be the wish-granter and can fix anything. Children embrace religious holidays and rituals during this stage.

KEY TERMS

Altruistic—Thinking of others.

Anthropomorphic—Taking on human characteristics or looking like humans.

Cognition—The act or process of knowing or perceiving.

Flat affect—Showing no emotion.

Moral choice—Deciding whether to act in the morally right way.

Moral judgment—Perceiving an act as morally right or wrong.

In the Intermediate Stage during pre-adolescence, children are considered to be in the pre-religious stage. The anthropomorphized divinity is pictured as being very old and wise. God is also thought of as doing supernatural things: having a halo, floating over the world, or performing miracles. Children in this stage understand the panoply of religious or divine beings within the religious belief system. For example, Christian children will distinguish between God and Jesus and the disciples or saints.

The last stage in adolescence focuses on personalizing religious rituals and drawing closer to a divine being. Teenagers begin to think of God in abstract terms and look at the mystical side of the religious experience. They may also rebel against organized religion as they begin to question the world and the rules around them.

Some adults who are considered highly religious consider God to be an anthropomorphized divine being or may reject the supernatural or mystical religious experience. This does not mean that these adults have somehow been arrested in their religious development. This just means that the variation among these stages is great and is determined by the particular religious community in which the individual is involved.

Parental concerns

When to call the doctor

Every child misbehaves and will sometimes act selfishly and hurtfully. It is when these acts increase, impulses cannot be controlled, or authority defiance becomes troublesome, that parents may need to seek professional help. Lack of impulse control and authority defiance can be symptoms of medical conditions and psychological disorders. Self-centered behavior, coupled with lack of acceptance of wrongdoing that continues into older childhood and adolescence, may be a problem that requires family or individual counseling.

Risky behaviors such as speeding, drinking, **smoking**, doing drugs, or engaging in sexual behavior may be related to **peer pressure** and wanting to conform to the group or may be a way to defy authority. These behaviors, though deemed morally wrong by most societies, may also be symptoms of deeper psychological troubles.

Of extreme concern is the rare child who acts with no remorse, and appears to have to conscience. This is usually signaled by early violent outbursts, destructive behavior, or by acts of cruelty to pets or other children. After each incident, the child has a flat affect (no emotion) or fails to admit that there was anything wrong with the his or her actions. These children need intervention immediately. Behaviors such as these may be indicators of sociopathic disorders.

Resources

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ORGANIZATIONS

Association for Moral Education Dr. James M. Dubois Center for Health Care Ethics. Saint Louis University, Salus Center 3545 Lafayette Ave. St. Louis, MO 63104.

Developmental Studies Center 2000 Embarcadero, Suite 305 Oakland, CA 94606-5300. (510) 533-0213 or (800) 666-7270.

Center for the Advancement of Ethics and Character. Boston University School of Education. 605 Commonwealth Ave., Room 356 Boston, MA 02215. (617) 353-3262. Fax: (617) 353-3924.

Educators for Social Responsibility (ESR). 23 Garden St. Cambridge, MA 02138. (800) 370-2515.

The Heartwood Institute. 425 N. Craig St., Suite 302 Pittsburgh, PA 15213. (800) 432-7810.

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Morphine *see* **Narcotic drugs**

Mother-child relationships *see* **Parent-child relationships**

Motion sickness

Definition

Motion sickness is uncomfortable **dizziness**, **nausea**, and **vomiting** that people experience when their sense of balance and equilibrium is disturbed because their brain cannot make sense of conflicting information about their body's location in space and motion in their environment.

Description

Motion sickness is connected to the role of the sensory organs. The sensory organs control a body's sense of balance by telling the brain what direction the body is pointing, the direction it is moving, and if it is standing still or turning. These messages are relayed by the inner ear (or labyrinth); the eyes; the skin pressure receptors (such as in those in the feet), the muscle and joint sensory receptors, which track what body parts are moving to the central nervous system (the brain and spinal cord). The brain then is responsible for processing all incoming information and making sense out of it. Riding in a car, being on a ship, or taking an amusement park ride can cause conflicting stimulation of the different sense organs. The result is motion sickness.

For example, when reading a book in the back seat of a moving car, the inner ears and skin receptors sense the motion, but the eyes register only the stationary pages of the book. This conflicting information may cause the usual motion sickness symptoms of dizziness, nausea, and vomiting. While motion sickness can be bothersome, it is not a serious illness, and it can be prevented.

Demographics

Although nearly 80 percent of the general population experiences motion sickness at one time in their lives, children between the ages of four and ten are most vulnerable. Children often out grow motion sickness. Toddlers under age two are rarely motion sick. Adults who frequently get migraine headaches are more likely than others to have recurrent episodes of motion sickness.

Researchers at the Naval Medical Center in San Diego, California, reported in 2003 that 70 percent of research subjects with severe motion sickness had abnormalities of the vestibular system. Research also suggests that some people inherit a predisposition to motion sickness. This predisposition is more marked in some ethnic groups than in others. One study published in 2002 found that persons of Chinese or Japanese ancestry are significantly more vulnerable to motion sickness than persons of British ancestry.

Causes and symptoms

While all of the body's sensory organs contribute to motion sickness, excess stimulation to the vestibular system within the inner ear (the body's balance center) has been shown to be one of the primary reasons for this condition. Balance problems (vertigo) are often caused by a conflict between what is seen and how the inner ear perceives it, leading to confusion in the brain. This confusion may result in higher heart rates, rapid breathing, nausea and sweating, along with dizziness and vomiting.

Additional factors that may contribute to the occurrence or severity of motion sickness include the following:

- poor ventilation
- anxiety or **fear** (Both have been found to lower a person's threshold for experiencing motion sickness symptoms.)
- food (A heavy meal of spicy and greasy foods before traveling is thought to increase motion sickness symptoms.)
- alcohol consumption
- genetic factors
- pregnancy (Susceptibility in women to vomiting during pregnancy appears to be related to motion sickness, although the precise connections are not well understood as of 2004.)

Often viewed as a minor annoyance, some travelers are temporarily immobilized by motion sickness, and a few continue to feel its effects for hours and even days after a trip.

When to call the doctor

Most cases of motion sickness are mild and self-limiting. Parents should call the doctor before giving young children over-the-counter medications for motion sickness. Some remedies are recommended only for older children.

Diagnosis

Most cases of motion sickness are self-diagnosed. If symptoms such as dizziness become chronic, a doctor may be able to help alleviate the discomfort by looking further into a patient's general health. Questions regarding medications, head injuries, recent infections, and other questions about the ear and neurological system will be asked. An examination of the ears, nose, and throat, as well as tests of nerve and balance function, may also be performed.

Severe cases of motion sickness or those that become progressively worse may require additional, specific tests. Diagnosis in these situations deserves the attention and care of a doctor with specialized skills in diseases of the ear, nose, throat, equilibrium, and neurological system.

Treatment

Medications to help ease the symptoms of motion sickness are available without a prescription (over-the-counter or OTC). Normally these are taken 30 to 60 minutes before traveling to prevent motion sickness symptoms, as well as during extended trips.

Over-the-counter drugs

The following OTC drugs contain ingredients that are considered by the United States Food and Drug Administration (FDA) to be safe and effective for the treatment of motion sickness:

- Marezine (and others) includes the active ingredient cyclizine and is not for use in children under six years of age.
- Benadryl (and others) includes the active ingredient diphenhydramine and is not for use in children under age two without a doctor's permission.
- Dramamine (and others) includes the active ingredient dimenhydrinate and is not for use in children under two years of age.
- Bonine (and others) includes the active ingredient meclizine and is not for use in children under age 12.

The FDA recommends that people with emphysema, chronic **bronchitis**, glaucoma, or difficulty urinating due to an enlarged prostate do not use OTC drugs for motion sickness unless directed by their doctor. Children should not be given OTC motion sickness medication without first checking with a healthcare professional.

Prescription drugs

Longer trips may require a prescription medication called scopolamine available in the form of a skin patch or gel that is rubbed on the skin. Another prescription drug that is sometimes given for motion sickness is ondansetron (Zofran), which was originally developed to treat nausea associated with **cancer chemotherapy**. It appears to be safe for use in children under the age of six. In March 2003, the FDA approved a new anti-emetic (anti-nausea) drug. Known as aprepitant, it is sold under the brand name Emend.

Alternative treatment

Ginger (*Zingiber officinale*) in its various forms is often used to calm the stomach, and the oils it contains (gingerols and shogaols) appear to relax the intestinal tract in addition to mildly depressing the central nervous system. Some of the most effective forms of ginger are the powdered, encapsulated form; ginger tea prepared from sliced ginger root; and candied pieces. All forms of ginger should be taken on an empty stomach.

Placing manual pressure on the Neiguan or Pericardium-6 acupuncture point (located about three finger-widths above the wrist on the inner arm), either by acupuncture, acupressure, or a mild, electrical pulse, has shown to be effective against the symptoms of motion sickness. Elastic wristbands sold at most drugstores are

also used as a source of relief due to the pressure they place in this area. Pressing the small intestine 17 (just below the earlobes in the indentations behind the jawbone) may also help in the functioning of the ear's balancing mechanism.

There are several homeopathic remedies that work specifically for motion sickness. They include *Cocculus*, *Petroleum*, and *Tabacum*.

Prognosis

Motion sickness is not a serious disorder and almost always resolves once the conflicting motion messages have stopped.

Prevention

Because motion sickness is easier to prevent than treat once it has begun, the best treatment is prevention. The following steps may help deter the unpleasant symptoms of motion sickness before they occur:

- Avoid reading while traveling.
- Ride in a location that allows the eyes to see the same motion that the body and inner ears feel. Safe positions include the front seat of the car (for older children) while looking at distant scenery; the deck of a ship where the horizon can be seen; and sitting by the window of an airplane. The least motion on an airplane is in a seat over the wings.
- Maintain a fairly straight-ahead view.
- Eat a light meal before traveling, or if already nauseated, avoid food altogether.
- Avoid watching or talking to another traveler who is having motion sickness.
- Take motion sickness medicine at least 30 to 60 minutes before travel begins or as recommended by a physician.

Even those who frequently endure motion sickness can learn to travel by anticipating the conditions of their next trip. Research also suggests that increased exposure to the stimulation that causes motion sickness may help decrease symptoms on future trips.

Parental concerns

Parental concerns center primarily on making the child comfortable and anticipating the logistics of traveling with a child who is motion sick. Rarely do children vomit to the point of becoming dehydrated. Prevention and practical steps such as taking something for the child to vomit into and carrying a change of clothes can ease parents' worries about motion sickness.

KEY TERMS

Acupressure—Often described as acupuncture without needles, acupressure is a traditional Chinese medical technique based on theory of *qi* (life energy) flowing in energy meridians or channels in the body. Applying pressure with the thumb and fingers to acupressure points can relieve specific conditions and promote overall balance and health. Also known as dian xue.

Acupuncture—Based on the same traditional Chinese medical foundation as acupressure, acupuncture uses sterile needles inserted at specific points to treat certain conditions or relieve pain.

Anti-emetic—A preparation or medication that relieves nausea and vomiting. Cola syrup, ginger, and motion sickness medications are examples of antiemetics.

Vertigo—A feeling of dizziness together with a sensation of movement and a feeling of rotating in space.

Vestibular system—The brain and parts of the inner ear that work together to detect movement and position.

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Motor skills, fine *see* **Fine motor skills**
Motor skills, gross *see* **Gross motor skills**
Mouth sores *see* **Stomatitis**

Movement disorders

Definition

Movement disorders are a group of diseases and syndromes affecting the ability to produce and control bodily movements.

Description

It seems simple and effortless, but normal movement requires an astonishingly complex system of control. Disruption of any portion of this system can cause a person to produce movements that are too weak, too forceful, too uncoordinated, or too poorly controlled for the task at hand. Unwanted movements may occur at rest. Intentional movement may become impossible. Such conditions are called movement disorders.

Abnormal movements are symptoms of underlying disorders. In some cases, the abnormal movements are the only symptoms. Childhood disorders or conditions that may cause abnormal movements include:

- **cerebral palsy**
- choreoasthetosis
- encephalopathies
- essential tremor
- inherited ataxias (**Friedreich's ataxia**, Machado-Joseph disease, and spinocerebellar ataxias)
- multiple sclerosis
- parkinsonism and juvenile Parkinson's disease
- **poisoning** by carbon monoxide, cyanide, methanol, or manganese
- psychogenic disorders
- restless legs syndrome
- **spasticity**
- **stroke**
- **Tourette syndrome** and other tic disorders
- Wilson disease

Demographics

The incidence rates and demographics vary for different types of movement disorders. Restless legs syndrome (RLS) affects approximately 12 million people in the United States. The disorder can affect males and females and can begin at any age, although it may become worse as a person gets older. The most common ataxia is Friedreich's ataxia; in the United States, it affects one in 50,000 people, both male and female.

Causes and symptoms

Causes

Movement is produced and coordinated by several interacting brain centers, including the motor cortex, the cerebellum, and a group of structures in the inner portions of the brain called the basal ganglia. Sensory information provides critical input on the current position and velocity of body parts, and spinal nerve cells (neurons) help prevent opposing muscle groups from contracting at the same time.

To understand how movement disorders occur, it is helpful to consider a normal voluntary movement, such as reaching to touch a nearby object with the right index finger. To accomplish the desired movement, the arm must be lifted and extended. The hand must be held out to align with the forearm, and the forefinger must be extended while the other fingers remain flexed.

THE MOTOR CORTEX Voluntary motor commands begin in the motor cortex, located on the outer wrinkled surface of the brain. Movement of the right arm is begun by the left motor cortex, which generates a large volley of signals to the involved muscles. These electrical signals pass along upper motor neurons through the midbrain to the spinal cord. Within the spinal cord, they

connect to lower motor neurons, which convey the signals out of the spinal cord to the surface of the muscles involved. Electrical stimulation of the muscles causes contraction, and the force of contraction pulling on the skeleton causes movement of the arm, hand, and fingers.

Damage to or death of any of the neurons along this path causes weakness or paralysis of the affected muscles.

ANTAGONISTIC MUSCLE PAIRS The previous description of movement is too simple, however. One important refinement to it comes from considering the role of opposing, or antagonistic, muscle pairs. Contraction of the biceps muscle, located on the top of the upper arm, pulls on the forearm to flex the elbow and bend the arm. Contraction of the triceps, located on the opposite side, extends the elbow and straightens the arm. Within the spine, these muscles are normally wired so that willed (voluntary) contraction of one is automatically accompanied by blocking of the other. In other words, the command to contract the biceps provokes another command within the spine to prevent contraction of the triceps. In this way, these antagonist muscles are kept from resisting one another. Spinal cord or brain injury can damage this control system and cause involuntary simultaneous contraction and spasticity, an increase in resistance to movement during motion.

THE CEREBELLUM Once the movement of the arm is initiated, sensory information is needed to guide the finger to its precise destination. In addition to sight, the most important source of information comes from the ''position sense'' provided by the many sensory neurons located within the limbs (proprioception). Proprioception is what allows a person to touch a finger to his or her nose, even with eyes closed. The balance organs in the ears provide important information about posture. Both postural and proprioceptive information are processed by a structure at the rear of the brain called the cerebellum. The cerebellum sends out electrical signals to modify movements as they progress, ''sculpting'' the barrage of voluntary commands into a tightly controlled, constantly evolving pattern. Cerebellar disorders cause inability to control the force, fine positioning, and speed of movements (ataxia). Disorders of the cerebellum may also impair the ability to judge distance so that a person under- or over-reaches the target (dysmetria). Tremor during voluntary movements can also result from cerebellar damage.

THE BASAL GANGLIA Both the cerebellum and the motor cortex send information to a set of structures deep within the brain that help control involuntary components of movement (basal ganglia). The basal ganglia send output messages to the motor cortex, helping to initiate movements, regulate repetitive or patterned movements, and control muscle tone.

Circuits within the basal ganglia are complex. Within this structure, some groups of cells begin the action of other basal ganglia components and some groups of cells block the action. These complicated feedback circuits are not entirely understood. Disruptions of these circuits are known to cause several distinct movement disorders. A portion of the basal ganglia called the substantia nigra sends electrical signals that block output from another structure called the subthalamic nucleus. The subthalamic nucleus sends signals to the globus pallidus, which in turn blocks the thalamic nuclei. Finally, the thalamic nuclei send signals to the motor cortex. The substantia nigra, then, begins movement and the globus pallidus blocks it. This complicated circuit can be disrupted at several points.

Disruptions in other portions of the basal ganglia are thought to cause **tics**, tremors, dystonia, and a variety of other movement disorders, although the exact mechanisms are not well understood.

Some movement disorders, including Huntington's disease and inherited ataxias, are caused by inherited genetic defects. Some diseases that cause sustained muscle contraction limited to a particular muscle group (focal dystonia) are inherited, but others are caused by trauma. The cause of most cases of Parkinson's disease is unknown, although genes have been found for some familial forms.

Symptoms

Abnormal movements are broadly classified as either hyperkinetic (too much movement) and hypokinetic (too little movement). Hyperkinetic movements include:

- Dystonia—sustained muscle contractions, often causing twisting or repetitive movements and abnormal postures. Dystonia may be limited to one area (focal) or may affect the whole body (general). Focal dystonias may affect the neck (cervical dystonia or torticollis); the face (one-sided or hemifacial spasm, contraction of the eyelid or blepharospasm, contraction of the mouth and jaw or oromandibular dystonia, simultaneous spasm of the chin and eyelid or Meige syndrome); the vocal cords (laryngeal dystonia); or the arms and legs (writer's cramp or occupational cramps). Dystonia may be painful as well as incapacitating.
- Tremor—uncontrollable (involuntary) shaking of a body part. Tremor may occur only when muscles are relaxed or only during an action or while holding an active posture.

- Tics—involuntary, rapid, non-rhythmic movement or sound. Tics can be controlled briefly.
- Myoclonus—a sudden, brief, jerky, shock-like involuntary muscle contraction. Myoclonic jerks may occur singly or repetitively. Unlike tics, myoclonus cannot be controlled even briefly.
- Spasticity—an abnormal increase in muscle tone. It may be associated with involuntary **muscle spasms**, sustained muscle contractions, and exaggerated deep tendon reflexes that make movement difficult or uncontrollable.
- Chorea—rapid, non-rhythmic, uncontrolled jerky movements, most often in the arms and legs. Chorea also may affect the hands, feet, trunk, neck, and face. Choreoathetosis is a syndrome of continuous random movements that usually occur at rest and may appear to be fidgety, dancing, or writhing.
- Ballism—like chorea, but the movements are much larger, more explosive and involve more of the arm or leg. This condition, also called ballismus, can occur on both sides of the body or on one side only (hemiballismus).
- Akathisia—restlessness and a desire to move to relieve uncomfortable sensations. Sensations may include a feeling of crawling, **itching**, stretching, or creeping, usually in the legs.
- Athetosis—slow, writhing, continuous, uncontrollable movement of the arms and legs.

Hypokinetic movements include:

- Bradykinesia—extreme slowness and stiffness of movement.
- Freezing—inability to begin a movement or involuntary stopping of a movement before it is completed.
- Rigidity—an increase in muscle tension when an arm or leg is moved by an outside force.
- Postural instability—loss of the ability to maintain upright posture caused by slow or absent righting reflexes.

Diagnosis

Diagnosis of movement disorders requires a careful medical history and a thorough physical and neurological examination.

The medical history helps the physician evaluate the presence of other conditions or disorders that might contribute to or cause the disorder. Records of previous diagnoses, surgeries, and treatments are reviewed. The child's **family** medical history is evaluated to determine if there is a history of muscular or neurological disorders. Genetic testing is available for some forms of movement disorders.

The physical and neurological exams may include an evaluation of the child's motor reflexes, including muscle tone, mobility, strength, balance, and endurance; heart and lung function; cranial nerve function; and an examination of the child's abdomen, spine, throat, and ears. The child's height, weight, and blood pressure also are checked and recorded. Routine blood and urine analyses are performed.

Brain imaging studies are usually performed. Imaging techniques include **computed tomography** scan (CT scan), positron emission tomography (PET), or **magnetic resonance imaging** (MRI) scans. A lumbar puncture (spinal tap) may be necessary. Video recording of the abnormal movement is often used to analyze movement patterns and track progress of the disorder and its treatment.

Other tests may include **x rays** of the spine and hips or diagnostic blocks with local anesthetics to provide information on the effectiveness of potential treatments.

To aid diagnosis, a multi-disciplinary team may be consulted so the proper treatment can be planned. Occupational and physical therapy evaluations may be helpful to determine upper and lower extremity movement patterns and passive range of motion.

In some cases, nerve conduction studies with electromyography of the affected muscles may be performed to evaluate the child's muscular activity and provide a comprehensive **assessment** of nerve and muscle function.

In both tests, the examiner uses a computer, monitor, amplifier, loudspeaker, stimulator, and high-tech filters to see and hear how the muscles and nerves are responding during the test. In the nerve conduction study, small electrodes are placed on the skin over the muscles to be examined. A stimulator delivers a very small electrical current (that does not cause damage to the body) through the electrodes, causing the nerves to fire. In the electromyogram, a very thin, sterilized needle is inserted into various muscles. The needle is attached by wires to a recording machine. The patient is asked to relax and contract the muscles being examined. The electrical signals produced by the nerves and muscles during these tests are measured and recorded by a computer and displayed as electrical waves on the monitor. The test results are interpreted by a specially trained physician.

An EEG (**electroencephalogram**) may be performed to detect seizures, analyze general brain

functioning, and measure brain activity associated with movement or sensation. This test measures the electrical signals from the brain. Surface electrodes attached to the scalp measure voltages in the brain. The electrical activity can be measured while the child is resting or, in some cases, when the child is moving. An evoked potentials study may be part of the EEG test. Evoked potentials record the response of the brain to a sensory, visual, or auditory stimulus.

Treatment

Treatment of a movement disorder begins with a proper diagnostic evaluation. Treatment options include physical and occupational therapies, medications, surgery, or a combination of these treatments.

The goals of treatment are to increase the child's comfort, decrease **pain**, ease mobility, help with activities of daily living such as hygiene, ease rehabilitation procedures, and prevent or decrease the risk of developing a joint contracture. The type of treatment recommended will depend upon the severity of the disorder; the child's overall health; the potential benefits, limitations, and side effects of the treatment; and the impact of the treatment on the child's quality of life.

Clinicians should work with the child and parents or caregivers to develop an individual treatment plan. Specific treatment goals will vary from one person to the next. Treatment should be provided by a movement disorders specialist or specially trained pediatric neurologist and a multi-disciplinary team of specialists that may include a physiatrist, physical therapist, occupational therapist, gait and movement specialists, social worker, and surgical specialists as applicable, such as a pediatric orthopedic surgeon or pediatric neurosurgeon.

In some cases, treatment is not recommended or desired, because it would actually interfere with the patient's current mobility and it would not improve function. For example, some people with multiple sclerosis who experience significant leg weakness find that spasticity makes their legs more rigid, helping them to stand, transfer to a chair or bed, or walk.

Physical and occupational therapies

Physical therapy includes stretching exercises, muscle group strengthening exercises, and range of motion exercises to prevent muscles from shortening (contracture), preserve flexibility and range of motion, and reduce the severity of symptoms. Exercises should be practiced daily, as recommended by the physical therapist. Prolonged stretching can lengthen muscles, and strengthening exercises can restore the proper strength to affected muscles. Aquatic therapy also may be recommended, since there is less stress on the body when in the water.

A physical therapist can instruct the patient on proper posture guidelines. Proper posture is critical, especially while sitting and sleeping, to maintain proper alignment of the hips and back. Balancing rest and **exercise** is also important.

Occupational therapy may include splints, casts, or braces on the affected arm or leg to enable proper limb positioning, and maintain flexibility and range of motion. The therapy may include training for proper limb positioning while seated in a wheelchair or lying in bed.

Physical and occupational therapists can provide guidelines on how to adapt the child's environment to ensure **safety** and comfort.

Medications

Medications can help compensate for some imbalances of the basal ganglionic circuit. Drugs to treat movement disorders include oral medications, injected medications, and continuous delivery medications. These medications work by preventing nerves from signaling the muscles to contract, thereby preventing muscle contractions.

If treatment with a single medicine fails to effectively treat the disorder, a different medicine may be tried or an additional medicine may be prescribed. The most important medication guidelines are to ensure that the child takes the medication exactly as prescribed, and to never discontinue any medication without first talking to the child's doctor, even if the medication does not seem to be working or is causing unwanted side effects.

ORAL MEDICATIONS Baclofen (Lioresal) is a muscle relaxant that works on nerves in the spinal cord to reduce spasticity. The benefits of baclofen include decreased stretch reflexes, improved passive range of motion, and reduced muscle spasms, pain, and tightness. Side effects include drowsiness and sedation, as well as weakness, decreased muscle tone, confusion, fatigue, **nausea**, and **dizziness**. Baclofen should not be taken with central nervous system depressants or alcohol.

Levodopa (L-dopa) is a medication that is converted to dopamine in the brain. Dopamine is a chemical that aids in the transmission of nerve signals. Sinemet is a combination medication containing levodopa and carbidopa. Carbidopa enables L-dopa to be converted to dopamine after the L-dopa enters the brain, thereby lowering

the oral dose and decreasing side effects. Side effects include nausea, **diarrhea**, and low blood pressure.

Anticholinergics, including trihexyphenidyl (Artane) and benztropine (Benztrop MES, Cogentin), block acetylcholine receptors in the brain. Acetylcholine receptors are integral proteins that respond to the neurotransmitter acetylcholine by opening a pathway in the membrane for ion diffusion across the cell membrane. Side effects include dry mouth, blurred vision, **constipation**, urinary retention, and rapid heart rate. These side effects are usually much less frequent in children than adults; therefore, much higher doses are usually prescribed in children.

Benzodiazepines, such as diazepam (Valium), clonazepam (Klonopin, Rivotril), and lorazepam (Ativan) act on the central nervous system to improve passive range of motion, reduce muscle overactivity and painful spasms, and provide overall relaxation. These medications are often taken at night because they cause drowsiness, but they also can relieve muscle spasms that interrupt **sleep**. Side effects include unsteadiness, loss of strength, low blood pressure, gastrointestinal symptoms, memory problems, confusion, and behavioral problems.

Dantrolene sodium (Dantrium) acts on the muscles to directly interfere with the chemistry of the muscle contraction. It is generally used when other medications are not effective. Benefits may include improved passive movement, decreased muscle tone, and reduced muscle spasms, tightness, and pain. Side effects include generalized weakness—including weakness of the respiratory muscles—as well as drowsiness, fatigue, diarrhea, and sensitivity to the sun. Liver problems may occur with this medication, and frequent lab tests are performed to evaluate liver function.

Tizanidine (Zanaflex) acts on the central nervous system. It does not usually cause reduced muscle strength. The most common side effect is sedation, and other side effects include low blood pressure, dry mouth, dizziness, and hallucinations. Liver problems may occur with this medication, and frequent lab tests are performed to evaluate liver function.

A variety of other medications may be used to treat movement disorders, including antiepileptic drugs that stimulate GABA receptors in the brain's basal ganglia; neuroleptics that block dopamine D2-like receptors; Clonidine (Catapres) and selective serotonin reuptake inhibitors (SSRIs, such as fluoxetine, commonly known as Prozac) for the treatment of tics; and channel modulators that affect the behavior of channels that transport small molecules such as potassium, sodium, or calcium across cell membranes.

INJECTED MEDICATIONS Botulinum-toxin type A (Botox, Dysport) or type B (Myobloc) is injected locally into the affected muscle group to relax the muscles in dystonia or spasticity. It works by preventing nerves from sending signals to the muscles that cause them to contract. Although the treatment takes one to two weeks to reach its full effectiveness, the beneficial effects last three to four months. Botulinum-toxin allows more normal limb positioning and improved mobility. In some patients, the injections also decrease pain. Injections may be used to make casting easier, ease the adjustment of a new brace, or delay surgery.

Botulinum-toxin is made by the bacteria that cause **botulism**. However, the amount of botulinum-toxin injected to treat spasticity is such a small amount that it would not cause botulism poisoning. This treatment is very safe, and the injections can be given in a doctor's office without the use of sedation or anesthesia. Injections can be repeated, but should be spaced from three to six months apart to avoid exceeding the recommended dose. Botulinum-toxin injections may be used in combination with other treatments.

Botulinum-toxin injections are typically expensive and may not be covered by insurance. A Reimbursement Hotline established by Allergan, the manufacturer of Botox, is a resource for reimbursement questions: (800) 530-6680 or online at <www.botox.com>. Elan, the manufacturer of Myobloc, also has resources available to answer questions about reimbursement. Interested persons may call (888) 461-2255 or go online at <www.elan.com>.

Alcohol and phenol are injected in combination, but are less common treatments. The medications are injected directly onto nerves that supply spastic muscles to destroy them. The injections cut off the signals to those muscles, allowing them to relax. This treatment may be used to treat spasticity in larger muscle groups closer to the trunk, such as the thigh muscles. Although this treatment is generally less expensive than botulinum-toxin injections, there are more serious side effects.

Short-term medications such as lidocaine, a local anesthetic, can be used to assess the potential benefit of botulinum-toxin or alcohol and phenol injections.

CONTINUOUS DELIVERY MEDICATIONS Baclofen usually is taken as an oral medication but also can be delivered directly into the spinal fluid when the oral medication does not effectively control symptoms. An intrathecal baclofen delivery system, surgically placed by a neurosurgeon, continuously releases prescribed amounts of baclofen in small doses directly into the spinal fluid via a small catheter and pump. This type of

delivery system causes fewer and less severe side effects than the oral baclofen.

Pump refills and medication adjustments are generally made once every two to three months after the initial dosage is established. The pump system lasts from three to five years, at which time it needs to be replaced.

Surgery

Surgery is only recommended when all other treatments have been tried and have not effectively controlled the child's symptoms.

Selective dorsal rhizotomy surgery, also called selective posterior rhizotomy, involves a surgical resection of part of the spinal nerve. By cutting the sensory nerve rootlets that cause the spasticity, muscle stiffness is decreased while other functions are maintained. Potential benefits of this surgical procedure include pain relief, reduced spasticity to improve walking or aid sitting in a wheelchair, increased ability to bend at the waist, and improved use of the hands. Sometimes rhizotomy results in improved breathing and better control of the arms, legs, and head.

Thalamotomy is a surgical procedure used to destroy part of the thalamus, which is thought to produce abnormal brain activity that causes tremor. Pallidotomy is a surgical procedure used to destroy part of the globus pallidus, which is thought to become overactive with certain disorders, such as Parkinson's disease. Although effective, these surgeries have significant risks, including paralysis, loss of vision, or loss of speech if the precise location of the brain is not targeted during surgery. With the advent of a less invasive approach called deep brain stimulation, these surgeries have become less common.

Deep brain stimulation (DBS) is a way to inactivate the parts of the brain thought to cause overactivity or tremor in certain muscles, without destroying a part of the brain. It is currently a treatment option for adult patients with Parkinson's disease, but research is underway to determine if the procedure can benefit children with movement disorders.

During the DBS procedure, an electrode placed in a precise area of the brain delivers small, electrical shocks to interrupt the abnormal brain activity that leads to symptoms. The electrode has four metal contacts that can be used in different combinations. A few patients may have stimulators implanted on both sides of the brain, but this increases the risk for complications. The electrode is connected by a wire to a pacemaker-like device implanted under the skin in the chest. This device generates the electrical shocks. The electrical stimulation can be adjusted as the patient's condition progresses over time, and the stimulator can be turned off in the event that other beneficial therapies, such as brain cell transplantation, are performed.

Orthopedic surgery may be performed to correct a contracture. During contracture release surgery, the tendon of a contractured muscle is cut, the joint repositioned to a more normal angle, and a cast is applied. Regrowth of the tendon to this new length occurs over several weeks following surgery. After the cast is removed, physical therapy can help strengthen the muscles and improve range of motion. This procedure is most commonly performed on the Achilles tendon but may also be performed on the knees, hips, shoulders, elbows, and wrists. Tendon transfer surgery is another technique to treat contractures. During this procedure, the tendon attached to a spastic muscle is cut and transferred to a different site, preventing the muscle from being pulled into an abnormal position. The disadvantages of these orthopedic procedures are that they are irreversible and they may need to be repeated.

Other orthopedic surgeries that may accompany contracture release surgery include osteotomy, in which a small wedge is removed from a bone to allow repositioning. A cast is applied while the bone heals in a more natural position. Osteotomy is more commonly performed on the bones in the hips or feet. Arthrodesis is a fusing of bones that normally move independently, to limit the ability of a spastic muscle to pull the joint into an abnormal position. Arthrodesis is more commonly performed on the bones in the ankle.

Other treatments

Transplantation of fetal cells into the basal ganglia has produced mixed results in Parkinson's disease and is being researched for application in other movement disorders.

Brief application (about 10 minutes) of cold packs to spastic muscles may help ease pain and improve function for a short period of time.

Electrical stimulation may be used to stimulate a weak muscle to counteract the action of a stronger, spastic muscle.

Alternative treatment

Alternative and complementary therapies include approaches that are considered to be outside the mainstream of traditional health care. Among the therapies that may be helpful are acupuncture, homeopathy, touch therapies, postural alignment therapies, and biofeedback.

The progress made will depend on the individual and his or her condition.

Biofeedback training may be used to teach the patient how to consciously reduce muscle tension. Biofeedback uses an electrical signal that indicates when a spastic muscle relaxes. The patient may be able to use biofeedback to learn how to consciously reduce muscle tension and possibly reduce symptoms.

Coenzyme Q10 supplements may be beneficial, as some people with movement disorders may have low levels of this substance. Coenzyme Q10 is a natural substance produced by the body that transports electrons during cellular respiration, or the process in which cells get their energy from oxygen.

Initial trials of cannabinoids, the active ingredient in marijuana, have shown promise in the treatment of muscle stiffness and limb straightening associated with multiple sclerosis. Further research is needed to determine the beneficial effects of marijuana-derived substances on neuromuscular symptoms associated with movement disorders. Researchers caution that smoking marijuana is dangerous, especially since there may be other harmful substances mixed in with the illegal drug.

Before learning or practicing any particular technique, it is important for the parent or caregiver and child to learn about the therapy, its safety and effectiveness, potential side effects, and the expertise and qualifications of the practitioner. Although some practices are beneficial, others may be harmful to certain patients. Alternative therapies should not be used as a substitute for medical therapies prescribed by a doctor. Parents should discuss these alternative treatments with the child's doctor to determine the techniques and remedies that may be beneficial for the child.

Nutritional concerns

Dietary guidelines are individualized, based on the child's age, diagnosis, overall health, severity of disability, and level of functioning. Specific nutritional problems, such as swallowing or feeding difficulties, may be a concern in some patients and should be managed by a team of specialists, including a speech therapist. Early identification, treatment, and correction of specific feeding problems will improve the health and nutritional status of the patient.

A well-balanced and carefully planned diet will help maintain general good health for people with movement disorders. Specialists recommend that people with multiple sclerosis and other movement disorders adhere to the same low-fat, high fiber diet that is recommended for the general population. A diet rich in fresh fruits and vegetables will ensure adequate intake of antioxidants, substances that help protect against free radical damage.

Children with movement disorders may have different energy needs, depending on their condition. One study indicated that ambulatory and non-ambulatory adolescents with cerebral palsy had decreased energy needs compared with a control group of normal adolescents. Therefore, a child's specific calorie needs should be evaluated by a registered dietitian who can work with the parents to develop an individualized meal plan. The child's weight should be obtained once a week or at least once a month to determine if caloric intake is adequate.

A child's self-feeding skills can impact his or her health outcome. One study indicated that 90 percent of children with good to fair motor and feeding skills reached adulthood. In contrast, a lack of self-feeding skills was associated with a six-fold increase in mortality (rate of death).

Maintaining a healthy weight is important to prevent the development of chronic diseases such as diabetes, high blood pressure (**hypertension**), and heart disease.

Tube feedings may be required in some patients with **failure to thrive**, aspiration **pneumonia**, difficulty swallowing, or an inability to ingest adequate calories orally to maintain nutritional status or promote growth.

Prognosis

The prognosis for a patient with a movement disorder depends on the specific disorder. There is no cure for movement disorders. However, they can be well-managed with the proper combination of physical and occupational therapies, medication, and surgery. The long-term outlook depends on the severity of the disorder.

Prevention

Prevention depends on the specific disorder.

Parental concerns

Parents should work closely with the child's therapists and doctors to create an effective treatment plan. It is important for parents to communicate their treatment goals with the health care team. Parents should take an active role in the child's exercise program and help the child practice the exercises, as prescribed, every day.

There are many tips to make the home and school environments safer for a child with a movement disorder. An occupational therapist can work with parents to assess the home environment and provide resources for

KEY TERMS

Active motion—Spontaneous; produced by active efforts. Active range of motion exercises are those that are performed by the patient without assistance.

Activities of daily living (ADL)—The activities performed during the course of a normal day, for example, eating, bathing, dressing, toileting, etc.

Acupuncture—Based on the same traditional Chinese medical foundation as acupressure, acupuncture uses sterile needles inserted at specific points to treat certain conditions or relieve pain.

Anoxia—Lack of oxygen.

Ataxia—A condition marked by impaired muscular coordination, most frequently resulting from disorders in the brain or spinal cord.

Autonomic nervous system—The part of the nervous system that controls so-called involuntary functions, such as heart rate, salivary gland secretion, respiratory function, and pupil dilation.

Biofeedback—A training technique that enables an individual to gain some element of control over involuntary or automatic body functions.

Botulinum toxin—A potent bacterial toxin or poison made by *Clostridium botulinum*; causes paralysis in high doses, but is used medically in small, localized doses to treat disorders associated with involuntary muscle contraction and spasms, in addition to strabismus. Commonly known as Botox.

Bradykinesia—Extremely slow movement.

Central nervous system—Part of the nervous system consisting of the brain, cranial nerves, and spinal cord. The brain is the center of higher processes, such as thought and emotion and is responsible for the coordination and control of bodily activities and the interpretation of information from the senses. The cranial nerves and spinal cord link the brain to the peripheral nervous system, that is the nerves present in the rest of body.

Cerebral palsy—A nonprogressive movement disability caused by abnormal development of or damage to motor control centers of the brain.

Chorea—Involuntary movements in which the arms or legs may jerk or flail uncontrollably.

Choreoathetosis—Involuntary rapid, irregular, jerky movements or slow, writhing movements that flow into one another.

Clonic—Referring to clonus, a series of muscle contractions and partial relaxations that alternate in some nervous diseases in the form of convulsive spasms.

Computed tomography (CT)—An imaging technique in which cross-sectional x rays of the body are compiled to create a three-dimensional image of the body's internal structures; also called computed axial tomography.

Contraction—A tightening of the uterus during pregnancy. Contractions may or may not be painful and may or may not indicate labor.

Contracture—A tightening or shortening of muscles that prevents normal movement of the associated limb or other body part.

Encephalopathy—Any abnormality in the structure or function of brain tissues.

Essential tremor—An uncontrollable (involuntary) shaking of the hands, head, and face. Also called familial tremor because it is sometimes inherited, it can begin in the teens or in middle age. The exact cause is not known.

Fasciculations—Small involuntary muscle contractions visible under the skin.

Fetal tissue transplantation—A method of treating Parkinson's and other neurological diseases by grafting brain cells from human fetuses onto the basal ganglia. Human adults cannot grow new brain cells but developing fetuses can. Grafting fetal tissue stimulates the growth of new brain cells in affected adult brains.

General anesthesia—Deep sleep induced by a combination of medicines that allows surgery to be performed.

Hereditary ataxia—One of a group of hereditary degenerative diseases of the spinal cord or cerebellum. These diseases cause tremor, spasm, and wasting of muscle.

Homeopathy—A holistic system of treatment developed in the eighteenth century. It is based on the idea that substances that produce symptoms of sickness in healthy people will have a curative effect when given in very dilute quantities to sick people who exhibit those same symptoms. Homeopathic remedies are believed to stimulate the body's own healing processes.

Huntington's disease—A rare hereditary disease that causes progressive chorea (jerky muscle movements) and mental deterioration that ends in dementia. Huntington's symptoms usually appear in patients in their 40s. Also called Huntington's chorea.

KEY TERMS (contd.)

Hyperactive reflexes—Reflexes that persist too long and may be too strong. For example, a hyperactive grasp reflex may cause the hand to stay clenched in a tight fist.

Hypermobility—Unusual flexibility of the joints, allowing them to be bent or moved beyond their normal range of motion.

Hypertonia—Having excessive muscular tone or strength.

Levodopa (L-dopa)—A substance used in the treatment of Parkinson's disease. Levodopa can cross the blood-brain barrier that protects the brain. Once in the brain, it is converted to dopamine and thus can replace the dopamine lost in Parkinson's disease.

Local anesthesia—Pain-relieving medication used to numb an area while the patient remains awake. Also see general anesthesia.

Magnetic resonance imaging (MRI)—An imaging technique that uses a large circular magnet and radio waves to generate signals from atoms in the body. These signals are used to construct detailed images of internal body structures and organs, including the brain.

Mask—An expressionless look, caused by reduced movements of the face.

Motor neuron—A nerve cell that specifically controls and stimulates voluntary muscles.

Multiple sclerosis—A progressive, autoimmune disease of the central nervous system characterized by damage to the myelin sheath that covers nerves. The disease, which causes progressive paralysis, is marked by periods of exacerbation and remission.

Muscle spasm—Localized muscle contraction that occurs when the brain signals the muscle to contract.

Myoclonus—Involuntary contractions of a muscle or an interrelated group of muscles. Also known as myoclonic seizures.

Neurologist—A doctor who specializes in disorders of the nervous system, including the brain, spinal cord, and nerves.

Neurosurgeon—Physician who performs surgery on the nervous system.

Nocturnal leg cramps—Cramps that may be related to exertion and awaken a person during sleep.

Occupational therapist—A healthcare provider who specializes in adapting the physical environment to meet a patient's needs. An occupational therapist also assists patients and caregivers with activities of daily living and provide instructions on wheelchair use or other adaptive equipment.

Orthopedist—A doctor specializing in treatment of the musculoskeletal system.

Parkinsonism—A set of symptoms originally associated with Parkinson disease that can occur as side effects of neuroleptic medications. The symptoms include trembling of the fingers or hands, a shuffling gait, and tight or rigid muscles.

Parkinson's disease—A slowly progressive disease that destroys nerve cells in the basal ganglia and thus causes loss of dopamine, a chemical that aids in transmission of nerve signals (neurotransmitter). Parkinson's is characterized by shaking in resting muscles, a stooping posture, slurred speech, muscular stiffness, and weakness.

Passive movement—Movement that occurs under the power of an outside source such as a clinician. There is no voluntary muscular contraction by the individual who is being passively moved.

Periodic limb movement disorder—A disorder characterized by involuntary flexion of leg muscles, causing twitching and leg extension or kicking during sleep.

Peripheral nerves—Nerves outside the brain and spinal cord that provide the link between the body and the central nervous system.

Physiatrist—A physician who specializes in physical medicine and rehabilitation.

Physical therapist—A healthcare provider who teaches patients how to perform therapeutic exercises to maintain maximum mobility and range of motion.

Positron emission tomography (PET)—A computerized diagnostic technique that uses radioactive substances to examine structures of the body. When used to assess the brain, it produces a three-dimensional image that shows anatomy and function, including such information as blood flow, oxygen consumption, glucose metabolism, and concentrations of various molecules in brain tissue.

Progressive supranuclear palsy—A rare disease that gradually destroys nerve cells in the parts of the brain that control eye movements, breathing, and muscle coordination. The loss of nerve cells causes palsy, or paralysis, that slowly gets worse as the disease progresses. The palsy affects ability to move the eyes, relax the muscles, and control balance. Also called Steele-Richardson-Olszewski syndrome.

KEY TERMS (contd.)

Psychogenic disorders—A variety of unusual, involuntary movements that occur in children with psychiatric disorders or in response to anxiety, stress, depression, anger, or grief. Psychogenic movements are thought to represent the physical expression of an intolerable mental conflict.

Range of motion (ROM)—The range of motion of a joint from full extension to full flexion (bending) measured in degrees like a circle.

Restless legs syndrome (RLS)—A disorder in which the patient experiences crawling, aching, or other disagreeable sensations in the calves that can be relieved by movement. RLS is a frequent cause of difficulty falling asleep at night.

Rigidity—A constant resistance to passive motion.

Scissoring—Involuntary crossing of the legs.

Spinal cord injury—Injury to the spinal cord, via blunt or penetrating trauma.

Stroke—Interruption of blood flow to a part of the brain with consequent brain damage. A stroke may be caused by a blood clot or by hemorrhage due to a burst blood vessel. Also known as a cerebrovascular accident.

Tourette syndrome—A neurological disorder characterized by multiple involuntary movements and uncontrollable vocalizations called tics that come and go over years, usually beginning in childhood and becoming chronic. Sometimes the tics include inappropriate or obscene language (coprolalia).

Wilson disease—A rare, inherited disorder that causes excess copper to accumulate in the body. Steadily increasing amounts of copper circulating in the blood are deposited primarily in the brain, liver, kidneys, and the cornea of the eyes. It can cause psychiatric symptoms resembling schizophrenia.

adaptive equipment that may be helpful. Some of these tips include:

- All throw rugs should be removed unless they are firmly attached to the floor.
- There must be proper lighting. Nightlights should be placed along key pathways of the home.
- The top and bottom of stairs should be highlighted with a contrasting color or texture to distinguish them.
- The floor should be free of clutter, to prevent tripping or falling.
- Handrails should be installed, especially along stairways and in the bathroom.
- All electrical cords and other cords should be kept out of the way.

Raising a child with a movement disorder can be challenging. Support groups are available to provide information and assistance.

See also Tics; Tourette syndrome.

Resources

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ORGANIZATIONS

Brain Injury Association of America. 8201 Greensboro Dr., Ste. 611, McLean, VA 22102. (800) 444-6443 or (703) 761-0750. Web site: &;lt;http://www.biausa.org>.

Movement Disorders Society. 555 East Wells St., Suite 1100, Milwaukee, WI 53202-3823. (414) 276-2145. Web site: <www.movementdisorders.org>.

National Center on Birth Defects and Developmental Disabilities. Centers for Disease Control. 4770 Buford Highway., NE, Ste. F-35, Atlanta, GA 30341. (770) 488-7080. Web site: <http://cdc.gov/ncbddd/dh>.

National Institute on Disability and Rehabilitation Research. Office of Special Education and Rehabilitative Services. U.S. Department of Education, 400 Maryland Ave., SW,

Washington, DC 20202-7100. (202) 245-7640. Web site: <www.ed.gov/about/offices/list/osers/nidrr>.

National Institute of Neurological Disorders and Stroke (NINDS). National Institutes of Health. P.O. Box 5801, Bethesda, MD 20824. (800) 352-9424 or (301) 496-5751. Web site: <www.ninds.nih.gov/about_ninds/>.

National Rehabilitation Information Center (NARIC). 4200 Forbes Blvd., Ste. 202, Lanham, MD 20700. (800) 346-2742 or (301) 459-5900. Web site: <www.naric.com>.

National Spinal Cord Injury Association. 6701 Democracy Blvd., #300-9, Bethesda, MD 20817. (800) 962-9629 or (301) 214-4006. info@spinalcord.org. Web site: <www.spinalcord.org>.

WE MOVE (Worldwide Education and Awareness for Movement Disorders). 204 W. 84th St. New York, NY 10024. (800) 437-MOVE. Web site: <www.wemove.org>.

WEB SITES

Spinal Cord Injury Information Network. Available online at: <www.spinalcord.uab.edu>.

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MRI *see* **Magnetic resonance imaging**

Mucopolysaccharidoses

Definition

Mucopolysaccharidosis (MPS) is a general term for many different related inherited disorders that are caused by the accumulation of mucopolysaccharides in body tissues. This accumulation interferes with the individual's development.

Description

Mucopolysaccharides are long chains of sugar molecules that are essential for building the bones, cartilage, skin, tendons, and other tissues in the body. Another name for mucopolysaccharides is glycosaminoglycans (GAGs). Normally, the human body continuously breaks down and rebuilds cells that contain GAGs. There are many different types of GAGs, and different GAGs are unable to be broken down in each of the MPS conditions. Several enzymes are involved in breaking down each GAG, and a deficiency or absence of any of the essential enzymes can cause the GAG not to be broken down completely. This condition results in the accumulation of GAGs in the tissues and organs in the body. The accumulating GAGs are stored in cellular structures called lysosomes, and these disorders are known as lysosomal storage diseases. When too many GAGs accumulate, organs and tissues become damaged or do not function properly.

Before specific deficient enzymes were identified, MPS disorders were diagnosed by the signs and symptoms seen in an individual. The discovery of individual enzyme deficits resulted in a reclassification of some of the MPS disorders. Types of MPS disorders are MPS I, MPS II, MPS III, MPS IV, MPS VI, MPS VII, and MPS IX. However, these conditions are also referred to by their original names, which are Hurler, Hurler-Scheie, Scheie (all MPS I), Hunter (MPS II), Sanfilippo (all MPS III), Morquio (all MPS IV), Maroteaux-Lamy (MPS VI), Sly (MPS VII), and Hyaluronidase deficiency (MPS IX).

Demographics

MPS disorders are rare, and the frequency with which they occur varies depending on the type of the disorder. For all MPS types combined, the disorder occurs in only about one of every 25,000 people. Except for MPS II, individuals of both genders are affected equally. Because of its inheritance pattern, MPS II is found only in males. All MPS disorders are present at birth, although symptoms appear at different times, depending on the type of disorder. There appears to be no race or ethnic component in the distribution of MPS disorders.

Causes and symptoms

All MPS disorder except MPS II are inherited in an autosomal recessive manner. An individual with an autosomal recessive disorder inherits one non-working genes from each parent. The parents are called carriers of the disorder. If the parent has one good copy of the gene and one defective copy, the parent will not have MPS and may be unaware that he or she has a defective gene. MPS only occurs when both of an individual's genes that produce the same enzyme contain a mutation or defect, causing them not to function properly. As a result, either no enzyme is produced, or the amount produced is inadequate. When two people are carriers for an autosomal recessive disorder, they have a 25 percent chance with each pregnancy to have a child with the disorder. Some individuals who have MPS are able to have children. Children of MPS parents are all carriers of the disorder, because they inherit one bad copy of the gene from the affected parent. However, these children are not at risk to develop the disorder unless the other parent is a carrier or affected with the same autosomal recessive condition.

Unlike the other MPS conditions, MPS II is inherited in an X-linked recessive manner, which means that the gene causing the condition is located on the X chromosome, one of the two sex chromosomes. A male child inherits an X chromosome from his mother and a Y chromosome from his father. He will have the disorder if the X chromosome inherited from his mother carries the defective gene, since he has only one (nonfunctioning) copy of the gene. Females inherit one X chromosome from their mother and a second X chromosome from their father. Because they have two X chromosomes, they are carriers of the disorder if one of their X chromosomes has the gene that causes the condition, while the other X chromosome does not.

Although MPS are all inherited disorders, each type is caused by a deficiency of one particular enzyme involved in breaking down GAGs. The accumulation of the GAGs in the tissues and organs in the body causes the symptoms characteristic of the MPS disorders. Symptoms and their time of onset vary widely depending on which form of the disorder the individual inherits.

MPS I

MPS I is caused by a deficiency of the enzyme alpha-L-iduronidase. Three conditions, Hurler, Hurler-Scheie, and Scheie syndromes, are caused by a deficiency of this enzyme. Initially, these three conditions were believed to be separate, because each was associated with different physical symptoms and prognoses. However, once the underlying cause of these conditions was identified, it was realized that these three conditions are variants of the same disorder.

MPS I H (Hurler syndrome)

About one child in 100,000 is born with Hurler syndrome. This tends to be the most severe form of MPS I. Symptoms of Hurler syndrome are often evident within the first year or two after birth. Often these infants initially grow faster than expected, but then reach a point where they begin to lose the skills that they have learned. Their growth slows and typically stops by age three.

Facial features begin to coarsen. These children develop a short nose, flatter face, thicker skin, and a protruding tongue. Their heads become larger, and they develop more hair on their bodies, with the hair becoming coarser. Their bones are also affected, and they usually develop joint contractures (stiff joints), kyphosis (a specific type of curve to the spine), and broad hands with short fingers. Many of these children have breathing difficulties, and respiratory infections are common. Other common problems include heart valve dysfunction, thickening of the heart muscle (cardiomyopathy), enlarged spleen and liver, clouding of the cornea, hearing loss, and carpal tunnel syndrome. These children typically do not live past age 12.

MPS I H/S (Hurler-Scheie syndrome)

Hurler-Scheie syndrome an intermediate form of MPS I, meaning that the symptoms are not as severe as those in individuals who have MPS I H but not as mild as those in MPS I S. Approximately one baby in 115,000 is born with Hurler-Scheie syndrome. These individuals tend to be shorter than expected. They can have normal **intelligence**; however, some individuals with MPS I H/S experience learning difficulties. These individuals may develop some of the same physical features as those with Hurler syndrome, but usually they are not as severe. The prognosis for children with MPS I H/S is variable with some individuals dying during childhood, while others live to adulthood.

MPS I S (Scheie syndrome)

Scheie syndrome is the mild form of MPS I. About one baby in 500,000 is born with Scheie syndrome. Individuals with MPS I S usually have normal intelligence, although there have been some reports of individuals with MPS I S developing psychiatric problems. Common physical problems include corneal clouding, heart abnormalities, and orthopedic difficulties involving their hands and back. Individuals with MPS I S do not develop the facial features seen with MPS I H and usually these individuals have a normal life span.

MPS II (Hunter syndrome)

Hunter syndrome is caused by a deficiency of the enzyme iduronate-2-sulphatase. All individuals with Hunter syndrome are male, because the gene that causes the condition is located on their single X chromosome. Like many MPS conditions, Hunter syndrome is divided into two forms, mild and severe. About one in 110,000 males are born with Hunter syndrome, with the severe form being three times more common than the mild form.

The severe form of MPS II is associated with progressive **mental retardation** and physical disability, with most individuals dying before age 15. In the milder form, most of these individuals live to adulthood and have normal intelligence or only mild mental impairments. Males with the mild form of Hunter syndrome develop physical differences similar to the males with the severe form, but not as quickly. Males with mild Hunter syndrome can have a normal life span and some have had children. Most males with Hunter syndrome develop joint stiffness, chronic **diarrhea**, enlarged liver and spleen, heart valve problems, hearing loss, and

kyphosis. They also tend to be shorter than expected. These symptoms progress at different rates depending on whether the individual has the mild or severe form of MPS II.

MPS III (Sanfilippo syndrome)

MPS III, like the other MPS conditions, was initially diagnosed by the individual having certain physical signs and symptoms. It was later discovered that the physical symptoms associated with Sanfilippo syndrome could be caused by a deficiency in one of four enzymes. MPS III is in the early 2000s subdivided into four groups, labeled A through D, based on the specific enzyme that is deficient. All four of these enzymes are involved in breaking down the same GAG, heparan sulfate. Heparan sulfate is mainly found in the central nervous system and accumulates in the brain when it cannot be broken down because one of those four enzymes is deficient or missing.

MPS III is a variable condition, with symptoms beginning to appear between two and six years of age. Because of the accumulation of heparan sulfate in the central nervous system (CNS), the CNS is severely affected. In MPS III, signs that the CNS is degenerating usually become evident between six and ten years of age. Many children with MPS III develop seizures, sleeplessness, thicker skin, joint contractures, enlarged tongues, cardiomyopathy, behavior problems, and mental retardation. The life expectancy in MPS III is also variable. On average, individuals with MPS III live until they are teenagers, with some living longer and others not that long.

MPS IIIA (Sanfilippo syndrome type A) is caused by a deficiency of the enzyme heparan N-sulfatase. Type IIIA is the most severe of the four types of MPS III. Symptoms appear and death occurs at an earlier age than in other subtypes. A study in British Columbia estimated that one in every 325,000 babies is born with MPS IIIA. MPS IIIA is the most common of the four types in Northwestern Europe. The gene that causes MPS IIIA is located on the long arm of chromosome 17.

MPS IIIB(Sanfilippo syndrome type B) is due to a deficiency in N-acetyl-alpha-D-glucosaminidase (NAG). This type of MPS III is not as severe as type IIIA, and the characteristic signs and symptoms vary. Type IIIB is the most common of the type III disorders in southeastern Europe. The gene associated with MPS IIIB is also located on the long arm of chromosome 17.

MPS IIIC (Sanfilippo syndrome type C) is caused by a deficiency in the enzyme acetyl-CoA-alpha-glucosaminide acetyltransferase. This is a rare form of MPS III. The gene involved in MPS IIIC is believed to be located on chromosome 14.

MPS IIID (Sanfilippo syndrome type D) is caused by a deficiency in the enzyme N-acetylglucosamine-6-sulfatase. This form of MPS III is also rare. The gene involved in MPS IIID is located on the long arm of chromosome 12.

MPS IV A (Morquio syndrome type A)

MPS IV A is the severe form of the disorder and is caused by a deficiency in the enzyme galactosamine-6-sulphatase. The gene involved with MPS IV A is located on the long arm of chromosome 16. The major organs affected by MPS IV are the cornea and the cartilage, particularly the cartilage of the neck. Bowel and bladder function also can be impaired. Respiratory problems and **sleep** apnea are common. Individuals with MPS IV appear healthy at birth but show skeletal deformities and growth retardation by age three. Death often occurs early in individuals with the severe form of this disorder.

MPS IV B (Morquio syndrome type B) is the milder form of the disorder. The enzyme, beta-galactosidase, is deficient in MPS IV B. The gene that produces beta-galactosidase is located on the short arm of chromosome 3. Individuals with the MPS IV B can have normal lifespans (into their 70s).

MPS VI (Maroteaux-Lamy syndrome)

MPS VI, which is another rare form of MPS, is caused by a deficiency of the enzyme N-acetylglucosamine-4-sulphatase. This condition is also variable; individuals may have a mild or severe form of the disorder. Typically, the nervous system or intelligence of an individual with MPS VI is not affected. Individuals with a more severe form of MPS VI can have airway obstruction, develop **hydrocephalus** (accumulation of fluid in the brain), and exhibit bone changes. Individuals with a severe form of MPS VI are more likely to die while in their teens. With a milder form of the disorder, individuals tend to be shorter than expected for their age, develop corneal clouding, and live longer. The gene involved in MPS VI is believed to be located on the long arm of chromosome 5.

MPS VII (Sly syndrome)

MPS VII is an extremely rare form of MPS and is caused by a deficiency of the enzyme beta-glucuronidase. It is also highly variable, but symptoms are generally similar to those seen in individuals with Hurler syndrome. The gene that causes MPS VII is located on the long arm of chromosome 7.

MPS IX (Hyaluronidase deficiency)

MPS IX, a condition first described in 1996, is caused by a deficiency of the enzyme hyaluronidase. In the few individuals described with this condition, the symptoms are variable. Some individuals develop soft tissue masses (growths) under the skin. Also, these individuals are shorter than expected for their age. The gene involved in MPS IX is believed to be located on the short arm of chromosome 3.

When to call the doctor

Parents should inform the doctor immediately if MPS runs in their **family**, so that early testing can be done on their children. In addition, any time they have questions about their child's growth and development, they should talk to their pediatrician.

Diagnosis

While a diagnosis for each type of MPS can be made based on the physical signs described above, several of the conditions have similar features. Therefore, enzyme analysis is used to determine the specific MPS disorder. Enzyme analysis often cannot accurately determine if an individual is a carrier for an MPS disorder, because the enzyme levels in individuals who are not carriers overlaps the enzyme levels seen in those individuals who are carrier for MPS. With many of the MPS conditions, several mutations have been found in each gene involved that can cause symptoms of each condition. If the specific mutation is known in a family, DNA analysis may be possible.

Once a couple has had a child with MPS, prenatal testing is available to them to help determine if another fetus is affected with the same MPS as their previous other child. This can be accomplished using procedures such as an **amniocentesis** or chorionic villus sampling (CVS), after which parents can explore their options relating to the pregnancy.

Treatment

As of 2004 there was no cure for MPS, although several types of experimental therapies are being investigated in the early 2000s. Typically, treatment involves trying to relieve the symptoms and improve quality of life. For MPS I and VI, bone marrow transplantation has been attempted as a treatment option. For those types of MPS, bone marrow transplantation has sometimes helped slow down the progression or reverse some of symptoms of the disorder in some children. The benefits of bone marrow transplantation are more likely to be noticed when performed on children less than two years of age. However, bone marrow transplantation is not thought to be helpful in other MPS disorders. Availability of donors is limited, and as a result, very few bone marrow transplantations are done for MPS. There are risks as well as benefits with this procedure, and mortality resulting from the procedure is high.

Another experimental treatment for MPS I involves extended treatment with recombinant human alpha-L-iduronidase. Some individuals treated with this technique show an improvement in some symptoms. Additionally, there is ongoing research involving gene replacement therapy (the insertion of normal copies of a gene into the cells of patients whose gene copies are defective), although this was as of 2004 still highly experimental.

Prognosis

The course of this disorder varies with the specific type of MPS the individual has. MPS I H is often fatal in childhood, with individuals rarely living past age 12. Individuals with MPS I H/S may die in childhood or live to adulthood. Individuals with MPS I H have health problems but usually have a normal lifespan. Individuals with mild MPS II live relatively normal lives, while individuals with the severe form of the disorder usually die in their teens. The life expectancy in MPS III and MPS IV is also variable, depending on the severity of the disorder. Individuals with MPS VI often have shorter than average life spans. As of 2004 MPS IX had been diagnosed so recently that little information is available.

Prevention

No specific measures can prevent the gene mutations that cause MPS. For some of the MPS diseases, biochemical tests may be able to identify healthy individuals who are carriers of the defective gene, allowing them to make informed reproductive decisions. Prenatal testing can also diagnose MPS in the fetus, but this testing is normally done only when there is some reason to expect to find the disorder (e.g. family history of the disease).

Parental concerns

Many individuals with an MPS condition have problems with airway constriction. This constriction may be so serious as to create significant difficulties in administering general anesthesia. Therefore, it is recommended that surgical procedures be performed under local anesthesia whenever possible.

KEY TERMS

Cardiomyopathy—A disease of the heart muscle.

Enzyme—A protein that catalyzes a biochemical reaction without changing its own structure or function.

Joint contractures—Stiffness of the joints that prevents full extension.

Kyphosis—An extreme, abnormal outward curvature of the spine, with a hump at the upper back.

Lysosome—A membrane-enclosed compartment in cells, containing many hydrolytic enzymes, where large molecules and cellular components are broken down.

Mucopolysaccharide—A complex molecule made of smaller sugar molecules strung together to form a chain. It is found in mucous secretions and intercellular spaces.

Recessive gene—A type of gene that is not expressed as a trait unless inherited by both parents.

X-linked gene—A gene carried on the X chromosome, one of the two sex chromosomes.

Resources

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Kakkis, E. D., et al. "Enzyme-Replacement Therapy in Mucopolysaccharidosis I." *The New England Journal of Medicine* 344 (2001): 182–8.

ORGANIZATIONS

National MPS Society. PO Box 736, Bangor, ME 04402–0736. Web site: <www.mpsspciety.org>.

National Organization for Rare Disorders Inc. 55 Kenosia Ave, PO Box 1968, Danbury, CT 06813–1968. Web site: <www.rarediseases.org>.

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Multicultural education/ curriculum

Definition

Multicultural education describes a system of instruction that attempts to foster cultural pluralism and acknowledges the differences between races and cultures. It addresses the educational needs of a society that contains more than one set of traditions, that is a mixture of many cultures.

Description

The goal of multicultural education is to help students understand and appreciate cultural differences and similarities and to recognize the accomplishments of diverse ethnic, racial, and socioeconomic groups. It is a practice that hopes to transform the ways in which students are instructed by giving equal attention to the contributions of all the groups in a society. Special focus may be placed on minority groups that have been underrepresented in the past. A multicultural curriculum strives to present more than one perspective of a cultural phenomenon or an historical event. The old American melting pot metaphor is challenged as no longer being valid. Adherents of multicultural educational theory believe that the idea that students should be Americanized, in reality, assumed they should conform to a white, Eurocentric cultural model. In its place, multiculturalists believe school curricula should embrace a whole host of voices that exist in multicultural U.S. society. Their belief is that this transformation in the methods of learning is a start in addressing inequities in U.S. society. They believe this is increasingly important because of the changing population mix in the United States. For example, demographers estimate that by the year 2020, 46 percent of all public school students will be children of color.

The roots of multicultural education lie in the civil rights movements of various groups, including African Americans and women. In addition, the rise in ethnic consciousness and a more critical analysis of textbooks and other materials played a role. Community leaders, activists, and parents began to demand curricula that were more supportive and consistent with the cultural and racial diversity in the United States. In the late 1960s and 1970s, the concepts of multicultural education begin to emerge, and by the 1980s, an entire body of scholarship addressing multiculturalism existed.

One of the pioneers of multicultural education was James Banks, who believed all aspects of education needed to be transformed in order to create a multicultural school environment. These aspects include teaching methods, instructional materials, teacher attitudes, as well as the way the performance of students is assessed. Banks described five areas of multicultural education in which teachers and researchers are involved:

- Content integration: Concepts, values, and materials from a variety of cultures are included in teaching.
- Knowledge construction: This belief asserts that all knowledge is created in the minds of human beings and can, therefore, be challenged. A critical part of multicultural education, the idea that knowledge is a human construct challenges teachers to alter their own perceptions of the world before they can teach multiculturally.
- Equity pedagogy: Teachers must modify their methods of instruction by allowing for students' cultural differences before they can encourage academic achievement.
- Prejudice reduction: Teachers must work to shift students' prejudices regarding race and ethnicity. Prejudice reduction may also encompass teaching the tolerance of various religions, sexual preferences, and disabilities.
- Empowering school culture: Schools must identify those aspects of education that hinder learning and then empower families and students from all backgrounds, so that the full development of students is achieved.

Types of multicultural education programs

As of the early 2000s, there is no universally agreed upon multicultural curriculum. Teachers tend, however, to take one of two approaches. Some use what has been called the multicultural festival approach, in which students are invited to celebrate ethnic diversity by being exposed to foods, holidays, and festivals of other cultures. Many critics say that this conveys the notion that diversity is only important during celebratory moments. Other teachers apply a transformative approach, weaving different perspectives on cultures throughout the curriculum. Multicultural education can also be roughly divided into three different categories:

- Content-focused: These are the most common types of multicultural educational programs. Their overall objective is to include subject matter in the curriculum about various cultural groups in order to cultivate students' knowledge about these groups. Content may include holiday celebrations, recognizing heroes from different racial and ethnic groups, and focusing on the achievements of women and minorities. It may also include single-group studies, for example, black, ethnic, or women's studies programs.
- Student-focused: Many programs go beyond changes in the curriculum and specifically address the academic needs of defined groups of students, usually minorities. In this type of approach, the curriculum may not be changed significantly. Instead, the focus may be on aiding students in making the transition into the mainstream of education. Student-focused programs can take many forms, including efforts to draw on culturally-based learning styles and bilingual programs.
- Socially focused: These programs seek to reduce bias and increase cultural and racial tolerance. Included here might be desegregation programs, programs designed to increase contact among different races and cultures. Also, having teachers who are themselves members of minorities would be encouraged.

In spite of the fact that there are a variety of approaches to multicultural education, supporters point to several shared ideals among those who practice this kind of education. Shared ideals include:

- Each student must have equal opportunities to achieve his or her full potential.
- Every student must be able to participate in an increasingly multicultural society.
- Teachers must be able to facilitate learning for every student, no matter how similar or different each student is from the teacher.
- Schools must actively work towards ending oppression of all types, by ending it within their own walls.
- Education must include the voices and experiences of all students.

Common problems

There are many people who are either opposed to multicultural education or believe it has numerous problems. Some feel that the idea of multicultural education tends to divide cultures instead of building tolerance

KEY TERMS

Eurocentric—Centered or focused on Europe or European peoples, especially in relation to historical or cultural influence.

Multicultural education—A social or educational theory that encourages interest in many cultures within a society rather than in only a mainstream culture.

between them. They believe that American students should be taught to think of themselves as part of a whole rather than as people from different places who just happen to live in the same country.

Others believe multicultural education interferes with a child expressing his or her own individuality, by placing too much emphasis on ethnic or racial backgrounds. Even supporters recognize that someone's culture may be influenced as much by their sex or socioeconomic status as their race or ethnicity. Culture is itself complex and varies from community to community, **family** to family, or from person to person. The dynamic and variable nature of culture makes teaching about multiple cultural influences a daunting if not impossible task.

Critics also point out that educating students about the formation of U.S. democracy inevitably focuses on its European origins. If students are not informed that the dominant participants in the formation of the United States were white males, these critics say, students will not receive an accurate picture of U.S. history. In addition, there is the belief that if citizens are not willing to subordinate some parts of their heritage to the present set of dominant cultural values, then these citizens may find it even harder to integrate the mainstream.

Parental concerns

Parents should feel free to speak up about any concerns they have with the curriculum in their child's classroom. Multicultural education came about in part because parents expressed a need for the unique cultures of their children to be acknowledged and honored in school.

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Multiple endocrine neoplasia syndromes

Definition

The multiple endocrine neoplasia (MEN) syndromes are three related inherited disorders affecting the thyroid and other hormone producing (endocrine) glands of the body. Before the early 2000s, MEN was called familial endocrine adenomatosis.

Description

There are three types of MEN: MEN 1 (Wermer's syndrome), MEN 2A (Sipple syndrome), and MEN 2B (previously known as MEN 3). All MEN types are the result of inherited genetic mutations that predispose the individual to excessive growth of cells (hyperplasia) and tumor formation in multiple endocrine glands. For all types of MEN, the children of an affected individual have a 50 percent chance of inheriting the defective gene that causes the disorder.

Demographics

MEN 1 is uncommon and occurs in only about one of every 30,000 individuals. The disorder runs in families, and males are twice as likely to develop the disorder as females. Individuals with MEN 1 can show symptoms of excessive parathyroid secretion by age five, and almost all individuals with MEN 1 show parathyroid symptoms by age 40.

MEN 2 affects about one in every 40,000 individuals. MEN 2A is ten to 20 times more common than MEN 2B.

Causes and symptoms

MEN 1 is caused by a mutation at the PYGM gene on chromosome 11. PYGM is one of a group of genes known as tumor suppressor genes that help to control cell division. An individual who inherits one defective copy of a tumor suppressor gene from either parent has a strong likelihood of developing MEN 1, because there is a high probability of another mutation developing in the other copy of the PYGM gene at some point during the thousands of cell divisions that occur with growth and development. When a second mutation occurs, the cell that contains the mutation no longer has any normal copy of the tumor suppressor gene. When both copies are defective, tumor suppression fails and tumors develop.

As a result, individuals with MEN 1 have uncontrolled cell growth and develop tumors in several endocrine glands, including the parathyroid glands (80–95% of patients), the pancreas (about 50% of patients) and the pituitary (around 25% of patients). The most frequent symptom of MEN 1 is hyperparathyroidism, which is excessive growth of the parathyroid gland and excessive secretion of parathyroid hormone. This condition leads to increased amounts of calcium in the blood, kidney stones, weakened bones, and nervous system depression. Children with MEN 1 can show signs of hyperparathyroidism as young as age five.

Tumors of the pancreas, known as gastrinomas, are also common in MEN 1. Excessive secretion of gastrin (a hormone secreted into the stomach to aid in digestion) by these tumors can cause upper gastrointestinal ulcers. The anterior pituitary gland and the adrenal glands can also be affected. Unlike MEN 2, the thyroid gland is rarely involved in MEN 1 symptoms. Children with MEN1 rarely develop tumors of the pancreas until they reach adulthood.

There are two types of MEN 2. Both MEN 2A and MEN 2B are caused by mutations in another gene, known as RET. A mutation in only one copy of the RET gene is sufficient to cause disease. A number of different mutations can lead to MEN 2A, but only one specific genetic alteration causes MEN 2B.

Patients with both MEN 2A and MEN 2B experience two main symptoms, medullary thyroid **cancer** (MTC) and a tumor of the adrenal gland medulla known as pheochromocytoma. MTC is a slow-growing cancer, but one that can be cured in less than 50 percent of cases. Pheochromocytoma is usually a benign (noncancerous) tumor that causes excessive secretion of adrenal hormones. This, in turn, can cause life-threatening high blood pressure (**hypertension**) and irregular heart beat (cardiac arrhythmia).

The two forms of MEN 2 are distinguished by other symptoms. Individuals with MEN 2A have a predisposition to develop tumors of the parathyroid gland. Although similar to MEN 1, less than 20 percent of MEN 2A patients show parathyroid involvement.

Individuals with MEN 2B show a variety of additional conditions: a characteristic facial appearance with swollen lips; tumors of the mucous membranes of the eye, mouth, tongue, and nasal cavity; enlarged colon; and skeletal abnormalities. Symptoms develop early in life (often before five years of age) in cases of MEN 2B and the medullary thyroid cancer is much more aggressive and may develop in patients who are one year old.

When to call the doctor

Since MEN is inherited and runs in families, the doctor should be informed of this history when the child is born, so that genetic testing can be done immediately.

Diagnosis

In the past, classical diagnosis of MEN was based on clinical features and on testing for elevated hormone levels. For MEN 1, the relevant hormone was parathyroid hormone. For both types of MEN 2, the greatest concern is development of medullary thyroid cancer. MTC

can be detected by measuring levels of the thyroid hormone, calcitonin. Numerous other hormone levels can be measured to assess the involvement of the various other endocrine glands.

Diagnosis of MEN 2B can be made by physical examination alone. However, MEN 2A shows no distinct physical features and must be identified by measuring hormone levels or by finding endocrine tumors.

Since 1994, genetic screening using DNA technology has been available for both MEN 1 and MEN 2. This methodology allows diagnosis before the onset of symptoms. Before the development of genetic testing, there was no way to definitively identify which children had inherited the defective gene. As a result, all offspring of individuals with MEN had to be considered at risk. In the case of MEN 2A and MEN 2B, children would undergo frequent calcitonin testing. Molecular techniques as of the early 2000s allow a positive distinction to be made between children who are and are not carrying the defective genes that cause MEN.

Treatment

As of 2004 no comprehensive treatment is available for genetic conditions such as MEN. However, some of the consequences of MEN can be symptomatically treated. Pheochromocytoma in both types of MEN 2 can be cured by surgical removal of this slow growing tumor.

Treatment of MTC is by surgical removal of the thyroid. After thyroidectomy, the patient receives normal levels of thyroid hormone by mouth or by injection. Even when thyroid surgery is performed early, metastatic spread of the cancer may have already occurred. Since MTC is slow growing, metastasis may not be obvious. Metastasis is very serious in MTC because **chemotherapy** and radiation therapy are not effective in controlling its spread.

Prognosis

Diagnosed early through genetic testing, the prognosis for the MEN diseases is reasonably good, even for MEN 2B, the most dangerous of the three forms. Even in the absence of treatment, a few individuals with MEN 2A mutations never show any symptoms at all. Analysis of at-risk **family** members using molecular genetic techniques leads to earlier treatment and improved outcomes.

Prevention

As of 2004 there is no way to block the occurrence of genetic mutations that cause MEN. One of the most serious consequences of MEN is MTC. Children who are identified as carriers of the RET gene can be offered total thyroidectomy as a preventative (prophylactic) measure to prevent the development of MTC.

KEY TERMS

Adrenal glands—A pair of endocrine glands (glands that secrete hormones directly into the bloodstream) that are located on top of the kidneys. The outer tissue of the glands (cortex) produces several steroid hormones, while the inner tissue (medulla) produces the hormones epinephrine (adrenaline) and norepinephrine.

Endocrine—Refers to glands that secrete hormones circulated in the bloodstream or lymphatic system.

Medullary thyroid cancer—A slow-growing tumor associated with multiple endocrine neoplasia syndromes.

Neoplasm—An abnormal formation of new tissue. A neoplasm may be malignant or benign.

Pancreas—A five-inch-long gland that lies behind the stomach and next to the duodenum. The pancreas releases glucagon, insulin, and some of the enzymes which aid digestion.

Parathyroid gland—A pair of glands adjacent to the thyroid gland that primarily regulate blood calcium levels.

Parathyroid hormone—A chemical substance produced by the parathyroid glands. This hormone plays a major role in regulating calcium concentration in the body.

Pheochromocytoma—A tumor that originates from the adrenal gland's chromaffin cells, causing overproduction of catecholamines, powerful hormones that induce high blood pressure and other symptoms.

Pituitary gland—The most important of the endocrine glands (glands that release hormones directly into the bloodstream), the pituitary is located at the base of the brain. Sometimes referred to as the "master gland," it regulates and controls the activities of other endocrine glands and many body processes including growth and reproductive function. Also called the hypophysis.

Thyroid gland—An endocrine gland in the neck overlying the windpipe (trachea) that regulates the speed of metabolic processes by producing a hormone, thyroxin.

Parental concerns

MEN is an inherited disorder. Individuals who have MEN in their families may wish to get genetic counseling before attempting a pregnancy.

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Multiple pregnancy

Definition

Multiple pregnancy, usually referred to as multiple gestation, is one in which more than one fetus develops simultaneously in the mother's womb.

Description

The frequency of multiple births in the United States has been steadily increasing with advances in reproductive technologies. It is estimated that pregnancies resulting from assisted technologies have a 25–30 percent incidence of **twins** and a 5 percent incidence of triplets. The frequency of naturally occurring twins is approximately one in 80 births; however the frequency of multiple births in the United States for 2002 was as follows:

- twins, one in 32
- triplets, one in 583
- quadruplets, one in 9,267
- quintuplets and up, one in 58,286

Twin birth is by far the most common multiple birth. There are two types of twin pregnancy: fraternal and identical. Fraternal twins develop from two separate ova released at the same time and fertilized by two separate sperm. Fraternal twins are referred to as dizygotic twins, meaning that two unions of two gametes or male/female sex cells occurred to produce two separate embryos. Characteristically, with fraternal twins, each has its own placenta and amniotic sac. They may be the same or different sex, occur twice as frequently as identical twins, and have a mortality rate of 11.5 percent. Identical twins represent the splitting of a single fertilized zygote (union of two gametes or male/female sex cells to produce a developing embryo) into two separate individuals. Identical twins will have the same DNA, genetic material (genotype), but it may be expressed differently (phenotype). There are three ways identical twins can exist in the uterus: dichorionic-diamniotic twins; monochorionic-diamniotic twins; monochorionic-monoamniotic twins. In the instance of dichorionic-diamniotic twins, division of the fertilized egg occurs within 72 hours past fertilization, before the inner cell mass has developed. About 30 percent of identical twins have this classification, and each twin has its own chorion, amnion, and placenta. The mortality rate for this type of twinning is 9 percent. With monochorionic-diamniotic twins, division occurs in the range of four to eight days after fertilization, and the inner cell mass divides in two. The placenta has one chorion and two amnions, so each twin has its own amniotic sac. Approximately 68 percent of identical twins are in this classification, and they have a mortality rate of 25 percent. Thirdly, monochorionic-monoamniotic twins are contained in the same amniotic sac. The division of the fertilized egg in this case occurs nine to 13 days past fertilization or near the time of implantation in the uterus. Since they share an amniotic sac, they have an increased risk of their umbilical cords becoming entangled or knotted. Only 2 percent of identical twins are in this classification, and they have a mortality rate of greater than 50 percent. If a complete separation does not take place during the division process, the result is Siamese (or conjoined) twins.

The human female typically releases only one egg every menstrual cycle. A hormone called progesterone, released by the first egg to be produced, prevents any other egg from maturing during that cycle. When this control fails, fertilization of more than one egg is possible. Fertility drugs inhibit these controls, allowing multiple gestation to occur. It seems as if, however, that more pregnancies start out naturally with twins than was originally believed. The development of improved technology, such as ultrasound, has made it possible to determine more accurately the early pregnancy loss rate of

twins to include both complete pregnancy loss and spontaneous resorption of one twin, frequently referred to as the vanishing twin phenomenon. Recent research suggests that 75 percent of twin pregnancies are lost before the end of the first trimester. Moreover, only about 50 percent of pregnancies diagnosed in the first trimester with twins result in the birth of two live infants.

An old adage related to multiple gestation is the human female was not meant to have more than twins because she only had two breasts for feeding. Of course, pregnancies with more than two babies have occurred throughout history. However, once the number of babies reaches three, overexpansion of a woman's uterus begins to cause difficulties. The implantation of several embryos and placentas in the endometrium of the uterus results in a competition for space and inevitably some implant in an area without good circulation. During a pregnancy, it is essential that the uterus be well perfused to sustain the fetus with nutrients and oxygen. A lack of oxygen can cause central nervous system damage in the fetuses that implanted in a less than desirable area. Since the human female was not made to carry an indefinite number of fetuses, multiple gestations can have many of the following complications:

- increased rate of spontaneous abortion
- two to three times greater risk of developing severe **hypertension** or preeclampsia (increased blood pressure)
- maternal anemia due to increased fetal demands
- premature rupture of membranes (bag of water)
- incompetent cervix (cervix opens due to pressure)
- intrauterine growth restriction of one or more fetuses
- preterm labor due to overstretched uterus
- abnormal fetal presentations
- need for **cesarean section**
- rare complications with twins, such as twin-to-twin-transfusion syndrome (one fetus receives more nutrients than the other due to more blood vessels perfusing one baby)
- conjoined twins
- postpartum hemorrhage

Causes and symptoms

Twinning seems to run in some families, is mainly confined to fraternal (dizygotic twins) and seems to be entirely a property of the mother, not the father. The primary cause is an increased chance of multiple ovulation, when a woman releases two or more eggs. Another major factor is maternal age; a woman who gives birth at 37 is four times more likely to have fraternal twins than at age 18. The 37-year-old is also more likely to be unable to conceive, since many women's ovaries are already starting to fail at that age. The third major factor is race; West Africans are ten times more likely to have fraternal twins than Chinese or Japanese, with Caucasians intermediate. This increased chance is also seen in African Americans. In addition, the more pregnancies a woman has had, the greater her chances of having twins. In fact, by the fourth or fifth pregnancy, the likelihood of having twins is four times higher than it was for the first pregnancy.

The use of assisted reproduction techniques, particularly ovarian stimulation, has caused a dramatic increase in the number of twin and higher multiple births. The normal process of single ovulation is interrupted because fertility drugs permit more than one egg at a time to mature and be released. The first drug to be used for this was clomiphene (Clomid). This was followed by the development of two natural hormones, follicle-stimulating hormone and chorionic gonadotrophin (Pergonal) to produce multiple eggs ovulation. The chance of multiple gestation with in vitro fertilization (IVF) is about the same as with the use of fertility drugs, because several embryos are inserted into the womb to increase the odds of conception. Similarly, other fertility techniques such as gamete intrafallopian transfer (GIFT) and zygote intrafallopian transfer (ZIFT) are also more likely to result in multiple gestations. The use of intrauterine insemination or artificial insemination (the injection of sperm into a woman's uterus with a syringe) is the only fertility treatment that does not increase the chances of conceiving multiples—of course, the woman is usually taking fertility drugs with this procedure also.

Diagnosis

If a multiple pregnancy occurred spontaneously, the obstetrician would suspect a problem with the dates because the uterus would grow faster than usual. The gestational age of a pregnancy is determined from the first day of the last menstrual period (LMP). In a multiple gestation, the uterine measurements would be larger than dates, which normally correspond. If multiple gestation is suspected, an ultrasound may be performed to determine the gestational age of the fetus or to check for more than one fetus. With the use of assisted reproductive technology, an ultrasound is usually performed with ten days to see if any of the embryos were successful with implantation, and a multiple gestation would be revealed at that time. Following the birth of multiples, the placenta is carefully examined to determine if they are fraternal or identical. One placenta indicates identical twins.

A multiple pregnancy almost always means increased monitoring and surveillance for complications. This often means more frequent visits to the healthcare provider, serial ultrasounds to make sure that the babies are growing satisfactorily, **amniocentesis** to check for lung development, and close monitoring for preterm labor.

Treatment

Ultrasound examinations play an important role in the care and treatment of multiple gestations. It assists with dating the pregnancy, determining the number of fetuses, detecting fetal anomalies, following the growth and development of each fetus, and serves to monitor the length of the cervix in anticipation of preterm labor. Premature birth is the constant threat of multiple gestation, and the primary threat of **prematurity** is related to lung development. Premature infants lack a substance, called surfactant, that permits their lungs to expand and breathe normally. If it becomes apparent that a multiple gestation is going to have a preterm delivery, the mother will be given an injection of a steroid, beta-methasone, to help the lungs mature. It is more beneficial if the steroid can be given twice in a 48 hour period; however, if there is not time for this, surfactant has been developed that can be administered into the lungs of a premature infant to facilitate breathing. An additional problem with the premature infant involves the lack of body fat. In a normal pregnancy the fetus spends the last four to six weeks growing and gaining weight, primarily body fat. This fat helps a newborn maintain his or her body temperature. Since premature infants do not have this fat, they use energy they cannot afford to stay warm.

Although research has shown that bed rest is not effective, a woman is often placed on bed rest during a multiple pregnancy to try to prevent pre-term labor and delivery. If preterm labor is impossible to control at home, the mother may be hospitalized and medication used to attempt to control contractions and dilatation of the cervix. Multiple gestations greater than twins in number are almost always delivered via cesarean section.

Alternative treatment

There are no specific treatments to alleviate medical difficulties caused by multiple pregnancies; however, there are supportive measures that may help both mother and children recover from the birthing process. There are treatments to encourage breast milk production and to combat postpartum difficulties. Various homeopathic remedies and massage can be helpful to both mother and children during the early adjustment period after birth.

KEY TERMS

Amnion—Thin, tough, innermost layer of the amniotic sac.

Amniotic membrane—The thin tissue that creates the walls of the amniotic sac.

Chorion—The outer membrane of the amniotic sac. Chorionic villi develop from its outer surface early in pregnancy. The villi establish a physical connection with the wall of the uterus and eventually develop into the placenta.

Gene—A building block of inheritance, which contains the instructions for the production of a particular protein, and is made up of a molecular sequence found on a section of DNA. Each gene is found on a precise location on a chromosome.

Gestation—The period from conception to birth, during which the developing fetus is carried in the uterus.

Ova—The plural of ovum, it is the female reproductive cell.

Ovulate—To release a mature egg for fertilization.

Placenta—The organ that provides oxygen and nutrition from the mother to the unborn baby during pregnancy. The placenta is attached to the wall of the uterus and leads to the unborn baby via the umbilical cord.

Sequela—Plural, sequalae. An abnormal condition resulting from a previous disease or disorder.

Zygote—The result of the sperm successfully fertilizing the ovum. The zygote is a single cell that contains the genetic material of both the mother and the father.

Prognosis

Many multiple pregnancies reach fruition without difficulties; however, many do not. Despite medical advances, if the babies are born too early, they may survive but will have sequelae that limit the quality of life. If the babies are born prematurely, immediate medical care increases the chance of survival without any complications.

Parental concerns

Mothers with multiple pregnancy should be especially careful to get adequate prenatal care, including any necessary **vitamins** or recommended tests. Because

of the extra stress on the mother's body, increased rate of complications, and threat of prematurity, the mother should be vigilant in making sure she gets enough rest, reduces stress, and maintains a healthy diet.

See also Antepartum testing; Cesarean section.

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Mumps

Definition

Mumps is a relatively mild short-term viral infection of the salivary glands that usually occurs during childhood.

Description

Typically, mumps is characterized by a painful swelling of both cheek areas, although the person could have swelling on one side or no perceivable swelling at all. The salivary glands are also called the parotid glands; therefore, mumps is sometimes referred to as an inflammation of the parotid glands (epidemic parotitis). The word mumps comes from an old English dialect, meaning lumps or bumps within the cheeks.

Demographics

Mumps is a very contagious infection that spreads easily in such highly populated environments as daycare centers and schools. Although not as contagious as **measles** or **chickenpox**, mumps was once quite common. Prior to the release of a mumps vaccine in the United States in 1967, approximately 92 percent of all children had been exposed to mumps by the age of 15. In the pre-vaccine years, most children contracted mumps between the ages of four and seven. Mumps epidemics came in two to five year cycles. The greatest mumps epidemic was in 1941 when approximately 250 cases were reported for every 100,000 people. In 1968, the year after the live mumps vaccine was released, only 76 cases were reported for every 100,000 people. By 1985, fewer than 3,000 cases of mumps were reported throughout the entire United States, the equivalent of about one case per 100,000 people. The reason for the decline in mumps was the increased usage of the mumps vaccine. However, 1987 noted a five-fold increase in the incidence of the disease because of the reluctance of some states to adopt comprehensive school immunization laws. After that, state-enforced school entry requirements achieved student immunization rates of nearly 100 percent in kindergarten and first grade. In 1996, the Centers for Disease Control and Prevention (CDC) reported only 751 cases of mumps nationwide, that is, about one case for every 5 million people.

Causes and symptoms

The paramyxovirus that causes mumps is harbored in the saliva and is spread by sneezing, coughing, and other direct contact with another person's infected saliva. Once the person is exposed to the virus, symptoms generally occur in 14 to 24 days. Initial symptoms include chills, **headache**, loss of appetite, and a lack of energy. However, an infected person may not experience these initial symptoms. Swelling of the salivary glands in the face (parotitis) generally occurs within 12 to 24 hours of the above symptoms. Accompanying the swollen glands is **pain** on chewing or swallowing, especially with acidic

beverages, such as lemonade. A **fever** as high as 104°F (40°C) is also common. Swelling of the glands reaches a maximum on about the second day and usually disappears by the seventh day. Once individuals have contracted mumps, they become immune to the disease, despite how mild or severe their symptoms may have been.

While the majority of cases of mumps are uncomplicated and pass without incident, some complications can occur. Complications are, however, more noticeable in adults who get the infection. In 15 percent of cases, the covering of the brain and spinal cord becomes inflamed (**meningitis**). Symptoms of meningitis usually develop within four or five days after the first signs of mumps. These symptoms include a stiff neck, headache, **vomiting**, and a lack of energy. Mumps meningitis is usually resolved within seven days, and damage to the brain is exceedingly rare.

The mumps infection can spread into the brain causing inflammation of the brain (**encephalitis**). Symptoms of mumps encephalitis include the inability to feel pain, seizures, and high fever. Encephalitis can occur during the parotitis stage or one to two weeks later. Recovery from mumps encephalitis is usually complete, although complications, such as seizure disorders, have been noted. Only about one person in 100 with mumps encephalitis dies from the complication.

About one-fourth of all post-pubertal males who contract mumps can develop a swelling of the scrotum (orchitis) about seven days after the parotitis stage. Symptoms include marked swelling of one or both testicles, severe pain, fever, **nausea**, and headache. Pain and swelling usually subside after five to seven days, although the testicles can remain tender for weeks.

Girls occasionally suffer an inflammation of the ovaries (oophoritis) as a complication of mumps, but this condition is far less painful than orchitis in boys.

Diagnosis

When mumps reaches epidemic proportions, diagnosis is relatively easy on the basis of the physical symptoms. The doctor will take the child's temperature, gently palpate (touch) the skin over the parotid glands, and look inside the child's mouth. If the child has mumps, the openings to the ducts inside the mouth will be slightly inflamed and have a "pouty" appearance. With so many people vaccinated as of the early 2000s, a case of mumps must be properly diagnosed in the event the salivary glands are swollen for reasons other than viral infection. For example, in persons with poor **oral hygiene**, the salivary glands can be infected with bacteria. In these cases, **antibiotics** are necessary. Also in rare cases, the salivary glands can become blocked, develop tumors, or swell due to the use of certain drugs, such as iodine. A test can be performed to determine whether the person with swelling of the salivary glands actually has the mumps virus.

In late 2002, researchers in London reported the development of a bioassay for measuring mumps-specific IgG. This test would allow a doctor to check whether an individual patient is immune to mumps and allow researchers to measure the susceptibility of a local population to mumps in areas with low rates of **vaccination**.

Treatment

When mumps does occurs, the illness is usually allowed to run its course. The symptoms, however, are treatable. Because of difficulty swallowing, the most important challenge is to keep the patient fed and hydrated. The individual should be provided a soft diet, consisting of cooked cereals, mashed potatoes, broth-based soups, prepared baby foods, or foods put through a home food processor. Aspirin (only for individuals over the age of 20), **acetaminophen**, or ibuprofen can relieve some of the pain due to swelling, headache, and fever. Patients should void fruit juices and other acidic foods or beverages that can irritate the salivary glands. They should also avoid dairy products that can be hard to digest. In the event of complications, a physician should be contacted at once. For example, if orchitis occurs, a physician should be called. Also, supporting the scrotum in a cotton bed on an adhesive-tape bridge between the thighs can minimize tension. Ice packs are also helpful.

Prognosis

When mumps is uncomplicated, prognosis is excellent. However, in rare cases, a relapse occurs after about two weeks. Complications can also delay complete recovery.

Prevention

A vaccine exists to protect against mumps. The vaccine preparation (MMR) is usually given as part of a combination injection that helps protect against measles, mumps, and **rubella**. MMR is a live vaccine administered in one dose between the ages of 12 and 15 months, between four and six years of age, or 11 and 12 years of age. Persons who are unsure of their mumps history and/or mumps vaccination history should be vaccinated. Susceptible healthcare workers, especially those who work in hospitals, should be vaccinated. Because mumps is

A young child with mumps. *(Photo Researchers, Inc.)*

still prevalent throughout the world, susceptible persons over the age of one year who are traveling abroad would benefit from receiving the mumps vaccine.

The mumps vaccine is extremely effective, and virtually everyone should be vaccinated against this disease. There are, however, a few reasons why people should not be vaccinated against mumps:

- Pregnant women who contract mumps during pregnancy have an increased rate of miscarriage but not birth defects. As a result, pregnant women should not receive the mumps vaccine because of the possibility of damage to the fetus. Women who have had the vaccine should postpone pregnancy for three months after being vaccinated.
- Unvaccinated persons who have been exposed to mumps should not get the vaccine, as it may not provide protection. The persons should, however, be vaccinated if no symptoms result from the exposure to mumps.
- Persons with minor fever-producing illnesses, such as an upper respiratory infection, should not get the vaccine until the illness has subsided.
- Because mumps vaccine is produced using eggs, individuals who develop **hives**, swelling of the mouth or throat, **dizziness**, or breathing difficulties after eating eggs should not receive the mumps vaccine.
- Persons with immune deficiency diseases and/or those whose immunity has been suppressed with anti-cancer drugs, corticosteroids, or radiation should not receive the vaccine. **Family** members of immunocompromised people, however, should get vaccinated to reduce the risk of mumps.
- The CDC recommends that all children infected with human **immunodeficiency** disease (HIV) who are asymptomatic should receive an the **MMR vaccine** at 15 months of age.

KEY TERMS

Asymptomatic—Persons who carry a disease and are usually capable of transmitting the disease but who do not exhibit symptoms of the disease are said to be asymptomatic.

Autism—A developmental disability that appears early in life, in which normal brain development is disrupted and social and communication skills are retarded, sometimes severely.

Encephalitis—Inflammation of the brain, usually caused by a virus. The inflammation may interfere with normal brain function and may cause seizures, sleepiness, confusion, personality changes, weakness in one or more parts of the body, and even coma.

Epidemic parotitis—The medical name for mumps.

Immunoglobulin G (IgG)—Immunoglobulin type gamma, the most common type found in the blood and tissue fluids.

Meningitis—An infection or inflammation of the membranes that cover the brain and spinal cord. It is usually caused by bacteria or a virus.

Orchitis—Inflammation of one or both testes, accompanied by swelling, pain, fever, and a sensation of heaviness in the affected area.

Paramyxovirus—A genus of viruses that includes the causative agent of mumps.

Parotitis—Inflammation and swelling of one or both of the parotid salivary glands.

Parental concerns

The mumps vaccine has been controversial in the early 2000s because of concern that its use was linked to an increased rate of childhood **autism**. The negative publicity given to the vaccine in the mass media led some parents to refuse to immunize their children with the MMR vaccine. One result has been an increase in the number of mumps outbreaks in several European countries, including Italy and the United Kingdom.

In the fall of 2002, the *New England Journal of Medicine* published a major Danish study disproving the hypothesis of a connection between the MMR vaccine

and autism. A second study in Finland showed that the vaccine is also not associated with aseptic meningitis or encephalitis. Since these studies were published, U.S. primary care physicians have once again reminded parents of the importance of immunizing their children against mumps and other childhood diseases.

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Munchausen syndrome

Definition

Munchausen syndrome is a psychiatric disorder that causes an individual to self-inflict injury or illness or to fabricate symptoms of physical or mental illness in order to receive medical care or **hospitalization**. In a variation of the disorder, Munchausen by proxy (MSBP), an individual, typically a mother, intentionally causes or fabricates illness in a child or other person under her care.

Description

Munchausen syndrome takes its name from Baron Karl Friederich von Munchausen, an eighteenth century German military man known for his tall tales. The disorder first appeared in psychiatric literature in the early 1950s when it was used to describe patients who sought hospitalization by inventing symptoms and complicated medical histories, and/or inducing illness and injury in themselves. Categorized as a factitious disorder (a disorder in which the physical or psychological symptoms are under voluntary control), Munchausen syndrome seems to be motivated by a need to assume the role of a patient. Unlike malingering, there does not seem to be any clear secondary gain (e.g., money) in Munchausen syndrome.

Individuals with Munchausen by proxy syndrome use their child (or another dependent person) to fulfill their need to step into the patient role. The disorder most commonly victimizes children from birth to eight years old. Parents or caregivers with MSBP may only exaggerate or fabricate their child's symptoms, or they may deliberately induce symptoms through various methods, including **poisoning**, suffocation, starvation, or introducing bacteria into open **wounds**. They often display an extraordinary depth of medical knowledge and may even be in the medical profession themselves.

Demographics

Both Munchausen syndrome and Munchausen syndrome by proxy are thought to be rare, but there are no solid statistics on the frequency of either diagnosis. Data on Munchausen syndrome in children and adolescents specifically are very limited. In 2000 one review found that among the 42 cases reported in the medical literature, 71 percent were female and the mean age was 14 years of age. Children age 14 and younger were more likely to admit to falsifying symptoms when confronted than those between the ages of 15 and 18.

Munchausen syndrome by proxy is also hard to quantify due to the number of undetected or undiagnosed cases. The incidence of the condition in the United States is not known, but a 1996 study of children in Ireland and the United Kingdom estimated that Munchausen syndrome by proxy occurred annually in 0.5 of every 100,000 children under age 16, and in 2.8 of every 100,000 children under the age of one.

Causes and symptoms

The exact cause of Munchausen syndrome is unknown. It has been theorized that Munchausen patients are motivated by a desire to be cared for, a need for attention, dependency, an ambivalence toward doctors, or a need to suffer. Factors that may predispose an individual to Munchausen include a serious illness in childhood or an existing personality disorder. Some research indicates that children and adolescents who develop Munchausen syndrome are more likely to have been previous victims of Munchausen syndrome by proxy.

The Munchausen and Munchausen by proxy patient can appear to have a wide array of physical or psychiatric symptoms, usually limited only by their (or their caregiver's) medical knowledge. Many Munchausen patients are very familiar with medical terminology and symptoms. Some common complaints include fevers, **rashes**, abscesses, bleeding, and **vomiting**. Common Munchausen by proxy symptoms include apnea (cessation of breathing), **fever**, vomiting, and **diarrhea**. In both Munchausen and MSBP syndromes, the suspected illness does not respond to a normal course of treatment, and

diagnostic tests turn up nothing out of the ordinary. Patients or parents may push for invasive procedures and display an extraordinary depth of knowledge of medical therapies.

Diagnosis

Because Munchausen sufferers often go from doctor to doctor, gaining admission into many hospitals along the way, diagnosis can be difficult. They are typically detected rather than diagnosed. During a course of treatment, they may be discovered by a hospital employee who encountered them during a previous hospitalization. Their caregivers may also notice that symptoms such as high fever occur only when the patient is left unattended. Occasionally, medication used to induce symptoms is found with the patient's belongings. When the patient is confronted, they often react with outrage and check out of the hospital to seek treatment at another facility with a new caregiver.

A diagnosis of Munchausen syndrome may be even more difficult in children and adolescents. A physician may be able to recognize a pattern of symptoms (e.g., those that occur only when the child is alone or that begin only when the parent is present with the child) or the child may admit to fabricating or self-inflicting symptoms upon questioning. Surveillance video may record the child or the child's caregiver inducing symptoms.

Treatment

There is no clearly effective treatment for Munchausen syndrome. Extensive psychotherapy may be helpful with some Munchausen patients. If Munchausen syndrome coexists with other mental disorders, such as a personality disorder, the underlying disorder is typically treated first. Children who develop the syndrome may respond more favorably to therapy than adults, particularly if they are diagnosed at an early age.

Children who are victims of Munchausen syndrome by proxy are usually removed from the offending caregiver immediately and placed in protective custody. Therapy may also be beneficial to these children in recovering from the emotional trauma of MSBP.

Prognosis

The infections and injuries Munchausen patients self-inflict can cause serious illness. Patients often undergo countless unnecessary surgeries throughout their lifetimes. In addition, because of their frequent hospitalizations, they have difficulty holding down a job. Further, their chronic health complaints may damage interpersonal relationships with **family** and friends.

Children victimized by sufferers of MSBP are at a real risk for serious injury and possible death. A UK study published in 1998 found that although the majority of children with MSBP studied (90 percent) were placed in child protection care at diagnosis, at two-year follow up the number had fallen to 32 percent. A reported 17 percent of children who were victims of MSBP and who were eventually returned to an abusive caregiver suffered further abuse. Those who survive physically unscathed may suffer developmental and emotional problems.

KEY TERMS

Apnea—The temporary absence of breathing. Sleep apnea consists of repeated episodes of temporary suspension of breathing during sleep.

Factitious disorder—A mental condition in which symptoms are deliberately manufactured by patients in order to gain attention and sympathy. Patients with factitious diseases do not fake symptoms for obvious financial gain or to evade the legal system.

Malingering—Pretending to be sick in order to be relieved of an unwanted duty or obtain some other obvious benefit.

Prevention

Because the cause of Munchausen syndrome is unknown, formulating a prevention strategy is difficult. Some medical facilities and healthcare practitioners have attempted to limit hospital admissions for Munchausen patients by sharing medical records. While these attempts may curb the number of hospital admissions, they do not treat the underlying disorder and may endanger Munchausen sufferers that have made themselves critically ill and require treatment. Children who are found to be victims of persons with Munchausen by proxy syndrome should be immediately removed from the care of the abusing parent or guardian.

Parental concerns

Parents who suspect that their child may be deliberately hurting themselves or falsifying symptoms should contact their pediatrician immediately for assessment. Children who are thought to pose potentially life-threatening

danger to themselves may require hospitalization, and a referral to a child psychologist or therapist will be necessary. It is important to remember that properly treating the condition requires addressing the motives and emotions behind the disorder, not simply punishing the behavior.

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American Psychological Association (APA). 750 First St. NE, Washington, DC 20002–4242. Web site: <www.apa.org>.

National Alliance for the Mentally Ill (NAMI). Colonial Place Three, 2107 Wilson Blvd., Ste. 300, Arlington, VA 22201–3042. Web site: <www.nami.org>.

National Institute of Mental Health (NIMH). Office of Communications, 6001 Executive Boulevard, Room 8184, MSC 9663, Bethesda, MD 20892–9663. Web site: <www.nimh.nih.gov>.

Paula Ford-Martin

Muscle contractures *see* **Spasticity**

Muscle spasms and cramps

Definition

Muscle spasms and cramps are spontaneous, often painful muscle contractions.

Description

The rapid, uncontrolled muscle contraction, or spasm, happens unexpectedly, with either no stimulation or some trivially small one. The muscle contraction and **pain** last for several minutes and then slowly ease. Cramps may affect any muscle but are most common in the calves, feet, and hands. While painful, they are harmless and, in most cases, not related to any underlying disorder. Nonetheless, cramps and spasms can be manifestations of many neurological or muscular diseases.

The terms cramp and spasm can be somewhat vague, and they are sometimes used to include types of abnormal muscle activity other than sudden painful contraction. These include stiffness at rest, slow muscle relaxation, and spontaneous contractions of a muscle at rest (fasciculation). Fasciculation is a type of painless muscle spasm, marked by rapid, uncoordinated contraction of many small muscle fibers. A critical part of diagnosis is distinguishing these different meanings and allowing the patient to describe the problem as precisely as possible.

Demographics

The exact incidence of muscle cramps and spasms is not known. They are more likely to occur in older children and teenagers who are participating in organized, competitive **sports** and strenuous aerobic activities.

Causes and symptoms

Causes

Normal voluntary muscle contraction begins when electrical signals are sent from the brain through the spinal cord along nerve cells called motor neurons. These include both the upper motor neurons within the brain and the lower motor neurons within the spinal cord and leading out to the muscle. At the muscle, chemicals released by the motor neuron stimulate the internal release of calcium ions from stores within the muscle cell. These calcium ions then interact with muscle proteins within the cell, causing the proteins (actin and myosin) to slide past one another. This motion pulls their fixed ends closer, thereby shortening the cell and, ultimately, the muscle itself. Recapture of calcium and unlinking of actin and myosin allow the muscle fiber to relax.

Abnormal contraction may be caused by unusual activity at any stage in this process. Certain mechanisms within the brain and the rest of the central nervous system help regulate contraction. Interruption of these mechanisms can cause spasm. Motor neurons that are

overly sensitive may fire below their normal thresholds. The muscle membrane itself may be overly sensitive, causing contraction without stimulation. Calcium ions may not be recaptured quickly enough, causing prolonged contraction.

Structural disorders such as flat feet, hyperextended knees (genu recurvatum), and hypermobility syndrome (joints that can move beyond the normal range of motion) may predispose a person to developing leg cramps. Prolonged sitting, inappropriate leg positioning during sedentary activity, or standing on concrete flooring for prolonged periods may be associated with an increased incidence of leg cramps.

Interruption of brain mechanisms and overly sensitive motor neurons may result from damage to the nerve pathways. Possible causes include **stroke**, multiple sclerosis, **cerebral palsy**, neurodegenerative diseases, trauma, **spinal cord injury**, and nervous system poisons such as strychnine, **tetanus**, and certain insecticides. Nerve damage may lead to a prolonged or permanent muscle shortening called contracture.

Changes in muscle responsiveness may be due to or associated with the following:

- Prolonged **exercise**: Curiously, relaxation of a muscle actually requires energy to be expended. The energy is used to recapture calcium and to unlink actin and myosin. Normally, sensations of pain and fatigue signal that it is time to rest. Ignoring or overriding those warning signals can lead to such severe energy depletion that the muscle cannot be relaxed, causing a cramp. The familiar advice about not swimming after a heavy meal, when blood flow is directed away from the muscles, is intended to avoid this type of cramp.
- Exercising or participating in activities in high or humid temperatures: Copious sweating during prolonged exercise can lead to heat cramps, a condition associated with brief, painful cramps, especially in the legs, sweating, and mild **fever**, usually less than 102°F. Heat cramps are more likely to occur when the child has not taken in enough fluids before, during, and after the activity. Exercising in high temperatures without adequate fluid intake may increase the risk of **dehydration**.
- Dehydration and salt depletion: This condition may be brought on by repeated bouts of **vomiting** or **diarrhea** or by copious sweating during prolonged exercise. Loss of fluids, salts, and minerals—especially sodium, potassium, magnesium, and calcium—can disrupt ion balances in both muscle and nerves. This imbalance can prevent the muscles and nerves from responding and recovering normally and can lead to cramping.
- Metabolic disorders that affect the energy supply in muscle: These are inherited diseases in which particular muscle enzymes are deficient. They include deficiencies of myophosphorylase (McArdle's disease), phosphorylase b kinase, phosphofructokinase, phosphoglycerate kinase, and lactate dehydrogenase.
- Myotonia: Myotonias include **myotonic dystrophy**, myotonia congenita, paramyotonia congenita, and neuromyotonia. These conditions cause stiffness due to delayed relaxation of the muscle but do not cause the spontaneous contraction usually associated with cramps. However, many patients with myotonia do experience cramping from exercise. Symptoms of myotonia are often worse in cold temperatures.

Fasciculation may be due to fatigue, cold, medications, metabolic disorders, nerve damage, or neurodegenerative disease, including amyotrophic lateral sclerosis (ALS, also known as Lou Gehrig's disease). Most people experience brief, mild fasciculation from time to time, usually in the calves.

Symptoms

The pain of a muscle cramp is intense, localized, and often debilitating. Coming on quickly, it may last for minutes and fade gradually. Contractures develop more slowly, over days or weeks, and may be permanent if untreated. Fasciculation may occur at rest or after muscle contraction and may last several minutes.

Exercising in high temperatures can lead to dehydration. Dehydration should be suspected if these symptoms are present: dry mouth or tongue, increased or excessive thirst, few or no tears when crying, decreased urination, dark yellow urine, irritability, low energy, lightheadedness or fainting, severe weakness, and sunken abdomen, eyes and cheeks.

When to call the doctor

Prompt medical attention is required if the child has any of the symptoms associated with dehydration, as listed above. Prompt medical attention also is required if the child has a high fever—temperature of 102°F or 38.9°C, or above. Parents also should call the child's pediatrician if the following symptoms are present:

- acute pain associated with the muscle cramp or spasm
- prolonged muscle contractions
- cramps or spasms that cause twisting and repetitive movement or abnormal posture
- apparent development of muscle contractures (prolonged joint flexion in an abnormal position)

Diagnosis

A usual bout of muscle cramps should not require a visit to the doctor. However, medical treatment is essential if the child has any symptoms of dehydration associated with the muscle cramps. In addition, any abnormal contractions or frequent muscle cramps or spasms that cause concern should be evaluated by a physician. Abnormal muscle contractions are diagnosed through a careful medical history, as well as a physical and neurological examination. In some cases when a structural abnormality is suspected, x rays may be performed.

The medical history helps the physician evaluate the presence of other conditions or disorders that might contribute to or cause the abnormal contractions. Records of previous diagnoses, surgeries, and treatments are reviewed. The child's **family** medical history is evaluated to determine if there is a history of muscular or neurological disorders.

Questions about the child's medical history may include:

- When were the symptoms first noticed?
- How long have the symptoms lasted?
- Are the symptoms always present?
- What muscles are affected?
- What makes the symptoms improve?
- What specific treatments or techniques have been tried?
- What makes the symptoms worse?
- Do certain activities, emotions, or events seem to aggravate the symptoms?
- Are other symptoms present?

The physical and neurological exams may include an evaluation of the child's motor reflexes including muscle tone, mobility, strength, balance, and endurance; heart and lung function; cranial nerve function; and an examination of the child's abdomen, spine, throat, and ears. The child's height and weight and blood pressure also are checked and recorded.

When a neurological cause is suspected, a multi-disciplinary team may be consulted to provide an accurate diagnosis, so the proper treatment can be planned. Occupational and physical therapy evaluations may be helpful to determine upper and lower extremity movement patterns and passive range of motion.

In some cases, nerve conduction studies with electromyography of the affected muscles may be performed to evaluate an underlying neuromuscular disorder. These tests are useful in evaluating a child's muscular activity and provide a comprehensive **assessment** of nerve and muscle function.

In both tests, the examiner uses a computer, monitor, amplifier, loudspeaker, stimulator, and high-tech filters to see and hear how the muscles and nerves are responding during the test. In the nerve conduction study, small electrodes are placed on the skin over the muscles to be examined. A stimulator delivers a very small electrical current (that does not cause damage to the body) through the electrodes, causing the nerves to fire. In the electromyogram, a very thin, sterilized needle is inserted into various muscles, usually those affected most by **spasticity** symptoms. The needle is attached by wires to a recording machine. The patient is asked to relax and contract the muscles being examined. The electrical signals produced by the nerves and muscles during these tests are measured and recorded by a computer and displayed as electrical waves on the monitor. The test results are interpreted by a specially trained physician.

Treatment

Most cases of simple cramps require no treatment other than patience and stretching. When heat cramps occur, the child should stop the activity, move to a cool or shady place, remove excess clothing, drink cool water or a sports drink with electrolytes, such as Gatorade, and rest. If the child appears nauseous or is feeling dizzy, he should lie down, with feet slightly elevated. Directing a fan on the child will help cool the child. Gently and gradually stretching and massaging the affected muscle may ease the pain and hasten recovery.

Briefly applying cold packs to cramped muscles, for about ten minutes, may help ease pain.

Acetaminophen (such as Tylenol) or ibuprofen (such as Advil or Motrin) should be used sparingly for relief of discomfort. Ask the child's doctor for specific guidelines. More prolonged or regular cramps may be treated with prescribed medications.

If the child has any signs of dehydration, generous amounts of fluids and an oral rehydrating solution containing glucose and electrolytes should be given. Oral rehydrating solutions, including brands such as Pedialyte, Infalyte, Ceralyte, and Oralyte, are available at most grocery stores and drug stores. They are essential for replacing fluids, **minerals**, and salts. Dehydration can upset the body's electrolyte balance, leading to potentially life-threatening problems such as heart beat abnormalities (arrhythmia). Prolonged, severe dehydration requires medical treatment with intravenous (IV) fluids and may require **hospitalization**.

Treatment of underlying metabolic or neurologic diseases, when possible, may help relieve symptoms.

KEY TERMS

Active motion—Spontaneous; produced by active efforts. Active range of motion exercises are those that are performed by the patient without assistance.

Acupuncture—Based on the same traditional Chinese medical foundation as acupressure, acupuncture uses sterile needles inserted at specific points to treat certain conditions or relieve pain.

Anoxia—Lack of oxygen.

Ataxia—A condition marked by impaired muscular coordination, most frequently resulting from disorders in the brain or spinal cord.

Biofeedback—A training technique that enables an individual to gain some element of control over involuntary or automatic body functions.

Central nervous system—Part of the nervous system consisting of the brain, cranial nerves, and spinal cord. The brain is the center of higher processes, such as thought and emotion and is responsible for the coordination and control of bodily activities and the interpretation of information from the senses. The cranial nerves and spinal cord link the brain to the peripheral nervous system, that is the nerves present in the rest of body.

Cerebral palsy—A nonprogressive movement disability caused by abnormal development of or damage to motor control centers of the brain.

Clonic—Referring to clonus, a series of muscle contractions and partial relaxations that alternate in some nervous diseases in the form of convulsive spasms.

Contraction—A tightening of the uterus during pregnancy. Contractions may or may not be painful and may or may not indicate labor.

Contracture—A tightening or shortening of muscles that prevents normal movement of the associated limb or other body part.

Dystonia—Painful involuntary muscle cramps or spasms.

Fasciculations—Small involuntary muscle contractions visible under the skin.

Genu recurvatum—Hyperextension of the knee.

Hyperactive reflexes—Reflexes that persist too long and may be too strong. For example, a hyperactive grasp reflex may cause the hand to stay clenched in a tight fist.

Hypermobility—Unusual flexibility of the joints, allowing them to be bent or moved beyond their normal range of motion.

Hypertonia—Having excessive muscular tone or strength.

Idiopathic—Refers to a disease or condition of unknown origin.

Motor neuron—A nerve cell that specifically controls and stimulates voluntary muscles.

Multiple sclerosis—A progressive, autoimmune disease of the central nervous system characterized by damage to the myelin sheath that covers nerves. The disease, which causes progressive paralysis, is marked by periods of exacerbation and remission.

Muscle spasm—Localized muscle contraction that occurs when the brain signals the muscle to contract.

Myoclonus—Involuntary contractions of a muscle or an interrelated group of muscles. Also known as myoclonic seizures.

Neurologist—A doctor who specializes in disorders of the nervous system, including the brain, spinal cord, and nerves.

Neurosurgeon—Physician who performs surgery on the nervous system.

Nocturnal leg cramps—Cramps that may be related to exertion and awaken a person during sleep.

Occupational therapist—A healthcare provider who specializes in adapting the physical environment to meet a patient's needs. An occupational therapist also assists patients and caregivers with activities of daily living and provide instructions on wheelchair use or other adaptive equipment.

Orthopedist—A doctor specializing in treatment of the musculoskeletal system.

Passive movement—Movement that occurs under the power of an outside source such as a clinician. There is no voluntary muscular contraction by the individual who is being passively moved.

Periodic limb movement disorder—A disorder characterized by involuntary flexion of leg muscles, causing twitching and leg extension or kicking during sleep.

Peripheral nerves—Nerves outside the brain and spinal cord that provide the link between the body and the central nervous system.

Physiatrist—A physician who specializes in physical medicine and rehabilitation.

Physical therapist—A healthcare provider who teaches patients how to perform therapeutic exercises to maintain maximum mobility and range of motion.

KEY TERMS (contd.)

Range of motion (ROM)—The range of motion of a joint from full extension to full flexion (bending) measured in degrees like a circle.

Restless legs syndrome (RLS)—A disorder in which the patient experiences crawling, aching, or other disagreeable sensations in the calves that can be relieved by movement. RLS is a frequent cause of difficulty falling asleep at night.

Rigidity—A constant resistance to passive motion.

Spinal cord injury—Injury to the spinal cord, via blunt or penetrating trauma.

Stroke—Interruption of blood flow to a part of the brain with consequent brain damage. A stroke may be caused by a blood clot or by hemorrhage due to a burst blood vessel. Also known as a cerebrovascular accident.

Alternative treatment

Alternative and complementary therapies include approaches that are considered to be outside the mainstream of traditional health care. In adults, alternative treatments for cramps include gingko (*Ginkgo biloba*) or Japanese quince (*Chaenomeles speciosa*). Supplements of vitamin E, niacin, calcium, and magnesium may also help to relieve the likelihood of night cramps, especially when taken at bedtime. Indications for these treatments in children have not been documented.

There are several alternative therapies that can be useful when treating **movement disorders**. Among the therapies that may be helpful are acupuncture, homeopathy, touch therapies, postural alignment therapies, and biofeedback. The progress made will depend on the individual and his/her condition.

Biofeedback training may be used to teach older children how to consciously reduce muscle tension. Biofeedback uses an electrical signal that indicates when a spastic muscle relaxes. The patient may be able to use biofeedback to learn how to consciously reduce muscle tension and possibly reduce symptoms.

Before learning or practicing any particular technique, it is important for the parent or caregiver and child to learn about the therapy, its safety and effectiveness, potential side effects, and the expertise and qualifications of the practitioner. Although some practices are beneficial, others may be harmful to certain patients. Alternative therapies should not be used as a substitute for medical therapies prescribed by a doctor. Parents should discuss these alternative treatments with the child's doctor to determine the techniques and remedies that may be beneficial for the child.

Prognosis

Occasional cramps are common and have no special medical significance.

Prevention

The likelihood of developing cramps may be reduced with regular exercise to build up energy reserves in the muscles. Avoiding exercising in extreme heat helps prevent heat cramps. Heat cramps can also be avoided by drinking plenty of water before and during exercise in extreme heat. Practicing proper body mechanics while sitting (sitting with both feet on the floor, back straight and legs uncrossed) can help prevent the development of leg cramps. Taking a warm bath before bedtime may increase circulation to the legs and reduce the incidence of nighttime leg cramps.

Nutritional concerns

The likelihood of developing cramps may be reduced by eating a well-balanced, healthy diet with appropriate levels of minerals. A registered dietitian can work with parents to identify a child's specific calorie needs and develop an individualized meal plan.

Fluids should be encouraged during all strenuous activities, especially in warm weather. People should aim for two to four eight-ounce glasses of fluid per hour of activity.

If an underlying neurological disorder has been identified, dietary guidelines are individualized, based on the child's age, diagnosis, overall health, caloric and energy needs, and level of functioning. Early identification, treatment, and correction of specific feeding problems will improve the health and nutritional status of the patient.

Parental concerns

Occasional muscle cramps are common. The most important concern is preventing dehydration, especially when the child is exercising in high or humid temperatures. Make sure the child drinks enough fluids before, during, and after sports and other activities. Pack a water bottle and/or sports drink for the child to have at sports practices, games, and other physical activities. Make

sure the coach provides time out for water breaks. After the activity, encourage the child to continue drinking water to replace lost fluids.

If a movement disorder has been diagnosed, parents should work closely with the child's therapists and doctors to create an effective treatment plan. It is important for parents to communicate their treatment goals with the health care team. Parents should take an active role in the child's exercise program.

Raising a child with a movement disorder can be challenging. There are several support groups available to provide information and assistance.

Resources

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National Institute of Neurological Disorders and Stroke (NINDS). National Institutes of Health. PO Box 5801, Bethesda, MD 20824. Web site: <www.ninds.nih.gov/about_ninds>.

National Rehabilitation Information Center (NARIC). 4200 Forbes Blvd., Ste. 202, Lanham, MD 20700. Web site: <www.naric.com>.

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Muscular dystrophy

Definition

Muscular dystrophy is the name for a group of inherited disorders in which strength and muscle bulk gradually decline. Nine types of muscular dystrophies are generally recognized.

Description

The muscular dystrophies include:

- Duchenne muscular dystrophy (DMD), which affects young boys, causing progressive muscle weakness, usually beginning in the legs. It is the most severe form of muscular dystrophy.
- Becker muscular dystrophy (BMD), which affects older boys and young men, following a milder course than DMD
- Emery-Dreifuss muscular dystrophy (EDMD), which affects young boys, causing contractures and weakness in the calves, weakness in the shoulders and upper arms, and problems in the way electrical impulses travel through the heart to make it beat (heart conduction defects). Female carriers of EDMD are at risk for heart block.
- Limb-girdle muscular dystrophy (LGMD), which begins in late childhood to early adulthood and affects both men and women, causing weakness in the muscles around the hips and shoulders. It is the most variable of the muscular dystrophies, and there are as of 2004 several different forms of the disease recognized. Many people with suspected LGMD have probably been misdiagnosed in the past; therefore, the prevalence of the disease is difficult to estimate.
- Facioscapulohumeral muscular dystrophy (FSH), also known as Landouzy-Dejerine disease, which begins in late childhood to early adulthood and affects both men and women, causing weakness in the muscles of the face, shoulders, and upper arms. The hips and legs may also be affected.
- **Myotonic dystrophy**, also known as Steinert's disease, which affects both men and women, causing generalized weakness first seen in the face, feet, and hands. It is accompanied by the inability to relax the affected muscles (myotonia). Symptoms may begin any time from birth through adulthood.
- Oculopharyngeal muscular dystrophy (OPMD), which affects adults of both sexes, causing weakness in the eye muscles and throat
- Distal muscular dystrophy (DD), which begins in middle age or later, causing weakness in the muscles of the feet and hands
- Congenital muscular dystrophy (CMD), which is present from birth, results in generalized weakness, and usually progresses slowly. A subtype, called Fukuyama CMD, also involves **mental retardation**. Both are rare.

Demographics

DMD occurs in about one in 3,500 male births and affects approximately 8,000 boys and young men in the United States. A milder form occurs in very few female carriers.

BMD occurs in about one in 30,000 male births.

Fewer than 300 cases of EDMD have been identified.

The number of people affected with LGMD in the United States may be in the low thousands.

FSH occurs in about one out of every 20,000 people and affects approximately 13,000 people in the United States.

Myotonic dystrophy is the most common form of muscular dystrophy, affecting more than 30,000 people in the United States.

OPMD is most common among French Canadian families in Quebec and in Spanish-American families in the southwestern United States.

DD is most common in Sweden and rare in other parts of the world.

Fukuyama CMD is most common in Japan.

Causes and symptoms

Causes

Several of the muscular dystrophies, including DMD, BMD, CMD, and most forms of LGMD, are due to defects in the genes for a complex of muscle proteins. This complex spans the muscle cell membrane to unite a fibrous network on the interior of the cell with a fibrous network on the outside. As of 2004 the theory was that by linking these two networks, the complex acts as a "shock absorber," redistributing and evening out the forces generated by contraction of the muscle, thereby preventing rupture of the muscle membrane. Defects in the proteins of the complex lead to deterioration of the muscle. Symptoms of these diseases set in as the muscle gradually exhausts its ability to repair itself. Both DMD and BMD are caused by flaws in the gene for the protein called dystrophin. The flaw leading to DMD prevents the formation of any dystrophin, while that of BMD allows some protein to be made, accounting for the differences in severity and onset between the two diseases. Differences among the other diseases in the muscles involved and the ages of onset are less easily explained.

The causes of the other muscular dystrophies are not as well understood:

- One form of LGMD is caused by defects in the gene for a muscle enzyme, calpain. The relationship between this defect and the symptoms of the disease is unclear.
- EDMD is due to a defect in the gene for a protein called emerin, which is found in the membrane of a cell's nucleus, but whose exact function is unknown.
- Myotonic dystrophy is linked to gene defects for a protein that may control the flow of charged particles within muscle cells. This gene defect is called a triple repeat, meaning it contains extra triplets of DNA code. It is possible that this mutation affects nearby genes as well, and that the widespread symptoms of myotonic dystrophy are due to a range of genetic disruptions.
- The gene for OPMD appears to also be mutated with a triple repeat. The function of the affected protein may involve translation of genetic messages in a cell's nucleus.
- The cause of FSH is unknown. The genetic region responsible for it has been localized on its chromosome, however.
- The gene responsible for DD has not yet been found.

Genetics and patterns of inheritance

The muscular dystrophies are genetic diseases, meaning they are caused by defects in genes. Genes, which are linked together on chromosomes, have two functions. They code for the production of proteins, and they are the material of inheritance. Parents pass along genes to their children, providing them with a complete set of instructions for making their own proteins.

Because both parents contribute genetic material to their offspring, each child carries two copies of almost every gene, one from each parent. For some diseases to occur, both copies must be flawed. Such diseases are called autosomal recessive diseases. Some forms of LGMD and DD exhibit this pattern of inheritance, as does CMD. A person with only one flawed copy, called a carrier, will not have the disease but may pass the flawed gene on to children. When two carriers have children, the chances of having a child with the disease is one in four for each pregnancy.

Other diseases occur when only one flawed gene copy is present. Such diseases are called autosomal dominant diseases. Other forms of LGMD exhibit this pattern of inheritance, as do DM, FSH, OPMD, and some forms of DD. When a person affected by the disease has a child with someone not affected, the chances of having an affected child is one in two.

Because of chromosomal differences between the sexes, some genes are not present in two copies. The chromosomes that determine whether a person is male or female are called the X and Y chromosomes. A person with two X chromosomes is female, while a person with one X and one Y is male. While the X chromosome carries many genes, the Y chromosome carries almost none. Therefore, a male has only one copy of each gene on the X chromosome, and if it is flawed, he will have the disease that defect causes. Such diseases are said to be X-linked. X-linked diseases include DMD, BMD, and

EDMD. Women are not usually affected by X-linked diseases, since they will likely have one unaffected copy between the two chromosomes. Some female carriers of DMD suffer a mild form of the disease, probably because their one unaffected gene copy is shut down in some of their cells.

Women carriers of X-linked diseases have a one-in-two chance of passing the flawed gene on to each child born. Daughters who inherit the disease gene are carriers. A son born without the disease gene is free of the disease and cannot pass it on to his children. A son born with the defect has the disease. He will pass the flawed gene on to each of his daughters, who will then be carriers, but to none of his sons (because they inherit his Y chromosome).

Not all genetic flaws are inherited. As many as one-third of the cases of DMD are due to new mutations that arise during egg formation in the mother. New mutations are less common in other forms of muscular dystrophy.

Symptoms

All of the muscular dystrophies are marked by muscle weakness as the major symptom. The distribution of symptoms, age of onset, and progression differ significantly. **Pain** is sometimes a symptom of each, usually due to the effects of weakness on joint position.

DMD A boy with Duchenne muscular dystrophy usually begins to show symptoms as a preschooler. The legs are affected first, making walking difficult and causing balance problems. Most affected persons walk three to six months later than expected and have difficulty running. Later on, the boy with DMD will push his hands against his knees to rise to a standing position, to compensate for leg weakness. About the same time, his calves will begin to swell, though with fibrous tissue rather than with muscle and feel firm and rubbery; this condition gives DMD one of its alternate names, pseudohypertrophic muscular dystrophy. The boy will widen his stance to maintain balance and walk with a waddling gait to advance his weakened legs. Contractures (permanent muscle tightening) usually begin by age five or six, most severely in the calf muscles. This pulls the foot down and back, forcing the boy to walk on tip-toes, called equinus, and further decreases balance. Frequent falls and broken bones are common beginning at this age. Climbing stairs and rising unaided may become impossible by age nine or ten, and most boys use a wheelchair for mobility by the age of 12. Weakening of the trunk muscles around this age often leads to **scoliosis** (a side-to-side spine curvature) and kyphosis (a front-to-back curvature).

The most serious weakness of DMD is weakness of the diaphragm, the sheet of muscles at the top of the abdomen that perform the main work of breathing and coughing. Diaphragm weakness leads to reduced energy and stamina and increased lung infection because of the inability to **cough** effectively. Young men with DMD often live into their twenties and beyond, provided they have mechanical ventilation assistance and good respiratory hygiene.

About one third of boys with DMD experience specific learning disabilities, including trouble learning by ear rather than by sight and trouble paying attention to long lists of instructions. Individualized educational programs usually compensate well for these disabilities.

BMD The symptoms of BMD usually appear in late childhood to early adulthood. Though the progression of symptoms may parallel that of DMD, the symptoms are usually milder, and the course more variable. The same pattern of leg weakness, unsteadiness, and contractures occurs later for the young man with BMD, often allowing independent walking into the twenties or early thirties. Scoliosis may occur but is usually milder and progresses more slowly. Heart muscle disease (cardiomyopathy) occurs more commonly in BMD. Problems may include irregular heartbeats (arrhythmias) and congestive heart failure. Symptoms may include fatigue, shortness of breath, chest pain, and **dizziness**. Respiratory weakness also occurs and may lead to the need for mechanical ventilation.

EDMD This type of muscular dystrophy usually begins in early childhood, often with contractures preceding muscle weakness. Weakness affects the shoulder and upper arm originally, along with the calf muscles, leading to foot-drop. Most men with EDMD survive into middle age, although a defect in the heart's rhythm (heart block) may be fatal if not treated with a pacemaker.

LGMD While there are at least six genes that cause the various types of LGMD, two major clinical forms of LGMD are usually recognized. A severe childhood form is similar in appearance to DMD but is inherited as an autosomal recessive trait. Symptoms of adult-onset LGMD usually appear in a person's teens or twenties and are marked by progressive weakness and wasting of the muscles closest to the trunk. Contractures may occur, and the ability to walk is usually lost about 20 years after onset. Some people with LGMD develop respiratory weakness that requires use of a ventilator. Lifespan may be somewhat shortened. (Autosomal dominant forms usually occur later in life and progress relatively slowly.)

FSH FSH varies in its severity and age of onset, even among members of the same **family**. Symptoms most commonly begin in the teens or early twenties, though infant or childhood onset is possible. Symptoms tend to be more severe in those with earlier onset. The disease is named for the regions of the body most severely affected by the disease: muscles of the face (facio-), shoulders (scapulo-), and upper arms (humeral). Hips and legs may be affected as well. Children with FSH often develop partial or complete deafness.

The first symptom noticed is often difficulty lifting objects above the shoulders. The weakness may be greater on one side than the other. Shoulder weakness also causes the shoulder blades to jut backward, called scapular winging. Muscles in the upper arm often lose bulk sooner than those of the forearm, giving a "Popeye" appearance to the arms. Facial weakness may lead to loss of facial expression, difficulty closing the eyes completely, and inability to drink through a straw, blow up a balloon, or whistle. A person with FSH may not develop strong facial wrinkles. Contracture of the calf muscles may cause foot-drop, leading to frequent tripping over curbs or rough spots. People with earlier onset often require a wheelchair for mobility, while those with later onset rarely do.

MYOTONIC DYSTROPHY Symptoms of myotonic dystrophy include facial weakness and a slack jaw, drooping eyelids (ptosis), and muscle wasting in the forearms and calves. A person with this dystrophy has difficulty relaxing his grasp, especially if the object is cold. Myotonic dystrophy affects heart muscle, causing arrhythmias and heart block, and the muscles of the digestive system, leading to motility disorders and **constipation**. Other body systems are affected as well: myotonic dystrophy may cause cataracts, retinal degeneration, low IQ, frontal balding, skin disorders, testicular atrophy, **sleep** apnea, and insulin resistance. An increased need or desire for sleep is common, as is diminished motivation. Severe disability affects most people with this type of dystrophy within 20 years of onset, although most do not require a wheelchair even late in life.

OPMD OPMD usually begins in a person's thirties or forties, with weakness in the muscles controlling the eyes and throat. Symptoms include drooping eyelids, difficulty swallowing (dysphagia), and weakness progresses to other muscles of the face, neck, and occasionally the upper limbs. Swallowing difficulty may cause aspiration or the introduction of food or saliva into the airways. **Pneumonia** may follow.

DD DD usually begins in the twenties or thirties with weakness in the hands, forearms, and lower legs. Difficulty with fine movements such as typing or fastening buttons may be the first symptoms. Symptoms progress slowly, and the disease usually does not affect life span.

CMD CMD is marked by severe muscle weakness from birth, with infants displaying "floppiness" and very little voluntary movement. Nonetheless, a child with CMD may learn to walk, either with or without some assistive device, and live into young adulthood or beyond. In contrast, children with Fukuyama CMD are rarely able to walk and have severe mental retardation. Most children with this type of CMD die in childhood.

When to call the doctor

A doctor should be consulted whenever muscle development is thought to be abnormal or slow.

Diagnosis

Diagnosis of muscular dystrophy involves a careful medical history and a thorough physical exam to determine the distribution of symptoms and to rule out other causes. Family history may give important clues, since all the muscular dystrophies are genetic conditions (though no family history will be evident in the event of new mutations).

Lab tests may include the following:

- Blood level of the muscle enzyme creatine kinase (CK). CK levels rise in the blood due to muscle damage and may be seen in some conditions even before symptoms appear.
- Muscle biopsy, in which a small piece of muscle tissue is removed for microscopic examination. Changes in the structure of muscle cells and presence of fibrous tissue or other aberrant structures are characteristic of different forms of muscular dystrophy. The muscle tissue can also be stained to detect the presence or absence of particular proteins, including dystrophin.
- Electromyogram (EMG). EMG is used to examine the response of the muscles to stimulation. Decreased response is seen in muscular dystrophy. Other characteristic changes are seen in DM.
- Genetic tests. Several of the muscular dystrophies can be positively identified by testing for the presence of the mutated gene involved. Accurate genetic tests are available for DMD, BMD, DM, several forms of LGMD, and EDMD.
- Other specific tests as necessary. For EDMD and BMD, for example, an electrocardiogram may be nee-

MD patient Sarah Schwegel with Jerry Lewis at the Muscular Dystrophy Telethon, which raises money for research for the cure and treatment of muscular dystrophy. *(Muscular Dystrophy Association.)*

ded to test heart function, and hearing tests are performed for children with FSH.

For most forms of muscular dystrophy, accurate diagnosis is not difficult when done by someone familiar with the range of diseases. There are exceptions, however. Even with a muscle biopsy, it may be difficult to distinguish between FSH and another muscle disease, polymyositis. Childhood-onset LGMD is often mistaken for the much more common DMD, especially when it occurs in boys. BMD with an early onset appears very similar to DMD, and a muscle biopsy may be needed to accurately distinguish them. The muscular dystrophies may be confused with diseases involving the motor neurons, such as **spinal muscular atrophy**; diseases of the neuromuscular junction, such as myasthenia gravis; and other muscle diseases, as all involve generalized weakening of varying distribution.

Treatment

Drugs

As of 2004 there were no cures for any of the muscular dystrophies. Prednisone, a corticosteroid, has been shown to delay the progression of DMD somewhat, for reasons that as of 2004 are still unclear. Prednisone is also prescribed for BMD.

Treatment of muscular dystrophy is mainly directed at preventing the complications of weakness, including decreased mobility and dexterity, contractures, scoliosis, heart defects, and respiratory insufficiency.

Physical therapy

Physical therapy, in particular regular stretching, is used to maintain the range of motion of affected muscles and to prevent or delay contractures. Braces are used as well, especially on the ankles and feet to prevent equinus. Full-leg braces may be used in DMD to prolong the period of independent walking. Strengthening other muscle groups to compensate for weakness may be possible if the affected muscles are few and isolated, as in the earlier stages of the milder muscular dystrophies. Regular, nonstrenuous **exercise** helps maintain general good health. Strenuous exercise is usually not recommended, since it may damage muscles further.

Surgery

When contractures become more pronounced, tenotomy surgery may be performed. In this operation, the tendon of the contractured muscle is cut, and the limb is braced in its normal resting position while the tendon regrows. In FSH, surgical fixation of the scapula can help compensate for shoulder weakness. For a person with OPMD, surgical lifting of the eyelids may help compensate for weakened muscular control. For a person with DM, sleep apnea may be treated surgically to maintain an open airway. Scoliosis surgery is often needed in DMD but much less often in other muscular dystrophies. Surgery is recommended at a much lower degree of curvature for DMD than for scoliosis due to other conditions, since the decline in respiratory function in DMD makes surgery at a later time dangerous. In this surgery, the vertebrae are fused together to maintain the spine in the upright position. Steel rods are inserted at the time of operation to keep the spine rigid while the bones grow together.

When any type of surgery is performed in people with muscular dystrophy, anesthesia must be carefully selected. People with MD are susceptible to a severe reaction, known as malignant hyperthermia, when given halothane anesthetic.

Occupational therapy

The occupational therapist suggests techniques and tools to compensate for the loss of strength and dexterity. Strategies may include modifications in the home, adaptive utensils and dressing aids, compensatory movements and positioning, wheelchair accessories, or communication aids.

Nutrition

Good **nutrition** helps to promote general health in all the muscular dystrophies. No special diet or supplement has as of 2004 been shown to be of use in any of

KEY TERMS

Autosomal dominant—A pattern of inheritance in which only one of the two copies of an autosomal gene must be abnormal for a genetic condition or disease to occur. An autosomal gene is a gene that is located on one of the autosomes or non-sex chromosomes. A person with an autosomal dominant disorder has a 50 percent chance of passing it to each of their offspring.

Autosomal recessive—A pattern of inheritance in which both copies of an autosomal gene must be abnormal for a genetic condition or disease to occur. An autosomal gene is a gene that is located on one of the autosomes or non-sex chromosomes. When both parents have one abnormal copy of the same gene, they have a 25 percent chance with each pregnancy that their offspring will have the disorder.

Becker muscular dystrophy (BMD)—A type of muscular dystrophy that affects older boys and men and usually follows a milder course than Duchenne muscular dystrophy.

Contracture—A tightening or shortening of muscles that prevents normal movement of the associated limb or other body part.

Distal muscular dystrophy (DD)—A form of muscular dystrophy that usually begins in middle age or later, causing weakness in the muscles of the feet and hands.

Duchenne muscular dystrophy (DMD)—The most severe form of muscular dystrophy, DMD usually affects young boys and causes progressive muscle weakness, usually beginning in the legs.

Dystrophin—A protein that helps muscle tissue repair itself. Both Duchenne muscular dystrophy and Becker muscular dystrophy are caused by flaws in the gene that tells the body how to make this protein.

Facioscapulohumeral muscular dystrophy (FSH)—This form of muscular dystrophy, also known as Landouzy-Dejerine disease, begins in late childhood to early adulthood and affects both men and women, causing weakness in the muscles of the face, shoulders, and upper arms.

Limb-girdle muscular dystrophy (LGMD)—A form of muscular dystrophy that begins in late childhood to early adulthood and affects both men and women. It causes weakness in the muscles around the hips and shoulders.

Myotonic dystrophy—A form of muscular dystrophy, also known as Steinert's disease, that affects both men and women. It is characterized by delay in the ability to relax muscles after forceful contraction (myotonia) and wasting of muscles, as well as other abnormalities.

Oculopharyngeal muscular dystrophy—A type of muscular dystrophy that affects adults of both sexes, causing weakness in the eye muscles and throat.

the conditions. The weakness in the throat muscles seen especially in OPMD and later DMD may necessitate the use of a gastrostomy tube, inserted in the stomach to provide nutrition directly.

Cardiac care

The arrhythmias of EDMD and BMD may be treatable with antiarrhythmia drugs such as mexiletine or nifedipine. A pacemaker may be implanted if these do not provide adequate control. Heart transplants are increasingly common for men with BMD.

Respiratory care

People who develop weakness of the diaphragm or other ventilatory muscles may require a mechanical ventilator to continue breathing deeply enough. Air may be administered through a nasal mask or mouthpiece or through a tracheostomy tube, which is inserted through a surgical incision through the neck and into the windpipe. Most people with muscular dystrophy do not need a tracheostomy, although some may prefer it to continual use of a mask or mouthpiece. Supplemental oxygen is not needed. Good hygiene of the lungs is critical for health and long-term survival of a person with weakened ventilatory muscles. Assisted cough techniques provide the strength needed to clear the airways of secretions; an assisted cough machine is also available and provides excellent results.

Experimental treatments

Two experimental procedures aiming to cure DMD have attracted a great deal of attention. In myoblast transfer, millions of immature muscle cells are injected into an affected muscle. The goal of the treatment is to promote the growth of the injected cells, replacing the

defective host cells with healthy new ones. Despite continued claims to the contrary by a very few researchers, this procedure is widely judged a failure.

Gene therapy introduces good copies of the dystrophin gene into muscle cells. The goal is to allow the existing muscle cells to use the new gene to produce the dystrophin it cannot make with its flawed gene. Problems have included immune rejection of the virus used to introduce the gene, loss of gene function after several weeks, and an inability to get the gene to enough cells to make a functional difference in the affected muscle. Nonetheless, after a number of years of refining the techniques in mice, researchers began human trials in 1998. These trials are ongoing.

Prognosis

The expected life span for a male with DMD has increased significantly since the 1970s. Most young men live into their early or mid-twenties. Respiratory infections become an increasing problem as their breathing becomes weaker, and these infections are usually the cause of death.

The course of the other muscular dystrophies is more variable; expected life spans and degrees of disability are hard to predict but may be related to age of onset and initial symptoms. Prediction is made more difficult because, as new genes are discovered, it becomes clear that several of the dystrophies are not uniform disorders but rather symptom groups caused by different genes.

People with dystrophies with significant heart involvement (BMD, EDMD, Myotonic dystrophy) may nonetheless have almost normal life spans, provided that cardiac complications are monitored and treated aggressively. The respiratory involvement of BMD and LGMD similarly require careful and prompt treatment.

Prevention

As of 2004 there was no way to prevent any of the muscular dystrophies in a person who has the genes responsible for these disorders. Accurate genetic tests, including prenatal tests, are available for some of the muscular dystrophies. Results of these tests may be useful for purposes of family planning.

Nutritional concerns

There is no known link between nutrition and the onset of muscular dystrophy.

Parental concerns

Prospective parents with first-degree relatives (parents, siblings, or other children) who have been diagnosed with muscular dystrophy should consider including counseling in their family planning process.

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ORGANIZATIONS

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Muscular Dystrophy Association. National Headquarters, 3300 E. Sunrise Drive, Tucson, AZ 85718. Web site: <www.mdausa.org/

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Mutism

Definition

Mutism is a rare childhood condition characterized by a consistent failure to speak in situations where talking is expected.

Description

In mutism, the child has the ability to converse normally and does so, for example, in the home, but consistently fails to speak in specific situations such as at school or with strangers. The condition is also called selective mutism, to differentiate it from children who are physically unable to speak. Experts believe that this selective problem is associated with **anxiety** and **fear** in social situations such as in school or in the company of adults. It is, therefore, often considered a type of social phobia. This is not a communication disorder because the affected children can converse normally in some situations. It is not a developmental disorder because their ability to talk, when they choose to do so, is appropriate for their age level. This problem has been linked to anxiety, and one of the major ways in which both children and adults attempt to cope with anxiety is by avoiding whatever provokes the anxiety. Affected children are typically shy and are especially so in the presence of strangers and unfamiliar surroundings or situations. However, the behaviors of children with this condition go beyond **shyness**.

These children understand language and are able to talk normally in settings where they are comfortable, secure and relaxed. Over 90 percent of children with mutism also have social phobia or social anxiety, and some experts view mutism as a symptom of social anxiety. Others view it as a separate, but related, disorder. It is not yet understood why some individuals develop typical symptoms of social anxiety, like reluctance to speak in front of a group of people or feeling embarrassed easily, while others experience the inability to speak that characterizes mutism. What is clear is that children and adolescents with mutism have an actual fear of speaking and of social interactions where there is an expectation to talk. They may also be unable to communicate nonverbally, may be unable to make eye contact, and may stand motionless with fear as they are confronted with specific social settings. This can be quite heart wrenching to watch and is often very debilitating for the child as well as frustrating for parents and teachers.

A child meets the criteria for mutism if the following are true:

- The child does not speak in certain selected places such as school or at particular social events.
- The child speaks normally in at least one environment, usually in the home, but a small percentage of children with mutism are mute at home.
- The child's inability to speak interferes with his or her ability to function in school and/or social settings.
- The mutism has persisted for at least one month.
- The mutism is not caused by a communication disorder (such as **stuttering**) and does not occur as part of other mental disorders (such as **autism**).

Demographics

It is estimated that one in every 1,000 school-age children are affected by mutism.

Causes and symptoms

Mutism is believed to arise from anxiety experienced in social situations where the child may be called upon to speak. Refusing to speak or speaking in a whisper spares the child from the possible humiliation or embarrassment of saying the "wrong" thing. When asked a direct question by teachers, for example, the affected child may act as if they are unable to answer. Some children may communicate via gestures, nodding, or very brief utterances. Additional features may include excessive shyness, oppositional behavior, and impaired learning at school.

The majority of children with mutism have a genetic predisposition to anxiety. In other words, they have inherited the tendency to be anxious from **family** members and may be vulnerable to the development of an anxiety disorder. Very often, these children show signs of anxiety, such as difficulty separating from parents, moodiness, clinging behavior, inflexibility, **sleep** problems, frequent **tantrums** and crying, and extreme shyness starting in infancy. When they reach the age when they begin to interact socially outside the family environment, their persistent fear of speaking or communicating begins to manifest in symptoms like freezing, lack of response, stiff posture, blank facial expression, lack of smiling, and mutism. Studies have shown that some children are born with inhibited temperaments, which means that even as infants, they are more likely to be fearful and wary of new situations. There is reason to believe that many or most children with mutism were born with this inhibited personality type.

Research has also shown that these behaviorally inhibited children have a decreased threshold of

excitability in the area of the brain called the amygdala. The normal function of the amygdala is to receive and process signals of potential danger and set off a series of reactions that will help individuals protect themselves, such as the fight-or-flight response. In anxious individuals, the amygdala seems to overreact and set off these responses even when the individual is not really in danger. In the case of selectively mute children, the anxiety responses are triggered by social interactions in settings such as school, the playground, or social gatherings. Over time, a child with selective mutism becomes mute because of an inability to cope with fearful feelings that occur when he or she is expected to speak. When the child does not respond, the pressure is usually removed and the child feels relief from fear.

Besides genetics and biological factors, researchers believe that other factors may contribute to the development of selective mutism. A significant number of children with mutism also have expressive **language disorders**, and a fairly large number come from a bilingual environment, which may add to a child's vulnerability to mutism. Anxiety is still the root cause of the mutism, and it is theorized that these language difficulties may make the child more self-conscious about his or her speaking skills and thus may increase the fear of being judged by others. These risk factors are probably additive; in other words, if a child has genetic risk of anxiety, plus a bilingual environment or a speech disorder, the likelihood of that child developing selective mutism becomes higher with each added factor.

When to call the doctor

If selective mutism persists for more than a month, parents should discuss this pattern with their child's teachers, family physician, or pediatrician. The doctor may refer the child to a speech therapist, psychiatrist or psychologist.

Diagnosis

The diagnosis of mutism is fairly easy to make because the signs and symptoms are clear-cut and easily observable. However, other social disorders effecting social speech, such as autism or **schizophrenia**, must be considered in the diagnosis. The average age of diagnosis is between three and eight years of age; however, in retrospect many parents will say that their child displayed signs of excessive shyness and inhibition since infancy. It is not until children enter school, where there is an expectation to perform, interact, and speak, that mutism becomes more apparent. Often a parent suspects during the **preschool** years that there is a problem, but lack of knowledge about selective mutism makes it difficult to find help. It is all too common for parents to question their child's pediatrician about the child's inability to speak in public and be told that the child is just shy and will outgrow the behavior. Once a child enters school, though, teachers often point out the severity of the problem to the parents. Some parents are also reluctant to have their child evaluated and treated.

Treatment

Since selective mutism is an anxiety disorder, successful treatment focuses on methods to lower anxiety, increase **self-esteem**, and increase confidence and communication in social settings. The emphasis should never be on "getting a child to talk," nor should the goal of treatment be for the child to speak to the therapist. Progress outside the clinic or doctor's office is much more important than whether the child speaks during the therapy session. Initially, all expectations for verbalization should be removed. As the child's anxiety is lowered and confidence increases, verbalization usually follows. If it does not occur spontaneously, techniques can later be added to help encourage progress. A professional should devise an individualized treatment plan for each child and allow the child, family, and school to have a great deal of input into the treatment process. Therapy usually involves some combination of behavioral therapy, cognitive behavioral therapy, **play therapy**, or psychoanalytic therapy, medication, and in some cases, **family therapy**.

Behavioral therapy

The primary types of behavioral therapy used for selective mutism are desensitization, fading, and positive reinforcement techniques. Desensitization means exposing a child to something that is feared in a gradual way, in order to help the child overcome the fear. Fading therapy is a type of desensitization that creates a series of events or exposures that starts with a situation that is comfortable for the child, such as being alone in the classroom with a parent and playing a board game. New variables that are progressively more difficult are gradually added. For example, having the teacher walk past the room and overhear the child speaking to the parent, and then having the teacher enter the room, and eventually have the child interacting with the teacher in the classroom. Positive reinforcement, or the use of rewards for changes in behavior, should only be introduced after anxiety is lowered and the child is ready to begin working on goals. It is also important to realize that there are many intermediate steps between being mute and being verbal. During the early stages of treatment, nonverbal communication such as pointing, nodding, and use of pictures to express needs, can be encouraged and

rewarded. Though some may fear that allowing nonverbal communication will enable the mutism to continue, many therapists believe it is a necessary step for most children with mutism to overcome their communication anxiety in a step-by-step manner.

Cognitive behavioral therapy

Cognitive behavioral therapy (CBT) helps children change their thoughts (the cognitive part) and their actions (the behavioral part). CBT therapists recognize that anxious children tend to exaggerate the frightening aspects of certain situations, so they help the children gain a more realistic perspective in order to decrease anxiety. They also know that anxious children avoid situations they fear or (in the case of selectively mute children) avoid speech in anxiety-provoking situations. Avoidance makes anxiety worse. Therefore, CBT helps the child overcome avoidance by gradually facing what is feared with lots of praise and positive reinforcement for doing so. Parents, teachers, and other adults around the child can be very helpful in this process. Cognitive strategies for the selectively mute child aim to reduce the social anxiety that is often part of the disorder. Cognitive strategies help the child challenge negative expectations and replace them with more realistic ones. This process is combined with behavioral strategies that focus on helping the selectively mute child to talk in increasingly challenging situations. The therapist carefully collects information on where and with whom the child already speaks and then helps the child choose a goal to work on in a situation that is just slightly more challenging.

Play therapy

Play therapy is an adaptation of psychoanalytic therapy, which is a psychological treatment based on helping people understand their unconscious thoughts. This field of psychology includes Freudian theories but also many other modern theories about how our minds work. Play therapy refers to the use of play as communication; therapists who are trained in these techniques observe and participate in play activities with the child and interpret the child's actions as a form of subconscious communication. There is not a lot of evidence for play therapy being effective in the treatment of mutism; however, a well trained play therapist might be able to help a child with mutism better understand and express emotions and may be a part of an overall treatment plan. It may be especially useful when a stressful event or environment is a factor. For some children there may be contributing factors such as the death of a parent or other loved one, a **divorce**, or a move. Play therapists may be able to help a child to express and better understand the emotions that they are experiencing in these situations.

Family therapy

Since there is no evidence of family pathology being the cause of most cases of mutism, this type of therapy is not necessary in most cases. However, if there are unusual circumstances or a highly stressful family environment, then it may be advisable for families to participate in more intensive family therapy.

Prognosis

The prognosis for mutism is good. Sometimes it disappears suddenly on its own. The negative impact on learning and school activities may, however, persist into adult life.

Prevention

Mutism cannot be prevented because the cause is not known. However, family conflict or problems at school contribute to the seriousness of the symptoms.

Parental concerns

Parents should remove all pressure and expectations for the child to speak, conveying to their child that they understand he or she feels "scared" to speak or has difficulty speaking at times. Many parents report that simply removing the pressure and letting the child know that they understand can help to improvement the child's symptoms. Parents should also reassure their child that they will help him or her through this difficult time. The child's accomplishments and efforts should be praised, and support and understanding should be offered when the child has difficulties and frustrations. Parents should read as much information as they can to become well informed about selective mutism.

It is important for family members to be educated and informed about selective mutism and to be included in the child's treatment plan in order to provide a supportive environment for the child's recovery. The stress of dealing with the child's mutism may have created various imbalances in family dynamics, and parents may need help in coming to terms with their own emotions and becoming more consistent in their parenting styles. It is also common for parents to begin to recognize their own anxiety as they are learning to help their child. Many times they will seek help in overcoming social anxiety to improve their ability to advocate for their child's needs and to become a positive role model for their child.

KEY TERMS

Amygdala—An almond-shaped brain structure in the limbic system that is activated in stressful situations to trigger the emotion of fear. It is thought that the emotional overreactions in Alzheimer's patients are related to the destruction of neurons in the amygdala.

Autism—A developmental disability that appears early in life, in which normal brain development is disrupted and social and communication skills are retarded, sometimes severely.

Behavior modification—A form of therapy that uses rewards to reinforce desired behavior. An example would be to give a child a piece of chocolate for grooming appropriately.

Cognitive-behavioral therapy—A type of psychotherapy in which people learn to recognize and change negative and self-defeating patterns of thinking and behavior.

Family therapy—A type of therapy in which the entire immediate family participates.

Play therapy—A type of psychotherapy for young children involving the use of toys and games to build a therapeutic relationship and encourage the child's self-expression.

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Myers-Briggs Type Indicator

Definition

The Myers-Briggs Type Indicator (MBTI) is a widely used personality inventory, or test, employed in vocational, educational, and psychotherapy settings to evaluate personality type in adolescents and adults age 14 and older.

Purpose

In an educational setting, the MBTI may be performed to assess student learning style. In a classroom setting, the MBTI may be used to help teens and young adults better understand their learning, communication, and social interaction styles. Guidance counselors also might use the test to help teens determine which occupational field or college major they might be best suited for.

Because the MBTI is also a tool for self-discovery, mental health professionals may administer the test in counseling sessions to provide their patients with insight into their behavior. Among adults, the MBTI is also used in organizational settings to assess management skills and facilitate teamwork and problem solving.

Description

In 2000, an estimated two million people took the MBTI, making it the most frequently used personality inventory available. First introduced in 1942, the test was the work of mother and daughter Katharine C. Myers Briggs and Isabel Briggs. There are now several different versions of the test available. Form M, which contains 93 items and is a self-scoring **assessment**, is the most commonly used. It can be used in a classroom or other group setting, and takes approximately 15 to 25 minutes to complete.

The Myers-Briggs inventory is based on Carl Jung's theory of types, outlined in his 1921 work *Psychological Types*. Jung's theory holds that human beings are either introverts or extraverts, and their behavior follows from these inborn psychological types. He also believed that people take in and process information in different ways, based on their personality traits.

The Myers-Briggs evaluates personality type and preference based on the four Jungian psychological types:

- extraversion (E) or introversion (I)
- sensing (S) or intuition (N)
- thinking (T) or feeling (F)
- judging (J) or perceiving (P)

A derivative version of the MBTI, developed by Elizabeth Murphy and Charles Meisgeier, is available for children age seven through 13 (grades two through eight). The assessment, called the Murphy-Meisgeier Type Indicator for Children (MMTIC) uses the same four psychological types as the MBTI, but is written for a second grade reading level.

Precautions

The MBTI should only be administered, scored, and interpreted by a professional trained in its use (except in the case of Form M, which can be self-scored but should still be administered and interpreted by a professional). Cultural and language differences in the test subject may affect performance and may result in inaccurate test results. The test administrator should be informed before testing begins if the test taker is not fluent in English and/or he or she has a unique cultural background.

Preparation

Prior to the administration of the MBTI, the test subject should be fully informed about the nature of the test and its intended use. He or she should also receive standardized instructions for taking the test and any information on the confidentiality of the results.

KEY TERMS

Multi-tasking—Performing multiple duties or taking on multiple responsibilities and roles simultaneously.

Vocational—Relating to an occupation, career, or job.

Normal results

Myers-Briggs results are reported as a four-letter personality type (e.g., ESTP, ISFJ). Each letter corresponds to an individual's preference in each of the four pairs of personality indicators (i.e., E or I, S or N, T or F, and J or P). There are a total of sixteen possible combinations of personality types on the MBTI.

- Letter One: E or I: Extraverts focus more on people and things, introverts on ideas.
- Letter Two: S or N: Sensing-dominant personalities prefer to perceive things through sight, sound, taste, touch, and smell, while intuition-dominant types look to past experience and are more abstract in their thinking.
- Letter Three: T or F: The third subtype is a measure of how people use judgment. Thinking types use logic to judge the world, while feeling types tend to view things on the basis of what emotions they invoke.
- Letter Four: J or P: Everyone judges and perceives, but those who are judging dominant are said to be more methodical and results-oriented, while perceiving dominant personalities are good at multi-tasking and are flexible.

Risks

There are no risks involved with the Myers-Briggs Type Indicator test.

Parental concerns

When interpreting test results, the test administrator will review what the test evaluates, its precision in evaluation and any margins of error involved in scoring, and what the individual scores mean in the context of overall norms for the test and the background of the adolescent.

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Myopathies

Definition

Myopathies are diseases of skeletal muscle that are not caused by nerve disorders. These diseases cause the skeletal or voluntary muscles to become weak or shrunken (atrophied).

Description

There are many different types of myopathies. Some are inherited, some inflammatory, and some caused by endocrine or metabolic problems. Myopathies usually are not fatal. Typically they cause muscle weakness and movement problems. The shoulders and thigh muscles are usually, but not always, affected earlier than the muscles of the hands and feet. Most myopathies are degenerative, meaning they become more pronounced over time. Some weaknesses are transitory. Only rarely do individuals become dependent on a wheelchair. However, **muscular dystrophy** (technically a myopathy) is far more severe. Some types of muscular dystrophy are fatal in early adulthood.

Causes and symptoms

There is great variety among myopathies, but what they all share are effects on the skeletal muscles. The main causes of myopathies are genetic, inflammatory (caused by infection), endocrine (hormonal), and metabolic (errors in how cells function). Often the cause of the myopathy is not known (idiopathic disease).

Genetic myopathies

Among their many functions, genes are responsible for overseeing the production of proteins important in maintaining healthy cells. Muscle cells produce thousands of proteins. With each of the inherited myopathies, a genetic defect is linked to a lack of, or defect in, one of the proteins needed for normal muscle cell function.

There are several different kinds of myopathy caused by defective genes:

- central core disease
- centronuclear (myotubular) myopathy
- myotonia congenita
- nemaline myopathy
- paramyotonia congenita
- periodic paralysis (hypokalemic and hyperkalemic forms)
- mitochondrial myopathies

Most, but not all, of these genetic myopathies are inherited through an autosomal dominant pattern of inheritance. In this pattern of inheritance, one copy of each gene comes from each parent. Only one of these two copies needs to have the mutation (change) or defect in order for the child to have the disease. The parent with the defective gene has the disease, and each of this parent's children has a 50 percent chance of inheriting the disease. This percentage is not changed by results of other pregnancies. With this pattern of inheritance, male and female children are equally at risk of developing the disease.

However, for a child to have one type of myotonia congenita and some forms of nemaline myopathy, two defective genes must be inherited—one from each parents. This is called an autosomal recessive pattern of inheritance. Neither parent may have symptoms of the disease, but each carries a recessive defective gene for it. Each child of such parents has a 25 percent chance of inheriting both genes and showing signs of the disease, and a 50 percent chance of inheriting one defective gene

from only one parent. If the child has inherited just one defective gene, he or she will be a carrier of the disease and can pass the gene on to his or her offspring, while showing no signs of the disease himself.

A few forms of centronuclear myopathy develop primarily in males. Females who inherit the defective gene are usually carriers without symptoms, like their mothers, but they can pass on the disease to their sons. Mitochondrial myopathies are inherited only through the mother, since sperm do not contain mitochondria.

The major symptoms associated with the genetic myopathies are:

- Central core disease: mild weakness of voluntary muscles, especially in the hips and legs; hip displacement; delays in reaching developmental motor milestones; problems with running, jumping, and climbing stairs develop in childhood.
- Centronuclear myopathy: weakness of voluntary muscles, including those on the face, arms, legs, and trunk; drooping upper eyelids; facial weakness; foot drop; affected muscles almost always lack reflexes.
- Myotonia congenita: voluntary muscles of the arms, legs, and face stiff or slow to relax after contracting (myotonia); stiffness triggered by fatigue, stress, cold, or long rest periods, such as a night's **sleep**; stiffness can be relieved by repeated movement of the affected muscles.
- Nemaline myopathy: moderate weakness of voluntary muscles in the arms, legs, and trunk; mild weakness of facial muscles; delays in reaching developmental motor milestones; decreased or absent reflexes in affected muscles; long, narrow face; high-arched palate; jaw projects beyond upper part of the face.
- Paramyotonia congenita: stiffness of voluntary muscles in the face, hands, and forearms; attacks spontaneous or triggered by cold temperatures; stiffness made worse by repeated movement; episodes of stiffness last longer than those seen in myotonia congenita.
- Periodic paralysis: attacks of temporary muscle weakness (muscles work normally between attacks); in the hypokalemic (low potassium) form, attacks triggered by vigorous **exercise**, heavy meals high in carbohydrates, insulin, stress, alcohol, infection, pregnancy; in the hyperkalemic (high potassium) form, attacks triggered by vigorous exercise, stress, pregnancy, missing a meal, steroid drugs, high potassium intake.
- Mitochondrial myopathies: symptoms vary quite widely with the form of the disease and may include progressive weakness of the eye muscles (ocular myopathy), weakness of the arms and legs, or multi-system problems primarily involving the brain and muscles.

Endocrine-related myopathies

In some cases, myopathies can be caused by a malfunctioning endocrine gland that produces either too much or too little of the chemical messengers called hormones. Hormones travel through the bloodstream. One of their many functions is to help regulate muscle activity. Problems in producing hormones can lead to muscle weakness.

Hyperthyroid myopathy and hypothyroid myopathy affect different muscles in different ways. Hyperthyroid myopathy occurs when the thyroid gland produces too much of the hormone thyroxine, leading to muscle weakness, some muscle wasting in hips and shoulders, and, sometimes, problems with eye muscles. The hypothyroid type of myopathy occurs when too little hormone is produced, leading to stiffness, cramps, and weakness of arm and leg muscles.

Inflammatory myopathies

Some myopathies are caused by inflammation. Inflammation is a protective response of injured tissues characterized by redness, increased heat, swelling, and/or **pain** in the affected area. Examples of this type of myopathy include **dermatomyositis**, polymyositis, and myositis ossificans.

Dermatomyositis is a disease of the connective tissue that also involves weak, tender, inflamed muscles. Muscle tissue loss may be so severe that the individual may be unable to walk. Skin inflammation is also present. The cause of dermatomyositis is as of 2004 unknown, but viral infection and antibiotic use are associated with the condition. In some cases, dermatomyositis is associated with rheumatologic disease or **cancer**. Polymyositis involves inflammation of many muscles, usually accompanied by deformity, swelling, sleeplessness, pain, sweating, and tension. It, too, may be associated with cancer. Myositis ossificans is a rare inherited disease in which muscle tissue is replaced by bone, beginning in childhood.

Muscular dystrophies

While considered a separate group of diseases, the muscular dystrophies also involve muscle wasting and can be described as myopathies. Symptoms of muscular dystrophy (MD) diseases usually appear during childhood and **adolescence**. These are genetic disorders that result in defects in the production of specific proteins. The forms of muscular dystrophy differ according to the way they are inherited, the age at which symptoms begin, the muscles they affect, and how fast they progress.

Demographics

Myopathies are not common. About 14 percent of myopathies are inherited. Worldwide the rate of inflammatory myopathies is about five to ten individuals per 100,000. These myopathies are more often seen in women. MD is found in about 63 of every 1 million individuals, but the rates vary widely depending on the type of MD. The most common type is Duchenne MD, affecting one in every 3,300 boys. Other more common types of MD are Becker's, **myotonic dystrophy**, limb-girdle MD, and facioscapulohumeral MD. MD is more common in boys. The rate of metabolic and endocrine myopathies was, as of 2004, not known.

When to call the doctor

Parents should let the doctor know as soon as possible if there is a **family** history of muscle weakness or muscle wasting disease. Otherwise, they should contact their pediatrician if the child is showing any signs of delayed or abnormal growth or unexplained muscle weakness.

Diagnosis

Early diagnosis of myopathy is important in order to provide the best care possible. An experienced physician can diagnose a myopathy by evaluating a child's medical history and by performing a thorough physical examination. Diagnostic tests can help differentiate among the different types of myopathies, as well as between myopathy and other neuromuscular disorders. If the doctor suspects a genetic myopathy, a thorough family history will also be taken. Genetic tests are available for a few myopathies.

Diagnostic tests the doctor may order include: measurements of potassium, (K) creatine kinase,(CK) lactic dehydrogenase (LDH) and pyruvate kinase (PK) and certain antibodies in the blood; muscle tissue biopsy; and electromyogram (EMG).

Treatment

As of 2004, there was no cure for many myopathies. Treatment depends on the specific type of myopathy the person has and is aimed at controlling symptoms. Specific treatment approaches for specific forms of myopathies are as follows:

- periodic paralysis: medication and dietary changes
- hyperthyroid or hypothyroid myopathy: treatment of the underlying thyroid abnormality
- myositis ossificans: medication to help prevent abnormal bone formation, but there is no cure following onset

KEY TERMS

Electrooculography (EOG)—A diagnostic test that records the electrical activity of the muscles that control eye movement.

Hyperkalemia—An abnormally high level of potassium in the blood.

Hypokalemia—A condition characterized by a deficiency of potassium in the blood.

Inflammation—Pain, redness, swelling, and heat that develop in response to tissue irritation or injury. It usually is caused by the immune system's response to the body's contact with a foreign substance, such as an allergen or pathogen.

Mitochondria—Spherical or rod-shaped structures of the cell. Mitochondria contain genetic material (DNA and RNA) and are responsible for converting food to energy.

Voluntary muscles—Muscles that can be moved by conscious thought.

- central core disease: no treatment
- nemaline myopathy: no treatment
- centronuclear (myotubular) myopathy: no treatment
- paramyotonia congenita: treatment often unnecessary
- myotonia congenita: drug treatment (if necessary), but drugs do not affect the underlying disease, and attacks may still occur

General treatments aim at supporting the individual's functioning and independence. Physical therapy can help preserve or increase strength and flexibility in muscles. Ankle and wrist braces can support weakened limbs. Occupational therapy is used to develop tools and techniques to compensate for loss of strength and dexterity. A speech-language pathologist can provide retraining for weakness in the muscles controlling speech and swallowing.

Prognosis

The prognosis for patients with myopathy depends on the type and severity of the individual's disease. In most cases, the myopathy symptoms can be successfully treated, but in others, the disease can be fatal in childhood or adolescence.

Muscular dystrophy is generally a more serious disease than many other types of myopathies. Duchenne's

MD is usually fatal by the late teens; Becker's MD is less serious and may not be fatal until the 50s.

Prevention

As of 2004 there is no way to prevent the genetic mutations that cause myopathies, nor are there ways to prevent metabolic and endocrine failures that result in myopathies. Inflammatory myopathies often occur as a result of exposure to viruses or drugs, but it is almost impossible to predict their development.

Parental concerns

Individuals with known myopathies who wish to become parents may want to seek genetic counseling before attempting to have children.

See also Muscular dystrophy; Myotonic dystrophy.

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Myopia

Definition

Myopia, or nearsightedness, is a condition in which objects in the distance are blurred either because the eye is too long or too strong. It is the result of both environmental and genetic factors.

Description

The degree to which one is myopic depends on the powers of the cornea and the lens of the eye and the length of the eyeball. In a normal eye the incoming visual images meet on the retina in the back of the eye. If these visual images converge in front of the retina instead of on the retina, then one is myopic.

There are several types of myopia, of which simple myopia is the most common. Individuals with simple myopia have eyes that are either too long or too powerful. Congenital myopia develops in infants. Individuals with high myopia, greater than six diopters, can develop pathological changes in the retina, called degenerative myopia. Nocturnal myopia, another type of myopia sometimes referred to as "night blindness," is blurred vision only in darkness. Myopia can also be induced by co-existing medical problems and drug exposure.

A child's refractive status or power of the eye when he or she begins school is a good indicator of whether the child will become nearsighted. Most children are hyperopic, or far-sighted, at birth and experience a decrease in far-sightedness throughout early childhood. Myopia is less likely to develop by age 13 if a child still has at least 0.75 diopters of hyperopia at age eight. But if a child has become at least 0.25 diopters myopic at this age then there is a 60 percent chance that the child will require spectacle correction for nearsightedness by age 13.

Myopia does decreases in later life. This appears not to be related to a decrease in close work as is often suggested but rather due to some factor intrinsically related to ageing. It has been hypothesized the power of the lens of the eye changes in later life.

High myopia has been associated with various syndromes: Ehlers-Danlos, Marfan, Down, and Stickler syndromes. Myopia is often observed in retinopathy of **prematurity** (ROP). ROP is seen in 68 percent of infants with low birth weights and over 80 percent of infants born with ROP will be myopic. The myopia associated with ROP increases through age five, after which it stabilizes.

Transmission

Although no gene for myopia has been isolated, heredity is believed to play a role in myopia. If both parents are myopic, then the odds that the child will be myopic are as high as 60 percent. This drops to at most 40 percent when only one parent is nearsighted, and for 15 percent of myopic children, neither parent has myopia. High myopia is especially likely to have a genetic component.

Demographics

Overall, 25 percent of those living in the United States are myopic. Myopia is slightly more prevalent among females than males, and among those with advanced academic training. Less than 5 percent of five year olds are myopic, but this percentage increases to 25 percent by late teens and to approximately 35 percent for young adults and to 42 percent of those middle-aged. These percentages decrease to 20 percent by age 65 and to less than 13 percent by age 80.

Myopia is more prevalent in Asian countries; as much as 70 percent of the Chinese population is nearsighted.

Causes and symptoms

Congenital myopia develops because of an obstruction along the visual pathway such as cataract. The eye becomes elongated in response to blur these causes, creating a myopic eye.

A first sign that a child might have myopia is difficulty in seeing things in the distance, such as the chalkboard. The child may not see things in the distance as well as a classmate or sibling.

For the 2 percent of the population who are extremely nearsighted, an inherently weak sclera, whose fibers are not held together tightly, causes the eye to stretch. This stretching can continue into adulthood, increasing myopia.

Other causes of increasing nearsightedness include difficulty with converging, the process through which the eyes move inwardly together when reading, and esophoria, the condition in which the eyes are more comfortable positioned close to the nose. Doing a lot of close work, such as playing **video games** and using the computer for extended periods, may increase myopia for these children.

Causes of induced myopia include cataracts and elevations of blood sugar in diagnosed or undiagnosed diabetics. Some drugs, such as corticosteroids, **antihistamines**, and some **antibiotics**, including **sulfonamides**, can induce myopia.

Another cause of increasing myopia is the over wearing of **contact lenses**. Swelling of the cornea can occur if the eye does not have sufficient oxygen causing a transient increase in myopia. Silicone contact lenses allow a marked increase in oxygen to reach the eye decreasing the probability of myopic increases.

For the child with diabetes, fluctuations in blood sugars can cause swelling of the cornea of the eye, leading to transient increases in myopia, which stabilize once the diabetes is controlled. But the child may independently become more myopic later in life.

In the early 2000s, it has been suggested that insulin resistance, which accompanies type 2 diabetes and prediabetes, may increase myopia in children and adolescents. The level of insulin-like growth factor binding protein 3 (IGFBP-3), a hormone that works with insulin to lower blood glucose levels, is low in individuals who are insulin resistant. This decreased level, in turn, decreases the sensitivity of ocular tissue to another compound called retinoic acid, which prevents increases in the length of the eye. Thus, if insulin levels are higher than normal, the risk of myopia may be increased.

Asthenopia or a feeling of eye strain is not common in myopia. If a child complains of eye strain, then usually there is another cause of the eye strain, including an astigmatism, a condition in which the eye is football shaped; anisometropia, a condition in which the eyes are of different powers; or difficulty with focusing.

When to call the doctor

There are many reasons why a child cannot see well in the distance. Myopia is the most common cause of distance blur, and since much of what a child learns comes from vision and visual cues, correction of myopia is important. Also, problems with vision may be a sign of a more serious ocular problem, such as cataracts, or of a medical problem, such as diabetes.

Diagnosis

Myopia is diagnosed by determining a child's unaided vision and is confirmed objectively by the eye care practitioner with various techniques, including retinoscopy and refraction.

The type and extent of myopia is determined by additional testing. These tests include an evaluation of the child's binocular vision, his eye movements, his ability to converge and focus on objects close-up, and his ocular health. Dilation of the eye allows the doctor to check for complications of ROP, diabetes, or degenerative myopia. Since children are capable of over focusing, dilation can help the eye care practitioner determine a child's true prescription because the drugs used to dilate also impair this tendency to over focus. Over focusing can cause a child to appear to be myopic when he or she is actually not.

Treatment

Myopia is most commonly treated with spectacles or glasses. Myopia in **preschool** children does not need to be corrected with glasses, unless either anisometropia, a condition in which there is a difference of more than 1.00 diopters between the two eyes, or **amblyopia**, a condition in which a child cannot be corrected to 20/20 with spectacles, is present. As the child enters school, distance vision becomes critical for learning, and children with prescriptions of at least 1.00 diopter of myopia or who have 20/40 vision or worse should be given glasses. Once a child is diagnosed with myopia, he or she should be examined every six months to a year, and each eye should be corrected to 20/20 at each visit. The glasses are then usually worn full time, except for children with difficulty with convergence (esophoria), who may remove their glasses for close work.

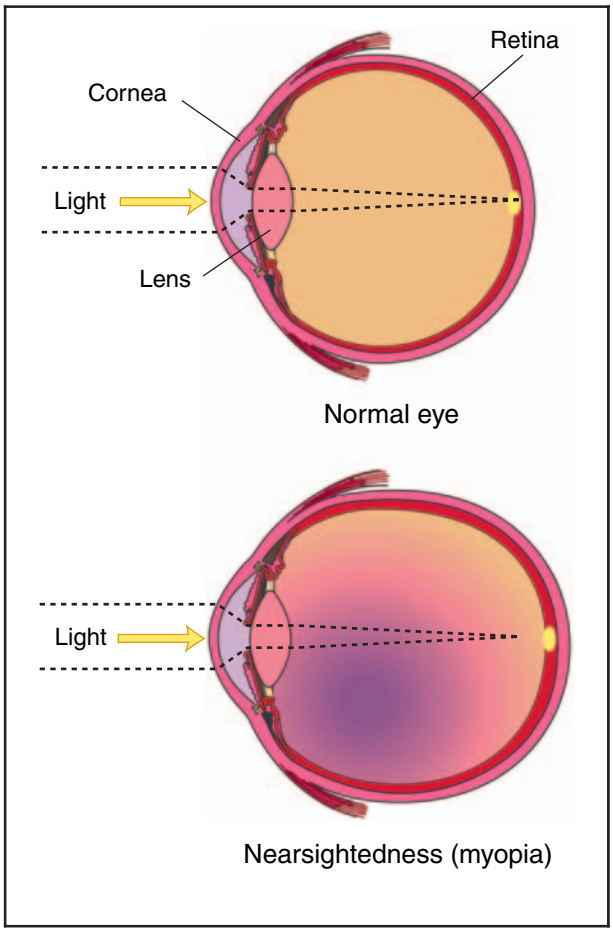
Some adolescents may want to wear contact lenses. Wearing contact lenses can improve appearance. Peripheral vision is improved with contact lenses, especially for those with high degrees of myopia or who have anisometropia.

Rigid gas permeable (RGPs) contact lenses are used to correct myopia. The rate at which myopia increases may be slowed by RGP lenses. RGPs are also employed in orthokeratology, a technique in which RGP lenses of gradually decreasing flatness are worn for specified amounts of time. These lenses, called ortho-K lenses, flatten the cornea, changing the power of the cornea over time and decreasing myopia. This effect of ortho-K lenses is not permanent and an ortho-K lens must be worn periodically or the original myopia reappears. For some individuals—for example, those with keratoconnus, a disease of the cornea—RGPs may offer the only way to correct vision.

For some children the development of myopia may be slowed with reading glasses or bifocals. If bifocals are prescribed, then either progressive or no-lines, or a lined bifocal may be given. If a lined bifocal is prescribed, then the line is always placed higher for the child than for the adult. This is done to encourage use of the power of the bifocal.

Refractive surgery is also used to correct myopia, but only on fully grown individuals. A child's eyes change and the **safety** of these procedures have not been established in the growing eye.

The most common surgical procedure performed to correct myopia is laser in situ keratomileusis (LASIK). Other techniques to correct myopia include photorefractive keratectomy (PRK), radial keratomy (RK), laser epithelial keratomileusis (LASEK), intraocular lens implants and intrastromal corneal rings. Inflammation of the eye, increased dryness of the eye, and cataracts are some of the risks associated with refractive surgery.



Myopia, or nearsightedness, is a condition of the eye in which objects are seen more clearly when close to the eye while distant objects appear blurred or fuzzy. *(Illustration by Electronic Illustrators Group.)*

Alternative treatment

Cycloplegic drugs, such as atropine, may decrease myopia, but they may hinder the child's ability to see up close. As of 2004 pirenzepine, which has shown to decrease the rate of myopia in children without sacrificing the ability of the child to do close work, is in clinical trial.

Prognosis

Most infants are born far-sighted and eventually reach emmetropia or normal vision, by age six. Over one third of children go on to become myopic as adults.

KEY TERMS

Accommodation—The ability of the lens to change its focus from distant to near objects and vice versa. It is achieved through the action of the ciliary muscles that change the shape of the lens.

Anisometropia—An eye condition in which there is an inequality of vision between the two eyes. There may be unequal amounts of nearsightedness, farsightedness, or astigmatism, so that one eye will be in focus while the other will not.

Astigmatism—An eye condition in which the cornea doesn't focus light properly on the retina, resulting in a blurred image.

Cataract—A condition in which the lens of the eye turns cloudy and interferes with vision.

Convergence—The natural movement of the eyes inward to view objects close-up.

Cornea—The clear, dome-shaped outer covering of the eye that lies in front of the iris and pupil. The cornea lets light into the eye.

Diopter (D)—A unit of measure for describing the refractive power of a lens.

Emmetropia—Normal vision.

Retina—The inner, light-sensitive layer of the eye containing rods and cones. The retina transforms the image it receives into electrical signals that are sent to the brain via the optic nerve.

Sclera—The tough, fibrous, white outer protective covering of the eyeball.

Patients with high myopia, greater than 6.00 diopters, have an increased risk of developing a retinal tear, hole, or detachment; a posterior staphyloma; a posterior vitreous detachment; or glaucoma. Rarely are these pathological changes of myopia seen in children or adolescents. Retinal detachments and tears are possible, however, in highly myopic children or adolescents who play contact **sports**. If a retinal problem is diagnosed or suspected, referral to a retinal specialist is necessary.

Prevention

For individuals who have difficulty with convergence or focusing or who are esophoric, close work may increase myopia. Children diagnosed with these problems would benefit from frequent breaks while doing close work. Increases in myopia for these children may be slowed with bifocals and/or removal of glasses for reading and homework.

Nutritional concerns

Since elevated levels of insulin may be associated with increased myopia; a diet low in those foods that increase insulin secretion, such as refined carbohydrates, may help decrease myopia.

Parental concerns

Children rely on their vision in their learning processes; if they have difficulty seeing this handicap affects academic performance. Thus, any vision problem should be corrected promptly. Once myopia is diagnosed, it typically increases through childhood and vision correction is needed for classroom work and for sports. If a child is not corrected fully and continues to have blurred vision, the eye may elongate in response to blur, perpetuating the myopia.

Myopia cannot be diagnosed by school screenings or by simply reading eye chart at the pediatrician's office. A comprehensive eye exam as given by an ophthalmologist or an optometrist is needed, if myopia is suspected. Most cases of myopia result from changes within the eye, but the condition can be a manifestation of other more serious problems, such as cataract or diabetes.

See also Diabetes mellitus; Ehlers-Danlos syndrome; Marfan's syndrome.

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Myotonic dystrophy

Definition

Myotonic dystrophy is a progressive disease in which the muscles are weak and slow to relax after contraction.

Description

Myotonic dystrophy (DM), also called dystrophia myotonica, myotonia atrophica, or Steinert disease, is a common form of **muscular dystrophy**. DM is an inherited disease. It causes general weakness, usually beginning in the muscles of the hands, feet, neck, or face. It slowly progresses to involve other muscle groups, including the heart and a wide variety of other organ systems.

There are four types of DM as determined by when symptoms appear. These are:

- Congenital: Severe symptoms are apparent at birth.
- Juvenile: Symptoms appear between birth and adolescence.
- Adult: Symptoms appear in individuals ages 20–40.
- Late onset: Mild symptoms appear after age 40.

Transmission

DM is an inherited disease. It is passed from parent to child through an autosomal dominant pattern of inheritance. In the case of DM, one copy of each gene is inherited from each parent. In an autosomal dominant pattern of inheritance, only one of these two copies needs to have the mutation (change) or defect in order for the child to have DM. Therefore, there is a 50 percent chance that a parent who has DM will pass it onto each child. This percentage is not changed by results of other pregnancies. In each pregnancy, a parent with DM has a 50% chance of having a child with DM.

Demographics

Myotonic dystrophy is an uncommon disease occurring in about one out of every 8,000 individuals. It is found worldwide. The congenital form of DM is much rarer, occurring in only about one out of every 100,000 births. DM affects males and females approximately equally.

Causes and symptoms

The most common type of DM is called DM1, which is caused by a mutation in a gene called myotonic dystrophy protein kinase (DMPK). The DMPK gene is located on chromosome 19. The specific mutation that causes DM1 is called a trinucleotide repeat expansion. In people who have DM1, a particular unit of the gene is repeated too many times—more than the normal range of five to 38 times—and thus this section of the gene is too big and is unstable. The enlarged section of the gene is called a trinucleotide repeat expansion.

People who have repeat numbers in the normal range will not develop DM1 and cannot pass it to their children. Having more than 50 repeats causes DM1. People who have 38–49 repeats have what is called a premutation. They do not develop DM1, but can pass DM1 on to their children.

Myotonic dystrophy has an effect called "anticipation." This means that when a person with repeat numbers in the affected or premutation range (above 38) has children, the expansion grows larger, and the child has more of the repeated genetic unit (a higher repeat number). As a result, symptoms of the disease tend to appear at an earlier age in children than in their affected parent. Anticipation happens more often when a mother, rather than the father, passes DM1 to children. Occasionally, repeat sizes stay the same or even get smaller when they are passed to a person's children.

In general, the more repeats above 38 an individual has, the earlier the age of onset of symptoms and the more severe the symptoms. Having repeat numbers greater than 1,000 causes congenital myotonic dystrophy. However, this is a general rule. It is not possible to look at a person's repeat number and predict at what age

he or she will begin to have symptoms or how the condition will progress.

Some families with symptoms of DM do not have a mutation in the DMPK gene. Instead, they have a mutation in a gene on chromosome 3 that causes four units within the gene to be repeated. This genetic defect is called DM2 or proximal myotonic myopathia (PROMM). Symptoms of DM2 are almost never apparent at birth. This defect has only been decoded since 2001; therefore, less is known about how it functions.

Symptoms of DM vary in severity, and not everyone will have all of the symptoms. In general, myotonic dystrophy causes weakness and delayed muscle relaxation called myotonia. Exactly how the repeat of genetic information causes myotonia, the inability to relax muscles, is not yet understood. The disease somehow blocks the flow of electrical impulses across the muscle cell membrane. Without proper flow of charged particles, the muscle cannot return to its relaxed state after it has contracted.

The most severe form of DM, congenital myotonic dystrophy, may appear in newborns of mothers who have DM1. Congenital myotonic dystrophy is marked by severe weakness, poor sucking and swallowing responses, respiratory difficulty, delayed motor development, and **mental retardation**. Death in infancy is common in babies with congenital DM.

Symptoms of juvenile and adult onset DM include facial weakness and a slack jaw, drooping eyelids called ptosis, and muscle wasting in the forearms and calves. A person with DM has difficulty relaxing his or her grasp, especially in the cold. DM affects the heart muscle, causing irregularities in the heartbeat. It also affects the muscles of the digestive system, causing **constipation** and other digestive problems. DM may cause cataracts in the eye, retinal degeneration, low IQ, early frontal balding, skin disorders, atrophy of the testicles, and diabetes. It can also cause **sleep** apnea, a condition in which normal breathing is interrupted during sleep. DM increases the need for sleep and decreases motivation. Often, severe disabilities do not set in until about 20 years after symptoms begin. Most people with myotonic dystrophy maintain the ability to walk, even late in life.

Some people who have a trinucleotide repeat expansion in their DMPK gene do not have DM symptoms or have very mild symptoms that go unnoticed. It is not unusual for a woman to be diagnosed with DM after she has an infant with congenital myotonic dystrophy.

When to call the doctor

Parents should let the doctor know as soon as possible if there is a **family** history of DM. Otherwise, they should contact their pediatrician if the child shows any signs of delayed or abnormal growth, or unexplained muscle weakness.

Diagnosis

Diagnosis of DM is not difficult once the disease is considered. However, the diagnosis may be masked because symptoms can begin at any age, can be mild or severe, and can occur with a wide variety of associated complaints. Diagnosis of DM begins with a careful medical history and a thorough physical examination to determine the distribution of symptoms and to rule out other causes. A family history of DM or unexplained weakness helps to establish the diagnosis.

Genetic testing, usually using a blood sample, establishes a definitive diagnosis of DM. The DNA in the blood cells is examined and the number of repeats in the affected gene is determined. Other tests may be done to help establish the diagnosis, but only rarely would other testing be needed. An electromyogram (EMG) is a test used to examine how muscles respond to stimulation. Characteristic changes revealed by this test, and seen in DM, help distinguish it from other muscle diseases. Removing a small piece of muscle tissue for microscopic examination is called a muscle biopsy. DM is marked by characteristic changes in the structure of muscle cells that can be seen on a muscle biopsy. An electrocardiogram could be performed to detect abnormalities in heart rhythm associated with DM. These symptoms often appear later in the course of the disease.

If genetic testing in a family has identified a DMPK mutation, it is possible to test a fetus during pregnancy. Testing can be done at 10–12 weeks gestation by a procedure called chorionic villus sampling (CVS) that involves removing a tiny piece of the placenta and analyzing DNA from its cells. It can also be done by **amniocentesis** after 14 weeks gestation by removing a small amount of the amniotic fluid surrounding the fetus and analyzing the cells in the fluid. Each of these procedures carries a small risk of miscarriage. Those who are interested in learning more should check with their doctor or genetic counselor.

Treatment

Myotonic dystrophy cannot be cured, and no treatment can delay its progression. However, many of its symptoms can be treated. Physical therapy can help

KEY TERMS

Electrocardiagram (ECG, EKG)—A record of the electrical activity of the heart, with each wave being labeled as P, Q, R, S, and T waves. It is often used in the diagnosis of cases of abnormal cardiac rhythm and myocardial damage.

Electromyography (EMG)—A diagnostic test that records the electrical activity of muscles. In the test, small electrodes are placed on or in the skin; the patterns of electrical activity are projected on a screen or over a loudspeaker. This procedure is used to test for muscle disorders, including muscular dystrophy.

Muscular dystrophy—A group of inherited diseases characterized by progressive wasting of the muscles.

Sleep apnea—A sleep disorder characterized by periods of breathing cessation lasting for 10 seconds or more.

Trinucleotide repeat expansion—A sequence of three nucleotides that is repeated too many times in a section of a gene.

preserve or increase strength and flexibility in muscles. Ankle and wrist braces can support weakened limbs. Occupational therapy is used to develop tools and techniques to compensate for loss of strength and dexterity. A speech-language pathologist can provide retraining for weakness in the muscles controlling speech and swallowing.

Irregularities in heartbeat may be treated with medication or a pacemaker. A yearly electrocardiogram is usually recommended. **Diabetes mellitus** in DM is treated in the same way that it is in the general population. A high-fiber diet can help prevent constipation. Sleep apnea may be treated with surgical procedures to open the airways or with nighttime ventilation. Treatment of sleep apnea may reduce drowsiness. Lens replacement surgery is available when cataracts develop.

Prognosis

The course of myotonic dystrophy varies. When symptoms appear earlier in life, disability tends to become more severe. Occasionally people with DM may require a wheelchair later in life. Children with congenital DM often die in infancy. If they survive, they usually require special educational programs and physical and occupational therapies. Respiratory infections pose a danger if weakness becomes severe.

Prevention

There is no way to prevent the genetic mutations that cause DM. However, it is possible to test someone who is at risk for developing DM1 before symptoms arise, to see whether he or she inherited an expanded trinucleotide repeat. This is called predictive testing. Predictive testing cannot determine the age at which someone will begin to have symptoms or the course of the disease.

Another procedure, called preimplantation diagnosis, allows a couple to have a child that does not have the genetic condition. This procedure is still experimental. Those interested in learning more about the procedure should check with their doctor or genetic counselor.

Parental concerns

Pregnant woman should be cared for by an obstetrician familiar with the particular problems of DM because complications can occur during pregnancy, labor, and delivery.

It is advisable for children or adults with DM to wear a medical alert bracelet. Some emergency medications may have dangerous effects on the heart rhythm in a person with DM. Adverse reactions to general anesthesia may also occur.

See also Muscular dystrophy.

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Myringotomy and ear tubes

Definition

Myringotomy is a surgical procedure in which a small incision is made in the eardrum (the tympanic membrane), usually in both ears. The word comes from *myringa*, modern Latin for drum membrane, and *tomē*, Greek for cutting. It is also called myringocentesis, tympanotomy, tympanostomy, or paracentesis of the tympanic membrane. The doctor can withdraw fluid from the middle ear through the incision.

Ear tubes, or tympanostomy tubes, are small tubes open at both ends that are inserted into the incisions in the eardrums during a myringotomy. The tubes come in various shapes and sizes and are made of plastic, metal, or both. They are left in place until they fall out by themselves or until they are removed by a doctor. Ear tubes are also sometimes called ventilation tubes.

Purpose

Myringotomy with the insertion of ear tubes is an optional treatment for inflammation of the middle ear with fluid collection (effusion), also called glue ear, that lasts more than three months (chronic **otitis media** with effusion) and does not respond to drug treatment. Myringotomy is the recommended treatment if the condition lasts four to six months. Effusion is the collection of fluid that escapes from blood vessels or the lymphatic system. In this case, the effusion collects in the child's middle ear.

Initially, acute inflammation of the middle ear with effusion is treated with one or two courses of antibiotic drugs. **Antihistamines** and **decongestants** have also been used to treat otitis media, but they have not been proven effective unless the child also has hay fever or some other allergic inflammation that contributes to the ear problem. Myringotomy with or without the insertion of ear tubes is *not* recommended as the initial treatment for otherwise healthy children with middle ear inflammation with effusion.

In about 10 percent of children, the ear effusion lasts for three months or longer; at that point the condition is considered chronic. Systemic steroids may help children with chronic ear infections, but the evidence that these drugs are beneficial is not clear, and there are risks associated with steroid use.

Myringotomy with insertion of ear tubes becomes an option when medical treatment does not stop the effusion after three months in a child who is one to three years old, is otherwise healthy, and has hearing loss in both ears. If the effusion lasts for four to six months, myringotomy with insertion of ear tubes may be recommended. Although doctors in the past sometimes removed the child's tonsils or adenoids to treat recurrent otitis media with effusion, this practice is not recommended as of the early 2000s.

Myringotomy may be performed to relieve the **pain** and other symptoms of otitis media; to restore the child's hearing; to take a sample of the fluid to examine in the laboratory in order to identify any microorganisms present; or to insert ventilation tubes.

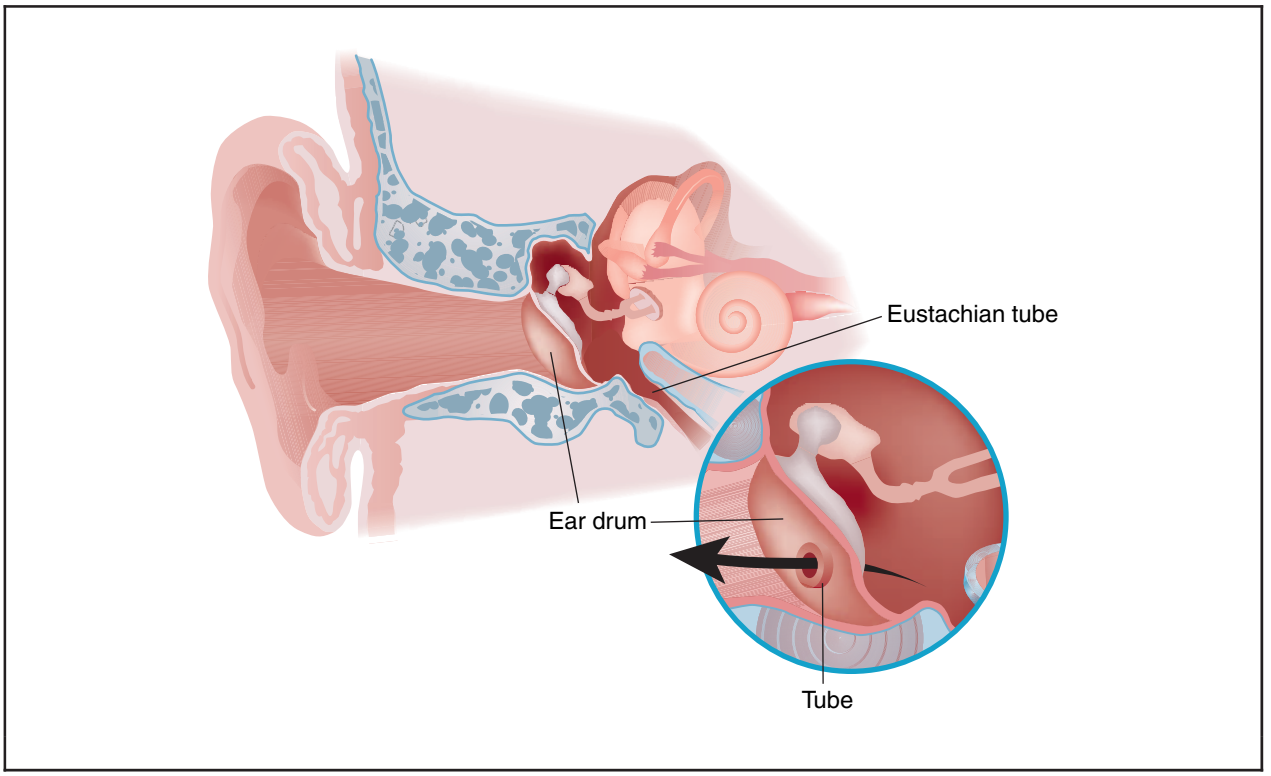
Ear tubes can be inserted into the incision during a myringotomy and left there. The eardrum heals around them, securing them in place. They usually fall out on their own in six to 12 months or are removed by a doctor.

While in place, the tubes keep the incision from closing, forming an open channel between the middle ear and the outer ear. This channel allows fresh air to reach the middle ear, allows fluid to drain out, and prevents pressure from building up in the middle ear. The patient's hearing returns to normal immediately and the risk of recurrence diminishes.

Parents often report that children talk better, hear better, are less irritable, **sleep** better, and behave better after myringotomy with the insertion of ear tubes.

Description

The procedure is usually performed in an ambulatory surgical unit under general anesthesia, although some physicians do it in the office with sedation and local anesthesia, especially in older children. Most primary care physicians prefer to refer children who need a myringotomy and tube placement to an otolaryngologist.



The insertion of ear tubes in the eardrum helps to alleviate chronic middle ear infections. *(Illustration by Argosy, Inc.)*

The ear is washed, a small incision made in the eardrum, the fluid sucked out, a tube inserted, and the ear packed with cotton to control bleeding.

Carbon dioxide lasers may also be used to perform the myringotomy. Laser-assisted myringotomy can be performed in a doctor's office with only a local anesthetic. It has several advantages over the older technique: it is less painful; less frightening to children; and minimizes the need for tube insertion because the hole in the eardrum produced by the laser remains open longer than an incision made with a scalpel. On the other hand, laser-assisted myringotomies have a higher rate of recurrence of infection.

Another technique to keep the incision in the eardrum open without the need for tube insertion is application of a medication called mitomycin C, which was originally developed to treat bladder **cancer**. The mitomycin prevents the incision from sealing over. As of the early 2000s, however, this technique is still in its experimental stages.

Some researchers have designed ear tubes that are easier to insert or to remove or that stay in place longer.

Precautions

As of 2004 clinical practice guidelines emphasized the importance of watchful waiting and medical treatment before performing a myringotomy and the importance of distinguishing between children at risk for speech or hearing problems from otitis media from others with chronic ear infections.

Preparation

A child scheduled for a myringotomy should not have food or water for four to six hours before being given anesthesia. **Antibiotics** are usually not needed before the procedure.

Aftercare

The use of antimicrobial drops after a myringotomy is controversial. Water should be kept out of the ear canal until the eardrum is intact. A doctor should be notified if the tubes fall out.

An additional element of postoperative care is the recommendation by many doctors that the child use ear plugs to keep water out of the ear during bathing or swimming to reduce the risk of infection and discharge.

KEY TERMS

Acute otitis media—Inflammation of the middle ear with signs of infection lasting less than three months.

Chronic otitis media—Inflammation of the middle ear with signs of infection lasting three months or longer.

Effusion—The escape of fluid from blood vessels or the lymphatic system and its collection in a cavity.

Middle ear—The cavity or space between the eardrum and the inner ear. It includes the eardrum, the three little bones (hammer, anvil, and stirrup) that transmit sound to the inner ear, and the eustachian tube, which connects the inner ear to the nasopharynx (the back of the nose).

Tympanic membrane—The eardrum, a thin disc of tissue that separates the outer ear from the middle ear. It can rupture if pressure in the ear is not equalized during airplane ascents and descents.

Tympanostomy tube—An ear tube. A tympanostomy tube is small tube made of metal or plastic that is inserted during myringotomy to ventilate the middle ear.

Risks

The risks of a myringotomy and ear tube placement include the following:

- cutting the outer ear
- formation of granular nodes due to inflammation at the site of the myringotomy
- formation of a cholesteatoma, which is a mass of skin cells and cholesterol in the middle ear that can grow and damage the surrounding bone
- permanent perforation of the eardrum
- hearing loss in late **adolescence** or early adulthood
- a 13 percent risk of persistent discharge from the ear (otorrhea)

If the procedure is repeated, structural changes in the eardrum can occur, such as loss of tone (flaccidity), shrinkage or retraction, or hardening of a spot on the eardrum (tympanosclerosis). The risk of hardening is 51 percent; its effects on hearing were not known as of 2004, but they are probably insignificant.

It is also possible that the incision will not heal properly, leaving a permanent hole in the eardrum, which can cause some hearing loss and increases the risk of infection.

It is also possible that the ear tube will move inward and get trapped in the middle ear rather than move out into the external ear, where it either falls out on its own or can be retrieved by a doctor. The exact incidence of tubes moving inward is not known, but this possibility could increase the risk of further episodes of middle-ear inflammation, inflammation of the eardrum or the part of the skull directly behind the ear, formation of a mass in the middle ear, or infection due to the presence of a foreign body.

The surgery may not be a permanent cure. As many as 30 percent of children undergoing myringotomy with insertion of ear tubes need to undergo another procedure within five years.

The other risks include the usual risks associated with sedatives or general anesthesia.

Parental concerns

Parental concerns with regard to a myringotomy and tube insertion are usually related to the risks associated with the procedure itself, such as the child's reaction to the anesthetic, the possibility that the procedure will have to be repeated at a later date, and the risk of eventual mild hearing loss. These potential complications against the risks of **language delay**, possible learning problems, or hearing loss resulting from chronic otitis media.

See also Ear exam with an otoscope; Hearing impairment; Otitis media.

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Myxedema *see* **Hypothyroidism**

N

Nail-patella syndrome

Definition

Nail-patella syndrome is a genetic disease of the connective tissue that produces defects in the fingernails, bone joints, and kidneys.

Description

Patients who have nail-patella syndrome may show a variety of physical defects. The most common features of this syndrome are missing or poorly developed fingernails, toenails, and patellae (kneecaps). Other common abnormalities include elbow deformities, abnormally shaped pelvis bone (hip bone), and kidney (renal) disease.

Less common medical findings include defects of the upper lip, the roof of the mouth, and unusual skeletal abnormalities. Skeletal abnormalities may include poorly developed shoulder blades (scapulae), sideways bent fingers (clinodactyly), **clubfoot**, **scoliosis**, and unusual neck bones. There are also other effects, such as thickening of the basement membrane in the skin and tiny clusters of capillaries (glomeruli) in the kidney. Nail-patella syndrome is associated with open-angle glaucoma, which, if untreated, may lead to blindness. Patients may also have cataracts, drooping eyelids (ptosis), or corneal problems such as glaucoma. In addition, scientists have recognized an association between nail-patella syndrome and colon **cancer**.

People with nail-patella syndrome may display only a few or many of the recognized signs of this disease, with symptoms varying widely from person to person. Symptoms even vary within a single **family** with multiple affected members.

Nail-patella syndrome is also known as Fong disease, hereditary onycho-osteodysplasia (HOOD), Iliac Horn syndrome, and Turner-Kieser syndrome.

Demographics

Nail-patella syndrome is a rare genetic disease. The incidence of nail-patella syndrome is approximately one in 50,000 births. This disorder affects males and females equally. It is found throughout the world and occurs in all ethnic groups. The strongest risk factor for nail-patella syndrome is a family history of the disease.

Causes and symptoms

Nail-patella syndrome has been recognized as an inherited disorder for over a hundred years. It is caused by mutations in a gene known as LIM Homeobox Transcription Factor 1-Beta (LMX1B), located on the long arm of chromosome 9. The LMX1B gene codes for a protein that is important in organizing embryonic limb development. Mutations in this gene have been detected in many unrelated people with nail-patella syndrome. Scientists have also been able to interrupt this gene in mice to produce defects similar to those seen in human nail-patella syndrome.

Nail-patella syndrome is inherited in an autosomal dominant manner. This means that possession of only one copy of the defective gene is enough to cause disease. When a parent has nail-patella syndrome, each of the children has a 50 percent chance to inherit the disease-causing mutation.

A new mutation causing nail-patella syndrome can also occur, causing disease in a child with no family history of the syndrome. This is called a sporadic occurrence and accounts for approximately 20 percent of cases of nail-patella syndrome. The children of a person with sporadic nail-patella syndrome are also at a 50 percent risk of inheriting the disorder.

Medical signs of nail-patella syndrome vary widely between patients. Some patients with this disorder do not display symptoms. These patients are discovered to have the nail-patella syndrome only when genetic studies are conducted to trace their family history.

The most obvious sign associated with nail-patella syndrome is absent, poorly developed, or unusual fingernails. Fingernail abnormalities are found in about 98 percent of children with this disorder. Abnormalities may be found in one or more fingernails. Only rarely are all fingernails affected. This disease most commonly affects the fingernails of the thumbs and index fingers. The pinky fingernail is least likely to be affected. Fingernails may be small and concave with pitting, ridges, splits, and/or discoloration. Toenails are less often affected.

Kneecap abnormalities, present in about 92 percent of children with this disorder, are the second most common sign associated with this disorder. Either or both kneecaps may be missing or poorly formed. If present, kneecaps are likely to be dislocated. The knees of people with nail-patella syndrome may have a square appearance. Besides the kneecap, other support structures including bones, ligaments, and tendons may also be malformed. Since these support structures stabilize the knee, patients with some leg malformations may have difficulty in walking. There may also be some hip bone anomaly or other skeletal symptoms, for example, clubfoot.

Kidney disease is present in about 30 to 50 percent of people with nail-patella syndrome. Kidney disease has been reported in children with nail-patella syndrome, but renal involvement more commonly develops during adulthood, usually during the fourth decade of life.

Eye problems may be present and vary from person to person. Nail-patella syndrome is thought to be associated with open-angle glaucoma, a condition caused by blockage of the outflow of fluid (aqueous humor) from the front chamber of the eyes. Nail-patella syndrome has also been associated with abnormalities of the cornea, cataracts, and astigmatism. Additionally, the irises of the eye may be multicolored.

When to call the doctor

The doctor should be called if a parent detects the symptoms of nail-patella syndrome in a child previously not diagnosed with the syndrome.

Diagnosis

Genetic testing for nail-patella syndrome is usually available only at research institutions that are working to further characterize this disorder. However, genetic testing cannot predict which signs of the disease will develop nor predict the severity of disease symptoms.

Diagnosis of this disease is most often made on visual medical symptoms such as the characteristic abnormalities of the fingernails and kneecaps. Diagnosis is confirmed by x-ray images of the affected bones and, when indicated, kidney biopsy. The bony pelvic spurs found in children with nail-patella syndrome are not associated with any other disease.

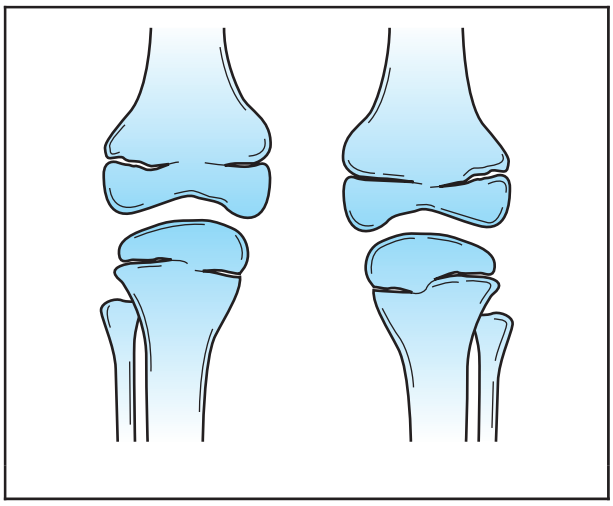

Illustration of bones around the knees showing absence of the patella in nail-patella syndrome. *(Illustration by Argosy, Inc.)*

Treatment

Treatment for children affected by nail-patella syndrome depends on the child's specific symptoms.

A wheelchair may be required if walking becomes painful due to bone, tendon, ligament, or muscle defects. Orthopedic surgery may be necessary for congenital clubfoot deformity. Manipulation or surgery may be required to correct hip dislocation. Cataracts are also surgically treated. Medical treatment at early signs of glaucoma prevents progression of the disease to blindness.

Controlling blood pressure may slow the rate of deterioration of kidney function. Severe kidney disease can be treated with dialysis or a kidney transplant. Children receiving kidney transplants do not develop nail-patella-type renal complications in their new kidney.

Because many possible manifestations of nail-patella syndrome exist, parents are advised to obtain extra medical care for their child with nail-patella syndrome, including regular urinalysis to monitor blood and protein levels to detect kidney disease as well as eye exams to detect glaucoma. Children with nail-patella syndrome should be periodically screened for scoliosis and lordosis.

KEY TERMS

Chorionic villus sampling—A procedure used for prenatal diagnosis at 10–12 weeks gestation. Under ultrasound guidance a needle is inserted either through the mother's vagina or abdominal wall and a sample of the chorionic membrane. These cells are then tested for chromosome abnormalities or other genetic diseases.

Glaucoma—A common eye disease characterized by increased fluid pressure in the eye that damages the optic nerve, which carries visual impulses to the brain. Glaucoma can be caused by another eye disorder, such as a tumor or congenital malformation, or can appear without obvious cause, but if untreated it generally leads to blindness.

Glomerulus—Plural, glomeruli; a network of capillaries located in the nephron of the kidney where wastes are filtered from the blood.

Hematuria—The presence of blood in the urine.

Hypnogogic hallucination—A vivid, dream-like hallucination, such as the sensation of falling, that occurs at the onset of sleep.

Patella—The kneecap.

Proteinuria—Abnormally large quantites of protein in the urine.

Prognosis

Symptoms vary from person to person and for one person through time. The long-term prognosis is extremely variable. One person may exhibit mild symptoms, while another person may become wheelchair-bound or require a kidney transplant.

Survival among patients with nail-patella syndrome is not decreased unless they exhibit renal complications. It is estimated that 8 percent of individuals with nail-patella syndrome who come to medical attention eventually die of kidney disease.

Prevention

Genetic counseling can be offered to persons who have the disease. Parents with this disease have a 50 percent chance of passing it to each of their children.

Parental concerns

Families may wish to seek counseling regarding the effects on relationships within the family after the birth of a child with nail-patella syndrome, for many people respond with guilt, **fear**, or blame when a genetic disorder is manifested within a family. Support groups are often good sources of information about nail-patella syndrome and can offer emotional and psychological support.

Resources

BOOKS

Nail Patella Syndrome: A Medical Dictionary, Bibliography, and Annotated Research Guide to Internet References. San Diego, CA: Icon Health Publications, 2004.

ORGANIZATIONS

Nail Patella Syndrome Networking/Support Group. 67 Woodlake Dr., Holland, PA 18966. Web site: <www.hometown.aol.com/pacali/npspage.html>.

National Organization for Rare Disorders Inc. 55 Kenosia Ave., PO Box 1968, Danbury, CT 06813–1968. Web site: <www.rarediseases.org>.

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Narcolepsy

Definition

Narcolepsy is a disorder of the nervous system marked by excessive daytime sleepiness, uncontrollable **sleep** attacks, and cataplexy (a sudden loss of muscle tone, usually lasting up to half an hour). The American Psychiatric Association (APA) classifies narcolepsy as a sleep disorder in the fourth edition of the *Diagnostic and Statistical Manual of Mental Disorders*, or *DSM-IV*. The National Institute of Neurological Disorders and Stroke (NINDS) defines narcolepsy as a "disorder caused by the brain's inability to regulate sleep-wake cycles normally." The disorder is sometimes called Gélineau's syndrome because it was first identified in 1880 by the French neurologist Jean-Baptiste Gélineau. The word *narcolepsy* itself comes from two Greek words that together mean "seized by sleepiness."

Description

Narcolepsy is the second-leading cause of excessive daytime sleepiness (after obstructive sleep apnea). Persistent sleepiness and sleep attacks are the hallmarks of this condition. The sleepiness has been compared to the

feeling of trying to stay awake after not sleeping for two or three days. It is not correct, however, to describe people with narcolepsy as sleeping longer or spending more time asleep in a 24-hour period than people without the disorder. Although patients with narcolepsy experience drowsiness and sleep attacks during the daytime, they also wake up frequently during the nighttime hours. For this reason, narcolepsy is more accurately described as a disorder of the normal boundaries between sleep and wakefulness.

People with narcolepsy fall asleep suddenly—anywhere, at any time, maybe even in the middle of a conversation. These sleep attacks can last from a few seconds to more than an hour. Depending on where they occur, they may be mildly inconvenient or even dangerous. Some people continue to function outwardly during the sleep episodes, such as talking or putting things away. But when they wake up, they have no memory of the event.

Narcolepsy is related to the deep, dreaming part of sleep known as rapid eye movement (REM) sleep. Normally when people fall asleep, they experience 80 to 100 minutes of non-REM sleep, which is then followed by about 20 minutes of REM sleep. People with narcolepsy, however, enter REM sleep immediately. In addition, REM sleep occurs inappropriately throughout the day in patients with narcolepsy.

Demographics

There has been considerable debate in the early 2000s over the incidence of narcolepsy. Some researchers think the disorder is underdiagnosed. According to NINDS, the disorder affects one American in every 2000, or about 135,000 people in the general United States population. However, the rates in other countries vary considerably, from one in 600 people in Japan to one in 500,000 in Israel. The reasons for these variations in different ethnic groups are not yet fully understood.

Males and females seem to experience this disorder at about the same rate.

Narcolepsy is a somewhat unusual disorder in terms of age distribution. Although the disorder has been identified in children as young as three years of age, most patients with narcolepsy are diagnosed either between the ages of ten and 25 or between the ages of 40 and 45. It is uncommon for a person to develop the signs of narcolepsy for the first time after age 55.

Causes and symptoms

Causes

In 1999 researchers identified the gene that causes narcolepsy on chromosome 12. The gene allows cells in the hypothalamus (the part of the brain that regulates sleep behavior) to receive messages from other cells. When this gene is abnormal, cells cannot communicate properly, and abnormal sleeping patterns develop. However, not everyone who has the gene develops narcolepsy; between 12 percent and 35 percent of the United States population is thought to carry the gene but only 0.02 percent develop the disorder. Narcolepsy sometimes clusters in families; first-degree relatives of a person diagnosed with the disorder have a 1 percent to 2 percent risk of developing narcolepsy themselves, or about 10 to 40 times the risk of a person in the general population.

In the late 1990s, three independent research groups discovered a neuropeptide system in the hypothalamus, the part of the brain that regulates body temperature and appetite. The newly discovered system, which has been called the hypocretinergic system, regulates sleep and wakefulness. The nerve cells, or neurons, in this part of the hypothalamus secrete substances known as hypocretins or orexins, which regulate the sleep/wake cycle in humans. There are two of these compounds, known as orexin-A and orexin-B, or as hypocretin-1 and hypocretin-2. As of the early 2000s, narcolepsy is thought to be an orexin deficiency syndrome; that is, it develops when a person's hypothalamus does not secrete enough orexins to keep the person from falling asleep at inappropriate times. Samples of cerebrospinal fluid taken from patients with narcolepsy contain little or no orexins. MRI scans of these patients indicate that there is some loss of brain tissue in the hypothalamus itself, suggesting that the neurons responsible for secreting orexins have died.

In a few cases, the first signs of narcolepsy are triggered by traumatic damage to the part of the brain that governs REM sleep or from a rapidly growing tumor that puts pressure on this region of the brain. It is also thought that the hormonal changes of **puberty** may affect this region of the brain in some people.

Symptoms

Narcolepsy is defined by four major symptoms:

- Excessive daytime sleepiness (EDS).
- Cataplexy, the most dramatic symptom of narcolepsy, affecting 75 percent of people with the disorder. During an attack of cataplexy, the person's knees buckle and the neck muscles go slack. In extreme cases, the person may become paralyzed and

fall to the floor. This loss of muscle tone is temporary, lasting from a few seconds to half an hour, but it is frightening to other **family** members or friends. Attacks of cataplexy can occur at any time but are often triggered by such strong emotions as anger, joy, or surprise.

- Hypnagogic hallucinations, intense and sometimes terrifying experiences that occur as the person is falling asleep. The hallucinations may be either visual or auditory. They are thought to represent an intrusion of REM sleep/dreaming into the wakeful state.
- Sleep paralysis, a frightening inability to move shortly after awakening or dozing off.

When to call the doctor

The symptoms of narcolepsy in children below the age of ten are somewhat different from the classical signs of the disorder in adolescents and adults. They may include the following (in addition to cataplexy and daytime sleepiness):

- unexplained falls or dropping of objects
- night terrors
- moodiness and abrupt episodes of irritability
- restlessness and hyperactivity
- difficulty waking up in the morning

Children between the ages of ten and 12 frequently report falling asleep in school or being unable to pay attention during class. In some cases a sudden drop in the child's academic performance is the first indication of narcolepsy.

Parents who suspect that their child may have narcolepsy should consult a specialist (usually a pediatric neurologist) and have the child tested in a sleep clinic. Children with narcolepsy have often been misdiagnosed as having attention-deficit hyperactivity disorder, while adolescents have sometimes been misdiagnosed as having substance abuse or **personality disorders**. The sooner narcolepsy is correctly identified, the better the child's chances of maintaining normal academic and social development.

Diagnosis

Narcolepsy is a complex disorder, and it is not always easy to identify. It takes ten years on average for an individual to be correctly diagnosed. The diagnosis of younger patients is additionally complicated by the fact that children with narcolepsy rarely have all four of the classical symptoms of the disorder as described in adults. Most often, the first symptom in children is an overwhelming feeling of fatigue. After several months or years, cataplexy and the other classical symptoms of the disorder may appear.

The child's doctor will not be able to diagnose narcolepsy on the basis of a routine physical examination. If the child has experienced both excessive daytime sleepiness and cataplexy, a tentative diagnosis may be made on the basis of the patient's history. In addition, the doctor may give the child or adolescent a short self-administered list of eight questions known as the Epworth Sleepiness Scale (ESS). First published by an Australian doctor in 1991, the ESS asks the person to rate how likely they are to doze off or fall asleep in eight different situations from everyday life. A score above ten (maximum score is 24) generally indicates that the doctor should consider laboratory testing for narcolepsy.

Imaging studies are not helpful in diagnosing narcolepsy, although in some cases the doctor may order an MRI or CT scan to rule out a brain tumor or other abnormality in brain structure. Laboratory tests used to evaluate a person for narcolepsy include an overnight polysomnogram (a test in which sleep is monitored with electrocardiography, a video camera, and respiratory parameters). A multiple sleep latency test, which measures sleep latency (onset) and how quickly REM sleep occurs, may also be used. People who have narcolepsy usually fall asleep much more rapidly that people without the disorder, often in less than five minutes.

If the diagnosis is still questionable, a genetic blood test can reveal the existence of certain substances in people who have a tendency to develop narcolepsy. Positive test results suggest but do not prove that the person has narcolepsy. As of the early 2000s, the diagnosis of narcolepsy also can be confirmed by taking a sample of the patient's cerebrospinal fluid by a spinal tap and testing it for the presence of hypocretin-1. Patients with narcolepsy have no hypocretin-1 in their spinal fluid.

Treatment

There is no cure for narcolepsy. The disorder is neither progressive nor fatal, but it is chronic. The symptoms, however, can be managed with a combination of medications and lifestyle adjustments. Amphetamine-like **stimulant drugs** are often prescribed to control drowsiness and sleep attacks. Patients who do not like taking high doses of stimulants may choose to take smaller doses and make adjustments in their lifestyles, such as napping every couple of hours, to relieve daytime sleepiness. **Antidepressants** are also often effective in treating symptoms of abnormal REM sleep.

Newer nonamphetamine wake-promoting drugs are available to treat narcolepsy. These medications lack the unpleasant side effects of amphetamines, particularly jitteriness and **anxiety**. Modafinil (Provigil) is the most commonly prescribed of the newer psychostimulants. As of 2004, however, researchers do not know exactly how modafinil prevents the drowsiness associated with narcolepsy. Its most common side effect is **headache**. A study published in 2003 reported that modafinil appears to be safe for use in children.

With discovery of the gene that causes narcolepsy, researchers are hopeful that therapies can eventually be designed to relieve the symptoms of the disorder.

Alternative treatment

The botanical remedy yohimbe (*Pausinystalia yohimbe*) may be useful in promoting alertness. As with any herbal preparation or medication, however, individuals should check with their healthcare professional before taking the remedy to treat narcolepsy.

Nutritional concerns

Children with narcolepsy sometimes fall asleep while eating, but the disorder itself does not cause or lead to **malnutrition**. Adolescents should be advised to avoid tobacco, **caffeine**, and alcoholic beverages, as these substances can increase daytime sleepiness in patients with narcolepsy.

Prognosis

The symptoms of narcolepsy are more severe when they develop in children than when they emerge in adult life. Narcolepsy is not a degenerative disease, however, and patients do not develop other neurologic symptoms. In fact, older patients often report that their symptoms decrease in severity after age 60. Apart from falls or other accidents, narcolepsy does not affect a person's life expectancy. It can, however, severely interfere with a young person's ability to study, **play**, participate in **sports** and other social activities, and develop close relationships with others. The sooner it is diagnosed and treated, the better the child's outlook for a happy and productive adult life.

Prevention

There is no way to prevent narcolepsy as of the early 2000s.

KEY TERMS

Cataplexy—A symptom of narcolepsy in which there is a sudden episode of muscle weakness triggered by emotions. The muscle weakness may cause the person's knees to buckle, or the head to drop. In severe cases, the patient may become paralyzed for a few seconds to minutes.

Hypocretins—Chemicals secreted in the hypothalamus that regulate the sleep/wake cycle.

Hypothalamus—A part of the forebrain that controls heartbeat, body temperature, thirst, hunger, body temperature and pressure, blood sugar levels, and other functions.

Orexin—Another name for hypocretin, a chemical secreted in the hypothalmus that regulates the sleep/wake cycle. Narcolepsy is sometimes described as an orexin deficiency syndrome.

Sleep paralysis—An abnormal episode of sleep in which the patient cannot move for a few minutes, usually occurring on falling asleep or waking up. Often found in patients with narcolepsy.

Parental concerns

Narcolepsy can affect a family in a number of ways before it is diagnosed. Younger children are at risk of injuring themselves by falling, and adolescents with driving privileges are at high risk of automobile accidents. Poor performance in school and difficulty making friends as a result of irritability or embarrassment over sleep attacks can have a lasting impact on a child's chances of preparation for college or the choice of a challenging and satisfying line of work. In many cases the child is accused of being lazy or stupid, which can have devastating effects on his or her **self-esteem**. Misdiagnoses can lead to inappropriate treatment and psychological depression for the affected child. In addition, sleep attacks and cataplexy can be frightening to other family members who witness them.

Narcolepsy can be particularly stressful for a family when the affected child reaches **adolescence**, because of **peer pressure** to experiment with **smoking** and recreational drugs, and because of resentment about restrictions on learning to drive or use of the family car. Families with a child diagnosed with narcolepsy should consider joining a support group for people affected by the disorder.

Resources

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ORGANIZATIONS

American Sleep Disorders Association. 1610 14th St. NW, Suite 300, Rochester, MN 55901. Web site: <www.sleepapnea.org>.

Narcolepsy Network. PO Box 42460, Cincinnati, OH 45242. Web site: <www.narcolepsynetwork.org>.

National Center on Sleep Disorders Research. Two Rockledge Centre, 6701 Rockledge Dr., Bethesda, MD 20892. Web site: <www.nhlbi.nih.gov/health/public/sleep>.

National Institute of Neurological Disorders and Stroke (NINDS). National Institutes of Health. 9000 Rockville Pike, Bethesda, MD 20892. Web site: <http://www.ninds.nih.gov>.

National Sleep Foundation. 1522 K St., NW, Suite 500, Washington, DC 20005. Web site: <www.sleepfoundation.org>.

Stanford Center for Narcolepsy. 1201 Welch Rd-Rm P-112, Stanford, CA 94305. Web site: <http://blackdogstudieos.com/portfolio/Web/narcolepsy>.

University of Illinois Center for Narcolepsy Research. 845 S. Damen Ave., Chicago, IL 60612. Web site: <www.uic.edu/depts./cnr/>

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Michelle Lee Brandt

Narcotic drugs

Definition

Narcotics are addictive drugs that reduce the user's perception of **pain** and induce euphoria (a feeling of exaggerated and unrealistic well-being). The English word narcotic is derived from the Greek *narkotikos*, which means "numbing" or "deadening." Although the term can refer to any drug that deadens sensation or produces stupor, it is commonly applied to the opioids—that is, to all natural or synthetic drugs that act like morphine.

Description

Historical background

Narcotics are the oldest as well as the strongest **analgesics**, or pain-relieving drugs, known to humans. Ancient Sumerian and Egyptian medical texts dated as early as 4000 B.C. mention the opium poppy (*Papaver somniferum*) as the source of a milky fluid (opium latex) that could be given to relieve coughs and insomnia as well as ease pain. Traditional Chinese medicine recommended the opium poppy, known to Chinese physicians as *ying su ke*, for the treatment of **asthma**, severe **diarrhea**, and dysentery as well as chronic pain and insomnia. Opium latex contains between 10 and 20 percent morphine, which in its purified form is a white crystalline powder with a bitter taste.

Narcotics are central nervous system depressants that produce a stuporous state in the person who takes them. These drugs often induce a state of euphoria or feeling of extreme well-being, and they are powerfully addictive. The body quickly builds a tolerance to narcotics in as little as two to three days, so that greater doses are required to achieve the same effect. Because of the addictive qualities of these drugs, most countries in the

twenty-first century have strict laws regarding the production and distribution of narcotics. These laws became necessary when opium **addiction** in the nineteenth century became a widespread social problem in the developed countries. Opium, which was the first of the opioids to be widely used, had been a common folk remedy for centuries that often led to addiction for the user; in fact, many popular Victorian patent medicines for ''female complaints'' actually contained opium. The invention of the hypodermic needle in the mid-nineteenth century, however, increased the number of addicts because it allowed opioids to be delivered directly into the bloodstream, thereby dramatically increasing their effect.

Classification of narcotics

As of the early 2000s, narcotics are commonly classified into three groups according to their origin:

- Natural derivatives of opium: Narcotics in this group include morphine itself and codeine.
- Partially synthetic drugs derived from morphine: These drugs include heroin, oxycodone (OxyContin), hydromorphone (Dilaudid), and oxymorphone (Numorphan).
- Synthetic compounds that resemble morphine in their chemical structure: Narcotics in this group include fentanyl (Duragesic), levorphanol (Levo-Dromoran), meperidine (Demerol), methadone, and propoxyphene (Darvon).

Narcotics are available in many different forms, ranging from oral, intramuscular, and intravenous preparations to patches that can be applied to the skin (fentanyl). Illegal street heroin can be taken by inhalation as well as by injection.

How narcotics work

The central nervous system in humans and other mammals contains five different types of opioid receptor proteins, located primarily in the brain, spinal cord, and digestive tract. When a person takes an opioid medication, the drug attaches to these opioid receptors in the brain and spinal cord and decreases the person's perception of pain. Narcotics do not, however, reduce or eliminate the cause of the pain.

Some of the opioid receptors (known as mu and sigma receptors) influence a person's perception of pleasure. When a narcotic medication stimulates these receptor proteins, the person typically experiences intense sensations of euphoria or well-being. The speed with which these drugs take effect depends on the method of administration; IV narcotics reach their peak effectiveness within ten minutes, while oral narcotics take about an hour and a half, and skin patches take between two and four hours.

Overdoses of narcotics can cause drowsiness, unconsciousness, and even death because these drugs suppress respiration.

General use

Narcotics have several legitimate uses:

- Analgesic: Doctors frequently prescribe oral codeine and propoxyphene (alone or in combination with aspirin) for pain control after oral surgery, for severe menstrual cramps, and for temporary pain relief after other outpatient surgical procedures. Intravenous narcotics may be given for several days after major surgery to relieve the patient's discomfort. Subsequent methods of administering opioids following surgery include a sustained-release injected form of morphine sulfate (DepoMorphine) and a patient-controlled transdermal system (E-TRANS) that releases doses of fentanyl when the patient pushes a button attached to the arm or upper chest. Intravenous narcotics may also be used for palliative care, to relieve the pain of patients diagnosed with terminal **cancer**.
- Antitussive: Antitussives are medications given to control coughing. Codeine is often effective in relieving severe coughs and is a common ingredient in prescription **cough** mixtures.
- Antidiarrheal: Paregoric, a liquid preparation containing powdered opium, anise oil, and glycerin, is sometimes prescribed for severe diarrhea. The opium in paregoric works to control diarrhea because it slows down the rhythmic contractions of the intestines that ordinarily move food through the digestive tract. Lomotil, another antidiarrheal medication, contains a synthetic opioid known as diphenoxylate; it is often recommended for treating cancer patients with diarrhea caused by radiation therapy.

Precautions

In the United States, opioids are as of 2004 classified as Schedule II drugs under the Controlled Substances Act of 1970. Drugs in this category are described by the government as having a high potential for abuse and a liability for dependence and yet an approved medical use in **pain management**. The corresponding Canadian legislation, the Controlled Drugs and Substances Act of 1997, classifies medications containing any narcotic under the heading (N) but specifies varying levels of regulation ranging from strict controls for highly addictive single-drug products to lesser controls on drugs combin-

ing a narcotic with non-narcotic substances. As both countries' legal controls indicate, narcotics should be used cautiously, for as short a period of time as possible, and only under a doctor's supervision. In particular, they should never be used together with certain other categories of prescription drugs or herbal preparations.

Side effects

In addition to the risk of dependency or addiction, narcotics have a number of physical side effects, including the following:

- **constipation**
- drowsiness
- withdrawal symptoms after extended use (tearing, sweating, diarrhea, **vomiting**, gooseflesh, muscle twitching, runny nose, loss of appetite, and hot or cold flashes)

Interactions

Narcotics can be dangerous because of their potential for deadly interactions with other medications as well as their potential for dependence and addiction. Narcotics should never be combined with other types of drugs that depress the central nervous system. These categories of drugs include the following:

- alcohol
- benzodiazepine tranquilizers, including such drugs as diazepam (Valium), alprazolam (Xanax), and chlordiazepoxide (Librium)
- barbiturates, used to treat insomnia and **anxiety**, including such medications as pentobarbital (Nembutal) and mephobarbital (Mebaral)
- antihistamines, even over-the-counter cold or allergy medications, which can interact with narcotics to intensify drowsiness and repress breathing

Narcotics can also interact with certain herbal preparations to cause central nervous system depression. Anyone taking narcotics for pain relief should avoid using herbal preparations containing kava kava (*Piper methysticum*), valerian (*Valeriana officinalis*), chamomile (*Matricaria recutita*), or lemon balm (*Melissa officinalis*), as these herbs intensify the tendency of opioids to cause drowsiness and slow down breathing. Ginseng (*Panax ginseng*) should also be avoided because it interferes with the pain-relieving qualities of opioid medications.

Prevention

Scientists have attempted to develop ways to use the pain-killing properties of narcotics while counteracting their addictive qualities. Substances known as narcotic or opioid antagonists are drugs that block the actions of narcotics and are used to reverse the side effects of narcotic abuse or an overdose. A class of drugs, a mixture of opioids and opioid antagonists, has been developed so that patients can be relieved of pain without the addictive or other unpleasant side effects associated with narcotics.

Parental concerns

One minor concern that parents may have if the doctor prescribes narcotic medications for their child is that such side effects as constipation or sleepiness are more common and more severe in children younger than 18 years of age.

Far more seriously, narcotic drugs are among those substances used illegally or abused by adolescents. Some researchers estimate that as many as 90 percent of adult drug addicts began a pattern of substance abuse during **adolescence**. Teenagers are particularly likely to begin experimenting with narcotics in the form of prescription cough syrup and such pain relievers as Darvon or OxyContin and to combine narcotics with alcohol or other drugs of abuse. Moreover, although opioid medications account for fewer cases of drug abuse than cocaine, alcohol, or several other drugs, they still account for 4 to 5 percent of emergency room visits. In addition, the death rate of opioid abusers is proportionately significantly higher than the mortality of people who abuse PCP or cocaine.

The American Academy of Child and Adolescent Psychiatry (AACAP) lists opiates as common drugs of abuse among teenagers and notes that children as young as 12 may be using narcotics. In many cases, children can obtain these drugs at home in the form of medications prescribed for other **family** members. Children who are abusing opioid medications may show the following signs:

- euphoria or "feeling no pain"
- constipation
- slurred speech
- shallow breathing
- itching or flushing of the skin
- mental confusion and poor judgment
- bloodshot eyes with small pupils
- nausea and vomiting
- unusual drowsiness

KEY TERMS

Analgesics—A class of pain-relieving medicines, including aspirin and Tylenol.

Antitussive—A drug used to suppress coughing.

Euphoria—A feeling or state of well-being or elation.

Morphine—The principal alkaloid derived from the opium poppy for use as a pain reliever and sedative. In its purified form, it is a white, bitter-tasting crystalline powder.

Narcotic—A drug derived from opium or compounds similar to opium. Such drugs are potent pain relievers and can affect mood and behavior. Long-term use of narcotics can lead to dependence and tolerance. Also known as a narcotic analgesic.

Opium latex—The milky juice or sap of the opium poppy, used to produce morphine.

Palliative—Referring to a drug or a form of care that relieves pain without providing a cure. Persons in severe pain from terminal cancer are often prescribed narcotics as palliative care.

Stupor—A trance-like state that causes a person to appear numb to their environment.

Parents who suspect that their children are abusing opioids, either alone or in combination with other drugs, should get help as soon as possible.

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Rebecca Frey, PhD

Nasal trauma

Definition

Nasal trauma is defined as any injury to the nose or related structure that may result in bleeding, a physical deformity, a decreased ability to breathe normally

because of obstruction, or an impaired sense of smell. The injury may be either internal or external.

Description

The human nose is composed of bone, soft tissue, and cartilage. It serves as a passageway for air to flow from the outside environment into the lower respiratory tract and lungs. At the same time the nasal passages warm and humidify the air that enters the body.

Internal injuries to the nose typically occur when a foreign object is placed in the nose or when a person takes in drugs of abuse (inhalants or cocaine) through the nose. External injuries to the nose are usually blunt force injuries related to **sports** participation, criminal violence, **child abuse**, or automobile or bicycle accidents. This type of injury may result in a nasal fracture. The nasal bones are the most frequently fractured facial bones due to their position on the face, and they are the third most common type of bone fracture in general after **fractures** of the wrist and collarbone. A force of only 30 g is required to break the nasal bones, compared to 70 g for the bones in the jaw and 200 g for the bony ridge above the eyes. The pattern of the fracture depends on the direction of the blow to the nose, whether coming from the front, the side, or above the nose. Although not usually life-threatening by itself, a fractured nose may lead to difficulties in breathing as well as facial disfigurement.

Fractures resulting from trauma to the nose may involve the bones of the septum (the partition of bone and cartilage dividing the two nostrils) as well as the bones surrounding the eyes. These bones include the nasal, maxilla, lacrimal, and frontal bones. Direct trauma to the bridge of the nose may also result in damage to a part of the base of the skull known as the cribriform plate. This injury in turn may allow cerebrospinal fluid to leak out of the skull and leave the body through the nose. Fractures may also damage the membranes that line the nasal passages, leading to possible formation of scar tissue, obstruction of the airway, and damage to the child's sense of smell.

In addition to fractures, external injuries of the nose include soft-tissue injuries resulting from **bites** (human and animal), insect **stings**, cuts, or scrapes. Penetrating injuries to the nasal area caused by air gun or BB pellets were as of 2004 also reported with increasing frequency in older children and adolescents. When fired at close range, these pellets can penetrate the skin and cheekbone and lodge in the nasal septum or the sinuses near the nose.

Lastly, nose **piercing** as a fashion trend is a type of intentional injury to the nose that has several possible complications, including infections of the cartilage and soft tissues in the nose; blockage of the airway due to a loosened stud or other nose ornament; and gastrointestinal emergencies caused by accidental swallowing of nose jewelry.

If a patient's nasal trauma leads doctors to suspect child abuse, the incident must be reported to the police.

Demographics

The demographics of nasal trauma vary according to the type of injury. Internal nasal injuries are unusual in infants but occur fairly frequently in toddlers and young children as a result of playfulness or curiosity. Children often insert small hard objects (buttons, coins, watch batteries, dried peas or beans, plastic parts from **toys**, etc.) in their nostrils. One Japanese study of children brought to the emergency room for removal of foreign bodies from the respiratory and digestive tracts found that the nose was the most common location (39.4% of patients) of these objects. Sixty-seven percent of the children treated were between one and four years of age, with two-year-olds the most common age group. Another common cause of injury to the nasal passages in children is scratching or picking the inside of the nose, often as a reaction to dry and **itching** nasal membranes during the heating season in colder climates.

In older children and adolescents, however, the single most common cause of internal nasal injuries is inhalant abuse or ingesting cocaine through the nose ("snorting"). Inhalants include such substances as toluene (paint thinner, nail polish remover, rubber cement, airplane glue), butane (lighter fluid, spray paint, room fresheners, hair spray), chlorinated hydrocarbons (dry cleaning fluid, spot removers, typewriter correction fluid), and acetone (rubber cement, permanent markers, nail polish remover). According to the American Academy of Family Practice (AAFP) and the U.S. Department of Health and Human Services, nearly 20 percent of children in the United States have used inhalants at least once by the time they are in eighth grade. The average age of children experimenting with inhalants is 13, with Hispanic and Caucasian youth more likely to abuse these substances than African Americans. With regard to cocaine, figures from the National Institute on Drug Abuse (NIDA) from the late 1990s indicate that that 3.2 percent of all eighth graders in the United States have used cocaine at least once, although young adults between the ages of 18 to 25 are the age group with the highest usage of the drug.

Nose piercing as a fashion statement is most common among adolescents and young adults. One study of

undergraduates at a university in upstate New York found that 51 percent had body piercing, with the nose and ears the most common sites. Seventeen percent of these students reported medical complications from the piercing, ranging from skin or cartilage infections to periodic bleeding from the nose. A less common cause of internal injuries to the nose in older children and adolescents is the use of magnetized jewelry as a substitute for body piercing. The external piece of jewelry is held in place on the outside of the nostril by a small magnet placed inside the nose. Displacement of these magnets has been reported to cause bleeding and perforation of the nasal septum, while accidental swallowing of these magnets may require emergency surgery. One British hospital reported no fewer than 24 such cases over an eight-week period.

With regard to nasal fractures, one group of American researchers estimates that they account for about 24 percent of all facial fractures. The most common single cause is assault, accounting for 41 percent of nasal fractures. The second most common cause is automobile accidents (27%), followed by **sports injuries** (11%). Falls account for most other nasal fractures in children, although dog bites are reported with increasing frequency as a cause of nasal fractures in children below 16 years of age. Adolescents who have had plastic surgery on the nose (rhinoplasty) are at increased risk of nasal fractures in later life.

As many as 10 percent of nasal injuries in younger children, however, result from physical abuse. Doctors in the early 2000s are advised to consider abuse as a possible diagnosis when evaluating nasal fractures in children under six years of age. Suspected child abuse must be reported to police.

Causes and symptoms

Causes

External trauma to the nose may be accidental (transportation accidents, animal bites, air gun injuries, and sports injuries) or intentional (fights, criminal assault, domestic violence, nose piercing). Nasal injuries from athletic activities may result from contact with equipment (being hit in the face by a baseball, hockey ball, or other small ball hit at high speed, or by the bat or stick itself) or the bodies of other players (football, boxing, martial arts, rugby). Nasal injuries from piercing include bacterial infections of the skin and nasal cartilage, allergic reactions to the jewelry, tissue damage, and periodic bleeding. Direct trauma and/or delayed type hypersensitivity reaction to nickel may occur from nasal rings and jewelry, facial adornments which as of 2004 are increasingly popular.

In a few cases, external trauma to the nose may also be iatrogenic, or caused by medical care. Most of these injuries result from medical examination of the nose—particularly in emergency circumstances—or as complications of plastic surgery.

Internal injuries to the nose may be either mechanical (caused by **foreign objects** in the nose or by picking or scratching the tissues lining the nose) or chemical (caused by environmental irritants or substance abuse).

Chemical injuries to the nose are caused by accidental or purposeful breathing or sniffing of irritating substances. These may include tobacco smoke; household cleaners (ammonia and chlorine bleach) and furniture polish; ozone and other air pollutants; cocaine; and glue, paint thinners, solvents, and similar household products that produce toxic vapors. An increasingly common form of chemical injury to the nasal membranes in toddlers is alkali **burns** caused by leakage from small batteries placed in the nose. While chemical damage to the nose is usually accidental in younger children, it is more often the result of substance abuse in adolescents. Taking cocaine through the nose (''snorting'') or inhalant abuse (''sniffing'' or ''huffing'') are the most common causes of chemical damage to the nose in older children or teenagers.

Symptoms

The symptoms of physical trauma to the nose may include the following:

- flattening or other deformation of the shape of the nose
- infections of the cartilage or soft tissue
- epistaxis, or bleeding from the nose
- crepitus, or the crackling or crunching sound heard when the ends of a fractured bone are rubbed together
- pain and tissue swelling
- airway blockage from bleeding, fluid discharge, or tissue swelling
- **rhinitis** or inflammation of the mucous membranes lining the nose (In the case of a fracture, rhinitis may lead to increased tear production in the eyes and a runny nose.)
- septal hematoma, a mass of blood from torn tissue that may collect within the cartilage that divides the two nostrils (It may become infected and form an abscess that eventually destroys the cartilage.)
- bruising or discoloration (ecchymosis) of the tissues around the eye

- leakage of cerebrospinal fluid through the nostrils

Chemical trauma to the nose may result in the following:

- runny nose and watering of the eyes
- pain
- loss of the sense of smell
- nasal congestion and sneezing
- reddening and swelling of the mucous membranes lining the nose
- eventual destruction of the cartilage in the nasal septum and the tissues lining the nose

When to call the doctor

Parents should call the doctor at once in the event of a nose injury when the following conditions are apparent:

- The child is bleeding profusely from the nose.
- The child is having difficulty breathing normally.
- The injury involves an air gun, BB gun, or animal or human bites.
- The child is seeing double, has other visual disturbances, or staggers when trying to walk.
- The child's nasal discharge is watery as well as bloody.
- The child is known to have inserted a battery into the nose or to have swallowed such a battery or piece of nose jewelry.
- The child's appearance or behavior suggests inhalant or cocaine abuse. Some danger signals are: a chemical odor on the child's breath; constant runny nose; unusual or excessive use of nose drops or **decongestants**; sores inside or around the mouth; stains on the fingernails; dazed appearance; **anxiety**, **sleep** disturbances, **nausea and vomiting**; slurred speech; visual disturbances; and loss of physical coordination.

Diagnosis

History and physical examination

In many cases the diagnosis of an injury to the child's nose is obvious to the doctor from taking a history, particularly if the parent witnessed the accident or saw the child putting something in his nose. The physical examination depends in part on the history. If the child's nose is bleeding without a history of a fall, blow to the face, or other obvious cause, the doctor gently examines inside the nose with a handheld speculum of the type used to examine the ears, in order to see where the nasal bleeding originates. Bleeding from the lower part of the nose is more common and usually less serious. In most cases the doctor is able to tell whether there is a foreign object in the nose or whether the child has been scratching or picking at the nose. Bleeding from the upper part of the nose closer to the throat is more serious because it can block the airway and because it may indicate that the child has a bleeding disorder rather than a traumatic injury. The doctor may then examine the child's throat for signs of blood from the upper nose trickling down into the throat. He or she will remove any blood clots from the nose with suction.

In the case of a known accident, sports injury, or assault, the doctor begins with the ABCs, which means that he or she will check the child's *airway*, *breathing*, and *circulation*. The doctor will usually have the child sit upright or lie on one side, and will remove blood clots, broken teeth, or other foreign bodies from the nose or throat. He or she will then carry out a systematic examination of the child's face and head. The most common pattern of examination moves from the inside of the nose and mouth to the outside of the face and from the bottom of the face to the top.

The doctor looks for signs of bruising and tissue swelling as well as bleeding and gently palpates, or touches, the various facial bones for movement and stability. If the doctor suspects that the nose itself is fractured, he or she will listen for crepitus when the nose is gently moved and will look for evidence of a dislocated septum or a septal hematoma, which will appear as a bluish bulging mass within the nasal septum. The child's teeth will be examined for looseness, and the muscles and nerves of the face will be evaluated. If there is a discharge from the nose, the doctor will look at it to see whether it contains cerebrospinal fluid, which would indicate damage to the bones of the skull as well as the nasal bones. Lastly, the doctor examines the child's eyes to make sure that the pupils are responding normally to light and that the child is not seeing double or having other visual problems that might indicate nerve damage or damage to the eye itself.

In cases involving animal bites or other tearing or crushing injuries to the skin and external tissues of the nose, the doctor carefully cleanses the broken skin with soap and water or disinfectants such as benzalkonium chloride as well as checks for fractured facial bones.

Many of the early signs of inhalant or cocaine abuse are not specific to these disorders; however, a careful history-taking and examination of the child's eyes, nose, and throat may lead the doctor to ask the child or adolescent appropriate questions about his or her use of inhalants or cocaine.

Imaging studies and laboratory tests

Computed tomography (CT) scans are the type of imaging study most commonly done to evaluate suspected nasal fractures. X-ray studies may be ordered to identify the location of a foreign body if it is metal, such as a pellet from a BB gun or air gun, or to evaluate the nasal area for evidence of repeated fractures when abuse is suspected. A blood test will be performed prior to surgery in order to determine the child's blood type, clotting time, and complete blood count. In some cases the doctor may order a filter paper or glucose content test of nasal secretions to check for the presence of cerebrospinal fluid.

In some cases, the physician may ask the child's parents for photographs taken prior to the injury in order to determine the extent of deformity or other injuries to the nose. Photographs may also be taken for documentation if abuse is suspected and also for documentation of injuries for later plastic surgery.

Drug-specific blood or urine tests may be ordered for children or adolescents suspected of abusing inhalants or cocaine.

Treatment

Timing

Nasal injuries should be treated as promptly as possible to lower risk of infection. If the child has been bitten by an animal, the injury must be cleansed as soon as possible to lower the risk of **rabies**. Batteries placed in the nose should be removed as soon as possible, preferably within four hours to avoid serious burns from their contents. If a septal hematoma has developed, the doctor must remove it as quickly as possible to prevent infection or eventual death of the tissues in the nasal septum.

Treatment of nasal fractures is best performed during the first three hours after the injury. If this is impossible, management of a nasal fracture should be done within three to seven days. Timing is of utmost importance when treating nasal fractures because delays longer than seven to 10 days may allow the broken bones to set without proper alignment or lead to such complications as scar tissue formation and airway obstruction. Poorly set nasal fractures usually require surgical correction.

Specific procedures

Foreign objects in the nose can be removed by nasal suction in most cases. Most nosebleeds are treated by five to 30 minutes of direct pressure on the nostrils, with the child's head placed in an upright position. The doctor may also pack the child's nose with gauze coated with petroleum jelly. If the bleeding does not stop or if it appears to originate in the upper nose, the doctor will consult a head and neck surgeon or an otolaryngologist for specialized evaluation of the bleeding.

Air gun or BB pellets that have penetrated the nose or nearby sinuses are generally removed with the help of an endoscope, which is a slender tubular instrument that allows the doctor to examine the inside of a body cavity.

Treatment of nasal fractures depends on the extent of the injury; the most difficult fractures to treat are those that involve the nasal septum. The doctor will usually reduce the fracture, which means that he or she will restore the damaged bones to their proper position and alignment. Although local anesthesia is usually sufficient for treating nasal fractures in adults and older teenagers, general anesthesia is usually given when treating these injuries in younger children.

Reductions of nasal fractures may be either open or closed. A closed reduction involves manipulation of the bones without cutting into the overlying skin. This type of reduction is performed for fractures of the nasal bones that are limited in size and complexity. Open reductions are performed for more complex nasal fractures. In an open reduction, the nasal bones are moved back to their original location after the surgeon has made an incision in the overlying skin. This procedure is done for fractures involving dislocation of the septum as well as the nasal bones. In addition, an open reduction is necessary if the child has a septal hematoma or an open fracture in which the skin has been perforated. If a septal hematoma is present, the doctor will drain it and pack the nose to prevent subsequent accumulation of blood. The nasal bones are held in the proper position with external splints as well as the internal packing, and the splints are kept in place for seven to ten days. The child is given **antibiotics** to lower the risk of infection and may be referred to an otolaryngologist or plastic surgeon for further evaluation. Ice packs or cold compresses can be applied at home to reduce swelling and ease the child's discomfort.

In the case of animal bites, the child may be given passive or active immunization against rabies if there is a chance that the dog or other animal is rabid. This precaution is particularly important for animal bites on the nose or other parts of the face, as the incubation period of the rabies virus is much shorter for bites on the head and neck than for bites elsewhere on the body.

Prognosis

Most types of nasal trauma have a good prognosis. Nosebleeds or tissue damage caused by scratching or picking at the nose usually clear completely once the

child stops these habits. Infections or allergic reactions caused by foreign objects in the nose or piercing usually clear up promptly once the object or piece of jewelry is removed. Nasal fractures that do not involve the nasal septum or other facial bones and receive prompt treatment generally heal without deformities of the nose, cartilage destruction, or other complications. More extensive facial fractures, however, may require a second operation to correct the positioning of the bones and restore the appearance of the nose.

The prognosis for soft-tissue injuries to the nose depends on the cause and extent of the injuries. Such tearing or crushing injuries as those caused by bites take longer to heal than simple cuts and may require plastic surgery at a later date to restore the appearance of the nose.

Damage to the tissues lining the nose caused by exposure to tobacco smoke or other irritants in the environment is usually reversible once the child is removed from contact with the irritating substance. Erosion or destruction of the nasal cartilage as a result of inhalant or cocaine abuse, however, usually requires surgical treatment.

Prevention

Preventive strategies for nasal trauma depend on the child's age group. For younger children, parents should take the following precautions:

- Keep such small objects as coins, disk batteries, and buttons in childproof drawers or cabinets and throw out broken toys or toy parts.
- Use a humidifier during heating season to prevent drying and itching of the nasal membranes and coat the inside of the child's nose with petroleum jelly.
- Quit **smoking** completely or stop smoking inside the house.
- Open windows or otherwise ventilate the room when using ammonia, chlorine bleach, oven cleaner, degreasers, spray paints, dry cleaning fluid, furniture polish, or other household products that give off strong vapors at room temperature. Keep all such products in a childproof cabinet or closet.
- Teach the child basic rules of **safety** in playing with household pets as well as in dealing with large dogs and other animals outside the house. Have pet dogs or cats immunized against rabies.
- Drive safely and make sure the child is using an age-appropriate protective seat or seat belt.
- Make sure that the child understands basic safety precautions and traffic laws before allowing him or her to ride a bicycle in the street.

KEY TERMS

Crepitus—A crackling sound.

Dorsum—The medical term for the bridge of the nose.

Ecchymosis—The medical term for a bruise, or skin discoloration caused by blood seeping from broken capillaries under the skin.

Epistaxis—The medical term used to describe a bleeding from the nose.

Hematoma—A localized collection of blood, often clotted, in body tissue or an organ, usually due to a break or tear in the wall of blood vessel.

Iatrogenic—A condition that is caused by the diagnostic procedures or treatments administered by medical professionals. Iatrogenic conditions may be caused by any number of things including contaminated medical instruments or devices, contaminated blood or implants, or contaminated air within the medical facility.

Otolaryngologist—A doctor who is trained to treat injuries, defects, diseases, or conditions of the ear, nose, and throat. Also sometimes known as an otorhinolaryngologist.

Reduction—The restoration of a body part to its original position after displacement, such as the reduction of a fractured bone by bringing ends or fragments back into original alignment. The use of local or general anesthesia usually accompanies a fracture reduction. If performed by outside manipulation only, the reduction is described as closed; if surgery is necessary, it is described as open. Also describes a chemical reaction in which one or more electrons are added to an atom or molecule.

Rhinitis—Inflammation and swelling of the mucous membranes that line the nasal passages.

Rhinoplasty—Plastic surgery of the nose to repair it or change its shape.

Septum—A wall or partition. Often refers to the muscular wall dividing the left and right heart chambers or the partition in the nose that separates the two nostrils. Also refers to an abnormal fold of tissue down that center of the uterus that can cause infertility.

- Check the home for safety hazards that might lead to falls (for example, loose carpeting, poorly lit stairwells, and toys allowed to lie on the floor after play).
- Teach the child to deal with quarrels with other children without physical fighting and set the child a good example in relationships with others.

For older children and adolescents, parents should take the following steps:

- Set a good example of safe driving and make sure that teenagers have a mature attitude toward driving before they acquire a driver's license.
- Inform themselves about such problems as drug abuse, bullying, or violence in dating relationships, and learn to identify the signs of these problems in their children.
- Make sure that their child's sports teams use the appropriate safety equipment, that the equipment is in good condition, and that the teams have appropriate adult supervision.
- Discourage the child from nose piercing and similar fads or at least make certain that he or she has the procedure done at a reputable business that follows Food and Drug Administration (FDA) guidelines for cleanliness and sterilization of equipment.
- Teach safe and responsible use of BB guns and air guns.

Parental concerns

Parental concerns regarding nasal trauma depend on the cause and severity of the injury. Minor nosebleeds and uncomplicated fractures of the nose caused by accidents generally heal without problems and are quickly absorbed into the family's routine. Complex fractures or other injuries requiring a second operation may require explanation or discussion with the child. Nasal injuries related to the neighborhood environment (street crime, chemical pollution), lifestyle choices (body piercing, smoking in the home), or family dysfunction (substance abuse, domestic violence), however, suggest the need for professional counseling and changes in the family's structure, geographical location, or increased level of functioning.

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Rebecca Frey, PhD

Nausea and vomiting

Definition

Nausea is the sensation of being about to vomit. Vomiting, or emesis, is the expelling from the stomach of undigested food through the mouth.

Description

Nausea is a reaction to a number of causes that include overeating, infection, or irritation of the throat or stomach lining. Persistent or recurrent nausea and vomiting should be checked by a doctor.

A doctor should be called if nausea and vomiting occur in the following instances:

- after eating rich or spoiled food or taking a new medication
- repeatedly or for 48 hours or longer
- following intense dizziness

It is important to see a doctor if nausea and vomiting are accompanied by the following:

- yellowing of the skin and whites of the eyes
- **pain** in the chest or lower abdomen
- trouble with swallowing or urination
- **dehydration** or extreme thirst
- drowsiness or confusion
- constant, severe abdominal pain
- a fruity breath odor

Demographics

Nausea and vomiting are commonly experienced. There are no distinctive patterns of age, gender, or race.

Causes and symptoms

Persistent, unexplained, or recurring nausea and vomiting can be symptoms of a variety of serious illnesses. They can be caused by overeating or drinking too much alcohol. These symptoms can be due to stress, certain medications, or illness. For example, people who are given morphine or other opioid medications for pain relief after surgery sometimes feel nauseated by the drug. Such poisonous substances as arsenic and other heavy metals cause nausea and vomiting. Morning sickness is a consequence of pregnancy-related hormone changes. **Motion sickness** can be induced by traveling in a vehicle, plane, or on a boat. Many people experience nausea after eating spoiled food or foods to which they are allergic. Individuals who suffer migraine **headache** often experience nausea. **Cancer** patients receiving **chemotherapy** are often nauseated. Gallstones, **gastroenteritis**, and stomach ulcer may cause nausea and vomiting. These symptoms should be evaluated by a physician.

Nausea and vomiting may also be psychological in origin. Some people vomit under such conditions of emotional stress as **family** arguments, academic tests, airplane travel, losing a job, and similar high-stress situations. In addition, some eating disorders are characterized by self-induced vomiting.

When to call the doctor

A doctor should be notified if vomiting is heavy and/or bloody, if the vomitus looks like feces, or if the affected person has been unable to keep food down for 24 hours. Most vomiting episodes should stop in eight to 12 hours of onset. The pediatrician should be consulted if vomiting continues beyond that time, if the child shows signs of dehydration, seems extremely lethargic, or if the child is a very young infant.

An ambulance or emergency response number should be called immediately if the following occurs:

- The child's mouth and tongue are very dry.
- The child has very rapid heartbeat and breathing.
- The child cries but has no tears.
- The child has sunken eyes.
- Diabetic shock is suspected.
- Nausea and vomiting continue after other symptoms of viral infection have subsided.

- The person has a severe headache.
- The person is sweating and having chest pain and trouble breathing.
- The person is known or suspected to have swallowed a drug overdose or poisonous substance.
- The person has a high body temperature, **muscle cramps**, and other signs of heat exhaustion or heat stroke.
- Nausea, vomiting, and breathing problems occur after exposure to a known allergen.

Diagnosis

Diagnosis is based on the severity, frequency, and duration of symptoms, and other factors that could indicate the presence of a serious illness.

Diagnosis is based on a careful medical history that includes foods recently eaten, travel, and occupation. In some cases, the doctor may order laboratory tests or imaging studies to determine the presence of drugs or poisonous substances in the person's blood or urine, or evidence of head injuries or abnormalities in the digestive tract. If the nausea and vomiting appear to be related to **anxiety**, stress, or an eating disorder, the doctor may refer the person to a psychiatrist for further evaluation.

Treatment

Getting a breath of fresh air or getting away from whatever is causing the nausea can solve the problem. Eating olives or crackers or sucking on a lemon can calm the stomach by absorbing acid and excess fluid. Cola syrup is another proven remedy.

Vomiting relieves nausea quickly but can cause dehydration. Sipping clear juices, weak tea, and some sports drinks helps replace lost fluid and **minerals** without irritating the stomach. Infants and small children under age two do best with an oral rehydration solution like Pedialyte. The solution should be given a teaspoon at a time, at frequent intervals, starting 30–60 minutes after vomiting has ceased. Food should be reintroduced gradually, several hours after vomiting stops, beginning with small amounts of dry, bland food like crackers and toast.

Medications that are given to relieve nausea and vomiting are called antiemetics. Meclizine (Bonine), a medication for motion sickness, also diminishes the feeling of queasiness in the stomach. Dimenhydrinate (Dramamine), another motion-sickness drug, is not effective on other types of nausea and may cause drowsiness.

Other drugs that have been developed to treat post-operative or post-chemotherapy nausea and vomiting include ondansetron (Zofran) and granisetron (Kytril). Intravenous administration of supplemental fluid before the operation can lower the risk of nausea after surgery.

Alternative treatment

Advocates of alternative treatments suggest biofeedback, acupressure and the use of herbs to calm the stomach. Biofeedback uses **exercise** and deep relaxation to control nausea. Acupressure (applying pressure to specific areas of the body) can be applied by wearing a special wristband or by applying firm pressure to the following:

- the back of the jawbone
- the webbing between the thumb and index finger
- the top of the foot
- the inside of the wrist
- the base of the rib cage

Acupuncture is an alternative treatment found to be effective in relieving nausea. A few people, however, experience nausea as a side effect of acupuncture.

Chamomile (*Matricaria recutita*) or lemon balm (*Melissa officinalis*) tea may relieve symptoms. Ginger (*Zingiber officinale*), another natural remedy, can be ingested as tea or taken as candy, cookies, or powdered capsules.

Prognosis

Most instances of nausea and vomiting respond well to appropriate treatment, including removing any substance or condition that precipitates the nausea.

Prevention

Massage, meditation, **yoga**, and other relaxation techniques can help prevent stress-induced nausea. Anti-nausea medication taken before traveling can prevent motion sickness. Sitting in the front seat, focusing on the horizon, and traveling after dark can also minimize symptoms.

Food should be fresh, properly prepared, and eaten slowly. Overeating, tight-fitting clothes, and strenuous activity immediately after a meal should be avoided.

Vomiting related to emotional upsets may be avoided by forms of psychotherapy that teach people to manage stress in healthier ways.

KEY TERMS

Antiemetic drug—A medication that helps control nausea; also called an antinausea drug.

Dehydration—An excessive loss of water from the body. It may follow vomiting, prolonged diarrhea, or excessive sweating.

Diabetic coma—A life-threatening, reduced level of consciousness that occurs in persons with uncontrolled diabetes mellitus.

Emesis—An act or episode of vomiting.

Nutritional concerns

Prolonged vomiting can lead to fluid and electrolyte depletion. Nausea can curtail appetite. Over time, this can lead to nutritional problems.

Parental concerns

Parents should be especially concerned about prolonged vomiting in children younger than two years of age. This concern intensifies if the vomiting is accompanied by **diarrhea** that accelerates fluid and electrolyte depletion. Parents should consult a pediatrician for treatment options if an infant younger than six months of age vomits multiple times within several hours.

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American Academy of Family Physicians. 11400 Tomahawk Creek Parkway, Leawood, KS 66211–2672. Web site: <www.aafp.org/>.

American Academy of Pediatrics. 141 Northwest Point Boulevard, Elk Grove Village, IL 60007–1098. Web site: <www.aap.org/default.htm>.

American Academy of Physical Medicine and Rehabilitation. One IBM Plaza, Suite 2500, Chicago, IL 60611–3604. Web site: <www.aapmr.org/>.

American Association of Poison Control Centers. 3201 New Mexico Avenue NW, Washington, DC 20016. Web site: <www.aapcc.org/>.

American Board of Obstetrics and Gynecology. 2915 Vine Street Suite 300, Dallas TX. 75204. Web site: <www.abog.org/>.

American College of Gastroenterology. 4900 B South 31st Street, Arlington VA 22206. Web site: <www.acg.gi.org/>.

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Near-drowning

Definition

Near-drowning is the term for survival after suffocation caused by submersion in water or other fluid. Some experts exclude from this definition cases of temporary survival that end in death within 24 hours, which they prefer to classify as drownings.

Description

Drowning is always fatal, but near-drowning may result in survival with no long-lasting effects; survival with permanent damage, usually to the brain; or death after a 24-hour survival period. Near drowning sets into motion a collection of reactions in the body that ultimately can damage the lungs and lead to an absence of oxygen in tissues, even when individuals have been removed from the water and begun breathing either on their own or with mechanical help.

Near-drowning happens very quickly. Within three minutes of submersion, most people are unconscious, and within five minutes the brain begins to suffer from lack of oxygen. Abnormal heart rhythms (cardiac dysrhythmias) often occur in near-drowning cases, and the heart may stop pumping (cardiac arrest). The blood may increase in acidity (acidosis) and, under some circumstances, near drowning can cause a substantial increase or decrease in the volume of circulating blood. If not rapidly reversed, these events cause permanent damage to the brain.

Demographics

About 1,500 children drown every year in the United States. Drowning is the second leading cause of injury-related deaths in children ages one month to 14 years of age in the United States as a whole, and the first leading cause of injury-related deaths in California, Arizona, and Florida. The rate of near drowning is much higher, as not all near drownings are reported. It is estimated that for every drowning, there are four additional hospitalizations and 14 additional emergency room visits due to near drowning.

Children under age four and between 15 and 19 years of age are at highest risk of drowning or near drowning. Most young children drown in swimming pools and bathtubs, while teens drown in natural bodies of water. Teen drownings are often associated with boating accidents, alcohol consumption, and illicit drug use. Boys are 12 times more likely to drown than girls, especially during **adolescence**, when risk-taking behavior is more pronounced in males. However, even in younger age groups, except in bathtub drownings, substantially more boys drown than girls.

Causes and symptoms

The circumstances leading to near-drownings and drownings are varied. Rarely do they involve nonswimmers accidentally entering deep water. In older children and adults, near-drownings are often secondary to an event such as or a head or spinal injury or (in adults) a heart attack that causes unconsciousness and prevents a diver from resurfacing.

Near-drownings can occur in shallow as well as deep water. Small children have drowned or almost drowned in bathtubs, toilets, industrial-size cleaning buckets, and washing machines. Bathtubs are especially dangerous for infants six months to one year of age, who can sit up straight in a bathtub but may lack the ability to pull themselves out of the water if they slip under the surface. One 2004 study found that 88 percent of children who drowned were under the supervision of another person, usually a **family** member. Seventy-seven percent of these children were under age ten. The most common occurrence was that the supervising adult knew the child was in or near the water but was distracted long enough for the child to drown.

A reduced concentration of oxygen in the blood (hypoxemia) is common to all near-drownings. When drowning begins, the larynx (a part of the air passage) closes involuntarily, preventing both air and water from entering the lungs. In 10 to 20 percent of cases, hypoxemia results because the larynx spasms and stays closed.

This is called "dry drowning," and no water is breathed into the lungs. Hypoxemia also occurs in "wet drowning," when the larynx relaxes and water enters the lungs. Individuals who are close to drowning can also regurgitate their stomach contents and breathe these into the lungs.

The physiological mechanisms that produce hypoxemia in wet drowning are different for freshwater and saltwater, but only a small amount of either kind of water is needed to damage the lungs and interfere with lung's ability to remove oxygen from the air.

The signs and symptoms of near-drowning can differ from person to person depending in part on how long the individual has been submerged, the person's age, and the temperature of the water. Upon rescue, some victims are alert but agitated or disoriented, while others are comatose. Breathing and heartbeat may have stopped, or the victim may be gasping for breath. Bluish skin (cyanosis), coughing, **vomiting**, and frothy pink sputum (material expelled from the respiratory tract by coughing) are often observed. Rapid breathing (tachypnea) and a rapid heart rate (tachycardia) are common during the first few hours after rescue. The victim may experience hypothermia (drop in core body temperature).

When to call the doctor

Emergency medical aid should be sought with any near drowning incident. Even a child who appears to have recovered should be checked by a physician, since some internal reactions to near drowning can be delayed.

Diagnosis

Diagnosis relies on a physical examination of the victim, reports of observers, and a wide range of tests and other procedures. Blood is taken to measure oxygen levels and to determine electrolyte balances. Pulse oximetry, another way of assessing oxygen levels, involves attaching a device called a pulse oximeter to the patient's finger. An electrocardiograph is used to monitor heart activity. X rays can detect head and neck injuries and fluid in the lungs.

Treatment

Treatment begins with removing the victim from the water and performing **cardiopulmonary resuscitation** (CPR) as needed to restore heartbeat and provide oxygen until the individual is able to breath without assistance. When emergency medical help arrives, 100 percent oxygen is administered to the victim. If the victim's breathing has stopped or is otherwise impaired, a tube is inserted into the windpipe (trachea) to maintain the airway (endotracheal intubation). The victim is also checked for head, neck, and other injuries, and intravenously fluids may be started. Hypothermia from submersion in very cold water requires special handling to protect the heart.

On arriving at the emergency room, the individual continues receiving oxygen until blood tests show a return to normal. About one-third of near-drowning victims are intubated and initially need mechanical support to breathe. Treatment is administered as needed for cardiac arrest or cardiac dysrhythmias. Slow rewarming is undertaken when hypothermia is present. Individuals are observed for the development of acute **respiratory distress syndrome** (ARDS) or multi-organ failure, both of which can develop after near drowning. Lung problems can develop 12 or more hours after submersion.

Based on symptoms, individuals may be admitted to the hospital or discharged from the emergency department after four to six hours, if their blood oxygen level is normal and no signs or symptoms of near-drowning are present. Discharged individuals must understand that should complications arise, they must immediately seek additional medical care. Admission to a hospital for at least 24 hours for further observation and treatment is necessary for patients who do not appear to fully recover in the emergency department.

Prognosis

Recovery is directly related to the amount of time the body was without adequate oxygen (hypoxia). Brain damage is the major long-term concern in the treatment of near-drowning victims. Patients who arrive at an emergency department awake and alert usually survive with brain function intact, although they may initially have respiratory complications. **Pneumonia** is common following near drowning and often develops within the first 24 hours.

Death or permanent neurological damage is very likely when patients arrive at the emergency room comatose or without a heartbeat. Of these patients, 35 to 60 percent die in the emergency department, while almost all of those who survive have permanent disabilities. Early rescue of near-drowning victims (within five minutes of submersion) and prompt CPR (within less than ten minutes of submersion) seem to be the best guarantees of a complete recovery. However, in a phenomenon that is not well understood, extremely cold water (less than 41°F or 5°C) seems to protect individuals from some of the neurological damage that occurs with near drowning. Some hypothermic near-drowning victims

KEY TERMS

Cyanosis—A bluish tinge to the skin that can occur when the blood oxygen level drops too low.

Electrolytes—Salts and minerals that produce electrically charged particles (ions) in body fluids. Common human electrolytes are sodium chloride, potassium, calcium, and sodium bicarbonate. Electrolytes control the fluid balance of the body and are important in muscle contraction, energy generation, and almost all major biochemical reactions in the body.

Hypothermia—A serious condition in which body temperature falls below 95°F (35 °C). It is usually caused by prolonged exposure to the cold.

Hypoxemia—A condition characterized by an abnormally low amount of oxygen in the arterial blood. It is the major consequence of respiratory failure, when the lungs no longer are able to perform their chief function of gas exchange.

Hypoxia—A condition characterized by insufficient oxygen in the cells of the body

have been revived after they appeared dead and have experienced few permanent disabilities.

Prevention

Drowning and near drowning are almost always preventable. Prevention depends on educating adults and children about water **safety**. Children cannot be left in or near water without adult supervision even for a short time. Unsupervised young children are at risk around swimming pools, bathtubs, toilets, buckets, and natural bodies of water. Pools and spas need to be enclosed with a fence at least 5 ft (1.5 m) high and have a self-closing and self-locking gate. Adults and teens should consider learning CPR. No one should swim alone or **play** along flooded streams or streets. Teens and adults should be educated to understand that alcohol and illicit drug use substantially increase the chances of a drowning accident. Boat owners need to participate in boat safety classes, and children should wear approved life preservers when boating, water skiing, or riding on a jet ski.

Parental concerns

Parents should be aware that physicians are required to consider the possibility of **child abuse** in every drowning or near drowning involving a child under the age of one year.

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Nearsightedness *see* **Myopia**

Necrotizing enterocolitis

Definition

Necrotizing enterocolitis (NEC) is a serious bacterial infection in the intestine, primarily affecting sick or premature newborn infants. It can cause the death (necrosis) of intestinal tissue and progress to blood poisoning (septicemia).

Description

Necrotizing enterocolitis is a serious infection that can produce complications in the intestine itself such as ulcers, perforations or holes in the intestinal wall, and tissue necrosis. It can also progress to life-threatening septicemia. Necrotizing enterocolitis most commonly affects the ileum, the lower portion of the small intestine. It is less common in the colon and upper small bowel.

Demographics

It is estimated that narcotizing enterocolitis affects 2 percent of all newborns, but it is more frequently seen in very low birth weight infants, affecting as many as 13.3 percent of these babies. It has a high mortality rate, especially among very low birth weight babies. Some 20 to 40 percent of these infants die. It does not appear that male or females are more susceptible to this condition, and no one race or nationality has a higher incidence.

Causes and symptoms

The cause of necrotizing enterocolitis is not clear. It is believed that the infection usually develops after the bowel wall has already been weakened or damaged by a lack of oxygen, predisposing it to bacterial invasion. Bacteria grow rapidly in the bowel, causing a deep infection that can kill bowel tissue and spread to the bloodstream.

Necrotizing enterocolitis almost always occurs in the first month of life. Infants who require tube feedings may have an increased risk for the disorder. A number of other conditions also make newborns susceptible, including **respiratory distress syndrome**, congenital heart problems, and episodes of apnea (cessation of breathing). The primary risk factor, however, is **prematurity**. Not only is the immature digestive tract less able to protect itself, but premature infants are subjected to many stresses on the body in their attempt to survive.

Early symptoms of necrotizing enterocolitis include an intolerance to formula, distended and tender abdomen, **vomiting**, and blood (visible or not) in the stool. One of the earliest signs may also be the need for mechanical support of the infant's breathing. If the infection spreads to the bloodstream, infants may develop lethargy, fluctuations in body temperature, and may periodically stop breathing.

Diagnosis

The key to reducing the complications of this disease is early detection by the physician. A series of x rays of the bowel often reveals the progressive condition, and blood tests confirm infection.

Treatment

Over two-thirds of infants can be treated without surgery. Aggressive medical therapy with **antibiotics** is begun as soon as the condition is diagnosed or even suspected. Tube feedings into the gastrointestinal tract (enteral **nutrition**) are discontinued, and tube feedings into the veins (parenteral nutrition) are used instead until the condition has resolved. Intravenous fluids are given for several weeks while the bowel heals.

Some infants are placed on a ventilator to help them breathe, and some receive transfusions of platelets, which help the blood clot when there is internal bleeding. Antibiotics are usually given intravenously for at least 10 days. These infants require frequent evaluations by the physician, who may order multiple abdominal x rays and blood tests in order to monitor their condition during the illness.

Sometimes, necrotizing enterocolitis must be treated with surgery. This is often the case when an infant's condition does not improve with medical therapy or there are signs of worsening infection.

The surgical treatment depends on the individual patient's condition. Patients with infection that has caused serious damage to the bowel may have portions of the bowel removed. It is sometimes necessary to create a substitute bowel by making an opening (ostomy) into the abdomen through the skin, from which waste products are discharged temporarily. But many physicians avoid this and operate to remove diseased bowel and repair the defect at the same time.

Postoperative complications are common, including wound infections and lack of healing, persistent sepsis and bowel necrosis, and a serious internal bleeding disorder known as disseminated intravascular coagulation.

Prognosis

Necrotizing enterocolitis is the most common cause of death in newborns undergoing surgery. The average mortality is 30 to 40 percent, even higher in severe cases.

Early identification and treatment are critical to improving the outcome for these infants. Aggressive nonsurgical support and careful timing of surgical intervention have improved overall survival; however, this condition can be fatal in about one third of cases. With the resolution of the infection, the bowel may begin functioning within weeks or months. But infants need to be carefully monitored by a physician for years because of possible future complications.

About 10 to 35 percent of all survivors eventually develop a stricture, or narrowing, of the intestine that occurs with healing. This can create an intestinal obstruction that requires surgery. Infants may also be more susceptible to future bacterial infections in the gastrointestinal tract and to a delay in growth. Infants with severe cases may also suffer neurological impairment.

The most serious long-term gastrointestinal complication associated with necrotizing enterocolitis is short-bowel, or short-gut, syndrome. This refers to a condition that can develop when a large amount of bowel must be removed, making the intestines less able to absorb certain nutrients and enzymes. These infants gradually evolve from tube feedings to oral feedings, and medications are used to control the malabsorption, **diarrhea**, and other consequences of this condition.

Prevention

In very small or sick premature infants, the risk for necrotizing enterocolitis may be diminished by beginning parenteral nutrition and delaying enteral feedings for several days to weeks.

Breast-fed infants have a lower incidence of necrotizing enterocolitis than formula-fed infants; however, conclusive data showing that breast milk may be protective was as of 2004 not available. A large multicenter trial showed that steroid drugs given to women in preterm labor may protect their offspring from necrotizing enterocolitis.

Sometimes necrotizing enterocolitis occurs in clusters, or outbreaks, in hospital newborn (neonatal) units. Because there is an infectious element to the disorder, infants with necrotizing enterocolitis may be isolated to avoid infecting other infants. Persons caring for these infants must also employ strict measures to prevent spreading the infection.

Parental concerns

Approximately 75 percent of all babies with necrotizing enterocolitis survive. After discharge from the hospital, these infants return home still requiring special care. Many have an ostomy. This is an external opening for the intestinal contents to exit the body while the affected part of the intestine heals. Parents and caregivers need instruction on how to care for the ostomy. Many sources advise parents to room in with the baby prior to discharge from the hospital so that they can learn how to care for the special health needs of infants recovering from necrotizing enterocolitis. Additionally, many of these infants have a condition called short-gut syndrome, which results from the removal of a large part of the small intestine. The small bowel will grow in time, but for as long as two years in some cases, the child will require careful monitoring of his or her nutritional intake to insure that he is receiving adequate levels of **vitamins**, **minerals**, and calories. These children will require tube feedings, and parents will need proper instruction in this type of feeding.

KEY TERMS

Enteral nutrition—Liquid nutrition provided through tubes that enter the gastrointestinal tract, usually through the mouth or nose.

Necrosis—Localized tissue death due to disease or injury, such as a lack of oxygen supply to the tissues.

Ostomy—A surgically-created opening in the abdomen for elimination of waste products (urine or stool).

Parenteral nutrition—Liquid nutrition usually provided intravenously.

Sepsis—A severe systemic infection in which bacteria have entered the bloodstream or body tissues.

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Neisseria meningitidis disease *see* **Meningococcemia**

Neisseria meningitidis vaccine *see* **Meningococcal meningitis vaccine**

Neonatal jaundice

Definition

Neonatal **jaundice** is the term used when a newborn has an excessive amount of bilirubin in the blood. Bilirubin is a yellowish-red pigment that is formed and released into the bloodstream when red blood cells are broken down. Jaundice comes from the French word

jaune, which means yellow; thus a jaundiced baby is one whose skin color appears yellow due to bilirubin.

Description

Normally, small amounts of bilirubin are found in everyone's blood. It is formed and released into the bloodstream when red blood cells are broken down. It is then carried to the liver where it is processed and eventually excreted from the body. When too much bilirubin is made, the excess is discarded into the bloodstream and deposited in tissues for temporary storage. In the neonate, however, there is more bilirubin than can be handled due to immature liver functioning and extra red blood cells that break down. Thus, the extra bilirubin remains in the tissues. Neonatal jaundice affects 60 percent of full-term infants and 80 percent of preterm infants in the first three days after birth.

Demographics

Infants of East Asian and Native American descent have higher levels of bilirubin than white infants, who in turn have higher bilirubin levels than infants of African descent. There is an enzyme, glucose-6-phosphate dehydrogenase (G6PD), deficiency that is more prevalent in infants of East Asian, Greek, and African descent which causes neonatal jaundice to appear at approximately the same time as physiological jaundice. **Sickle cell anemia** does not predispose newborn infants to jaundice.

Causes and symptoms

Typically, neonatal jaundice occurs in otherwise healthy infants for two reasons. First, infants have too many red blood cells and it is a natural process for the body to break down these excess red blood cells to form a large amount of bilirubin. It is this bilirubin that causes the skin to take on a yellowish color. Second, the newborn's liver is immature and cannot process bilirubin as quickly as the infant will be able to when older. This slow processing of bilirubin has nothing to do with liver disease. It merely means that the baby's liver is not as fully developed as it will be; thus, there is some delay in eliminating the bilirubin.

Breastfeeding is an important risk factor for hyperbilirubinemia in healthy infants and is related to inadequate maternal milk supply in the first few days, decreased caloric intake and delayed passage of meconium. Nonetheless, this is not a reason to give formula or stop breastfeeding. The breastfeeding mother just needs to nurse the baby more frequently and for longer periods of time to enhance the production of breastmilk. Other factors that cause neonatal jaundice are ABO incompatibility and Rh incompatibility. Both of these conditions result in a very fast breakdown of red blood cells. It is also possible for jaundice to appear in infants with physical defects in the organs that work to eliminate bilirubin from the body. An abnormal increase in red blood cells is frequently seen in infants who are large or small for their gestational age, as well as in trisomy syndromes, twin-to-twin transfusion syndrome, maternal-fetal transfusion, use of oxytocin in labor, Asian male babies, presence of bruising and cephalohematoma, and a **family** history of neonatal jaundice.

As the excess bilirubin builds up in the newborn, jaundice appears first in the face and upper body and progresses downward toward the toes. Most babies with jaundice have physiologic jaundice, which is the type caused by the natural process of breaking down red blood cells. If the baby's jaundice is caused by any other conditions, however, the healthcare giver will provide the parents with additional information for caring for the baby.

When to call the doctor

With short neonatal hospital stays, jaundice will not have peaked or become apparent at the time of hospital discharge. Therefore, infants at risk for severe hyperbilirubinemia should be identified so they can be observed closely both while in the hospital and after discharge. The parents need to be instructed on how to evaluate the infant for jaundice. They should look for it first in the face and upper body and if it progresses downward this means the concentration is getting too high and it is time to call the pediatrician. If there is an area of their living quarters that gets sunlight, it helps to let the baby lie there in only a diaper for a short period of time each day.

Diagnosis

Jaundice can be observed with the naked eye, but it is too difficult to estimate the variation in levels of bilirubin in that manner. Thus, if an infant begins to appear jaundiced, bilirubin levels will be ordered to determine the severity. Jaundice usually becomes apparent when total bilirubin levels exceed 5 mg/dL; however, the clinical significance of bilirubin levels depends on postnatal age in hours. A bilirubin level of 12 mg/dL may be pathologic in an infant younger than 48 hours but is benign in an infant older than 72 hours. In the determination of cause, it is suggested that laboratory testing be reserved for infants with nonphysiologic jaundice. In up to 50 percent of infants with severe jaundice, breast-

feeding and lower gestational age were the only causes identified despite extensive workups.

Treatment

The mainstay in treatment of hyperbilirubinemia is phototherapy, which is safe and widely available. Its effectiveness was demonstrated in a study by the National Institute of Child Health and Human Development. Multiple factors can influence the effectiveness of phototherapy, including the type and intensity of the light and the extent of skin surface exposure. Special blue fluorescent light has been shown to be most effective, although many nurseries use a combination of daylight, white, and blue lamps. In the early 2000s, fiberoptic blankets have been developed that emit light in the blue-green spectrum, which is light at a wavelength of 425–475 nm. Light at this wavelength converts bilirubin to a water-soluble form that can be excreted in the bile or urine. The intensity of light delivered is inversely related to the distance between the light source and the skin surface. Since phototherapy acts by altering the bilirubin that is deposited in the tissue, the area of the skin exposed to phototherapy should be maximized. This has been made more practical with the development of fiberoptic phototherapy blankets that can be wrapped around an infant.

Home-based care for neonatal jaundice has become more prevalent than hospital care, and the availability of fiberoptic blankets has made it possible. Infants receiving home phototherapy need daily visits by a nurse, who performs a physical examination and measures the total serum bilirubin level. If bilirubin levels continue to rise, hospital readmission should be considered. Discontinuation of home phototherapy is safe once the total serum bilirubin level has decreased to less than 15 mg/dL in healthy full-term infants older than four days. Office evaluation within two to three days of discontinuing home phototherapy is recommended.

Potential side effects of phototherapy used for elevated bilirubin levels, include watery **diarrhea**, increased water loss, skin rash, and transient bronzing of the skin. Many infants who are readmitted to the hospital because of hyperbilirubinemia are mildly to moderately dehydrated. Breastfeeding should be increased to every two to two and a half hours. Increased feedings can increase peristalsis and meconium passage, decreasing bilirubin resorption into circulation.

Full-term infants rarely require an exchange transfusion if intense phototherapy is initiated in a timely manner. It should be considered if the total serum bilirubin level is approaching 20 mg/dL and continues to rise despite intense in-hospital phototherapy. Exchange transfusion corrects anemia associated with the destruction of red blood cells and is effective in removing sensitized red blood cells before they are destroyed. It also removes about 60 percent of bilirubin from the plasma, resulting in a clearance of about 30 percent to 40 percent of the total bilirubin. If a transfusion is not performed and bilirubin levels get higher, the infant progresses through three phases. In the first two to three days the infant is lethargic, has muscle weakness, and sucks weakly. Progression is marked by a tensing of the muscles, arching, **fever**, seizures, and high-pitched crying. In the final phase, the patient is hypotonic for several years.

A newborn baby undergoes phototherapy with visible blue light to treat his jaundice. *(Photograph by Ron Sutherland. Photo Researchers, Inc.)*

Prognosis

The prognosis for physiological neonatal jaundice is generally very good. Very few infants ever have bilirubin levels greater than 20 mg/dL, which is the level that is correlated with kernicterus (an abnormal accumulation

KEY TERMS

ABO incompatability—The reaction that occurs with blood groups that are of a different type.

Cephalohematoma—A benign swelling of the scalp in a newborn due to an effusion of blood beneath the connective tissue that surrounds the skull, often resulting from birth trauma.

Kernicterus—A potentially lethal disease of newborns caused by excessive accumulation of the bile pigment bilirubin in tissues of the central nervous system.

Meconium—A greenish fecal material that forms the first bowel movement of an infant.

Oxytocin—A hormone that stimulates the uterus to contract during child birth and the breasts to release milk.

Peristalsis—Slow, rhythmic contractions of the muscles in a tubular organ, such as the intestines, that move the contents along.

Rh incompatability—A factor of blood classified as negative or positive and related to the reaction that occurs between different types.

Trisomy—An abnormal condition where three copies of one chromosome are present in the cells of an individual's body instead of two, the normal number.

of bile pigment in the brain and other nerve tissue that causes yellow staining and tissue damage). It rarely occurs with bilirubin levels lower than 20 mg/dL but typically occurs when levels exceed 30 mg/dL. Levels between 20 and 30 mg/dL associated with **prematurity** and hemolytic disease may increase the risk of kernicterus. There are long-term neurological problems when this occurs. Affected children have marked developmental and motor delays in the form of **cerebral palsy** and **mental retardation** may also be present.

Prevention

Elevated bilirubin in the neonate is the most common reason for hospital readmission in the first two weeks of life. Kernicterus is still relatively uncommon but has been on the rise with the mandated early postnatal discharge policies. Bilirubin-induced complications can be prevented by introducing a neonatal jaundice protocol to identify infants at risk for significant bilirubin increases, by ensuring adequate parental education and providing for follow-up care.

Parental concerns

Parents of a newborn need to be vigilant in monitoring changes in their infant. If the mother is breastfeeding, she should nurse the baby at least once every three hours to ensure the onset of milk production and to maintain hydration, which can also be evaluated by the number of wet diapers. Many pediatricians recommend seeing the infant at two weeks but if the parents feel it should be sooner due to alterations in the newborn's physical status, they should take the infant in for a visit.

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Association of Women's Health, Obstetric and Neonatal Nursing. 2000 L Street, NW, Suite 740, Washington, DC 20036. Web site: <www.awhonn.org>.

American Academy of Pediatrics. 141 Northwest Point Blvd., Elk Grove Village, IL, 60007. Web site: <www.aap.org>.

American College of Obstetricians and Gynecologists. 409 12th Street, SW, PO Box 96920, Washington, DC 20090. Web site: <www.acog.org>.

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Neonatal reflexes

Definition

Neonatal reflexes or primitive reflexes are the inborn behavioral patterns that develop during uterine life. They should be fully present at birth and are gradually inhibited by higher centers in the brain during the first three to 12 months of postnatal life. These reflexes, which are essential for a newborn's survival immediately

after birth, include sucking, swallowing, blinking, urinating, hiccupping, and defecating. These typical reflexes are not learned; they are involuntary and necessary for survival.

Description

A normal birth is considered full term if the delivery occurs during the thirty-seventh to fortieth week after conception. Developmentally, the baby is considered a neonate for the first 28 days of life. At birth, the neonate must immediately make five major adjustments:

- Transition from an aquatic environment to a world of air. The first breath begins even before the umbilical cord is cut.
- Eat and digest his or her own food since the circulatory relationship between mother and baby stops with the severance of the umbilical cord.
- Excrete his or her own wastes.
- Maintain his or her own body temperature.
- Adjust to intermittent feeding since food is now only available at certain intervals.

Under normal developmental conditions, these neonatal reflexes represent important reactions of the nervous system and are only observable within a specific period of time over the first few months of life. The following reflexes are normally present from birth and are part of a normal newborn evaluation:

- The Moro reflex (or startle reflex) occurs when an infant is lying in a supine position and is stimulated by a sudden loud noise that causes rapid or sudden movement of the infant's head. This stimulus results in a symmetrical extension of the infant's extremities while forming a C shape with the thumb and forefinger. This is followed by a return to a flexed position with extremities against the body. Inhibition of this reflex occurs from the third to the sixth month. An asymmetrical response with this reflex may indicate a fractured clavicle or a birth injury to the nerves of the arm. Absence of this reflex in the neonate is an ominous implication of underlying neurological damage.
- Asymmetrical tonic neck reflex (sometimes called the tonic labyrinthine reflex) is activated as a result of turning the head to one side. As the head is turned, the arm and leg on the same side will extend while the opposite limbs bend, in a pose that mimics a fencer. The reflex should be inhibited by six months of age in the waking state. If this reflex is still present at eight to nine months of age, the baby will not be able to support its weight by straightening its arms and bringing its knees beneath its body.
- Symmetrical tonic neck reflex occurs with either the extension or flexion of the infant's head. Extension of the head results in extension of the arms and flexion of the legs, and a flexion of the head causes flexion of the arms and an extension of the legs. This reflex becomes inhibited by the sixth month to enable crawling.
- Grasping reflex occurs as the palmar reflex when a finger is placed in the neonate's palm and the neonate grasps the finger. The palmar reflex disappears around the sixth month. Similarly, the plantar reflex occurs by placing a finger against the base of the neonate's toes and the toes curl downward to grasp the finger. This reflex becomes inhibited around the ninth to tenth month.
- Rooting reflex is stimulated by touching a finger to the infant's cheek or the corner of the mouth. The neonate responds by turning the head toward the stimulus, opening the mouth and searching for the stimulus. This is a necessary reflex triggered by the mother's nipple during breastfeeding. It is usually inhibited by the third to fourth month.
- Sucking reflex is triggered by placing a finger or the mother's nipple in the infant's mouth. The neonate will suck on the finger or nipple forcefully and rhythmically and the sucking is coordinated with swallowing. Like the rooting reflex, it is inhibited by the third to fourth month.
- Babinski or plantar reflex is triggered by stroking one side of the infant's foot upward from the heel and across the ball of the foot. The infant responds by hyperextending the toes; the great toe flexes toward the top of the foot and the other toes fan outward. It generally becomes inhibited from the sixth to ninth month of post natal life.
- Blink reflex is stimulated by momentarily shining a bright light directly into the neonate's eyes causing him or her to blink. This reflex should not become inhibited.
- Pupillary reflex occurs with darkening the room and shining a penlight directly into the neonate's eye for several seconds. The pupils should both constrict equally; this reflex should not disappear.
- Galant reflex is stimulated by placing the infant on the stomach or lightly supporting him or her under the abdomen with a hand and, using a fingernail, gently stroking one side of the neonate's spinal column from the head to the buttocks. The response occurs with the neonate's trunk curving toward the stimulated side. This reflex can become inhibited at any time between the first and third month.
- Stepping reflex is observed by holding the infant in an upright position and touching one foot lightly to a flat

Neonatal reflexes

Reflex	Stimulation	Response	Duration
Babinski	Sole of foot stroked	Fans out toes and twists foot in	Disappears at nine months to a year
Blinking	Flash of light or puff of air	Closes eyes	Permanent
Grasping	Palms touched	Grasps tightly	Weakens at three months; disappears at a year
Moro	Sudden move; loud noise	Startles; throws out arms and legs and then pulls them toward body	Disappears at three to four months
Rooting	Cheek stroked or side of mouth touched	Turns toward source, opens mouth and sucks	Disappears at three to four months
Stepping	Infant held upright with feet touching ground	Moves feet as if to walk	Disappears at three to four months
Sucking	Mouth touched by object	Sucks on object	Disappears at three to four months
Swimming	Placed face down in water	Makes coordinated swimming movements	Disappears at six to seven months
Tonic neck	Placed on back	Makes fists and turns head to the right	Disappears at two months

SOURCE: Table after Child Development, 6th ed. Wm. C. Brown Communications, Inc., 1994.

(Table by GGS Information Services.)

surface, such as the bed. The infant responds by making walking motions with both feet. This reflex will disappear at approximately two months of age.

- Prone crawl reflex can be stimulated by placing the neonate prone (face down) on a flat surface. The neonate will attempt to crawl forward using the arms and legs. This reflex will be inhibited by three to four months of age.
- Doll's eye reflex can be noted with the infant supine (lying on the back) and slowly turning the head to either side. The infant's eyes will remain stationary. This reflex should disappear between three to four months of age.

Common problems

The presence and strength of a reflex is an important indication of neurological functioning. Within the first 24 hours after birth, a healthcare provider evaluates an infant's neurological functioning and development by testing and observing these reflexes. If a reflex is absent or abnormal in an infant, this may suggest significant neurological problems. In normal development, the primary reflex system is inhibited or transformed in the first year of life and a secondary or postural reflex system emerges. The secondary system forms the basis for later adult coordinated movement. Absence or presence of a reflex is a symptom, not a disorder.

Severe persistence of primary reflexes indicates predominantly persistent physical problems. Relatively milder persistence, however, is associated with less severe disorders that include specific reading difficulties.

The process of inhibition of these reflexes in the earliest months of life remains unknown but it has been assumed that this process cannot occur after early childhood because neonatal movement is largely stereotypical and follows the patterns of the primary reflex system. Thus, the early movements of the fetus and newborn were previously viewed as passive byproducts of the central nervous system. They are viewed as interactive and having a reciprocal effect on the underlying central nervous system structure and functioning. This implies that the actual rehearsal and repetition of primary reflex movements play a role in the inhibition process itself.

Parental concerns

An evaluation of neonatal reflexes is performed during well-baby examinations. The abnormal presence of infantile reflexes in an older child can be discovered during a neurological examination. **Assessment** of neonatal reflexes is a screening tool for at-risk children with neurological difficulties. Primary reflexes may persist for certain children beyond their normal time span causing a disruption in subsequent development. Children with neurological damage will have a common denomi-

KEY TERMS

Palmar—Referring to the palm of the hand.

Plantar—Relating to the sole of the foot.

Postural—Pertaining to the position of the head, neck, trunk and lower limbs in relation to the ground and the vertical.

Prone—Lying on the stomach with the face downward.

Supine—Lying on the back with the face upward.

Visuosensory—Pertaining to the perception of visual stimuli.

nator of prolonged neonatal reflexes. Since recent studies have demonstrated that repetition of these reflexes seems to eventually inhibit them, parents can work with the infant by assisting with the repetition of persistent reflexes.

When to call the doctor

Persistence of neonatal reflexes is not threatening to life and, therefore, can be discussed with the pediatrician during normal well-baby visits.

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The International Dyslexia Association. Chester Building, Suite 382;8600 LaSalle Road; Baltimore, MD 21286-2044. (410) 296-0232 or (800) 222-3123. Web site: <www.interdys.org>.

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Neonatal testing *see* **Apgar testing**

Nephroblastoma *see* **Wilms' tumor**

Neural tube defect *see* **Spina bifida**

Neurofibromatosis

Definition

Neurofibromatosis (NF) is a genetic disease in which multiple soft tumors (neurofibromas) develop under the skin and throughout the nervous system. Various sized tumors may grow on the nerves in or leading away from the brain and spinal cord (peripheral nerves) and in the vascular system (veins and arteries) and other organ systems. There are two types of NF: NF-1, also called vonRecklinghausen NF, and NF-2, also called acoustic NF (sometimes bilateral acoustic NF or BAN). NF-1 is more common, representing 90 percent of all cases, while NF-2 is diagnosed in 10 percent of NF cases.

Description

Neural crest cells are primitive cells that are present as part of the nervous system during fetal development. These cells eventually turn into the following:

- cells that form nerves throughout the brain, spinal cord, and body
- cells that serve as coverings around the nerves throughout the body
- pigment cells that provide color to body structures
- meninges, the thin, membranous coverings of the brain and spinal cord
- cells that ultimately develop into the bony structures of the head and neck

In both types of NF, a genetic defect causes these neural crest cells to develop abnormally, resulting in numerous tumors and malformations of the nerves, bones, and skin.

NF-1 affects nerves throughout the body, occurring as groups of soft, fibrous swellings that grow on nerves in the skin, brain, and spinal cord (central nervous system), muscles, and bone. Severe disfigurement can result from the development of these tumors as the disease progresses and bone deformities may occur as well.

NF-2 is a rare type of NF in which multiple tumors grow on the cranial (head) and spinal nerves and other growths can occur in the brain and spinal cord. Tumor growth (schwannoma) on the nerves to the ears (auditory nerves) is most characteristic of NF-2. Disfigurement does not occur although hearing and visual problems are typical.

Demographics

NF-1 is a common genetic disorder that occurs in about one of every 4,000 births worldwide. NF-2 is rare, occurring in one of every 40,000 births. Children with a **family** history of neurofibromatosis are at highest risk for having either form of the condition.

Causes and symptoms

Both forms of neurofibromatosis are caused by a defective gene. NF-1 is due to a defect on chromosome 17; NF-2 results from a defect on chromosome 22. Both of these disorders are inherited in a dominant fashion, which means that anyone who receives just one defective gene will have the disease. However, a family pattern of NF is only evident for about half of all cases of NF. The other cases of NF occur due to a spontaneous mutation (a permanent change in the structure of a specific gene). Once such a spontaneous mutation has been established in an individual, however, it can then be passed on to any offspring. The chance of a person with NF passing on the NF gene to a child is 50 percent.

NF-1 has a number of possible signs and can be diagnosed if any two of the following are present:

- The presence of coffee-colored spots. These are patches of tan or light brown skin, usually about 5 to 15 mm in diameter. Nearly all patients with NF-1 will display these spots.
- Multiple freckles in the armpit or groin area.
- Ninety percent of patients with NF-1 have tiny tumors called Lisch nodules in the iris (colored area) of the eye.
- Soft tumors (neurofibromas) are the hallmark of NF-1. They occur under the skin, often located along nerves or within the gastrointestinal tract. Neurofibromas are small and rubbery, and the skin overlying them may be somewhat purple in color.
- Skeletal deformities, such as a twisted spine (**scoliosis**), curved spine (humpback), or bowed legs.
- Tumors along the optic nerve, causing vision disturbances in about 20 percent of those affected.
- The presence of NF-1 in a child's parent or sibling.

Very high rates of speech impairment, learning disabilities, and attention deficit disorder occur in children with NF-1. Other complications include the development of a **seizure disorder** or the abnormal accumulation of fluid within the brain (**hydrocephalus**). A number of cancers are more common in individuals who have NF-1. These include various types of malignant brain tumors, as well as leukemia and cancerous tumors of certain muscles (rhabdomyosarcoma), the adrenal glands (pheochromocytoma), or the kidneys (**Wilms' tumor**).

Patients with NF-2 do not necessarily have the same characteristic skin symptoms that appear in NF-1. The characteristic symptoms of NF-2 are due to tumors along the acoustic nerve that result in nerve dysfunction and the loss of hearing. The tumor may also spread to neighboring nervous system structures, causing weakness of the muscles of the face, **headache**, **dizziness**, poor balance, and uncoordinated walking. Cloudy areas on the lens of the eye (cataracts) frequently develop at an unusually early age. As in NF-1, the chance of brain tumors developing is unusually high.

When to call the doctor

A history of either form of NF in the child's parent or sibling is reason to consult a physician. The presence of any of the symptoms associated with NF-1 or NF-2 should be investigated by a physician as well, particularly spots on the skin or small movable lumps under the skin and visual disturbances, memory loss, or difficulty maintaining balance. Hearing loss may be the first sign of NF-2 but can also be due to other unrelated conditions.

Diagnosis

Diagnosis is based on characteristic symptoms and physical examination. Diagnosis of NF-1 requires that at least two of the characteristic signs are present. Diagnosis of NF-2 requires the presence of either a nodule or mass (tumor) on the acoustic nerve or another distinctive nervous system tumor, which may only be identifiable

through imaging studies. An important diagnostic clue for either NF-1 or NF-2 is the known presence of the disorder in a child's parent or sibling. Gene studies may be done to detect abnormalities on chromosomes 17 and 22.

Diagnosis of NF-1 will be confirmed by manipulation of the skin to reveal moveable, small, solid lumps (nodules) and the presence of coffee-colored spots on the skin of the trunk and pelvis. The spots may appear in childhood and typically become more noticeable in young adults. Two or more nodules and six or more discolored spots are usually definitive for a diagnosis of NF-1. Curvature of the spin (scoliosis) may be present, elevated blood pressure, and abnormalities in height, weight, and head size may also be noticed on physical examination.

Diagnosis of NF-2 also relies on manipulation of the skin to indicate the presence of nodules and evaluation of hearing and vision to determine any impairment.

X rays, CT scans, and MRI scans are performed to track the development/progression of tumors in the brain and along the nerves. Auditory evoked potential testing (the electric response evoked in the cerebral cortex by stimulation of the acoustic nerve) may be helpful to determine involvement of the acoustic nerve, and EEG (**electroencephalogram**, a record of electrical currents in the brain) may be needed for children who have possible seizures. As the disease progresses, hearing and vision are carefully monitored and imaging studies of the bones are frequently done to watch for the development of deformities.

Treatment

There is no standard treatment for either type of neurofibromatosis. To some extent, the symptoms of NF-1 and NF-2 can be treated individually. Skin tumors can be surgically removed. Some brain tumors and tumors along the nerves, can be surgically removed or treated with chemotherapeutic drugs or x-ray treatments (radiation therapy). Twisting or curving of the spine and bowed legs may be corrected to some degree by surgical treatment or the wearing of a special brace. Social adjustment problems are common among young children with physical deformities caused by the condition.

Prognosis

NF of either type is progressive, and the clinical outcome is not predictable. Prognosis varies depending on the types of tumors that develop. As tumors grow, they begin to destroy surrounding nerves and structures. Ultimately, this destruction can result in blindness, deafness, increasingly poor balance, and increasing difficulty with the coordination necessary for walking. Deformities of the bones and spine can also interfere with walking and other kinds of movement. When cancers develop as a result of NF, prognosis worsens according to the specific type of **cancer**. Successful surgical removal of neurofibromas has a survival rate of 50 to 90 percent.

KEY TERMS

Chromosome—A microscopic thread-like structure found within each cell of the human body and consisting of a complex of proteins and DNA. Humans have 46 chromosomes arranged into 23 pairs. Chromosomes contain the genetic information necessary to direct the development and functioning of all cells and systems in the body. They pass on hereditary traits from parents to child (like eye color) and determine whether the child will be male or female.

Mutation—A permanent change in the genetic material that may alter a trait or characteristic of an individual, or manifest as disease. This change can be transmitted to offspring.

Neurofibroma—A soft tumor usually located on a nerve.

Tumor—A growth of tissue resulting from the uncontrolled proliferation of cells.

Prevention

There is no known way to prevent NF cases that occur as a result of spontaneous change in the genes (mutation). New cases of inherited NF can be prevented with careful genetic counseling. Parents with NF can be encouraged to understand that each of his or her offspring has a 50 percent chance of also having NF. When a parent has NF, and the specific genetic defect causing the parent's disease has been identified, tests can be performed on the fetus (developing baby) during pregnancy. Procedures such as **amniocentesis** or chorionic villus sampling allow small amounts of the baby's cells to be removed for examination. The tissue can then be examined for the presence of the parent's genetic defect. Some families choose to use this information in order to prepare for the arrival of a child with a serious medical problem. Other families may choose not to continue the pregnancy.

Parental concerns

Parents may worried about the development of deformities associated with NF-1. Social workers and psychologists can be consulted about possible counseling for children with the disease, helping them to cope with changes in their bodies that may be hard to accept. Hearing loss and visual disturbances associated with NF-2 are usually not reversible and specialists can be consulted about possible therapies to improve functioning in existing sight or hearing senses. Surgery to remove tumors may require the provision of educational information for both parents and children so that the procedure and possible complications are understood ahead of time.

Resources

ORGANIZATIONS

March of Dimes Birth Defects Foundation. Resource Center, 1275 Mamaroneck Ave., White Plains, NY 10605. Web site: <www.modimes.org>.

National Neurofibromatosis Foundation Inc. 95 Pine St., 16th Floor, New York, NY 10005. Web site: <nf.org>.

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Neurologic exam

Definition

A neurological examination is an essential component of a comprehensive physical examination. It is a systematic examination that surveys the functioning of nerves delivering sensory information to the brain and carrying motor commands (peripheral nervous system) and impulses back to the brain for processing and coordinating (central nervous system).

Purpose

A careful neurological evaluation can help to determine the cause of impairment and help a clinician begin to localize the problem. Symptoms that occur unexpectedly suggest a blood vessel or seizure problem. Those that are not so sudden suggest a possible tumor. Symptoms that have a waning course with recurrences and worsen over time suggest a disease that destroys nerve cells. Others that are chronic and progressive indicate a degenerative disorder. In cases of trauma, symptoms may be evident upon inspection and causes may be explained by third party witnesses. Some patients may require extensive neurological screening examination (NSE) and/or neurological examination (NE) to determine the cause. The NE will assist the clinician in diagnosing illnesses as diverse as seizure disorders, **narcolepsy**, migraine disorders, **dizziness**, and dementia.

Description

A neurological screening is an essential component of every comprehensive physical examination. In cases of neurological trauma, disease, or psychological disorders, patients are usually given an in-depth neurological examination. The examination is performed in a systematic manner, which means that there is a recommended order for procedures.

Neurological screening examination

The NSE is basic procedure, especially in patients who have a general neurological complaint or symptoms. The NSE consists of six areas of **assessment**:

- mental status: assessing normal orientation to time, place, space, and speech
- cranial nerves: checking the eyes with a special light source (ophthalmoscope) and also assessing facial muscles strength and functioning
- motor: checking for tone, drift, and heel and toe walking
- sensory: cold and vibration testing
- coordination: observing the patient walk and finger to nose testing
- reflexes: using a special instrument the clinician taps an area above a nerve to emit a reflex (usually movement of muscle groups)

Neurological examination

The NE should be performed on a patient suspected of having neurological trauma, or neurological or psychological diseases. The NE is performed in a systematic and comprehensive manner. It consists of several comprehensive and in-depth assessments of mental status, cranial nerves, motor abilities, reflexes, sensory acuity, and posture and walking (gait) abilities.

MENTAL STATUS EXAMINATION (MSE) There are two types of MSE, informal and formal. The informal MSE is usually done as clinicians are obtaining historical information from a patient. The formal MSE is performed for a patient suspected of a neurological problem. The patient is commonly asked his/her name, the location, the day, and date. Determining the number of digits that can be repeated in sequence can assess retentive memory capability and immediate recall. Recent memory is typically examined by testing recall potential of a series of objects after defined times, usually within five and 15 minutes. Asking the patient to review in a coherent and chronological fashion his or her illness or personal life events can provide the opportunity for assessment of remote memory. Patient recall of common historical or current events can be used to assess general knowledge. Brain processing capabilities can be assessed by spontaneous speech, repetition, reading, naming, writing, and comprehension. Modifications can be made based on the age and maturity of the child. The child may be asked to perform tasks such as identification of fingers, whistling, saluting, brushing teeth motions, combing hair, drawing, and tracing figures. These procedures allow for assessment of dominant (left-sided brain) functioning or higher cortical function.

The MSE is particularly important in psychotherapy. Psychotherapists recommend an in-depth MSE for all patients with possible organic (physiologica) or psychotic disorders. This examination is also performed in a systematic and orderly manner. It is divided into several categories:

- Appearance determines the child's presentation, i.e., how the child looks (clothes posture, grooming, and alertness).
- Behavior assesses the patient's motor activity (movements) such as walking, gestures, muscular twitching, and impulse control.
- Speech can be examined concerning volume, rate of speech, and coherence. Individuals who exhibit latent or delayed speech may be depressed, while those who have rapid or pressured speech may suffer from mania or **anxiety**.
- Mood and affect indicate attitude or feeling. Normal mood (euthymia) is healthy. Variations in mood include: flat, labile, blunted, constructed, or inappropriate mood. The child can also be euphoric (elevated) or dysphoric (on the down side).
- Thought processes and content is typically assessed by determining word usage (can indicate brain disease), thought stream (slow, restricted, blocked, or overabundant), continuity of thought (associations among ideas), and content of thought (delusional as opposed to reality-bound).
- Perception assessment examines the individual's sensory ability to hear, see, touch, taste, and smell. Certain psychological states may cause hearing and visual hallucinations. Impairments of smell and touch usually have medical (organic) causes or are side effects of certain medications.
- Attention and concentration assessment indicates the child's ability to focus on a specific task or activity. Abnormalities in attention and concentration can indicate problems related to anxiety or hallucinations.
- Orientation assessment determines if the child has a normal sense of time, place, and identification of self (can state his or her own name). Disturbances in orientation can be due to a medical condition (other than psychological), substance abuse, or to a side effect of certain medications such as those used to treat depression, anxiety, or psychosis, since these medications usually have a sedative affect.
- Memory assessment includes determining the child's remote, recent, and immediate memory capabilities. Remote and recent memory can be assessed by the patient's ability to recall historical and current events. Immediate memory can be tested by naming three objects and asking the child to repeat the named objects immediately, then after five and 15 minute intervals.
- Judgment assessment evaluates the individual's ability to exercise appropriate judgment. It also determines whether the individual has an understanding of consequences associated with their actions. This evaluation pertains primarily to older children.
- **Intelligence** and information measurement can be obtained by administering specialized intelligence tests, However, a preliminary assessment of intelligence can be made based on the child's fund of age-appropriate information, general knowledge, awareness of current events, and the ability for abstract thinking.
- Insight assessment pertains to determining the patient's awareness of their problem that prompted them to seek professional examination. Insight concerning the present illness can range from denial to fleeting admission of current illness.

CRANIAL NERVES (CN) Cranial nerves are nerves that originate in the brain and connect to specialized structures such as the nose, eyes, muscles in the face, scalp, ear, and tongue.

- CN I: This nerve checks for visual capabilities. Patients are usually given the Snellen Chart (a chart

with rows of large and small letters). Patients read letters with one eye at a time.

- CN III, IV, and VI: These nerves examine the pupillary (the circular center structure of the eye that light rays enter) reaction. The pupils get smaller, normally when exposed to the light. The eyelids are also examined for drooping or retraction. The eyeball is also checked for abnormalities in movement.
- CN V: The clinician can assess the muscles on both sides of the scalp muscles (the temporalis muscle). Additionally the jaw can be tested for motion resistance, opening, protrusion, and side-to-side mobility. The cornea located is a transparent tissue covering the eyeball and can be tested for intactness by lightly brushing a wisp of cotton directly on the outside of the eye.
- CN VII: Examination of CN VII assesses asymmetry of the face at rest and during spontaneous movements. The patient is asked to raise eyebrows, wrinkle forehead, close eyes, frown, smile, puff cheeks, purse lips, whistle, and contract chin muscles. Taste for the front and middle portions of the tongue can also be examined.
- CN VIII: Testing for this CN deals with hearing. The clinician usually uses a special instrument called a tuning fork and tests for air conduction and structural problems which can occur inside the ear.
- CN IX and X: These tests evaluate certain structures in the mouth. The clinician will usually ask the patient to say ''aah'' and can detect abnormal positioning of certain structures such as the palatel-uvula. The examiner will also assess the sensation capabilities of the pharynx, by stimulating the area with a wooden tongue depressor, causing a gag reflex.
- CN XI: This nerve is usually examined by asking the patient to shrug shoulders (testing a muscle called the trapezius) and rotating the head to each side (testing a muscle called the sternocleidomastoid). These muscles are responsible for movement of the shoulders and neck. The test is usually done with resistance, meaning the examiner holds the area while the patient is asked to move. This is done to assess patient's strength in these areas.
- CN XII: This nerve tests the bulk and power of the tongue. The examiner looks for tongue protrusion and/or abnormal movements.

MOTOR EXAMINATION The motor examination assesses the patient's muscle strength, tone, and shape. Muscles could be larger than expected (hypertrophy) or smaller due to tissues destruction (atrophy). It is important to assess if there is evidence of twitching or abnormal movements. Involuntary movements due to **tics** can be observed. Additionally, movements can be abnormal during maintained posture in some neurological disorders. Muscle tone is usually tested by applying resistance to passive motion of a relaxed limb. Power is assessed for movements at each joint. Decreases or increases in muscle tone can help the examiner localize the affected area.

REFLEXES The patient's reflexes are tested by using a special instrument that looks like a little hammer. The clinician taps the rubber triangular shaped end in several different areas in the arms, knee, and Achilles heel area. The clinician will ask the patient to relax and gently tap the area. If there is a difference in response from the left to right knee, then there may be an underlying problem that merits further evaluation. A difference in reflexes between the arms and legs usually indicates of a lesion involving the spinal cord. Depressed reflexes in only one limb, while the other limb demonstrates a normal response usually indicates a peripheral nerve lesion.

SENSORY EXAMINATION Although an essential component of the NE, the sensory examination is the least informative and least exacting since it requires patient concentration and cooperation. Five primary sensory categories are assessed: vibration (using a tuning fork), joint position (examiner moves the limb side-to-side and in a downward position), light touch, pinprick, and temperature. Patients who have sensory abnormalities may have a lesion above the thalamus. Spinal cord lesions or disease can possibly be detected by pinprick and temperature assessment.

COORDINATION The patient is asked to repetitively touch his nose using his index finger and then to touch the clinician's outstretched finger. Coordination can also be assessed by asking the patient to alternate tapping the palm then the back of one hand on the thigh. For coordination in the lower extremities (legs), the patient lies on his or her back and is asked to slide the heel of each foot from the knee down the shin of the opposite leg and to raise the leg and touch the examiner's index finger with the big toe.

WALKING (GAIT) Normal walking is a complex process and requires use of multiple systems such as power, coordination, and sensation, all working together in a coordinated fashion. The examination of gait can detect a variety of disease states. Decreased arm swinging on one side is indicative of corticospinal tract disease. A high-stepped, slapping gait may be the result of a peripheral nerve disease.

Precautions

A neurologic examination is not invasive and there are no risks or dangers associated with these tests. The results and validity of this exam may be affected by the child's age and ability to cooperate.

Preparation

The MSE is the first step in a continuous assessment to determine the diagnosis. A psychotherapist should take a detailed medical history in the process of ruling out a general medical condition. Little preparation is needed for this assessment, but parents should explain to young children what will happen in order to encourage their cooperation.

Aftercare

For suspected neurological diseases, the doctor uses information gained from the NE for ordering further tests. These tests may include a complete blood analysis, liver function tests, kidney function tests, hormone tests, and a lumbar puncture to determine abnormalities in cerebrospinal fluid. In trauma cases (e.g. car accident, **sports** injury), the NE is a quick and essential component of emergency assessment. Once a diagnosis is determined, emergency measures may include further tests and/or surgery.

In psychological cases the treatment may include therapy and/or medication. In cases of an acute insult such as trauma, the patient is usually admitted to the hospital for appropriate treatment. Some neurological diseases are chronic and require conservative medical treatment and frequent follow-up visits for monitoring and stability or progression of the disease.

Risks

The MSE and NE are good diagnostic tools. There are no risks associated with initial neurologic assessment.

Parental concerns

Parental concerns center on the cause of the medical disease or psychological disorder, rather than around the procedure, which is straightforward and non-threatening.

Resources

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KEY TERMS

Corticospinal tract—A tract of nerve cells that carries motor commands from the brain to the spinal cord.

Gait—Walking motions.

Reflex—An involuntary response to a particular stimulus.

Thalamus—A pair of oval masses of gray matter within the brain that relay sensory impulses from the spinal cord to the cerebrum.

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Newborn life support *see* **Extracorporeal membrane oxygenation**

Nicotine *see* **Smoking**

Niemann-Pick disease *see* **Lipidoses**

Night terrors

Definition

Childhood night terrors are a parasomnia, or partial-sleep disorder, common in young children. They occur in the deepest stage of **sleep** and are characterized by an abrupt arousal, usually within the first hour of sleep. The child may sit bolt upright in acute terror, screaming inconsolably. Night terrors are a confusional arousal resulting from immature sleep patterns with an intense activation of the flight or fight emotion.

Description

Night terrors are not a dream or typical nightmare. They occur in non-REM, slow-wave sleep. The panicked screaming, kicking, thrashing, and flailing is alarming in its intensity. Sleepwalking, another parasomnia disorder, may also occur in as many as one third of children with night terrors. While experiencing the night terror the child is extremely disoriented and may stare straight ahead, eyes wide open, with the dark centers (pupils) enlarged. There is profuse sweating, the heartbeat is rapid, the breathing fast, and the blood pressure is elevated. As the child is not fully awake, she is unable to see or recognize her parent or caretaker and cannot be easily awakened. The night terror may last from one to 15 minutes or more and is usually followed by a return to deep sleep. Afterwards the child may have no memory of the experience.

Night terrors appear to run in families, though there is no scientific evidence of genetic factors. They are a developmental process and not typically a result of mental or physical illness.

Demographics

Childhood night terrors occur more frequently in boys. Children between the ages of three and five years of age are most likely to experience such nocturnal episodes. Such confusional arousals rarely persist beyond childhood, and they are significantly less frequent or cease entirely after age 12.

Causes and symptoms

Childhood night terrors appear to be a normal physiological process of the immature and developing nervous system. These confusional arousals can be triggered by stressful circumstances such as when a child is overly tired, when there is a loud noise or other unusual disruption, a change in the child's regular sleep-wake schedule, or even a full bladder. Night terrors occurring in **adolescence** and adult life may be more severe and are often linked with trauma and post-traumatic stress disorders.

When to call the doctor

Consult a pediatrician for night terrors if any of the following occur:

- Episodes occur more than once a week.
- Episodes persist after a schedule of preventive awakenings.
- Episodes last more than 45 minutes.
- The child exhibits drooling, jerking, and stiffening of the body.
- The child is physically endangered during an episode.
- Episodes occur later during the sleep cycle, more than two hours after going to sleep.
- The child has fears that persist throughout the day.

Diagnosis

Diagnosis is based on observation of the following characteristic symptoms:

- recurring episodes of abrupt and partial awakening from deep sleep with panicked screaming and disorientation
- increased heart rate, rapid breathing, and profuse sweating during an episode
- child is unresponsive to efforts to arouse or console during an episode
- child has little or no memory of the event after a full awakening

Treatment

Parents should not attempt to awaken a child experiencing a night terror. Efforts to console may be futile, though holding the child firmly and speaking with soothing words may facilitate the return to deep sleep. The primary effort should be to protect the child from possible harm to herself and others and ease them back to sleep.

In some severe cases, a pediatrician may prescribe a benzodiazepine tranquilizer, such as diazepam, known to suppress the stage four level of deep sleep. Though tranquilizers may be used for short-term control of night terrors, the result is uncertain and not generally advised.

Alternative treatment

Hypnosis, biofeedback, and various relaxation techniques have been used with some success to reduce or eliminate occurrence of childhood night terrors. Calming music or bedtime stories can help lull a child into deep sleep. Maintaining a quiet home without sudden disruptive noise will minimize some of the external stimuli that may trigger night terrors.

Nutritional concerns

Unusually heavy or spicy meals should be avoided before bedtime as indigestion might act as a trigger for night terror arousals.

KEY TERMS

Benzodiazepine—One of a class of drugs that have a hypnotic and sedative action, used mainly as tranquilizers to control symptoms of anxiety. Diazepam (Valium), alprazolam (Xanax), and chlordiazepoxide (Librium) are all benzodiazepines.

Confusional arousal—A partial arousal state occurring during the fourth stage of deepest sleep. Childhood night terrors are a form of confusional arousal.

Parasomnia—A type of sleep disorder characterized by abnormal changes in behavior or body functions during sleep, specific stages of sleep, or the transition from sleeping to waking.

Rapid eye movement (REM) sleep—A phase of sleep during which the person's eyes move rapidly beneath the lids. It accounts for 20–25% of sleep time. Dreaming occurs during REM sleep.

Prognosis

Childhood night terrors are usually outgrown by the age of seven and rarely persist beyond adolescence.

Prevention

Some pediatricians suggest that parents maintain a sleep diary and observe the child throughout several night terror episodes, noting the amount of time following sleep when the night terror begins. After the sleep-wake pattern is determined, a series of 15–20 minutes prior to the usual occurrence of the night terror and keep the child awake and out of bed for a full five minutes. This may help to break the disruptive sleep pattern that has resulted in the night terrors.

Children often experience night terrors during the toilet-training years. The night terror might be triggered by a full bladder. Assisting the child to the toilet prior to bedtime and even during the course of a night-terror might be beneficial in reducing reoccurrence.

Parental concerns

Childhood night terrors are alarming to witness. Parents may find it particularly difficult when efforts to console the child fail and the child does not recognize them even though his or her eyes may be wide open. The screaming, flailing, and kicking that accompany a night terror may frighten parents who **fear** the child is having a seizure. It is not a seizure unless the behavior includes eyes rolling back in the head, stiffening of the body, and drooling. Most childhood night terrors will last about 10 minutes.

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Clare Hanrahan

Nightmares

Definition

Nightmares are a type of **sleep** disruption, or parasomnia, characterized by frightening psychological

content. Nightmares provoke a feeling of imminent physical danger with a sensation of being trapped or suffocated. These frightening dreams occur during rapid eye movement sleep (REM), or dream-time sleep, and trigger a partial or full awakening. Nightmares are a universal human experience occurring throughout the lifespan. They are especially common in early childhood and involve activation of the limbic brain, particularly the area that mediates negative emotion.

Description

Nightmares are greatly influenced by the particular stressors and anxieties present in the child's waking life. Typical childhood nightmares include dreams of **abandonment**; of being lost; of falling; or being chased, bitten, or eaten by a monster or hostile animal. Dream researchers have observed a developmental progression in the content and frequency of children's nightmares. A two-year-old dreamer may recall a fearful dream, but be unable to give form to the source of the threat. By the age of five, the frightened young dreamer may identify the attacker as a monster or wild animal. Older children who have developed more of an understanding of real-life dangers report dreams of pursuit by mean or bad people.

Children gradually develop the ability to understand the difference between dreams and reality. Very young children have great difficulty believing that the dream is not real. By three to four years of age, however, most children can distinguish between the nightmare content and their waking reality.

When a child is awakened by a nightmare she will soon become fully alert and able to remember the scary dream in elaborate detail, expressing emotions appropriate to the dream content. The frightened child will resist returning to bed and often seek the comfort and reassurance of a parent or caretaker. Nightmares are different than the non-dream sleep disturbance known as a night terror, which causes only a partial arousal from deep sleep and occurs during the first period of sleep known as slow-wave sleep (SWS). A child experiencing a night terror will be difficult to awaken or comfort, will not recognize her parent or caretaker, and will usually have no memory of the terrifying emotions that caused the sleep disturbance.

Demographics

Although infants spend most of their sleep time in the REM stage where dreams are known to occur, there is no reliable way to determine if dreaming actually takes place prior to the development of language and the reflective ability to think in images. Sleep researchers and developmental psychologists generally agree that nightmares first occur in children from 18 months to two years of age. By age three, more than half of all children will report having experienced a nightmare. The incidence of these frightening dreams increases considerably in elementary school children.

In a study of the dreams of four- to 12-year-old children published in the *Journal of Clinical Child Psychology* in 2000, researchers found that 67.7 percent of four to six year olds, 95.7 percent of seven to nine year olds, and 76.3 percent of ten to 12-year olds reported having had a nightmare experience. Nightmares are common throughout childhood, changing somewhat in content and frequency as children move through different developmental phases and acquire more skills to cope with the changing realities and stresses in their lives.

Causes and symptoms

Childhood nightmares are a normal maintenance function of the developing brain. They are a means of integrating recent and past learning and of establishing psychological equilibrium. Children who have not yet developed sufficient coping mechanisms to deal with many normal childhood fears and problems may feel overwhelmed and insecure. These distressing emotions provide the basis for nightmares. Common stressful events include moving to a different neighborhood or school, encountering a schoolyard bully, watching a horror movie or a violent television or video program, the birth of a sibling, ongoing conflict with siblings, parental marital problems, or any of numerous other stressful situations that may add to a child's sense of vulnerability or powerlessness.

Nightmares may increase and intensify following particularly traumatic events such as the death of a parent, a sibling, or other loved one; parental **divorce** or separation; an injury, illness, or other medical crises; or witnessing or being subjected to physical or verbal violence or sexual abuse. Children who have been traumatized may suffer ongoing post-traumatic stress and express it through recurring nightmares.

Certain medications used to treat **asthma**, **allergies**, and seizures can be a causal factor in the onset of sleep disturbances and nightmares. Abrupt withdrawal from drugs or medications, including barbiturates and benzodiazepines, can also induce these sleep disturbances. Illness with high **fever** may bring about delirium with frightening episodes of nighttime awakening. The following behaviors are usually present in a normal nightmare episode:

- Child awakens during the last third of her sleep period.
- Child is frightened and becomes fully alert.
- Child can describe the frightening dream in detail.
- Child seeks and responds to comfort and reassurance from a parent or caretaker.
- Child fears a recurrence of the frightful dream and may resist a return to bed.

When to call the doctor

If a child's nightmares increase in frequency and intensity, it is important to consult a physician or pediatrician to determine if the sleep disruption is due to any injury, illness, or infection, or if it might be caused by the use of or change in medications. Disturbed sleeping patterns in children are also present in some cases of juvenile rheumatoid arthritis, **autism**, and fibromyalgia.

Nightmares usually diminish in frequency and intensity over time. Recurring nightmares may indicate an ongoing problem that the child is having difficulty resolving. A child who is losing sleep and whose fears persist during day-time hours may benefit from the help of a pediatric psychotherapist in developing coping strategies for the stress and **anxiety** expressed through the dreams.

Diagnosis

Parental observation of the child's sleeping patterns and careful record-keeping of symptoms of any sleep disruption through use of a sleep journal will usually reveal any parasomnia patterns that may require professional **assessment**.

Treatment

Parents or caretakers should take into account the age and developmental maturity of their child when responding to the fears and anxieties that a nightmare brings to the surface. The child's ability to understand that the nightmare is not real and that it is an event happening only within their own mind increases with age. The nightmare reflects real fears and stressful circumstances present in the child's waking life. A parent's willingness to listen to and sympathize with a child's fears provides a necessary validation of the child's experience and helps to calm the child's anxiety. Very young children who lack the verbal skills to describe the frightening dream may require more reassurances that they are safe and more time in the comforting presence of a parent or caretaker before they are ready to return to bed.

Anne Sayre Wiseman, writing in her book *Nightmare Help: A Guide for Parents and Teachers*, suggests that parents approach the nightmare as a dream story with a problem to be solved. Parents who act as dream guides can help their children to find their own solution to the dream problem. The parents' goal, Wiseman counsels, is to "encourage autonomy so the child learns to empower themselves at whatever level they can handle."

Alan Siegel and Kelly Bulkeley, writing in their book, *Dreamcatching: Every Parent's Guide to Exploring and Understanding Children's Dreams and Nightmares*, suggest four beneficial remedies to help a child cope with disturbing nightmares. "The Four R's" of nightmare relief are:

- Reassurance: Provide physical and emotional reassurance and a listening ear so the frightened child will feel safe enough to share the dream images.
- Rescripting: Discuss the dream images with the child and work together with the frightened dreamer to imagine changes in the outcome. Encourage the child to express the images through artwork, fantasy, drama, and writing.
- Rehearsal: Encourage the child to imagine how the various alternative dream endings might change and assist the child in working through the different outcomes to find those that restore a sense of control and safety.
- Resolution: Work with the child to help her discover and acknowledge the life problems and stressful circumstances that may be reflected in the nightmare.

Nightmares are a common childhood parasomnia and medication is rarely indicated. Other parasomnias, such as **night terrors** and night walking, may call for medication if other interventions and treatments fail to relieve seriously disruptive symptoms.

Alternative treatment

Teaching the child simple relaxation skills through guided imagery will provide a valuable self-help resource that may minimize bed-time anxiety. Older children can be encouraged to connect with an "inner guide" as a source of strength when they are awakened by frightening dreams. Inner guides may take the form of a loving voice within that the child can listen for during times of **fear** or a beloved animal that they may call upon as a companionable inner helper when dealing with problems presented in scary dreams.

Nightmares

Preventing Nightmares

Have the child go to bed about the same time every day.
Avoid eating or exercising before bed.
Avoid scary books or movies before bed.
Put the child to sleep with a favorite stuffed toy or special blanket.
Keep a nightlight on in the child's room.
Keep the door to the child's room open.

(Table by GGS Information Services.)

Nutritional concerns

Nightmares are a sleep disturbance that is part of a normal adaptive mechanism of the developing child. They are usually not caused by a child's diet. However fatty foods or spicy meals that may bring about digestive distress at bedtime may trigger sleep disturbances and awaken a child out of an otherwise peaceful slumber.

Prognosis

Childhood nightmares are a normal process of coping with new challenges and integrating new life experiences into the child's understanding of the world. With guidance from a sensitive parent, a child's nightmares can provide an opportunity for parents and children to gain a deeper understanding of, and find solutions to childhood anxieties and insecurities.

Prevention

Nightmares are part of the normal developmental process that literally provide a "wake-up" call to both parent and child to pay attention to strong feelings and problems that may require some resolution. Nightmares diminish as children feel more confidence and control in their lives. If nightmares persist and intensify they may indicate a situation in the child's life that needs to be changed rather than worked through with dream problem-solving.

Establishing a regular bedtime schedule and routine, including a calming-down period prior to sleep, will help reduce the number of sleep disruptions. Eliminating the stimulation of television or **video games** at bedtime, particularly violent television shows, movies, and games may reduce the nightmares brought about by these unnecessary and sometimes disturbing stimulations.

Parental concerns

Sleep disruptions throughout childhood also cause disruptions for the entire **family**, particularly parents who must attend to the frightened child and soothe the child back to sleep. Childhood sleep disruptions that persist over time may interfere with the normal sleep-wake cycle of the caregivers. Loss of sleep has a cumulative impact on the well being of both children and adults. Parents may become anxious in sympathy with the child and lie awake in worry long after the frightened dreamer has returned to sleep. There are many good books available to help a parent understand the normal sleep-wake cycles of children and the common parasomnias that all children experience. This knowledge can bring much needed peace of mind and help the parent determine when professional assessment and assistance is appropriate.

KEY TERMS

Limbic system—A group of structures in the brain that includes the hypothalamus, amygdala, olfactory bulbs, and hippocampus. The limbic system plays an important part in regulation of human moods and emotions. Many psychiatric disorders are related to malfunctioning of the limbic system.

Parasomnia—A type of sleep disorder characterized by abnormal changes in behavior or body functions during sleep, specific stages of sleep, or the transition from sleeping to waking.

Rapid eye movement (REM) sleep—A phase of sleep during which the person's eyes move rapidly beneath the lids. It accounts for 20–25% of sleep time. Dreaming occurs during REM sleep.

Slow-wave sleep (SWS)—Stage of deepest sleep characterized by absence of eye movements, decreased body temperature, and involuntary body movements. Night terrors and sleepwalking occur during this stage of sleep.

Resources

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Clare Hanrahan

Nonsteroidal anti-inflammatory drugs

Definition

Nonsteroidal anti-inflammatory drugs are medicines that relieve **pain**, swelling, stiffness, and inflammation.

Description

Nonsteroidal anti-inflammatory drugs (NSAIDs) are prescribed for a variety of painful conditions, including arthritis, bursitis, tendonitis, gout, menstrual cramps, **sprains**, **strains**, and other injuries.

Although the NSAIDs are often discussed as a group, not all are approved for use in children. As of 2004, the following drugs are approved for pediatric use:

- Ibuprofen (Advil, Motrin, Nuprin).
- Indomethicin (Indocin), not recommended for children under the age of 14 except in circumstances that warrant the risk. Indomethicin has special application in some infants born with heart problems.
- Ketoprofen (Orudis, Oruvail), not given to children under the age of 16 unless directed by a physician.
- Ketorolac tromethamine (Toradol), not approved for use in children but has been reported safe by some pediatric authorities.
- Meclofenamate sodium, safety and efficacy in children under 14 years of age has not been established.
- Mefenamic acid (Ponstel), safety and efficacy in children under 14 years of age has not been established.
- Naproxen (Aleve, Anaprox, Naprosyn), safety and efficacy in children under two years of age has not been established.
- Tolmetin sodium (Tolectin), safety and efficacy in children under two years of age has not been established.

Other NSAIDs have been used in pediatric therapy, but should not be considered as first choice for treatment of children or adolescents.

A new class of NSAIDs, called COX-2 inhibitors, have a lower risk of causing ulcers than do the traditional NSAIDs. These drugs may be appropriate for use in older teenagers but have not been approved for use in younger children, and there is some evidence that they are inappropriate for infants.

General use

Nonsteroidal anti-inflammatory drugs relieve pain, stiffness, swelling, and inflammation, but they do not cure the diseases or injuries responsible for these problems. Two drugs in this category, ibuprofen and naproxen, also reduce **fever**. Some nonsteroidal anti-inflammatory drugs can be bought without a prescription; others are available only with a prescription from a physician or dentist.

Precautions

Children with certain medical conditions and those who are taking some other medicines can have problems if they take nonsteroidal anti-inflammatory drugs. Before giving children these drugs, parents need to let the physician know about any of the following conditions.

Allergies

The physician needs to know about any **allergies** to foods, dyes, preservatives, or other substances. For children who have had reactions to nonsteroidal anti-inflammatory drugs in the past, parents should check with a physician before having these drugs prescribed again.

Pregnancy

Teens and young women who are pregnant or who plan to become pregnant should check with their physicians before taking these medicines. Whether nonsteroidal anti-inflammatory drugs cause birth defects in people is unknown, but some do cause birth defects in laboratory animals. If taken late in pregnancy, these drugs may prolong pregnancy, lengthen labor time, cause problems during delivery, or affect the heart or blood flow of the fetus.

Breastfeeding

Some nonsteroidal anti-inflammatory drugs pass into breast milk. Women who are breastfeeding their babies should check with their physicians before taking these drugs.

Other medical conditions

A number of medical conditions may influence the effects of nonsteroidal anti-inflammatory drugs. Parents of children and teens who have any of the conditions listed below should tell their physician about the condition before having nonsteroidal anti-inflammatory drugs prescribed.

- stomach or intestinal problems, such as colitis or Crohn's disease
- liver disease
- current or past kidney disease or current or past kidney stones
- heart disease
- high blood pressure
- blood disorders, such as anemia, low **platelet count**, low white blood cell count
- bleeding problems
- diabetes mellitus
- hemorrhoids, rectal bleeding, or rectal irritation
- asthma
- epilepsy
- systemic lupus erythematosus
- diseases of the blood vessels, such as polymyalgia rheumatica and temporal arteritis
- fluid retention
- alcohol abuse
- mental illness

Side effects

The most common side effects are stomach pain or cramps, **nausea**, **vomiting**, indigestion, **diarrhea**, heartburn, **headache**, **dizziness** or lightheadedness, and drowsiness. As the patient's body adjusts to the medicine, these symptoms usually disappear. If they do not, the physician who prescribed the medicine should be contacted.

Serious side effects are rare, but do sometimes occur. If any of the following side effects occur, patients should stop taking the medicine and get emergency medical care immediately:

- swelling or puffiness of the face
- swelling of the hands, feet, or lower legs
- rapid weight gain
- fainting
- breathing problems
- fast or irregular heartbeat
- tightness in the chest

Other side effects do not require emergency medical care, but should have medical attention. If any of the following side effects occur, patients should stop taking the medicine and the physician who prescribed the medicine should be called as soon as possible:

- severe pain, cramps, or burning in the stomach or abdomen
- convulsions
- fever
- severe nausea, heartburn, or indigestion
- white spots or sores in the mouth or on the lips
- rashes or red spots on the skin
- any unusual bleeding, including nosebleeds and spitting up or vomiting blood or dark material
- black, tarry stool

- chest pain
- unusual bruising
- severe headaches

A number of less common, temporary side effects are also possible. They usually do not need medical attention and will disappear once the body adjusts to the medicine. If they continue or interfere with normal activity, the physician should be contacted. Among these side effects are:

- gas, bloating, or constipation
- bitter taste or other taste changes
- sweating
- restlessness, irritability, anxiety
- trembling or twitching

Interactions

Nonsteroidal anti-inflammatory drugs may interact with a variety of other medicines. When interaction occurs, the effects of the drugs may change, and the risk of side effects may be greater. Physicians prescribing this drug should know all other medicines the patient is already taking. Among the drugs that may interact with nonsteroidal anti-inflammatory drugs are:

- blood thinning drugs, such as warfarin (Coumadin)
- other nonsteroidal anti-inflammatory drugs
- heparin
- tetracyclines
- cyclosprorine
- digitalis drugs
- lithium
- phenytoin (Dilantin)
- zidovudine (AZT, Retrovir)

NSAIDs may also interact with certain herbal preparations sold as dietary supplements. Among the herbs known to interact with NSAIDs are bearberry (*Arctostaphylos uva-ursi*), feverfew (*Tanacetum parthenium*), evening primrose (*Oenothera biennis*), and gossypol, a pigment obtained from cottonseed oil and used as a male contraceptive. In most cases, the herb increases the tendency of NSAIDs to irritate the digestive tract. It is just as important for doctors to know which herbal remedies the patient is taking on a regular basis as it is for doctors to know the other prescription medications which are being taken.

KEY TERMS

Anemia—A lack of hemoglobin, the compound in blood that carries oxygen from the lungs throughout the body and brings waste carbon dioxide from the cells to the lungs, where it is released.

Bursitis—Inflammation of the tissue around a joint.

Colitis—Inflammation of the colon (large bowel).

COX-2 inhibitors—A class of newer NSAIDs that are less likely to cause side effects in the digestive tract. COX-2 inhibitors work by inhibiting the production of cyclooxygenase-2, an enzyme involved in inflammation.

Inflammation—Pain, redness, swelling, and heat that usually develop in response to injury or illness.

Salicylates—A group of drugs that includes aspirin and related compounds. Salicylates are used to relieve pain, reduce inflammation, and lower fever.

Tendonitis—Inflammation of a tendon, which is a tough band of tissue that connects muscle to bone.

Prevention

Many serious digestive system effects of NSAIDs can be prevented by taking mysoprostol (Cytotec), but this drug is only appropriate for patients with a high risk of ulcers. It is not called for when the NSAID is being used for a short period of time or in patients with other risk factors. Stomach upset can often be prevented by taking NSAIDs with food or milk.

Parental concerns

NSAIDs are very safe when used properly over a short period of time. They should not be used for longer periods or in larger doses than indicated on the label. If NSAIDs are to be used for prolonged periods, as in juvenile rheumatoid arthritis, there is a risk of potentially serious stomach and intestinal problems.

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Nontropical sprue *see* **Celiac disease**

Nose injuries *see* **Nasal trauma**

Nosebleed

Definition

A nosebleed, also called epistaxis, is a loss of blood from any blood vessel in the nose. It usually appears in only one nostril.

Description

Although unexpected or persistent bleeding from anywhere is a warning sign of **cancer** and should be investigated, nosebleeds are rarely a sign of serious illness. They are usually the result of minor injury or irritation.

Nosebleeds most often come from the front of the septum, the cartilage that separates the nose into two nostrils. These anterior nosebleeds comprise 80 percent of all nosebleeds. A mass of blood vessels, called Kiesselbach's plexus, lie on either side of the septum. These blood vessels are easy to injure and bleeding can occur.

Posterior nosebleeds, which come from the back of the nose, are less common and much harder to manage. Bleeding usually begins in the upper part of the nose and flows toward the throat and mouth where it is swallowed. It is difficult to determine how much blood is lost in these nosebleeds.

Demographics

Though it is a common misconception that children are more susceptible to nosebleeds than adults, research has found that nosebleeds are more prevalent in older adults and more often are a sign of other health problems. Nosebleeds do, however, occur frequently in childhood. About 30 percent of children up to five years of age have had spontaneous nosebleeds that appeared without apparent injury to the nose. Of children six to ten years of age, 56 percent have had them and 64 percent of preadolescents and adolescents from 11 to 15 have as well. Moreover, over half of adults with recurring nosebleeds had them as children. Only about 10 percent of children with frequent nosebleeds have been found to have a previously undiagnosed bleeding disorder.

Rarely, menstruating women, even adolescents, who have endometriosis, a condition in which tissues resembling the lining of the uterus occur abnormally in other parts of the pelvic cavity and sometimes in other parts of the body, can have cyclical nosebleeds with their menstrual periods.

Causes and symptoms

Causes

The most common cause of nosebleeds is injury from picking or blowing the nose. People with respiratory **allergies**, hay fever, and sinus infections have swollen nasal membranes that are fragile and more likely to bleed. Physical injury to the nose from falls, **sports**, or fighting can also cause nosebleeds. Chemical irritants such as cleaning products, aerosols, and paint can irritate the nose, sometimes resulting in nosebleeds. In addition, some drugs, such as cocaine, inflame the nose, causing it to bleed. Children with deviated septums or crooked noses are also prone to nosebleeds.

Nosebleeds occur more frequently in the winter when the air is cold outside and homes are filled with dry air from furnaces and other heating sources. Changes from cold to warm air or dry to humid air stress the delicate membranes of the nose and make it vulnerable to

injury and bleeding. Also, flu, colds, and other respiratory illnesses seem to occur more often in the winter. These also stress the nasal passages and make injury more likely. Bleeding from the nose, therefore, usually follows these seasonal stresses.

A nosebleed can also be an indication of illness. Certain blood disorders, such as **hemophilia**, can cause the nose to bleed. In this case, medical help should be sought immediately. Some head injuries produce nosebleeds. Uncontrolled high blood pressure, liver disease, leukemia, and tumors of the nasal passages and the brain can also cause the nose to bleed.

Even some medications can cause nosebleeds. Anticoagulants, medications used to thin the blood, including aspirin, can cause spontaneous bleeding from the nose. Overuse of nasal sprays can also produce nosebleeds. In addition, nosebleeds can be a side effect of alcohol abuse.

Nosebleeds in children can sometimes be caused by children putting objects into their noses. The object can tear the delicate membranes of the nose. Objects inserted into the nose can also cause obstruction of airflow and may need to be removed by a doctor.

Symptoms

Bleeding from one or both nostrils may be a trickle or a flood. Sometimes, it accompanies direct injury to the nose as in a sports injury or in picking the nose or too vigorous nose blowing. Children may experience frequent swallowing or a sensation of fluid in the back of the nose and throat.

When to call the doctor

If the bleeding does not stop after 20 minutes, it is necessary to seek medical help. Also, if there is a known or suspected **head injury** accompanying the nosebleed, there may be a skull fracture or brain disorder. In this case, the child should be taken to the emergency room immediately. In addition, if the nose is misshapen, especially after an accident, fall, or injury, it may be broken and will need to be evaluated by a doctor. Frequent nosebleeds, especially if they are occurring more often and are not due to colds, allergies, or trauma, will need to be seen by a doctor as well.

It may be a medical emergency if the bleeding is rapid or if there is a lot of blood. If the child feels faint or weak during a nosebleed, it may be do to blood loss and the child should see the doctor immediately.

Diagnosis

Bleeding from the nose is the obvious determinant of a nosebleed. The severity of it, however, may require blood work to look for bleeding disorders, diseases of the blood, or infections. X rays may be taken to determine if there has been a head injury or abnormalities within the structure of the nose. To further examine the nose, the doctor may perform a nasal endoscopy, a procedure that involves inserting a tiny camera into the nose to look at blood vessels and nasal structures.

Treatment

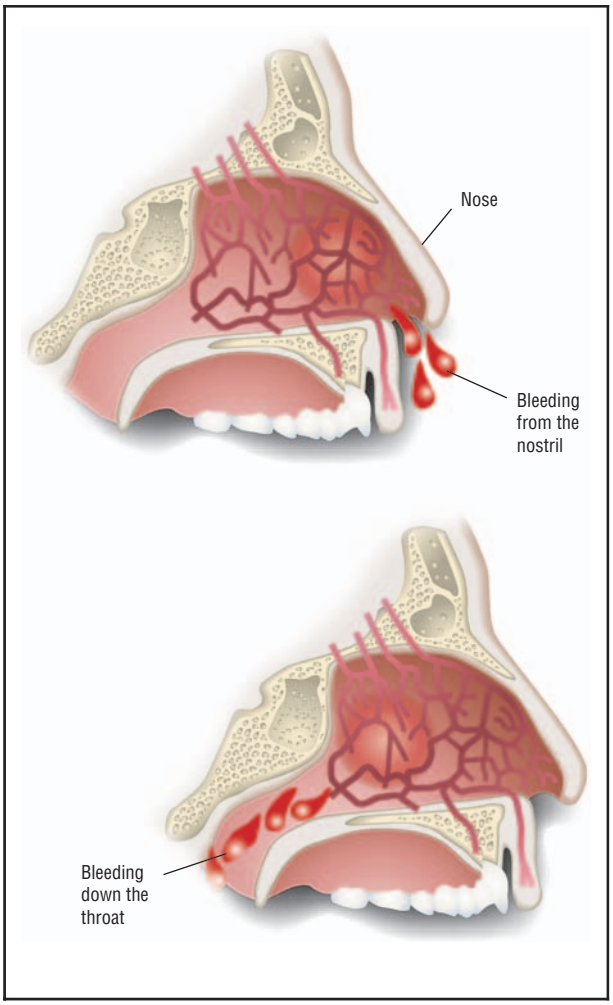
The first line of treatment is to gently pinch the nostrils together with the thumb and forefinger, while sitting upright and breathing through the mouth, for five to ten minutes. Leaning forward will prevent the child from swallowing blood. After at least five minutes, the parent or child can check to see if the bleeding has stopped. If it has not, then the pinching of the nose should be resumed and the child or parent should wait another five minutes. Most nosebleeds will stop within this time period, especially if the child is encouraged to remain calm. **Anxiety** or panic will cause blood to flow more rapidly and can hinder this self-healing process. It is very important for the child not to lie down while having a nosebleed.

Sometimes, a cold compress or crushed ice in a washcloth or plastic bag can be placed across the bridge of the nose and cheeks to encourage clotting. It is important not to pack the inside of the nose with gauze since this might further injure the nose.

In 2004, a new over-the-counter product was introduced for a quick home treatment for simple nosebleed. Called Nosebleed QR (Quick Relief), the product is composed of a hydrophilic polymer, a synthetic powder that absorbs blood, and potassium salt that aids in scab formation. The parent or child sprinkles the product onto a swab and coats the nostril then pinches the nose for 15 to 20 seconds. The product stops bleeding within one minute. However, it does sting and may not be a comfortable product to use with children.

Alternative treatment

Estrogen cream, the same preparation used to revitalize vaginal tissue, can toughen fragile blood vessels in the anterior septum and forestall the need for cauterization. Botanical medicines known as stiptics, which slow down and can stop bleeding, may be taken internally or applied topically. Some of the plants used are achillea (yarrow), trillium, geranium, and shepherd's purse (*Capsella bursa*).



Anatomical sideview of a nosebleed. *(Illustration by GGS Information Services)*

KEY TERMS

Deviated septum—A shift in the position of the nasal septum, the partition that divides the two nasal cavities.

Endometriosis—A condition in which the tissue that normally lines the uterus (endometrium) grows in other areas of the body, causing pain, irregular bleeding, and frequently, infertility.

Hemophilia—Any of several hereditary blood coagulation disorders occurring almost exclusively in males. Because blood does not clot properly, even minor injuries can cause significant blood loss that may require a blood transfusion, with its associated minor risk of infection.

Kiesselbach's plexus—The mass of blood vessels on either side of the septum.

Nasal endoscopy—A procedure that involves inserting a tiny camera into the nose in order to look at blood vessels and nasal structures.

Otorhinolaryngologist—An ear, nose, and throat specialist.

Septum—A wall or partition. Often refers to the muscular wall dividing the left and right heart chambers or the partition in the nose that separates the two nostrils. Also refers to an abnormal fold of tissue down that center of the uterus that can cause infertility.

Homeopathic remedies can be one of the quickest and most effective treatments for nosebleeds. One well known remedy is phosphorus.

Another natural treatment includes swabbing the nose with vitamin E oil for three days. If nosebleeds recur within that time, it is recommended to take 500 mg of bioflavonoids twice a day. Bioflavonoids are antioxidants found in citrus fruits that help strengthen blood vessels.

Prognosis

Most common nosebleeds are easily managed and can be prevented. Children usually recover quickly. Serious nosebleeds need further investigation but are usually controlled by treating the underlying cause.

Prevention

Keeping the home cooler in winter and using a humidifier help keep the nasal passages moist and reduce the risk of nosebleeds due to dry air. Some doctors also recommend using a nasal saline spray to enhance moisture in the nose.

Also, before sending children out to **play** on cold, dry days, parents can put a bit of petroleum jelly on a cotton swab and wipe it just inside their children's nostrils. This keeps the nose from drying out and producing surface cracks which can damage blood vessels in the lining of the nose and cause bleeding. Some parents routinely apply a thin layer of A & D ointment, petroleum jelly, or a mentholated salve around their children's noses at night as well as when they go outside.

Gently blowing the nose and not picking it will also prevent nosebleeds. This is especially important for several hours after nasal bleeding has stopped. In some cases,

strong sniffing is also discouraged to reduce further stress on the delicate tissues of the nose. Keeping the mouth open when sneezing can also reduce stress on the nose.

Treatment of hay fever and other respiratory allergies decreases sneezing and nasal inflammation. If a child is prone to sinus infections, some doctors may also use prophylactic or preventative treatment similar to allergy management.

Parental concerns

Nosebleeds can be frightening for children. Seeing any amount of blood may cause some children to panic or even faint. It is important to treat the nosebleed matter-of-factly as any parent would handle any other childhood scrape or wound. The parent's calm helps the child remain calm while waiting for the natural blood clotting mechanisms of the body to work. Parents should also be aware of any abnormal amount of blood during a nosebleed and make note of any recent falls or head injuries. These observations will inform parents about when to seek medical or emergency help.

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NSAIDs *see* **Nonsteroidal anti-inflammatory drugs**

Numbness and tingling

Definition

Numbness and tingling are decreased or abnormal sensations caused by altered sensory nerve function.

Description

The feeling of having a foot "fall asleep" is a familiar one. This same combination of numbness and tingling can occur in any region of the body and may be caused by a wide variety of disorders. Sensations such as these, which occur without any associated stimulus, are called paresthesias. Other types of paresthesias include feelings of cold, warmth, burning, **itching**, and skin crawling.

Demographics

People of all ages experience episodes of numbness and tingling. These generally become more common as people age. Episodes of numbness and tingling are more common among people with diabetes, **hypothyroidism**, **alcoholism**, **malnutrition**, or who experience mechanical trauma, especially to their limbs, neck or spine.

Causes and symptoms

Causes

Sensation is carried to the brain by neurons (nerve cells) running from the outer parts of the body to the spinal cord in bundles called nerves. In the spinal cord, these neurons make connections with other neurons that run up to the brain. Paresthesias are caused by disturbances in the function of neurons in the sensory pathway. This disturbance can occur in the central nervous system (the brain and spinal cord), the nerve roots that are attached to the spinal cord, or the peripheral nervous system (nerves outside the brain and spinal cord).

Peripheral disturbances are the most common cause of paresthesias. "Falling asleep" occurs when the blood supply to a nerve is cut off—a condition called ischemia. Ischemia usually occurs when an artery is compressed as it passes through a tightly flexed joint. Sleeping with the arms above the head or sitting with the legs tightly crossed frequently cause numbness and tingling.

Direct compression of the nerve also causes paresthesias. Compression can be short-lived, as when a heavy backpack compresses the nerves passing across the shoulders. Compression may also be chronic. Chronic nerve compression occurs in entrapment syndromes. The most common example is carpal tunnel syndrome, which occurs when the median nerve is compressed as it passes through a narrow channel in the wrist. Repetitive motion or prolonged vibration can cause the lining of the channel to swell and press on the nerve. Chronic nerve root compression, or radiculopathy, can occur in disk disease or spinal arthritis.

Other causes of paresthesias related to disorders of the peripheral nerves include the following:

- metabolic or nutritional disturbances, including diabetes, hypothyroidism (a condition caused by too little activity of the thyroid gland), alcoholism, malnutrition, and vitamin B_{12} deficiency
- trauma, including injuries that crush, sever, or pull on nerves
- inflammation
- connective tissue disease, including arthritis, systemic lupus erythematosus (a chronic inflammatory disease that affects many systems of the body, including the nervous system), polyarteritis nodosa (a vascular disease that causes widespread inflammation and ischemia of small and medium-size arteries), and Sjögren's syndrome (a disorder marked by insufficient moisture in the tear ducts, salivary glands, and other glands)
- toxins, including heavy metals (metallic elements such as arsenic, lead, and mercury which can, in large amounts, cause **poisoning**), certain medications **antibiotics** and **chemotherapy** agents, solvents, and overdose of pyridoxine (vitamin B_6)
- malignancy
- infections, including **Lyme disease**, human **immunodeficiency** virus (HIV), and leprosy
- hereditary disease, including **Charcot-Marie-Tooth disease** (a disorder that causes wasting of the leg muscles, resulting in malformation of the foot), porphyria (a group of disorders in which there is abnormally increased production of substances called porphyrins), and Denny-Brown's syndrome (a disorder of the nerve root)

Paresthesias can also be caused by central nervous system disturbances, including **stroke**, transient ischemic attack (TIA), tumor, trauma, multiple sclerosis, or infection.

Symptoms

Sensory nerves supply or innervate particular regions of the body. Determining the distribution of symptoms is an important way to identify the nerves involved. For instance, the median nerve innervates the thumb, the first two fingers, half of the ring finger, and the part of the hand to which they connect. The ulnar nerve innervates the other half of the ring finger, the little finger, and the remainder of the hand. Distribution of symptoms may also aid diagnosis of the underlying disease. Diabetes usually causes a symmetrical glove-and-stocking distribution in the hands and feet. Multiple sclerosis may cause symptoms in several, widely separated areas.

Other symptoms may accompany paresthesias, depending on the type and severity of the nerve disturbance. For instance, weakness may accompany damage to nerves that carry both sensory and motor neurons. (Motor neurons are those that carry messages outward from the brain.)

When to call the doctor

A healthcare professional should be consulted when instances of numbness or tingling last for more than a few hours.

Diagnosis

A careful history of the affected individual is needed for a diagnosis of paresthesias. The medical history should focus on the onset, duration, and location of symptoms. The history may also reveal current related medical problems and recent or past exposure to drugs, toxins, infection, or trauma. The **family** medical history may suggest a familial disorder. A work history may reveal repetitive motion, chronic vibration, or industrial chemical exposure.

The physical and neurological examination tests for distribution of symptoms and alterations in reflexes, sensation, or strength. The distribution of symptoms may be mapped by successive stimulation over the affected area of the body.

Lab tests for paresthesia may include blood tests and urinalysis to detect metabolic or nutritional abnormalities. Other tests are used to look for specific suspected causes. Nerve conduction velocity tests, electromyography, and imaging studies of the affected area may be employed. Nerve biopsy may be indicated in selected cases.

Treatment

Treatment of paresthesias depends on the underlying cause. For limbs that have "fallen asleep," restoring circulation by stretching, exercising, or massaging the affected limb can quickly dissipate the numbness and tingling. Physical therapy can also be helpful. If the paresthesia is caused by a chronic disease such as diabetes or occurs as a complication of treatments such as chemotherapy, most treatments are aimed at relieving symptoms. Anti-inflammatory drugs such as aspirin or ibuprofen are recommended if symptoms are mild. In more difficult cases, antidepressant drugs such as amitriptyline (Elavil) are sometimes prescribed. These drugs

are given at a much lower dosage for this purpose than for relief of depression. They are thought to help because they alter the body's perception of **pain**. In severe cases, opium derivatives such as codeine can be prescribed. In the early 2000s trials are being done to determine whether treatment with human nerve growth factor will be effective in regenerating the damaged nerves.

Several alternative treatments are available to help relieve symptoms of paresthesia. Nutritional therapy includes supplementation with B complex **vitamins**, especially vitamin B_{12} (intramuscular injection of vitamin B_{12} is most effective). Vitamin supplements should be used cautiously, however. Overdose of Vitamin B_6 is one of the causes of paresthesias. People experiencing paresthesia should also avoid alcohol. Acupuncture and massage are said to relieve symptoms. Self-massage with aromatic oils is sometimes helpful. The application of topical ointments containing capsaicin, the substance that makes hot peppers hot, provides relief for some. It may also be helpful to wear loosely fitting shoes and clothing. None of these alternatives should be used in place of traditional therapy for the underlying condition.

Prognosis

Treating the underlying disorder may reduce the occurrence of paresthesias. However, paresthesias resulting from damaged nerves may persist throughout or even beyond the recovery period. The overall prognosis depends on the cause.

Prevention

Preventing the underlying disorder may reduce the incidence of paresthesias. For those with frequent paresthesias caused by ischemia, changes in posture may help.

Nutritional concerns

Vitamin supplements should be used with caution as an overdose of vitamin B_6 is one of the causes of paresthesias. People experiencing paresthesia should avoid alcohol.

Parental concerns

Parents should monitor children who complain of numbness or tingling that lasts for more than a few minutes and fails to improve. They should recommend stretching and **exercise** and if relief does not come quickly, investigate other causes, such as clothing that is too tight.

KEY TERMS

Electrooculography (EOG)—A diagnostic test that records the electrical activity of the muscles that control eye movement.

Motor nerve—Motor or efferent nerve cells carry impulses from the brain to muscle or organ tissue.

Nerve condition velocity (NCV)—Technique for studying nerve or muscle disorders, measuring the speed at which nerves transmit signals.

Nerve growth factor—A protein resembling insulin that affects the growth and maintenance of nerve cells

Peripheral nervous system—The part of the nervous system that is outside the brain and spinal cord. Sensory, motor, and autonomic nerves are included.

Sensory nerves—Sensory or afferent nerves carry impulses of sensation from the periphery or outward parts of the body to the brain and spinal cord.

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American Academy of Neurology. 1080 Montreal Avenue, St. Paul, Minnesota 55116. Web site: <www.aan.com/>.

American Academy of Pediatrics. 141 Northwest Point Boulevard, Elk Grove Village, IL 60007–1098. Web site: <www.aap.org/default.htm>.

American Academy of Physical Medicine and Rehabilitation. One IBM Plaza, Suite 2500, Chicago, IL 60611–3604. Web site: <www.aapmr.org/>.

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Nursemaid's elbow

Definition

Nursemaid's elbow is an injury to the ligament (strong band of tissue) that keeps the two bones of the forearm in the correct place.

Description

The two bones in the forearm are the radius and the ulna. The radius is on the thumb side of the forearm. The upper end of the radius is called the radius head. The radial head is held in place by a ligament called the annular ligament. When the annular ligament is torn, a part of it slides upward and becomes trapped in the elbow joint, which is very painful condition. The child holds the arm with the palm facing inward and the elbow bent. This injury is uncommon after the ages of three to four, due to stronger joints and ligaments. Also, youngsters are less likely to be in situations where this injury might occur.

Nursemaid's elbow is also called subluxation of the radial head, pulled elbow, slipped elbow, or toddler elbow.

Demographics

Nursemaid's elbow is a common occurrence in children from the ages of one to four or five. It is rarely seen after the age of six.

Causes and symptoms

Nursemaid's elbow is caused by a strong force on the elbow, such as a sudden pulling or yanking on the hand or forearm. This can occur when a child falls, when an adult pulls up a child's arm, to assist the child up a curb or step or to hurry the child along, or when a child falls away from an adult while being held by the arm. Swinging toddlers from the arms while playing can cause this injury. It can also occur when an infant rolls himself or herself over.

Symptoms of nursemaid's elbow include immediate **pain** in the injured arm, refusal or inability to move the injured arm, creating a condition called pseudo-paralysis, persistent crying, refusal to **play**, and **anxiety**. However each child may experience symptoms differently. These symptoms may also resemble other conditions or medical problems, so a physician should be consulted for a diagnosis.

Swinging a young child by the arms can cause nursemaid's elbow. *(© Raoul Minsart/Corbis.)*

When to call the doctor

The doctor should be called or the child should be taken to an emergency room when a child does not use the arm that has been injured, when the child seems to be in a lot of pain, or when there is a suspicion that someone has deliberately harmed the child.

After treatment for nursemaid's elbow, the doctor should be called if the child still refuses to use the arm six hours after it is put back into place; the child still has pain after 24 hours; the child's fingers get numb and tingly; or the child's elbow comes out of the socket again.

Diagnosis

The diagnosis of nursemaid's elbow is made through a physical examination by the child's physician. X rays of the elbow are usually not necessary.

Treatment

The child's arm should not be straightened or its position changed before the doctor examines it. An ice pack can be used, and the arm splinted in the position in which it is found. The area both above and below the elbow should be immobilized, including the shoulder and wrist if possible.

The physician or health care provider will move the bone and ligament back to their correct positions. This is an easy procedure that can be done in the office by supinating (externally rotating) the forearm (turning the thumb out with palm up), and then gently flexing the arm at the elbow (pushing the forearm up into the biceps). The ligament needs time to heal, so the child's arm should be treated gently after the procedure. The arm may be immobilized in a sling for one or two days, if desired.

Once the ligament and the radial head are returned to their proper position, the child can usually begin to use his or her arm again within a few minutes. Sometimes there is a short period of crying for a minute or two after the release. The earlier after the injury that the child is treated, the more rapid will be the recovery. Alternatively, if the problem has existed for some days, then relief from pain may not be so rapid. Bracing and **immo-**

KEY TERMS

Elbow—Hinged joint between the forearm and upper arm.

Ligament—A type of tough, fibrous tissue that connects bones or cartilage and provides support and strength to joints.

Nursemaid's elbow—An injury to the ligament (strong band of tissue) that keeps the two bones of the forearm in the correct place.

Radius—The bone of the forearm which joins the wrist on the same side as the thumb.

Ulna—One of the two bones of the forearm. Two pivot joints join it to the radius, one near the elbow, one near the wrist.

bilization is usually not required if this is the first occurrence of this condition. For repeat injuries, however, a posterior splint may be applied for several days. A few children may have swelling or pain in the joint. Regular doses of ibuprofen or other over-the-counter pain killers for a few days will help with the swelling and the pain.

If the child fails to use the arm after 15 minutes, the elbow should be x rayed to determine if it was fractured. If no fracture is found, the arm should be splinted and put into a sling, and the condition re-evaluated after 24 hours.

Prognosis

The child should be able to use the elbow after the injury has healed. However, once an elbow has been injured, it is more likely that it will happen again in the future if care is not taken to prevent further injury.

Prevention

A child should not be lifted or swung by the arms or hands nor should a child's arm be pulled hard. A small child should be lifted from under the arms.

Parental concerns

Parents should be aware of the damage to the ligament structures of the elbow and should not lift the child by the arm, as the pulled-elbow condition can reoccur up. Recurrence is especially likely for three to four weeks following the injury.

Unfortunately, nursemaid's elbow commonly occurs when an obstinate child is forcibly pulled along or lifted by the forearm by a parent or older sibling. For example, the injury might occur in a shopping center where a young child, typically two or three years old, is intent on pursuing a course or selecting an item somewhat different from the intentions of the parent. Another scenario that might result in nursemaid's elbow is when a parent or caregiver and a child are crossing the street where the parent or caregiver is forced to pull the child along quickly to avoid an approaching vehicle or to make it to the other curb before the traffic lights change. Care should also be taken to control anger or impatience to prevent re-injury in the future.

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Judith Sims, MS

Nutrition

Definition

The process by which humans take in and use food in their bodies; also the study of diet as it relates to health.

Description

Good nutrition in childhood lays the foundation for good health throughout a person's lifetime. With the proliferation of fast food restaurants, the number of junk food commercials on television, and the increased trend toward eating out, it is more difficult than ever for parents to ensure that their children maintain a nutritious diet. Across the last decades of the twentieth century, increasing affluence and the widespread availability of vitamin-enriched foods have shifted the focus of nutritional

concerns in the United States from obtaining minimum requirements to cutting down on harmful elements in one's diet. Parents need to be as concerned about high levels of fat, cholesterol, sugar, and salt, as well as adequate intake of **vitamins**, **minerals**, and other nutrients.

The American Academy of Pediatrics, the National Academy of Sciences, the American Heart Association, and other health-care organizations agree that fat should not account for more than 30 percent of the calorie intake of children over the age of two, and saturated fat should account for under 10 percent. The main dietary sources in children's diets of saturated fat are whole milk, cheese, hot dogs, and luncheon meats. Recommendations for dietary change include switching to 1 percent or skim milk, low-fat cheese, and meats from which the fat can be trimmed. Since fat is important for growth, experts also caution that fat intake should not be under 25 percent of daily calorie intake and that parents of children under age two should not restrict fat in their diets.

The amount of refined sugar in children's diets—typically accounting for 14 percent of calorie intake by adolescence—is another cause for concern. Although sugar is known to cause **tooth decay** and also may be associated with behavior problems, the greatest danger in consuming foods high in added sugar is that these "empty calories" may replace the more nutritious foods that children need in order to maintain good health. (Soft drinks, perhaps the single greatest source of refined sugar in the diet of children and teenagers, get virtually all their calories from sugar and offer no nutrients.) This high intake of fat can lead to excess weight and, potentially, **obesity**.

Another element that needs to be restricted in children's diets is the intake of sodium through salted foods. Sodium has been closely linked to **hypertension** (high blood pressure), which increases a person's risk of heart disease and **stroke**. It has been determined that 18-year-olds need only 500 milligrams of sodium daily. In addition to limiting the amounts of fat, cholesterol, salt, and sugar in their children's diets, health authorities also recommend that parents concerned about nutrition ensure that children obtain a generous supply of complex carbohydrates (found in such foods as beans, potatoes, whole-grain products, and pasta) and have at least five servings of fresh fruits and vegetables daily.

Infancy

The first nutritional decision that must be made for a child by a parent or primary caregiver is whether to breastfeed or bottle feed. Breast milk is generally considered the best food for an infant up to the age of six to nine months. It has virtually all the nutrients that babies need and in the right balance. In addition, it contains important antibodies that help protect infants from infection at a time when their own immune systems are not yet fully developed.

The composition of breast milk actually changes during the first two weeks after a baby is born. Initially, it consists largely of colostrum, a substance that has more protein than complete breast milk and lower amounts of fat and sugar. It is also rich in the antibody immunoglobin A, which helps protect against infections. By the tenth day after birth, the regular breast milk, containing more carbohydrates and fat and less protein, is produced. The amounts of carbohydrates and fat gradually continue to increase, as will the quantity of the milk itself, to match the needs of the growing baby. Although most full-term infants get all the necessary nutrients from breastfeeding, some may need supplements of vitamins D and K.

Women who are either unable to breastfeed or who choose not to do so usually feed their babies formula made from processed cow's milk, generally reconstituted skim milk with vegetable oils added to substitute for the missing butterfat, which is difficult for infants to digest. Lactose (milk sugar) is also added, and some formulas contain whey protein as well. For infants who demonstrate sensitivity to cow's milk, formulas based on soy protein are available.

Breast milk or formula provides all the nutrients an infant needs up to the age of four to six months. Contrary to past beliefs, it has been found that not only do babies not need solid foods before then, introducing solids too early may lead to **food allergies** or overfeeding. Regular grocery-store cow's milk, which cannot be adequately digested by infants and can cause gastrointestinal bleeding, should not be introduced until a child is a year old. As the first solid food, pediatricians often recommend cereal made from a grain other than wheat, such as rice. The first solid foods may be either commercial baby food or strained foods prepared at home. Once solid foods have been introduced, infants still need to receive most of their nourishment from either breast milk or formula during their first year.

Toddlerhood

During children's second year, their growth rate slows dramatically compared to the prior period. In the first year, their birth weight triples, their length increases by 50 percent, and the size of their brain doubles. After that first year, it takes several years for their weight to even double. They will grow in spurts, with each spurt followed by a period of weight gain. This decreased

A child should eat a variety of foods every day for good health and proper growth. *(© Brian Leng/Corbis.)*

growth leads to a decreased demand for food, often manifested in a newfound pickiness. As long as a child consumes an adequate, varied diet over a period of several days, parents are cautioned against becoming unduly concerned over a single day of unbalanced eating. Toddlers need to eat more than three times a day, either five or six small meals or three major ones with snacks in between.

Preschool

Preschoolers are still growing relatively slowly. Their weight increases about 12 percent between the ages of three and five, although their appearance changes considerably as they lose the baby fat of infancy and toddlerhood. They are still picky eaters, generally eating less—and less consistently—than their parents would like. Although their fat requirement is not as high as that of infants, preschoolers still require more fat and fewer carbohydrates than adults. Fat is needed both for growth and for regulation of body temperature. Also, preschoolers need more than twice as much protein as adults. If the nutritional recommendations of the National Academy of Sciences are followed, a preschooler's diet will consist of 40 percent carbohydrates, 35 percent fats, 20 percent protein, and 5 percent fiber.

Between the ages of three and five, children's tastes expand considerably, and they are willing to consider foods they would have refused as toddlers. Four-year-olds can generally eat whatever foods the rest of the **family** is having. Preschoolers still cannot eat enough at three meals to meet their nutritional needs, and nutritious snacks are important. By this age, children's food choices can be strongly influenced by others. They will imitate good eating habits they see practiced by their parents, but they can also be easily swayed by television commercials for junk food.

School age

The diet of young school-age children, like that of preschoolers, should contain, in order of importance, carbohydrates, fat, and protein. A recommended proportion of these nutrients is 55 percent of the daily calorie intake from carbohydrates, 30 percent from fats, and 15 percent from protein. Once children begin spending a full day in school, a substantial, nutritious breakfast becomes more important than ever. Breakfast has been shown to affect the concentration and performance of elementary school children. Ideally, a balanced breakfast for a school-age child contains food high in protein as well as fruit and bread or another form of grain.

A major change affecting the nutrition of school-age children is the growth of opportunities to eat outside the home. The carefully packed homemade lunch may be traded for a salty snack or cupcake, and parts of it may be discarded. Vending machines and stores offer more temptations. In addition, school lunch programs differ widely in quality; even the nutritional value of a single food, such as a hamburger, can vary significantly depending on how it is prepared and what ingredients are used.

Adolescence brings its own set of nutritional needs and challenges. Beginning with the pre-teen years, children undergo their most intensive period of physical growth since infancy and need more food than at any other stage of life, particularly if they participate in **sports**. Teenagers, especially boys, are notorious for being able to empty the refrigerator of food, usually without gaining excess weight. Early adolescence in particular is a time of increased nutritional requirements for girls, who experience their greatest growth spurt at this time and also begin menstruating. It is difficult for weight-conscious teenage girls to eat enough to satisfy their minimum daily iron requirement of 18 milligrams, and they should try to eat either foods that are naturally rich in iron, such as turkey, beef, liver, and beans, or foods made from iron-enriched cereals. Adequate calcium intake is essential for the rapidly growing bones of teenagers, but milk has often been replaced by soft drinks as the beverage of choice among this age group. Parents should encourage adolescents, especially girls, to eat other foods rich in calcium, such as cheese, salmon, and broccoli.

As adolescents grow more independent, the number of meals and snacks eaten away from home increases as they spend more time with friends and take increased responsibility for arranging their own meals, with fast foods, soft drinks, and sweets often prominent on the menu. In addition to the natural appeal of these foods, **peer pressure** contributes to the choice of a diet soft drink over milk or juice, or pizza over broccoli. Although parents cannot control the eating habits of their teenagers, they can influence them by consistently making nutritious foods available at home and, at least in some cases, by discussing the benefits of good nutrition with them, especially if a relative or friend has had an illness, such as heart disease or colon **cancer**, that has known links to diet.

Common problems

A special problem that may affect childhood nutrition is the presence of food **allergies**, which are more common in children than in adults. They are most likely to begin when a child is very young and the immune system is still sensitive, usually in infancy. Food allergies also tend to run in families: if one parent has food allergies, a child has a 40 percent likelihood of developing one. This figure rises to 75 percent if both parents have food allergies. Common symptoms of food allergies include **hives** and **rashes**; swelling of the eyes, lips, and mouth; respiratory symptoms; and digestive problems. Foods that most often produce allergic reactions in infants are cow's milk, soy products, and citrus fruits. Other common childhood allergens include wheat, nuts, chocolate, strawberries, tomatoes, corn, and seafood. In time, childhood food allergies are often outgrown. Feeding a child with food allergies is a challenging but not impossible task for parents. A variety of foods can be substituted for those to which a child is allergic: soy products for milk and other dairy products; carob for chocolate; and, in the case of wheat allergies, products or flour made from grains such as rice or oats.

Parental concerns

Vegetarian kids

About 2 percent of Americans ages six to 17 (about 1 million) are vegetarian, the same percentage as among American adults, and 0.5 percent are vegan, according to a 2002 survey by the Vegetarian Resource Group (VRG). Six percent of six- to 17-year-olds do not eat meat but eat fish and/or poultry. Teens who follow a vegetarian diet are more likely to meet recommendations for total fat, saturated fat, and number of servings of fruits and vegetables as compared to non-vegetarians. They also have higher intakes of iron, vitamin A, fiber, and diet soda, and lower intakes of vitamin B12, cholesterol, and fast food. Most teens, whether they are vegetarian or not, do not meet recommendations for calcium, according to the VRG survey. The study concluded that rather than viewing adolescent **vegetarianism** as a phase or fad, the diet could be viewed as a healthy alternative

KEY TERMS

Colostrum—Milk secreted for a few days after birth and characterized by high protein and antibody content.

Essential fatty acid (EFA)—A fatty acid that the body requires but cannot make. It must be obtained from the diet. EFAs include omega-6 fatty acids found in primrose and safflower oils, and omega-3 fatty acids oils found in fatty fish and flaxseed, canola, soybean, and walnuts.

Immunoglobin A—A sugar protein with a high molecular weight that acts like an antibody and is produced by white blood cells during an immune response.

to the traditional American meat-based diet. The study also said that vegetarian diets in adolescence could lead to lifelong health-promoting dietary practices. The study was reported in the July-August 2002 issue of the VRG publication *Vegetarian Journal*.

Parents should closely monitor their vegetarian child's height, weight, and general health. A child who is not getting enough vitamins and nutrients may have symptoms such as skin rashes, fatigue, a painful and swollen tongue, irritability, pale skin, mental slowness, or difficulty breathing. The diets of vegetarian adolescents should be monitored closely to make sure they include a variety of foods, including fruits, vegetables, beans, whole grains, and non-meat protein sources. For vegetarians who do not eat fish, getting enough omega-3 essential fatty acids may be an issue, and supplements such as flaxseed oil should be considered, as well as walnuts and canola oil. Another essential fatty acid, omega-6, found in fish, can be obtained from borage oil or evening primrose oil supplements.

When to call the doctor

Parents should consult their child's pediatrician or physician if they are unsure the child's diet is nutritionally adequate. A doctor should also be consulted if a child's weight or height is not appropriate for their age.

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Ken R. Wells

Nystagmus

Definition

Nystagmus is a condition in which there is involuntary and rhythmic movement or oscillation of the eye. It is often caused by an underlying ocular or neurological disorder.

Description

The eye movements associated with nystagmus are varied. They can be either pendular, in which the oscillations are equal in all directions and or jerk, in which the movements may be faster in one direction than another. The frequency of the oscillation or movement and the amplitude of the oscillation also vary. The movements themselves may be vertical, horizontal, circular, or oblique in direction. Nystagmus can be sensory and develop as a result of poor vision, or it can be motor and develop as a result of a neurological problem.

Nystagmus may be congenital, or it may be acquired. Congenital, or infantile, nystagmus appears within the first few months of life. Congenital nystagmus is usually binocular and affects both eyes, is horizontal in direction, and does not occur while the child is sleeping. It decreases when the child's eyes converge or move inward. Most of these cases of nystagmus develop because of poor vision and do not have an underlying neurological cause.

Children with congenital nystagmus usually have a point in their eye movement in which the intensity of the nystagmus is decreased. This is called the null point, and the child may adopt a head tilt or rotation to help maintain his or her eyes at this position. This point is usually not in straight ahead or in a primary gaze position. Children with nystagmus who have their the null point located at a position in which the eyes are positioned inward may develop an esotropia, a form of **strabismus** or eye turn.

One variant of congenital nystagmus is spasmus nutans, which appears as a triad with accompanying head nodding and torticollis (head turn or tilt), and is seen between four months and three-and-a-half years of age and usually resolves without treatment within one to two years of onset. Rarely does it persist past age five. Usually spasmus nutans appears bilaterally, and the nystagmus is in a horizontal direction. When the nystagmus of spasmus nutans is vertical or rotary, the child does not have a head tilt.

There are various types of nystagmus. Downbeat nystagmus is characterized by a nystagmus that is more pronounced when the child looks down, especially when looking to the side, or in lateral gaze. An accentuated oscillation when looking up is seen in upbeat nystagmus. Seesaw nystagmus is an unusual type of in which one eye moves in and down and the other out and up. A periodic alternating nystagmus (PAN) is observed in primary gaze when the patient is looking straight ahead and is characterized by eye movements that continuously change direction and speed. Peripheral vestibular nystagmus may be accompanied by vertigo, **nausea**, and tinnitus, or ringing in the ears. This type of nystagmus is not always apparent but can be seen by a doctor when he or she looks in the back of the eye with a direct ophthalmoscope.

Latent nystagmus appears only when one eye is covered. This is a congenital nystagmus caused by an ocular motor disturbance rather than visual deprivation. It is often accompanied by strabismus or an eye turn. A child with latent nystagmus will not see well when one eye is covered.

Gaze evoked nystagmus occurs only when one is looking to the side in extreme lateral gaze. This type of nystagmus can be caused by ethanol and recreational drug use, but is seen in myasthenia gravis and thyroid disease as well.

Some types of nystagmus are normal. If one looks at an object in extreme gaze for a long period of time, endpoint nystagmus may be noted. Optokinetic nystagmus (OKN) is a nystagmus that can be elicited involuntarily when a rapidly moving striped object is passed in front of an individual's eyes.

Transmission

Congenital nystagmus may be transmitted genetically, either as an autosomal recessive or dominant, or as an X-linked recessive trait. It can also be associated with other conditions that are genetically transmitted. For example, Leber's amaurosis is an autosomal dominant trait and **albinism** is X-linked.

Demographics

Congenital nystagmus occurs twice as frequently in males than in females. The prevalence of nystagmus in the pediatric population is .015 percent. Eighty percent of nystagmus is congenital, and the remaining 20 percent is acquired.

Causes and symptoms

The eyes of an individual with nystagmus cannot remain still and oscillate in some position of gaze. Those

with nystagmus usually have decreased vision and poor depth perception, although those born with nystagmus, may not realize that their vision is poor. Those with acquired nystagmus may experience double vision or oscillopsia, or that objects in their visual space appear to move. An acquired nystagmus may be accompanied by other symptoms such **dizziness**, difficulty with balance, hearing loss, poor coordination, and **numbness**. If an individual with nystagmus experiences oscillopsia, then the nystagmus is acquired.

The primary cause of congenital nystagmus is visual deprivation, and the causes of visual deprivation in an infant include cataracts, oculoalbinism, glaucoma, retinal detachments, Leber's amaurosis, developmental abnormalities of the optic nerve such as a coloboma, and achromatopsia, a condition in which the infant cannot see color.

Acquired nystagmus can be caused by demyelination of nerve fibers, such as occurs in multiple sclerosis, lesions or tumors of the vestibular or visual pathways, strokes of the central nervous system, and drug use, both recreational as well as a side effect of prescribed drugs, such as those used to treat seizures and depression. Other causes of acquired nystagmus are Arnold-Chiari malformations, vitamin deficiencies, syphilis, Wernicke's encephalopathy, Behcet's syndrome, and Meniere's disease.

When to call the doctor

Since nystagmus can be caused by tumors, **stroke**, and trauma or neurological disorder, any type of nystagmus must be evaluated by a qualified practitioner. The nystagmus can be a sign of a serious problem. For example, a type of tumor called chiasmal glioma has signs and symptoms similar to spasmus nutans.

Diagnosis

Diagnosis of nystagmus is made primarily by patient history as reported by a parent, the age of onset, and observation of any accompanying signs such as a head turn, tilt or tremor, or oscillopsia. If possible, the infant or child's best visual acuity is determined. If the onset is acute, then usually the nystagmus is acquired.

The type of nystagmus can accurately be determined by eye movement recordings, which map direction, frequency, null point, and amplitude of the nystagmus. For the infant with congenital nystagmus, evoked response potential (EVR) and electroretinogram (ERG) give the doctor objective information about visual potential, and **magnetic resonance imaging** (MRI) can determine if and where a lesion is located. For the infant or young child, some of these tests may be done under anesthesia.

Treatment

The treatment for nystagmus, once the etiology is determined and treated, includes optical devices such as **contact lenses** and glasses, medication, and surgery.

For individuals with nystagmus correction of refractive error with glasses or contact lenses is the first step in treating the condition. For 85 percent of children with nystagmus, a spectacle prescription improves vision significantly. For those with congenital nystagmus, prism may be put in glasses to help position the eye at its null point or to help the eyes converge. For some people contact lenses are prescribed. Contact lenses slow down eye movements, and because the optical center of the prescription is always centered on the eye with the contact lens, vision improves. Low vision aids such as telescopes assist those whose vision cannot be fully corrected with spectacles and contact lenses alone. Tinting of the glasses or sunglasses may decrease the nystagmus of individuals with albinism. For the patient with oscillopsia, grinding prism into the spectacles may move the visual field to a point of decreased oscillopsia.

Congenital nystagmus, when due to a visual deprivation, is rarely improved by surgery. But when a head tilt or head turn accompanies nystagmus, surgery to correct a muscle imbalance may improve nystagmus and visual acuity. Surgery on the extraocular muscles of the eye may be helpful when the child's null point is in not in primary gaze but located at least 30 degrees from straight-ahead vision. When a tumor or stroke has caused an acquired nystagmus, then neurosurgery, if indicated for the underlying cause, may lead to resolution of the nystagmus. When surgery is considered, the risks of anesthesia must also be considered.

If oscillopsia is a co-existing symptom, then drugs can be given to reduce the ocular oscillations. Vestibular nystagmus can be treated by diazepam or scopolamine. Drugs called GABA agonists, such as baclofen and carbamazepine, are useful in treatment of seesaw nystagmus and PAN, if the nystagmus is acquired and not congenital. Baclofen cannot be given to children.

Botox (*Botulinum* toxin) injections can temporarily control the eye movements, but because of side effects such as double vision and ptosis or drooping of the eyelid, and because it is not a permanent solution, Botox is not used often.

KEY TERMS

Acupuncture—Based on the same traditional Chinese medical foundation as acupressure, acupuncture uses sterile needles inserted at specific points to treat certain conditions or relieve pain.

Albinism—An inherited condition that causes a lack of pigment. People with albinism typically have light skin, white or pale yellow hair, and light blue or gray eyes.

Autosomal—Relating to any chromosome besides the X and Y sex chromosomes. Human cells contain 22 pairs of autosomes and one pair of sex chromosomes.

Binocular—Affecting or having to do with both eyes.

Biofeedback—A training technique that enables an individual to gain some element of control over involuntary or automatic body functions.

Coloboma—A birth defect in which part of the eye does not form completely.

Lesion—A disruption of the normal structure and function of a tissue by an injury or disease process. Wounds, sores, rashes, and boils are all lesions.

Strabismus—A disorder in which the eyes do not point in the same direction. Also called squint.

X-linked—A gene carried on the X chromosome, one of the two sex chromosomes.

If the nystagmus is due to drug toxicity, then reducing or discontinuing the drug eventually resolves the problem.

Alternative treatment

Acupuncture and biofeedback and vision therapy have been successful for some patients.

Prognosis

Congenital nystagmus is usually a benign condition. It is not curable, but its symptoms can be diminished with spectacles or contact lenses. The best corrected vision for most individuals with congenital nystagmus is between 20/40 and 20/70, but correction to 20/20 is possible for some. Nystagmus associated with spasmus nutans resolves spontaneously before the child reaches school age.

The prognosis for an acquired nystagmus depends on its cause. If the condition is due to a side effect of a drug, then decreasing or changing the treatment drug eventually resolves the nystagmus.

Prevention

In general nystagmus cannot be prevented. Since the cause of acquired nystagmus can be due to a co-existing neurological condition, prompt attention to other neurological signs that may accompany nystagmus, such as dizziness, may prevent or decrease the severity of nystagmus itself. Careful monitor of dosage of those drugs with nystagmus as a side effect may prevent the condition.

Parental concerns

Because nystagmus can be associated with many medical problems, the child with this condition must undergo a complete ocular and neurological evaluation.

Children with nystagmus are not aware that they may have a visual deficiency and as they get older must be helped with the restrictions that nystagmus places on them. For instance, driving may be restricted or not permitted. Certain occupations for which good visual correction is a requirement may be not feasible. Every effort must be made to integrate the child with nystagmus into a normal school setting in order to prepare the child for adult life, even if cosmetic concerns may instinctively lead the parent to want to protect the child.

Support for families of those with nystagmus can be found through the American Nystagmus Network.

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Martha Reilly, OD

O

Obesity

Definition

Obesity is an abnormal accumulation of body fat, usually 20 percent or more over an individual's ideal body weight. Obesity is associated with increased risk of illness, disability, and death.

The branch of medicine that deals with the study and treatment of obesity is known as bariatrics. As obesity has become a major health problem in the United States, bariatrics has become a separate medical and surgical specialty.

Description

Childhood obesity is in the early 2000s a significant health problem in the United States. Obese children and adolescents are at increased risk for developing diabetes, **hypertension**, coronary artery disease, **sleep** apnea, orthopedic problems, and psychosocial disorders.

Obesity involves excessive weight gain and fat accumulation. For children and adolescents, obesity is defined in terms of body mass index (BMI) percentile. BMI is a formula that considers an individual's height and weight to determine body fat and health risk, and it is used differently for children and adolescents than it is for adults. In adults, BMI often misrepresents obesity because it does not consider healthy weight from muscle tissue; therefore, body fat percentage is considered a more accurate method for determining obesity in adults. In children and adolescents, because body fat changes as they mature, BMI is gender- and age-specific and plotted on gender-specific growth charts to determine BMI-for-age. Curved lines on the chart (percentiles) are used by healthcare professionals to identify children and adolescents at risk for overweight and obesity. Children and adolescents with a BMI-for-age in the 85th to 95th percentile are considered overweight and at risk for obesity, and those with a BMI-for-age greater than the 95th percentile are considered obese.

Demographics

According to the American Obesity Association and the Centers for Disease Control and Prevention, 30.3 percent of children aged six to 11 years are overweight and 15.3 percent are obese, and 30.4 percent of adolescents aged 12 to 19 years are overweight and 15.5 percent are obese. From 1980 to 2004, the prevalence of obesity among children quadrupled, and the prevalence of obesity in adolescents more than doubled. Overweight and obesity is more prevalent in boys (32.7%) than girls (27.8%). Obesity is more common in African American, Hispanic American, and Native American children and adolescents, than among Caucasians of the same ages.

Causes and symptoms

Although obesity can be a side effect of certain hormonal disorders or use of certain medications, the primary cause of obesity in children and adolescents is excess calorie consumption coupled with a sedentary lifestyle. Children and adolescents living in the twenty-first century are the most inactive generation ever. The majority of schools no longer offer daily physical education classes; and active leisure activities, such as bicycle riding, have been replaced by sedentary activities, such as television watching and playing computer games. Studies have documented dramatic changes in childhood food consumption from the 1970s to 2004. Fast foods and foods eaten at other restaurants have increased by 300 percent since 1977, and soft drink consumption has also increased significantly. In addition, standard meal portion sizes and snacking have increased.

Obesity is the result of a complex interaction of genetics and environmental factors. Genetics influence how the body regulates appetite and metabolism, while certain environmental factors encourage excess calorie

consumption. The body requires a certain amount of energy for basic metabolism and to support additional physical activity. When calories consumed from food and beverages equal calories expended during physical activity, body weight is maintained. When calories consumed exceed calories expended, weight gain results. To gain one pound, 3,500 additional calories must be consumed. In American society, excess calories are easily consumed just by drinking soft drinks and eating "supersized" fast food meals. A sedentary lifestyle results in far fewer calories being burned daily.

The major symptoms of obesity are excessive weight gain and the presence of large amounts of fatty tissue. Obesity can cause a number of other conditions, including type 2 diabetes, hypertension, **high cholesterol**, joint **pain**, **asthma**, **hypothyroidism**, and gallstones. Type 2 diabetes, previously referred to as adult-onset diabetes, has increased dramatically in children, and this increase has been directly linked to obesity.

When to call the doctor

Overweight and obese children should be evaluated by a physician for diabetes, hypertension, high cholesterol, and other medical conditions that are influenced by excessive weight gain. Primary care physicians can be consulted for weight management counseling to help children lose weight.

Diagnosis

Obesity in children and adolescents is diagnosed using the BMI-for-age formula described above, which is used to define obesity. Comorbid conditions, such as diabetes and high cholesterol, are diagnosed using medical laboratory tests.

Treatment

As of 2004, no weight loss drugs were approved for use in children, although some drugs used to treat obesity are approved for use in adolescents age 16 years and older. A few drugs are under investigation for use in children. Although no drugs are specifically approved for pediatric weight loss, some physicians may prescribe them "off-label." Because the side effects of these medications in children are unknown, children should not use adult weight loss drugs.

For extremely obese adolescents, surgical procedures—called bariatric surgery—may be performed, but only rarely. These procedures involve significant surgical alteration of the digestive tract and require substantial modification of diet after the surgery to much less than 1,000 calories per day. The long-term effects on growth and development from severe post-operative calorie restriction are not unknown, and weight loss surgery should only be performed on adolescents as a last resort.

The most effective treatment for obese children and adolescents is behavior and lifestyle modification under the guidance of a physician or weight management specialist experienced in dealing with children and adolescents. Behavior and lifestyle modification involves the following:

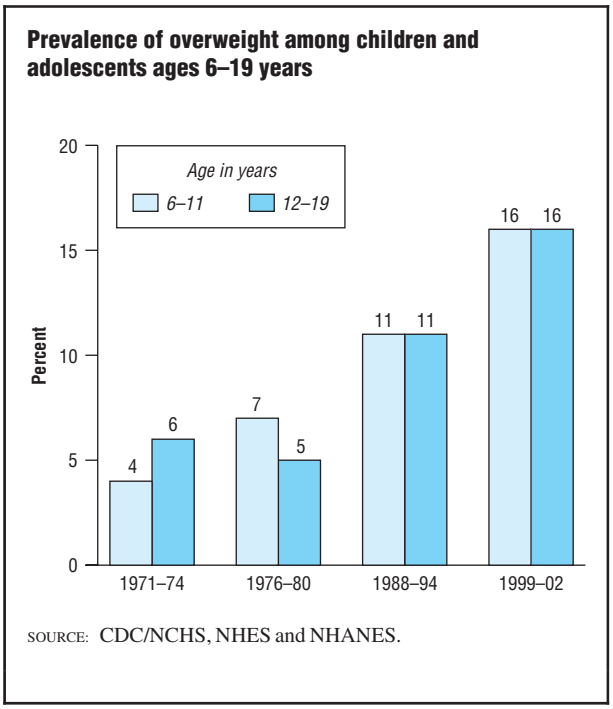
- **assessment** of child's and family's eating habits
- implementation of a regular, safe **exercise** program and increasing active leisure activities
- limiting television viewing and other sedentary activities
- setting reasonable goals and monitoring goal achievement using positive, non-food-related incentives
- counseling regarding how to keep a food/activity diary to track progress
- extensive support by involving entire **family** and/or joining a weight loss group of peers

Alternative treatment

Alternatives for weight loss involve the use of ephedra-containing drugs or herbal preparation or the use of diuretics and **laxatives**. Both of these practices are unsafe, especially for children and adolescents. Because ephedra can cause severe cardiac side effects, the Food and Drug Administration has issued warnings against its use. Diuretics and laxatives can result in severe **dehydration** and improper absorption of nutrients.

Acupressure and acupuncture can suppress food cravings. Visualization and meditation can create and reinforce a positive self-image that enhances the patient's determination to lose weight. By improving physical strength, mental concentration, and emotional serenity, **yoga** can provide the same benefits.

Given the drastic increase in childhood obesity, special summer programs and therapeutic schools have been formed to help children lose weight. Summer camp programs that focus on healthy eating and exercise habits are available for overweight and obese children. In addition, in early 2004, the first **alternative school** for overweight and obese children, which operates like other private and charter schools, but with a focus on healthy weight loss and maintenance, was established.



This graph shows the increasing numbers of overweight children in the United States. *(Illustration by GGS Information Services.)*

Prognosis

Obese and overweight children and adolescents are more likely to be obese or overweight as adults. According to the American Obesity Association, obese children aged 10 to 13 have a 70 percent chance of remaining obese for the rest of their lives. Obese individuals are at increased risk for many other diseases and early death. Behavior and lifestyle modification programs involving positive goal-setting, increased exercise, and group support can help children and adolescents successfully and safely lose weight.

Prevention

Obesity can be prevented by instilling healthy eating and regular exercise habits in children at an early age. Minimizing and structuring daily time for sedentary activities like television viewing and encouraging outdoor activities such as bicycle riding, walking, running, and active **play**, and active indoor activities such as dancing can help increase physical activity. Dietary modifications to help prevent obesity include limiting soft drink and fast food consumption, monitoring food portion sizes, and providing a well-balanced diet.

KEY TERMS

Adipose tissue—Fat tissue.

Bariatrics—The branch of medicine that deals with the prevention and treatment of obesity and related disorders.

Ghrelin—A peptide hormone secreted by cells in the lining of the stomach. Ghrelin is important in appetite regulation and maintaining the body's energy balance.

Hyperlipidemia—A condition characterized by abnormally high levels of lipids in blood plasma.

Hyperplastic obesity—Excessive weight gain in childhood, characterized by an increase in the number of new fat cells.

Hypertension—Abnormally high arterial blood pressure, which if left untreated can lead to heart disease and stroke.

Hypertrophic obesity—Excessive weight gain in adulthood, characterized by expansion of already existing fat cells.

Ideal weight—Weight corresponding to the lowest death rate for individuals of a specific height, gender, and age.

Leptin—A protein hormone that affects feeding behavior and hunger in humans. As of 2004 it is thought that obesity in humans may result in part from insensitivity to leptin.

Nutritional concerns

Nutrition is a primary factor for weight management of obese children and adolescents. Poor nutrition and dietary habits can lead to weight gain and obesity. Dietary modification is important for helping children lose weight and prevent obesity.

The following nutritional guidelines can help in the management of obesity:

- Limit soft drink consumption to one per day or less. One 12-ounce can of soda has 120 calories or more. Often, children and adolescents consume "super-size" sodas that may contain up to 1,000 calories.
- Limit fast food restaurant visits to one per week, and choose healthy options like grilled chicken and smaller sized portions of high-calorie items.
- Monitor food serving sizes.
- Increase consumption of fruits, vegetables, high-fiber foods, and whole-grain foods.

- Be aware that "low-fat" foods often substitute sugar for fat, and calories may actually be the same as the regular or high-fat version.

Parental concerns

Parents of obese children and adolescents should be concerned for their current and future health, since obesity can result in diabetes, hypertension, and coronary artery disease. Losing weight can be very difficult for obese children, and parental support is essential for success. Because children model behavior after their parents, obesity often affects both parents and children. Parents should strive to have healthy eating habits and exercise regularly to be effective role models for their children. Making healthy eating and exercise a family priority is better for everyone and helps reinforce positive changes in behavior for the obese child.

Obese children and adolescents are more susceptible to eating disorders, negative **self-esteem** and body image, and depression due to peer influences. Counseling, peer group therapy, and **family therapy** may be required to support lifestyle modifications for obese children and adolescents.

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American Obesity Association (AOA). 1250 24th Street NW, Suite 300, Washington, DC 20037. Web site: <www.obesity.org>.

American Society of Bariatric Physicians. 5453 East Evans Place, Denver, CO 80222–5234. Web site: <www.asbp.org>.

American Society for Bariatric Surgery. 7328 West University Avenue, Suite F, Gainesville, FL 32607. Web site: <www.asbs.org>.

National Institute of Diabetes and Digestive and Kidney Diseases. 31 Center Drive, USC2560, Building 31, Room 9A-04, Bethesda, MD 20892–2560. Web site: <www.niddk.nih/gov>.

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Obsessive-compulsive disorder

Definition

Obsessive-compulsive disorder (OCD) is a type of **anxiety** disorder. Anxiety disorder is the experience of prolonged, excessive worry about circumstances in one's life. OCD is characterized by distressing repetitive thoughts, impulses, or images that are intense, frightening, absurd, or unusual. These thoughts are followed by

ritualized actions that are usually bizarre and irrational. These ritual actions, known as compulsions, help reduce anxiety caused by the individual's obsessive thoughts. Often described as the "disease of doubt," the sufferer usually knows the obsessive thoughts and compulsions are irrational but, on another level, fears they may be true.

Description

Most people with obsessive-compulsive disorder have both obsessions and compulsions, but occasionally a person will have just one or the other. The degree to which this condition can interfere with daily living also varies. Some people are barely bothered, while others find the obsessions and compulsions to be profoundly traumatic and spend much time each day in compulsive actions. Because the symptoms are so distressing, sufferers often hide heir fears and rituals but cannot avoid acting on them. OCD sufferers are often unable to decide if their fears are realistic and need to be acted upon.

Obsessions are intrusive, irrational thoughts that keep popping up in a person's mind, such as the urgency to wash one's hands again. Typical obsessions include fears of dirt, germs, contamination, and violent or aggressive impulses. Other obsessions include feeling responsible or others' **safety** or an irrational **fear** of hitting a pedestrian with a car. Additional obsessions can involve excessive religious feelings or intrusive sexual thoughts. The patient may need to confess frequently to a religious counselor or may fear **acting out** the strong sexual thoughts in a hostile way. People with obsessive-compulsive disorder may have an intense preoccupation with order and symmetry or may be unable to throw anything out.

Compulsions usually involve repetitive rituals such as excessive washing (especially hand washing or bathing), cleaning, checking and touching, counting, arranging, and/or hoarding. As the person performs these acts, he may feel temporarily better, but there is no long-lasting sense of satisfaction or completion after the act is performed. Often, a person with obsessive-compulsive disorder believes that if the ritual is not performed, something dreadful will happen. While these compulsions may temporarily ease stress, short-term comfort is purchased at a heavy price—time spent repeating compulsive actions and a long-term interference with life.

The difference between OCD and other compulsive behavior is that while people who have problems with gambling, overeating, or with substance abuse may appear to be compulsive, these activities also provide pleasure to some degree. The compulsions of OCD, on the other hand, are never pleasurable.

OCD may be related to some other conditions, such as the continual urge to pull out body hair (**trichotillomania**); fear of having a serious disease (hypochondriasis); or preoccupation with imagined defects in personal appearance disorder (body dysmorphia). Some people with OCD also have **Tourette syndrome**, a condition featuring **tics** and unwanted vocalizations (such as swearing). OCD is often linked with depression and other anxiety disorders.

Demographics

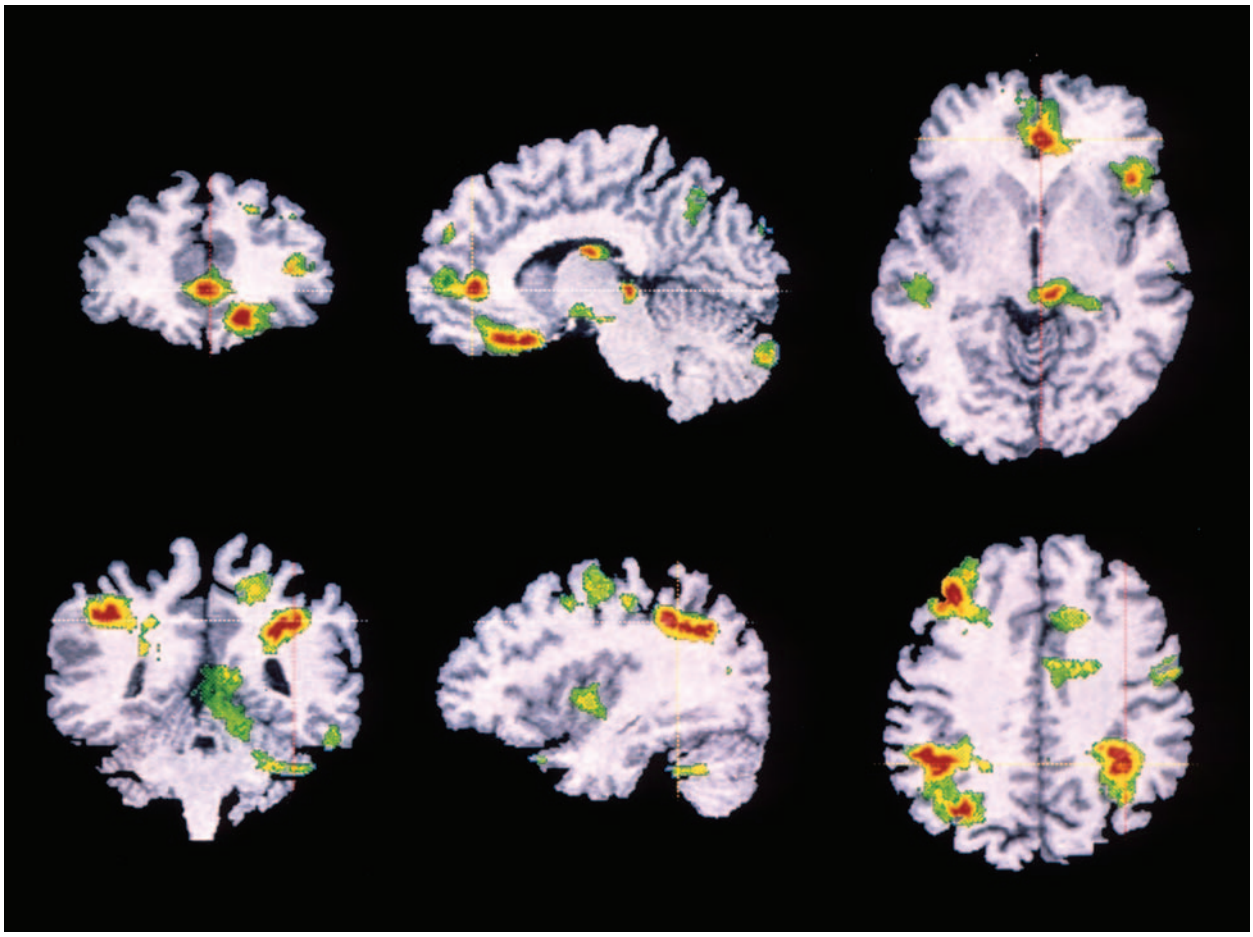
Almost one out of every 40 people suffers from obsessive-compulsive disorder at some time in their lives. The condition is two to three times more common than either **schizophrenia** or manic depression and strikes men and women of every ethnic group, age, and social level.

If one person in a **family** has obsessive-compulsive disorder, there is a 25 percent chance that another immediate family member has the condition. It also appears that stress and psychological factors may worsen symptoms, which usually begin during **adolescence** or early adulthood.

Causes and symptoms

Research suggests that the tendency to develop obsessive-compulsive disorder is inherited. There are several theories behind the cause of OCD. OCD may be related to a chemical imbalance within the brain that causes a communication problem between the front part of the brain (frontal lobe) and deeper parts of the brain responsible for the repetitive behavior. The orbital cortex located on the underside of the brain's frontal lobe is overactive in OCD patients. This may be one reason for the feeling of alarm that pushes the patient into compulsive, repetitive actions. It is possible that people with OCD experience overactivity deep within the brain that causes the cells to get "stuck," much like a jammed transmission in a car damages the gears. This could lead to the development of rigid thinking and repetitive movements common to the disorder. The fact that drugs which boost the levels of serotonin, a brain messenger substance linked to emotion and many different anxiety disorders, in the brain can reduce OCD symptoms may indicate that to some degree OCD is related to levels of serotonin in the brain.

There may also be a link between childhood episodes of **strep throat** and the development of OCD. In some vulnerable children, strep antibodies attack a certain part of the brain. Antibodies are cells that the body produces to fight specific diseases. That attack results in



PET scans of a brain showing active areas in obsessive-compulsive disorder; positive correlations (activity increases as symptoms get stronger), top row; negative correlation (activity decreases as symptoms strengthen), bottom. *(Wellcome Department of Cognitive Neurology/SPL/Photo Researchers, Inc.)*

the development of excessive washing or germ **phobias**. A phobia is a strong but irrational fear. In this instance the phobia is fear of disease germs present on commonly handled objects. These symptoms would normally disappear over time, but some children who have repeated infections may develop full-blown OCD. Treatment with **antibiotics**, immunoglobulin, or blood cleansing procedures can decrease the circulating anti-strep antibodies in the blood, thus lessening the OCD symptoms in some of these children.

Diagnosis

People with obsessive-compulsive disorder feel ashamed of their problem and often try to hide their symptoms. They avoid seeking treatment. Because they can be very good at keeping their problem from friends and family, many sufferers do not get the help they need until the behaviors are deeply ingrained habits and hard to change. As a result, the condition is often misdiagnosed or underdiagnosed. All too often, it can take more than a decade between the onset of symptoms and proper diagnosis and treatment.

OCD appears to be related to a disruption in serotonin levels, there is no blood test for the condition. Instead, doctors diagnose OCD after evaluating a person's symptoms and history.

Treatment

Obsessive-compulsive disorder can be effectively treated by a combination of cognitive-behavioral therapy and medication that regulates the brain's serotonin levels. Drugs that are approved to treat obsessive-compulsive disorder include fluoxetine (Prozac), fluvoxamine (Luvox), paroxetine (Paxil), and sertraline (Zoloft), all selective serotonin reuptake inhibitors (SSRIs) that

affect the level of serotonin in the brain. Older drugs include the antidepressant clomipramine (Anafranil), a widely studied drug in the treatment of OCD, but one that carries a greater risk of side effects. Drugs should be taken for at least 12 weeks before a person decides whether they are effective.

Cognitive-behavioral therapy (CBT) teaches patients how to confront their fears and obsessive thoughts by making the effort to endure or wait out the activities that usually cause anxiety without compulsively performing the calming rituals. Eventually their anxiety decreases. People who are able to alter their thought patterns in this way can lessen their preoccupation with the compulsive rituals. At the same time, the patient is encouraged to refocus attention elsewhere, such as on a hobby.

In a few very severe cases in which patients have not responded to medication or behavioral therapy, brain surgery may be tried as a way of relieving the unwanted symptoms. Surgery can help up to one third of patients with the most severe form of OCD. The most common operation involves removing a section of the brain called the cingulate cortex. The serious side effects of this surgery for some patients are seizures, personality changes, and less ability to plan.

Prognosis

Obsessive-compulsive disorder is a chronic disease that, if untreated, can last for decades, fluctuating from mild to severe and worsening with age. When treated by a combination of drugs and behavioral therapy, some patients go into complete remission. Unfortunately, not all patients have such a good response. About 20 percent of people cannot find relief with either drugs or behavioral therapy. **Hospitalization** may be required in some cases.

Despite the crippling nature of the symptoms, many successful doctors, lawyers, business people, performers, and entertainers function well in society despite their condition. Nevertheless, the emotional and financial cost of obsessive-compulsive disorder can be quite high.

Parental concerns

Some people have referred to obsessive-compulsive disorder as "the great pretender," because its symptoms can mimic a number of other disorders. Furthermore, children may become skilled at hiding the more embarrassing features of their condition. Because of these characteristics of the disorder, obsessive-compulsive disorder may go undiagnosed for some time.

KEY TERMS

Anxiety disorder—A mental disorder characterized by prolonged, excessive worry about circumstances in one's life. Anxiety disorders include agoraphobia and other phobias, obsessive-compulsive disorder, post-traumatic stress disorder, and panic disorder.

Cognitive-behavioral therapy—A type of psychotherapy in which people learn to recognize and change negative and self-defeating patterns of thinking and behavior.

Compulsion—A repetitive or ritualistic behavior that a person performs to reduce anxiety. Compulsions often develop as a way of controlling or "undoing" obsessive thoughts.

Obsession—A persistent image, idea, or desire that dominates a person's thoughts or feelings.

Selective serotonin reuptake inhibitors (SSRIs)—A class of antidepressants that work by blocking the reabsorption of serotonin in the brain, thus raising the levels of serotonin. SSRIs include fluoxetine (Prozac), sertraline (Zoloft), and paroxetine (Paxil).

Serotonin—A widely distributed neurotransmitter that is found in blood platelets, the lining of the digestive tract, and the brain, and that works in combination with norepinephrine. It causes very powerful contractions of smooth muscle and is associated with mood, attention, emotions, and sleep. Low levels of serotonin are associated with depression.

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National Alliance for the Mentally Ill (NAMI). Colonial Place Three, 2107 Wilson Blvd., Suite 300, Arlington, VA 22201–3042. Web site: <www.nami.org>.

National Anxiety Foundation. 3135 Custer Dr., Lexington, KY 40517. Web site: <www.lexington-on-line.com/naf.html>.

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Oligomenorrhea

Definition

Medical dictionaries define oligomenorrhea as infrequent or very light **menstruation**. But physicians typically apply a narrower definition, restricting the diagnosis of oligomenorrhea to women whose periods were regularly established before they developed problems with infrequent flow. With oligomenorrhea, menstrual periods occur at intervals of greater than 35 days, with only four to nine periods in a year.

Description

True oligomenorrhea cannot occur until a young woman's menstrual periods have been established. In the United States, 97.5 percent of women have begun normal menstrual cycles by age 16. The complete absence of menstruation (menstrual periods never started or they stopped after having been established) is called **amenorrhea**. Oligomenorrhea can be redefined as amenorrhea if menstruation stops for six months or more; however, there is no universally agreed-upon cutoff point or timeline.

It is quite common for women at the beginning and end of their reproductive lives to miss periods or have them at irregular intervals. This variation is normal and is usually the result of imperfect coordination between the hypothalamus, the pituitary gland, and the ovaries. For no apparent reason, a few women menstruate (with ovulation occurring) on a regular schedule as infrequently as once every two months. For them that schedule is normal and not a cause for concern.

Women with polycystic ovary syndrome (PCOS) are also likely to suffer from oligomenorrhea. PCOS is a condition in which the ovaries become filled with small cysts. Women with PCOS show menstrual irregularities that range from oligomenorrhea and amenorrhea to very heavy and irregular periods. PCOS affects about 6 percent of premenopausal women and is related to excess androgen production.

Other physical and emotional factors also cause a woman to miss periods. These include the following:

- emotional stress
- chronic illness
- poor nutritional status
- such eating disorders as **anorexia nervosa**
- excessive **exercise**
- estrogen-secreting tumors
- abnormalities in the structure of the uterus or cervix that obstruct the outflow of menstrual fluid
- illicit use of anabolic steroid drugs to enhance athletic performance

Professional ballet dancers, gymnasts, and ice skaters are especially at risk for oligomenorrhea because they combine strenuous physical activity with a diet intended to keep their weight down. Menstrual irregularities are known to be one of the three disorders comprising the so-called "female athlete triad," the other disorders being disordered eating and osteoporosis. The triad was first formally named at the annual meeting of the American College of Sports Medicine in 1993, but doctors were aware of the combination of bone mineral loss, stress **fractures**, eating disorders, and participation in women's **sports** for several decades before the triad was named. Women's coaches have become increasingly aware of the problem since the early 1990s and are encouraging female athletes to seek medical advice.

Demographics

By definition, oligomenorrhea is a health concern only for women. It is estimated that about 5 percent of women in the United States in their childbearing years experience an episode of oligomenorrhea each year. This percentage appears to be constant across racial and ethnic groups.

Oligomenorrhea related to the female athlete triad is more common in this group of women than in the general female population. One study at the University of California at San Francisco found that 11 percent of female

marathon runners had amenorrhea or oligomenorrhea. Although precise data are difficult to obtain because many athletes with the triad try to hide their symptoms from others, disordered eating and menstrual irregularities have been estimated to run as high as 62 percent of female athletes at the college level, with 4 percent to 39 percent meeting the criteria for anorexia nervosa or **bulimia nervosa** as defined by the fourth edition of the *Diagnostic and Statistical Manual of Mental Disorders*, or *DSM-IV*.

Causes and symptoms

Causes

Oligomenorrhea that occurs in adolescents is often caused by immaturity or lack of synchronization between the hypothalamus, pituitary gland, and ovaries. The hypothalamus is the part of the brain that controls body temperature, cellular metabolism, and such basic functions as appetite for food, the sleep/wake cycle, and reproduction. The hypothalamus also secretes hormones that regulate the pituitary gland.

The pituitary gland is then stimulated to produce hormones that affect growth and reproduction. At the beginning and end of a woman's reproductive life, some of these hormone messages may not be synchronized, resulting in menstrual irregularities.

Oligomenorrhea in PCOS is thought to be caused by inappropriate levels of both female and male hormones. Male hormones are produced in small quantities by all women, but in women with PCOS, levels of male hormone (androgens) are slightly higher than in other women. Some researchers hypothesize that the ovaries of women with PCOS are abnormal in other respects. In 2003, a group of researchers in London reported that there are fundamental differences between the development of egg follicles in normal ovaries and follicle development in the ovaries of women with PCOS.

In athletes, models, actresses, dancers, and women with anorexia nervosa, oligomenorrhea occurs because body fat drops too low compared to weight. Emotional stress related to performance anxiety may also be a factor in oligomenorrhea in these women.

Symptoms

Women with oligomenorrhea may have the following symptoms:

- menstrual periods at intervals of more than 35 days
- unusually light menstrual flow
- irregular menstrual periods with unpredictable flow
- difficulty conceiving

Young women whose oligomenorrhea is associated with the female athlete triad may have such other symptoms of the triad as frequent stress fractures, particularly in the bones of the hips, spine, or lower legs; abnormal eating patterns or extremely restrictive diets; and abnormal heart rhythms or low blood pressure.

When to call the doctor

A young woman should see her doctor as soon as she notices that a previously regular menstrual pattern has become irregular; it is not necessary to wait six months or longer to have oligomenorrhea investigated. A common rule is to consult the doctor after three missed periods.

Diagnosis

History and physical examination

Diagnosis of oligomenorrhea begins with the patient informing the doctor about infrequent periods. The doctor will ask for a detailed description of the problem and take a history of how long it has existed and any patterns the patient has observed. A woman can assist the doctor in diagnosing the cause of oligomenorrhea by keeping a record of the time, frequency, length, and quantity of bleeding. She should also tell the doctor about any recent illnesses, including longstanding conditions such as **diabetes mellitus**. The doctor may also inquire about the patient's diet, exercise patterns, sexual activity, contraceptive use, current medications, or past surgical procedures.

The doctor will then perform a physical examination to evaluate the patient's weight in proportion to her height, to check for signs of normal sexual development, to make sure the heart rhythm and other vital signs are normal, and to palpate (feel) the thyroid gland for evidence of swelling.

In the case of female athletes, the doctor may need to establish a relationship of trust with the patient before asking about such matters as diet, practice and workout schedules, and the use of such drugs as steroids or ephedrine. The presence of stress fractures in young women should be investigated. In some cases, the doctor may give the patients the Eating Disorder Inventory (EDI) or a similar screening questionnaire to help determine whether the patient is at risk for developing anorexia or bulimia.

Laboratory tests

After taking the young woman's history, the gynecologist or **family** practitioner does a pelvic examination and Pap smear. To rule out specific causes of oligomenorrhea, the doctor may also order a pregnancy test in sexually active women and blood tests to check the level of thyroid hormone. Based on the initial test results, the doctor may want to perform additional tests to determine the level of other hormones that play a role in reproduction.

As of 2003, more sensitive monoclonal assays had been developed for measuring hormone levels in the blood serum of women with PCOS, thus allowing earlier and more accurate diagnosis.

Imaging studies

In some cases the doctor may order an ultrasound study of the pelvic region to check for anatomical abnormalities or x rays or a bone scan to check for bone fractures. In a few cases the doctor may order an MRI to rule out tumors affecting the hypothalamus or pituitary gland.

Treatment

Treatment of oligomenorrhea depends on the cause. In adolescents and women near menopause, oligomenorrhea usually needs no treatment. For some athletes, changes in training routines and eating habits may be enough to return the woman to a regular menstrual cycle.

Most patients suffering from oligomenorrhea are treated with birth control pills. Other women, including those with PCOS, are treated with hormones. Prescribed hormones depend on which particular hormones are deficient or out of balance. When oligomenorrhea is associated with an eating disorder or the female athlete triad, the underlying condition must be treated. Consultation with a psychiatrist and nutritionist is usually necessary to manage an eating disorder. Female athletes may require physical therapy or rehabilitation as well.

Alternative treatment

As with conventional medical treatments, alternative treatments are based on the cause of the condition. If a hormonal imbalance is revealed by laboratory testing, hormone replacements that are more "natural" for the body (including tri-estrogen and natural progesterone) are recommended. Glandular therapy can assist in bringing about a balance in the glands involved in the reproductive cycle, including the hypothalamus, pituitary, thyroid, ovarian, and adrenal glands.

Since homeopathy and acupuncture work on deep, energetic levels to rebalance the body, these two forms of therapy may be helpful in treating oligomenorrhea. Western and Chinese herbal medicines also can be very effective. Herbs used to treat oligomenorrhea include dong quai (*Angelica sinensis*), black cohosh (*Cimicifuga racemosa*), and chaste tree (*Vitex agnus-castus*). Herbal preparations used to bring on the menstrual period are known as emmenagogues. For some women, meditation, guided imagery, and visualization can play a role in the treatment of oligomenorrhea by relieving emotional stress.

Nutritional concerns

Diet and adequate **nutrition**, including adequate protein, essential fatty acids, whole grains, and fresh fruits and vegetables are important for every woman, especially if deficiencies are present or if she regularly exercises very strenuously. Female athletes at the high school or college level should consult a nutritionist to make sure that they are eating a well-balanced diet that is adequate to maintain a healthy weight for their height. Girls participating in dance or in sports that emphasize weight control or a slender body type (gymnastics, track and field, swimming, and cheerleading) are at higher risk of developing eating disorders than those that are involved in such sports as softball, weight lifting, or basketball. In some cases the athlete may be given calcium or vitamin D supplements to lower the risk of osteoporosis.

Prognosis

Many women, including those with PCOS, are successfully treated with hormones for oligomenorrhea. They have more frequent periods and begin ovulating during their menstrual cycle, restoring their fertility.

For women who do not respond to hormones or who continue to have an underlying condition that causes oligomenorrhea, the outlook is less positive. Women who have oligomenorrhea as teenagers may have difficulty becoming pregnant and may receive fertility drugs. The absence of adequate estrogen increases the risk of osteoporosis, repeated bone fractures, and cardiovascular disease in later life. Female athletes who develop bone loss or osteoporosis in their late teens or early twenties are at increased risk of developing arthritis as they grow older. Women who do not have regular periods also are more likely to develop uterine **cancer**. Oligomenorrhea can become amenorrhea at any time, increasing the chance of having these complications.

KEY TERMS

Amenorrhea—The absence or abnormal stoppage of menstrual periods.

Anorexia nervosa—An eating disorder marked by an unrealistic fear of weight gain, self-starvation, and distortion of body image. It most commonly occurs in adolescent females.

Cyst—An abnormal sac or enclosed cavity in the body filled with liquid or partially solid material. Also refers to a protective, walled-off capsule in which an organism lies dormant.

Emmenagogue—A type of medication that brings on or increases a woman's menstrual flow.

Female athlete triad—A combination of disorders frequently found in female athletes that includes disordered eating, osteoporosis, and oligo- or amenorrhea. The triad was first officially named in 1993.

Osteoporosis—Literally meaning "porous bones," this condition occurs when bones lose an excessive amount of their protein and mineral content, particularly calcium. Over time, bone mass and strength are reduced leading to increased risk of fractures.

Prevention

Oligomenorrhea is preventable only in women whose low body fat to weight ratio is keeping them from maintaining a regular menstrual cycle. Adequate nutrition and less vigorous training schedules for female athletes will normally prevent oligomenorrhea. When oligomenorrhea is caused by hormonal factors, however, it is not preventable, but is usually treatable.

Parental concerns

Oligomenorrhea in teenagers who have only recently begun to menstruate is not usually a cause for parental concern, particularly if the girl's development during **puberty** has been otherwise normal or if there is a family history of oligomenorrhea. Oligomenorrhea in an adolescent should be investigated, however, if the girl is heavily involved in athletics or if she is otherwise at risk for developing an eating disorder. One way that parents can help college-age athletes is to be affectionate and emotionally supportive of their daughter, as girls who are away from home for the first time or who are "loners" are particularly at risk for developing the female athlete triad during their freshman year.

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Omphalocele *see* **Abdominal wall defects**

Oppositional defiant disorder

Definition

Oppositional defiant disorder (ODD) is a childhood mental disorder characterized by a pattern of angry, antagonistic, hostile, negative, irritable, and/or vindictive behavior lasting at least six months and occurring more frequently than is typically observed for the child's age and developmental stage. Children diagnosed with ODD do not meet the clinical diagnostic criteria for **conduct disorder**.

Description

Oppositional defiant disorder (ODD), a relatively new clinical classification, involves an ongoing pattern of antagonistic, defiant, and hostile behavior toward parents and other authority figures. Children and adolescents with ODD often have frequent temper **tantrums**, blame others for their misbehavior, argue excessively with adults, actively refuse to comply with adult rules and requests, deliberately defy adults and attempt to annoy or upset them, and are easily annoyed by others.

Demographics

Before **puberty**, ODD is more common in boys than girls; however, after puberty ODD occurrence rates are about equal in boys and girls. The disorder typically begins by the age of eight. According to the American Academy of Child and Adolescent Psychiatry, approximately 5 to 15 percent of all school-aged children have ODD.

Causes and symptoms

Although the specific causes of ODD are unknown, genetics and environment are thought to play a role in its development. As of 2004 several theories about the causes of oppositional defiant disorder are being investigated. ODD may be related to the following:

- the child's **temperament** and the family's response to that temperament
- an inherited predisposition to the disorder in certain families
- a neurological cause, such as a head injury
- a chemical imbalance in the brain (especially with the brain chemical serotonin)

ODD appears to be more common in families in which at least one parent has a history of a mood disorder, conduct disorder, attention deficit hyperactivity disorder (ADHD), **antisocial personality disorder**, or a substance abuse-related disorder. Children with one parent who is alcoholic or who has been in trouble with the law are almost three times more likely to have ODD. Additionally, some studies suggest that mothers with a depressive disorder are more likely to have children that develop ODD. ODD can also occur in conjunction with other conditions such as ADHD, learning disabilities, **anxiety** disorders, and **mood disorders**. About 50 percent to 65 percent of children with ADHD also have ODD.

Symptoms of ODD include a pattern of negative, hostile, and defiant behavior lasting at least six months. During this time four or more of the following must be present for a child to be diagnosed with ODD:

- often loses his/her temper
- often argues with adults
- often actively defies or refuses to comply with adults' requests or rules
- often deliberately annoys people
- often blames others for his/her mistakes or misbehavior
- is often touchy or easily annoyed by others
- is often angry and resentful
- is often spiteful or vindictive
- misbehaves frequently
- swears or uses obscene language
- has a low opinion of him/herself

Additional problems may be present, including the following:

- learning problems
- a depressed mood
- hyperactivity (although ADHD must be ruled out)

- substance abuse or dependence
- dramatic and erratic behavior

When to call the doctor

Parents of children and adolescents who exhibit symptoms of ODD should see a physician as soon as possible. Usually, a referral to a psychologist, psychiatrist, or therapist will be given.

Diagnosis

ODD is diagnosed by psychological and psychiatric evaluations; interviews with **family** members, teachers, and caregivers; and observation and interviews with the child or adolescent. Diagnosis is based on clinical criteria defined in the American Psychiatric Association's *Diagnostic and Statistical Manual of Mental Disorders*, fourth edition, text revision C (DSM-IV-TR).

ODD often has characteristics in common with other psychological disorders and often occurs in conjunction with other conditions, such as ADHD or mood disorders. Therefore, diagnosis of ODD usually depends on exclusion of other disorders. A diagnosis of ODD is not made if the symptoms occur exclusively in psychotic or mood disorders, or if the child meets clinical criteria for conduct disorder, or, if the adolescent is 18 years old or older and meets clinical criteria for antisocial personality disorder. Children and adolescents with ODD do not exhibit the more serious aggressive behaviors or physical cruelty that is common in other disorders.

Treatment

Treatment of ODD usually involves medication, and group, individual, and/or **family therapy**. Of these, individual therapy is the most common. The goal of therapy is to help provide a consistent daily schedule, support, rules, **discipline**, and limits, as well as to help train patients to get along with others by modifying behaviors. Therapy can occur in residential or day treatment facilities, in a medical setting, or on an outpatient basis. Therapy can instruct patients on how to effectively deal with ODD and help them learn how to do the following:

- use self time-outs
- identify what increases anxiety
- talk about feelings instead of acting on them
- find and use ways to calm themselves
- frequently remind themselves of their goals
- get involved in tasks and physical activities that provide a healthy outlet for energy
- learn how to talk with others
- develop a predictable, consistent, daily schedule of activity
- develop ways to obtain pleasure and feel good
- learn how to get along with other people
- find ways to limit stimulation
- learn to admit mistakes in a matter-of-fact way

Therapy can also involve the parents. Parent management training focuses on teaching parents specific and more effective techniques for handling the child's opposition and defiance. Research has shown that parent management training is more effective than family therapy.

Stimulant medication is used only when ODD co-occurs with ADHD. Occasionally, children and adolescents with ODD may also have depression or anxiety disorders, and treatment with **antidepressants** and anti-anxiety medications can help alleviate some symptoms of ODD.

Prognosis

The prognosis for ODD varies. In some children, ODD evolves into a conduct disorder or a mood disorder. ODD, if left untreated, has approximately an 80 percent chance of turning into conduct disorder as a child ages. Later in life, ODD can develop into passive-aggressive personality disorder or antisocial personality disorder. ODD can cause significant social, academic, and/or occupational impairment. Generally, with treatment and long-term participation in therapy, adjustment in social settings and in the workplace can be made in adulthood.

Prevention

As of 2004, ODD could not be prevented.

Parental concerns

Children and adolescents with ODD usually have difficulties in school and at home. In some cases, ODD can result in expulsion from school. Parents should investigate **alternative school** settings that may be able to provide counseling and group therapy integrated with academics. Assistance is available through county social or mental health services, educational consultants, and local school counselors. Family therapy may help alleviate stressful family situations and help other family members understand the disorder.

KEY TERMS

Alternative school—An educational setting designed to accommodate educational, behavioral, and/or medical needs of children and adolescents that cannot be adequately addressed in a traditional school environment.

Antisocial personality disorder—A disorder characterized by a behavior pattern that disregards for the rights of others. People with this disorder often deceive and manipulate, or their behavior might include aggression to people or animals or property destruction, for example. This disorder has also been called sociopathy.

Attention deficit hyperactivity disorder (ADHD)—A condition in which a person (usually a child) has an unusually high activity level and a short attention span. People with the disorder may act impulsively and may have learning and behavioral problems.

Conduct disorder—A behavioral and emotional disorder of childhood and adolescence. Children with a conduct disorder act inappropriately, infringe on the rights of others, and violate societal norms.

Television viewing and video/computer games can contribute to ODD behaviors. For children with ADHD or ODD, the American Academy of Pediatrics recommends limiting use of television and video/computer games to no more than two hours per day, monitoring children's use of television and computers, and viewing family-oriented television programs with their children.

Parents may find it helpful to track their child's moods and behaviors and to help children learn to track their own moods and behaviors to help identify possible stresses and causative factors.

Parents should actively participate in their child's therapy and learn positive parenting techniques that can help ODD behaviors. When parents are too restrictive, children and adolescents with ODD can rebel, and power struggles can frequently occur. Therapists specializing in ODD can help families become more effective in handling ODD behaviors in order to avoid such rebellion. The American Academy of Child and Adolescent Psychiatry recommends the following for parents with children who have ODD:

- Choose battles by setting priorities regarding child's behavior.
- Set reasonable, age-appropriate limits with consistently enforceable consequences.
- Work with teachers, coaches, and other family members for support in dealing with the child with ODD.
- Use positive reinforcement praise when the child displays desired behaviors.
- Take time to manage stress by exercising and/or relaxing away from the child.

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Oral contraceptives

Definition

Oral contraceptives are medicines taken by mouth to help prevent pregnancy. They are also known as the pill, OCs, or birth control pills.

Description

Oral contraceptives, or birth control pills, contain synthetic forms of two hormones produced naturally in the body. These hormones, estrogen and progestin, regulate the female menstrual cycle. Some types of oral contraceptives use only progestational hormones, but most use a combination of estrogen and progestin. As of 2004, there were three types of oral contraceptives marketed:

- Monophasic use a fixed dose of both estrogen and progestin during the entire cycle.
- Biphasic oral contraceptives use a constant amount of estrogen during the full cycle, but the amount of progestin is lower during the first half of the cycle and increases in the second half. This shift in dosage is intended to mimic the natural ovarian cycle.
- Triphasic oral contraceptives may vary both the estrogen and progestin levels at different times during the cycle.

The goal of the biphasic and triphasic formulations is to achieve adequate control of the menstrual cycle while using lower doses of both estrogens and progestins, thereby reducing the risk of adverse effects. Reviews of controlled studies have not demonstrated a clear advantage of the newer formulations over the older monophasic drugs.

General use

When taken in the proper amounts, following a specific schedule, oral contraceptives are very effective in preventing pregnancy. Studies show that fewer than one of every 100 females who use oral contraceptives correctly becomes pregnant during the first year of use.

These pills have several effects that help prevent pregnancy. For pregnancy to occur, an egg must become mature inside a woman's ovary, be released, and travel to the fallopian tube. Sperm must travel through the reproductive track to fertilize the egg in the fallopian tube. Then the fertilized egg must travel to the woman's uterus (womb), where it lodges in the uterus lining and develops into a fetus.

The main way that oral contraceptives prevent pregnancy is by keeping an egg from ripening fully. Eggs that do not ripen fully cannot be fertilized. In addition, birth control pills thicken mucus in the woman's body through which the sperm has to swim. Thus it is more difficult for the sperm to reach the egg. Oral contraceptives also change the uterine lining so that a fertilized egg cannot lodge there to develop.

Although **contraception** is the primary use of these medications, they may also be used to treat adolescent and post-adolescent **acne** in girls. Some products have this as part of their official indications, but others may be used as well.

Precautions

No form of birth control (except abstinence from sexual intercourse) is 100 percent effective. However, oral contraceptives can be highly effective when used properly. Teens and young women who anticipate having sexual intercourse should discuss the options with a healthcare professional.

Oral contraceptives do not protect against **AIDS** or other **sexually transmitted diseases**. For some protection against such diseases, teenage males and young men need to use a latex **condom**. Also, oral contraceptives are not effective immediately after a young woman begins taking them. Physicians recommend using other forms of birth control for the first one to three weeks. Then users should follow the instructions of the physician who prescribed the medicine.

Smoking cigarettes while taking oral contraceptives greatly increases the risk of serious side effects. Females who take oral contraceptives should not smoke cigarettes.

Seeing a physician regularly while taking this medicine is very important. The physician will note unwanted side effects, and patients should follow his or her advice on how often they should be seen.

Young women who take oral contraceptives should be sure to tell the healthcare professional in charge before they undergo surgical or dental procedures, laboratory tests, or emergency treatment.

This medicine may increase sensitivity to sunlight. Females using oral contraceptives should avoid too much sun exposure and should not use tanning beds, tanning booths, or sunlamps until they know how the medicine affects them. Some females taking oral contraceptives may get brown splotches on exposed areas of their skin. These usually go away over time after the women stop taking birth control pills.

Oral contraceptives may cause the gums to become tender and swollen or to bleed. Careful brushing and flossing, gum massage, and regular cleaning may help prevent this problem. Users should check with a physician or dentist if gum problems develop.

Side effects

Serious side effects are rare in healthy females who do not smoke cigarettes. In women with certain health problems, however, oral contraceptives may cause problems such as liver **cancer**, noncancerous liver tumors, blood clots, or **stroke**. Healthcare professionals can help prospective users weigh the benefits of being protected against unwanted pregnancy against the risks of possible health problems.

The most common minor side effects are **nausea**, **vomiting**, abdominal cramping or bloating, breast **pain**, tenderness or swelling, swollen ankles or feet, tiredness, and acne. These problems usually go away as the body adjusts to the drug and do not need medical attention unless they continue or they interfere with normal activities. Other side effects should be brought to the attention of the physician who prescribed the medicine. Teens and young women should check with the physician as soon as possible if any of the following side effects occur:

- menstrual changes, such as lighter periods or missed periods, longer periods, or bleeding or spotting between periods
- headaches
- vaginal infection, **itching**, or irritation
- increased blood pressure

Women who have any of the following symptoms should get emergency help right away. These symptoms may be signs of blood clots:

- sudden changes in vision, speech, breathing, or coordination
- severe or sudden **headache**
- coughing up blood
- sudden, severe, or continuing pain in the abdomen or stomach
- pain in the chest, groin, or leg (especially in the calf)
- weakness, **numbness**, or pain in an arm or leg

The adverse effects of oral contraceptives can be impossible to predict. Other than avoiding smoking, there are no effective means of preventing side effects. All observed adverse effects should be reported to a physician promptly.

Oral contraceptives may continue to affect the menstrual cycle for some time after a young woman stops taking them. Women who miss periods for several months after stopping this medicine should check with their physicians. Other rare side effects may occur. Anyone who has unusual symptoms while taking oral contraceptives should get in touch with her physician.

A doctor explains to a teenage girl how to use birth control pills. *(© LWA-Stephen Welstead/Corbis.)*

Interactions

Oral contraceptives may interact with a number of other medicines. When interaction occurs, the effects of one or both of the drugs may change or the risk of side effects may be greater. Anyone who takes oral contraceptives should let the physician know all other medicines she is taking and should ask whether possible interactions can interfere with drug therapy.

These drugs may make oral contraceptives less effective in preventing pregnancy. Anyone who takes these drugs should use an additional birth control method for the entire cycle in which the medicine is used:

- ampicillin
- penicillin V
- rifampin (Rifadin)
- tetracyclines
- griseofulvin (Gris-PEG, Fulvicin)

KEY TERMS

Cyst—An abnormal sac or enclosed cavity in the body filled with liquid or partially solid material. Also refers to a protective, walled-off capsule in which an organism lies dormant.

Endometriosis—A condition in which the tissue that normally lines the uterus (endometrium) grows in other areas of the body, causing pain, irregular bleeding, and frequently, infertility.

Fallopian tubes—The pair of narrow tubes leading from a woman's ovaries to the uterus. After an egg is released from the ovary during ovulation, fertilization (the union of sperm and egg) normally occurs in the fallopian tubes.

Fetus—In humans, the developing organism from the end of the eighth week to the moment of birth. Until the end of the eighth week the developing organism is called an embryo.

Fibroid tumor—A non-cancerous tumor of connective tissue made of elongated, threadlike structures, or fibers, which usually grow slowly and are contained within an irregular shape. Fibroids are firm in consistency but may become painful if they start to break down or apply pressure to areas within the body. They frequently occur in the uterus and are generally left alone unless growing rapidly or causing other problems. Surgery is needed to remove fibroids.

Hormone—A chemical messenger secreted by a gland or organ and released into the bloodstream. It travels via the bloodstream to distant cells where it exerts an effect.

Jaundice—A condition in which the skin and whites of the eyes take on a yellowish color due to an increase of bilirubin (a compound produced by the liver) in the blood. Also called icterus.

Migraine—A throbbing headache that usually affects only one side of the head. Nausea, vomiting, increased sensitivity to light, and other symptoms often accompany a migraine.

Mucus—The thick fluid produced by the mucous membranes that line many body cavities and structures. It contains mucin, white blood cells, water, inorganic salts, and shed cells, and it serve to lubricate body parts and to trap particles of dirt or other contaminants.

Ovary—One of the two almond-shaped glands in the female reproductive system responsible for producing eggs and the sex hormones estrogen and progesterone.

Pelvic inflammatory disease (PID)—Any infection of the lower female reproductive tract (vagina and cervix) that spreads to the upper female reproductive tract (uterus, fallopian tubes and ovaries). Symptoms include severe abdominal pain, high fever, and vaginal discharge. PID is the most common and most serious consequence of infection with sexually transmitted diseases in women and is a leading cause of female fertility problems.

Uterus—The female reproductive organ that contains and nourishes a fetus from implantation until birth. Also called the womb.

- corticosteroids
- barbiturates
- carbamazepine (Tegretol)
- phenytoin (Dilantin)
- primidone (Mysoline)
- ritonavir (Norvir)

In addition, taking the following medicines with oral contraceptives may increase the risk of side effects or interfere with the medicine's effects:

- Theophylline: Effects of this medicine may increase, along with the chance of unwanted side effects.
- Cyclosporine: Effects of this medicine may increase, along with the chance of unwanted side effects.
- Troleandomycin (TAO): Chance of liver problems may increase. Effectiveness of oral contraceptive may also decrease, raising the risk of pregnancy.

The list above does not include every drug that may interact with oral contraceptives. Women should be sure to check with a physician or pharmacist before combining oral contraceptives with any other prescription or nonprescription (over-the-counter) medicine. As with any medication, the benefits and risks should be discussed with a physician.

Parental concerns

Parents become concerned that teens who use oral contraceptives are at risk of becoming sexually active. Although studies have been limited, they have failed to

show that availability of oral contraceptives leads to an increase in sexual activity among adolescent girls.

Oral contraceptives do not protect against sexually transmitted diseases. When used for contraception, they should be limited to monogamous relationships.

Although the list of potential side effects and adverse effects is very long and contains some severe risks, the actual frequency of these risks is low. In most cases, oral contraceptives have a very high safety margin.

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Oral herpes *see* **Cold sore**

Oral hygiene

Definition

Oral hygiene is the practice of keeping the mouth clean and healthy by brushing and flossing to prevent **tooth decay** and gum disease.

Purpose

The purpose of oral hygiene is to prevent the build-up of plaque, the sticky film of bacteria and food that forms on the teeth. Plaque adheres to the crevices and fissures of the teeth and generates acids that, when not removed on a regular basis, slowly eat away, or decay, the protective enamel surface of the teeth, causing holes (cavities) to form. Plaque also irritates gums and can lead to gum disease, **periodontal disease**, and tooth loss. Brushing and flossing removes plaque from teeth, and antiseptic mouthwashes kill some of the bacteria that help form plaque. Fluoride, found in toothpaste, drinking water, or dental treatments, also helps to protect teeth by binding with enamel to make it stronger. In addition to such daily oral care, regular visits to the dentist promote oral health. Preventative services that the dentist can perform include fluoride treatments, sealant application, and scaling (scraping off the hardened plaque, called tartar). The dentist can also perform such diagnostic services as x-ray imaging and such treatments as filling cavities.

Description

The Centers for Disease Control and Prevention report that dental caries are perhaps the most prevalent of infectious diseases in children. More than 40 percent of all children have cavities by the time they reach kindergarten. It is, therefore, imperative that all parents learn the importance of early oral care and that they teach their children proper oral hygiene.

Good oral hygiene should start at the very beginning of a child's life. Even before his or her first teeth emerge, certain factors can affect their future appearance and health. Pregnant and nursing mothers should be careful about using medications, as some, like the antibiotic tetracycline, can cause tooth discoloration. Even before infants have teeth, they have special oral hygiene needs about which all parents should be aware. These include making certain the child receives adequate fluoride and guarding against baby bottle decay.

Fluoride in infancy

Fluoride is beneficial for babies even before their teeth erupt. It makes the tooth enamel stronger as the teeth are developing. In most municipal water supplies, the correct amount of fluoride is added for proper tooth development. If the water supply does not contain enough fluoride or if bottled water is used for drinking and cooking, the doctor or dentist should be informed. They may prescribe fluoride supplements for the baby.

Baby bottle decay

Baby bottle decay is caused by recurring exposure over time to sugary liquids. These include milk, formula, and fruit juices. These liquids pool for prolonged periods of time as the child sleeps. This exposure can lead to cavities forming, especially in the upper and lower front teeth. For this reason, children should not be allowed to fall asleep with a bottle of juice or milk in their mouths. An alternative is to give the child a bottle filled with water or a pacifier recommended by the dentist. Even breast-fed children are at risk. They should have their gums and teeth wiped with a clean, damp washcloth or gauze pad following each feeding.

Baby teeth, also known as primary teeth, are just as important as permanent teeth. They help the child to bite and chew food, help them speak correctly, save space for the child's permanent teeth, and help guide the permanent teeth into place. That is why it is so important to initiate a program of good oral hygiene for children early on.

Brushing

Once a baby has four teeth in a row, either on top or on the bottom, parents should begin using a toothbrush two times a day. When choosing a toothbrush, make sure the bristles are soft, polished, and made of nylon. Parents should administer only a pea-size amount of fluoride toothpaste that is made especially for children. Children tend to swallow, instead of spit out, toothpaste. If the child does not like the flavor of the toothpaste, using water alone is acceptable. Parents should also continue to wipe the toothless gum areas with a washcloth or gauze.

As the child gets older, parents should demonstrate proper brushing techniques. These include brushing the inside surface of each tooth first, where plaque tends to accumulate most. Then they should clean the outer surfaces of each tooth, angling the brush along the outer gum line. Next, they should brush the chewing surface of each tooth, then using the tip of the brush, clean behind each front tooth. They should use a gentle, back and forth motion when brushing and finish by brushing the tongue.

Children will, at some point, decide they would like to try brushing their teeth themselves. This is fine and should be encouraged, but parents should remain in charge of keeping children's teeth clean until they are between six to eight years old. Children do not have the dexterity or coordination to perform brushing well until this time. Even then, it is important that parents inspect their children's teeth each time they brush. They should pay special attention to the molars, as these teeth have lots of tiny grooves and crevices where food particles can hide.

A boy flosses his teeth as part of a daily regimen for good oral hygiene. *(© Joh Feingersh/Corbis.)*

Good oral hygiene remains important as children grow into **adolescence**. In fact, adolescence can often be a time when cavities and periodontal disease happen more frequently. This higher rate is usually caused by an increased intake of junk food and sugary foods such as soft drinks, as well as inattention to oral hygiene procedures. Add to that the fact that many older children and teens wear braces, making the cleaning of teeth even more challenging. Parents should talk to their children about how important good oral hygiene is in preventing not only cavities, but teeth stains, bad breath, and an assortment of other dental problems.

Flossing

Flossing once a day helps to prevent gum disease by removing food particles and plaque at and below the gum line, as well as between teeth. Parents do not need to initiate flossing until the child has teeth that touch each other, which normally occurs in the molar areas first. Parents should continue to floss their child's teeth until they are six or seven years old. They should continue to monitor the child's techniques and consistency thereafter.

Proper flossing technique is essential in removing as much plaque as possible in a safe manner. The following procedure is recommended by dental hygienists. Wind 18 inches (45 cm) of dental floss around the middle fingers of each hand. Pinch the floss between the thumbs and index fingers, leaving about 1–2 inches (3–5 cm) length in between. Use the thumbs to direct the floss between the upper teeth. Try to keep the floss taut

between the fingers. Use the index fingers to guide floss between lower teeth. Gently guide the floss between the teeth by using a zig-zag motion. Contour the floss around the side of each tooth. Slide the dental floss up and down against the tooth surface and under the gum line. Floss each tooth thoroughly with a clean section of floss.

Dental floss comes in many varieties (waxed, unwaxed, flavored, tape) and may be chosen based on personal preference. For those who have difficulty handling floss, floss holders and other types of interdental (between the teeth) cleaning aids are available. Some floss holders have animal and cartoon characters on them, which might make flossing more appealing to a child.

Precautions

It is important that younger children only use a very small amount of fluoridated toothpaste since using too much fluoride can be toxic to infants. Though brushing and flossing are important, neither should be performed too vigorously. The rough mechanical action may irritate or damage oral tissues. Parents should change their child's toothbrush three to four times a year and after every illness to avoid bacteria and germs.

Another factor that may affect a child's oral health is the increasingly popular practice among adolescents of oral piercings involving the tongue, lips, and cheeks. These piercings have been associated with infections, tooth fractures, periodontal disease, and nerve damage. Some life-threatening complications have occurred, including bleeding and airway obstruction. The American Academy of Pediatric Dentistry strongly opposes the practice of oral piercings.

Risks

The primary risks arise from a lack of proper oral hygiene practices. These major oral health problems are plaque, tartar, gingivitis, periodontitis, and tooth decay.

Parental concerns

Parents play an important role in both modeling and teaching good oral hygiene. Parents can make sure their child sees a dentist before the age of two. This can be a frightening experience for some children, but if parents exhibit a positive attitude, most children become comfortable with dentist visits. Children who learn proper oral care at a young age benefit from those good habits for the rest of their lives.

KEY TERMS

Calculus—Plural, calculi. Any type of hard concretion (stone) in the body, but usually found in the gallbladder, pancreas, and kidneys. They are formed by the accumulation of excess mineral salts and other organic material such as blood or mucous. Calculi (pl.) can cause problems by lodging in and obstructing the proper flow of fluids, such as bile to the intestines or urine to the bladder. In dentistry, calculus refers to a hardened yellow or brown mineral deposit from unremoved plaque, also called tartar.

Cavity—A hole or weak spot in the tooth surface caused by decay.

Gingivitis—Inflammation of the gums in which the margins of the gums near the teeth are red, puffy, and bleeding. It is most often due to poor dental hygiene.

Pediatric dentistry—The dental specialty concerned with the dental treatment of children and adolescents.

Plaque—A deposit, usually of fatty material, on the inside wall of a blood vessel. Also refers to a small, round demyelinated area that develops in the brain and spinal cord of an individual with multiple sclerosis.

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Orthodontics

Definition

Orthodontics is a specialized branch of dentistry that diagnoses, prevents, and treats dental and facial irregularities called malocclusions. Orthodontics includes dentofacial orthopedics, which is used to correct problems involving the growth of the jaw.

Purpose

Humans have attempted to straighten teeth for thousands of years before orthodontics became a dental specialty in 1900. Although orthodontic treatment often improves facial appearance and occasionally is performed for solely cosmetic reasons, it is used primarily to correct health problems and to ensure the proper functioning of the mouth. Properly aligned teeth, which close together correctly, simplify **oral hygiene** and enable children to chew their food efficiently. Orthodontic treatment provides the following:

- straightens teeth that are rotated, tilted, or otherwise improperly aligned
- corrects crowded or unevenly spaced teeth
- corrects bite problems
- aligns the upper and lower jaws

Malocclusions

Few children have perfectly symmetrical teeth and a perfect bite. In an ideal bite, the following are characteristics:

- All of the teeth fit easily without crowding or spacing.
- The teeth are not rotated, twisted, or leaning forward or backward.
- The teeth of the upper jaw slightly overlap those of the lower jaw.
- The points of the molars fit into the grooves of the opposite molars.

Types of malocclusions include the following:

- crowded, crooked, or misaligned teeth
- extra or missing teeth
- bite problems
- jaws that are out of alignment

Causes of malocclusion

Most malocclusions are caused by hereditary factors that affect the contours of the face and the size of the teeth and jaw. The most common cause of **malocclusion** is a disproportion in size between the jaw and teeth or between the upper and lower jaws. A child who inherits a mother's small jaw and a father's large teeth may have teeth that are too big for the jaw, causing overcrowding. Specific inherited malocclusions include:

- overcrowded teeth
- too much space between teeth
- extra or missing teeth
- various irregularities in the teeth, jaw, or face

Malocclusions can be acquired through the following:

- accidents such as a jaw fracture that causes misalignment
- prolonged sucking on thumbs, fingers, or pacifiers, particularly after the age of four
- fingernail or lip biting
- a lost tooth that causes nearby teeth to move into the empty space, throwing them out of alignment
- airways that are obstructed by tonsils or adenoids
- dental disease
- tumors in the mouth or jaw
- improperly fitted fillings, crowns, or braces
- premature loss of baby teeth or permanent teeth
- late loss of baby teeth

Symptoms of malocclusion

Occasionally children have mild, temporary symptoms of malocclusion resulting from a growth spurt. However, symptoms of malocclusion usually develop gradually beginning at the age of six. Symptoms may include the following:

- crowded or misaligned teeth
- abnormal spacing between teeth, most often occurring because teeth are small or missing or the dental arch—

the arch-shaped jawbone that supports the teeth—is very wide
- incisors (front teeth) that do not meet
- an open bite, occurring when the upper and lower incisors do not touch each other during biting, thereby putting all of the chewing pressure on the back teeth and resulting in inefficient chewing and excessive tooth wear
- an overbite or overjet, in which the upper incisors protrude, often caused by a lower jaw that is significantly shorter than the upper jaw
- a deep or closed bite, an excessive overbite in which the lower incisors bite too closely to or into the gum tissue or palate behind the upper teeth
- a crossbite, in which a protruding lower jaw that is longer than the upper jaw causes the upper front or back teeth to bite inside the lower teeth

Early intervention

Although orthodontic treatment can be performed at any age, children are easier, faster, and less expensive to treat than adults. Most often orthodontic treatment is used on older children and adolescents whose teeth are still developing. However some types of problems are corrected more readily before all of the permanent teeth have erupted and facial growth is complete. If a child's permanent lower incisors erupt behind each other, braces may be required at a young age. Crossbites are usually treated early because they can interfere with biting and chewing. Early treatment also is used when thumb- or finger-sucking has affected teeth positioning.

Early orthodontic intervention can provide the following:

- straighten crooked teeth
- preserve or create space for incoming permanent teeth
- guide erupting permanent teeth into the correct positions
- prevent impacted permanent teeth, those that remain partially covered by gum tissue or partially or completely buried in the jawbone
- correct harmful habits such as thumb- or finger-sucking
- lower the risk of accidents to protruding upper incisors

Other advantages of early orthodontic treatment include the following:

- correction of bite problems by guiding jaw growth and controlling the width of the upper and lower dental arches
- reduction or elimination of abnormal swallowing or speech problems
- shortening and simplification of later orthodontic treatment
- prevention of later tooth extractions
- improvements in appearance and self-esteem

Untreated malocclusions

Minor misalignment or crowding may not require treatment. However untreated malocclusions can cause the following:

- teeth that are partially impacted or fail to erupt
- lips, tongue, or cheeks that contact biting surfaces due to poor tooth alignment
- inefficient or uncomfortable biting, chewing, and digestion
- speech impairments
- teeth that are hard to clean, leading to cavities and gum disease
- abnormal wear of tooth surfaces
- chipped teeth
- loosening or fracturing of a misaligned tooth that is overstrained
- injury to a protruding upper incisor
- thinning and receding of bone and gums covering the roots of very crowded teeth
- accelerated gum disease and bone loss
- temporomandibular joint (TMJ) misalignments at the point where the lower jaw attaches to the skull
- stress and trauma to the teeth, gum tissue, ligaments, muscles, jawbone, and jaw joints
- premature loss of teeth
- adverse effects on facial development and appearance
- the need for surgery

Untreated malocclusions often worsen with time. TMJ problems can cause chronic headaches or **pain** in the face and neck. A deep overbite can cause significant pain and bone damage and may contribute to excessive wear on the incisors.

Description

Orthodontics in young children

Alignment problems usually become apparent as the permanent teeth begin erupting at about age six. Dentists monitor the development of a child's permanent teeth

and refer the child to an orthodontist if a problem is suspected. The American Association of Orthodontists recommends that all children be screened by an orthodontist by the age of seven.

Once a child's lower baby incisors have erupted, an orthodontist can measure the child's jaw and tooth size, project their growth rate, and possibly predict whether the child will have orthodontic problems with their permanent teeth. The orthodontist may be able to perform preventative or interceptive orthodontics that can reduce or eliminate the need for braces later.

In a procedure called selective serial extraction, the orthodontist removes one or more baby or permanent teeth. Doing so creates space for the permanent teeth, especially unerupted canine teeth that might become impacted or erupt in the wrong position. After the removal or loss of a tooth, braces or another orthodontic appliance may be used to prevent the remaining teeth from moving into the empty space. If a baby molar—that acts as a space-holder for later permanent teeth—is lost, a fixed orthodontic wire is inserted between the teeth to keep the space available.

Preparation

The orthodontist compiles pretreatment records that are used for diagnosis, determining the course of treatment, and measuring the progress of treatment. These records may include:

- a complete medical and dental history
- a clinical examination
- x rays revealing the positions of erupted and unerupted teeth, development of unerupted teeth, any missing or impacted teeth, shortened or damaged tooth roots, and the amount of bone supporting the teeth
- a facial-profile x ray or cephalometric film revealing the sizes, positions, and relationships of the teeth and jaw, as well as facial form, growth pattern, and the inclinations of tipped or tilted incisors
- plastic impressions of the bite and plaster models made from the impressions
- photographs and other measurements of the teeth and face

Based on the diagnosis the orthodontist develops a custom treatment plan and designs the appropriate corrective appliances that will gradually straighten or move the teeth. Severe overcrowding may necessitate the extraction of permanent teeth, usually the premolars, to create space prior to using braces to move teeth.

Braces and other orthodontic appliances

By applying constant gentle pressure in a specific direction, braces can slowly move teeth through the supporting bone to a new position. Springs and wires put pressure on teeth in order to straighten them. The pressure causes bone in the jaw to dissolve in front of the moving tooth as new bone grows behind the tooth. Braces and other appliances may be removable or fixed and are made of clear or colored metal, ceramic, or plastic. Removable appliances are often plastic plates that fit into the roof of the mouth and clip onto a tooth.

Fixed braces exert more pressure than removable braces and can achieve more complex movements. They consist of wires and springs that are held in place by small brackets glued to the outside surfaces of the incisors and sometimes the premolars. Lingual braces have brackets bonded to the back of the teeth. Bands encircling the molars also can be used for attachments. The wires, springs, and other devices attached to the brackets or bands put pressure on the teeth, gradually shifting them into new positions. The nickel-titanium wires are very light, and some are heat-activated. These are very flexible at room temperature and actively begin to move the teeth as they warm to body temperature. Elastic bands sometimes connect the upper and lower teeth to create tension.

Appliances used to direct jaw growth and development in growing children and adolescents include:

- Headgear attached to braces and usually worn for 10 to 12 hours at night puts pressure on the upper teeth and jaw and influences the direction and speed of upper jaw growth and upper teeth eruption.
- Herbst appliances attached to the upper and lower molars correct a severe overbite by holding the lower jaw forward, influencing jaw growth and tooth position; they force the jaw muscles to work in ways that promote forward development of the lower jaw; treatment with Herbst appliances must begin several years before the jaw stops growing and they must remain in place throughout the treatment.
- Palatal or upper jaw expansion devices can widen a narrow upper jaw and correct a crossbite within months.
- Removable bionators hold the lower jaw forward and guide tooth eruption while helping the upper and lower jaws to grow proportionately.

Headgear and Herbst appliances can significantly reduce protrusion of the four top incisors and enable the growing lower jaw to catch up with the upper jaw, eliminating swallowing problems.

Children with braces. *(Photograph by Robert J. Huffman/Field Mark Publications.)*

Duration of treatment

Orthodontic treatment usually continues until the desired outcome is reached. Active orthodontic treatment lasts an average of two years, with a range of one to three years. Some children respond to treatment faster than others and interceptive or early treatments may continue for only a few months. Appliances are adjusted periodically during treatment. Factors affecting the duration of treatment include:

- the growth of the mouth and face
- the severity of the problem
- the health of the teeth, gums, and supporting bones
- the child's level of cooperation

Precautions

Orthodontic appliances trap food, bacteria, and plaque, leading to **tooth decay**. Extra brushing with specially shaped and/or electric toothbrush and fluoride toothpaste is required around the areas where the braces or appliances attach to the teeth. Both the tops and bottoms of braces must be brushed and irrigated with a water jet directed from the top down and the bottom up. If possible, teeth should be flossed. A fluoride mouthwash may be recommended. Removable appliances should be brushed every time the teeth are brushed. Regular dental check-ups and cleanings must be continued.

Children with braces should eat raw fruits and vegetables and avoid soft, processed, and refined foods that attract bacteria, as well as hard or sticky foods, including gum, caramels, peanuts, ice chips, and popcorn. Chewing on hard items, such as fingernails or pencils, can damage braces. Children with braces should wear a protective mouth guard while playing contact **sports**.

Aftercare

After braces are removed the teeth must be stabilized in their new positions. This phase of treatment commonly takes two to three years. Occasionally it continues indefinitely. Types of retainers used for stabilization include:

KEY TERMS

Active treatment stage—The period during which orthodontic appliances or braces are used.

Bicuspid—Premolar; the two-cupped tooth between the first molar and the cuspid.

Canines—The two sharp teeth located next to the front incisor teeth in mammals that are used to grip and tear. Also called cuspids.

Crossbite—The condition in which the upper teeth bite inside the lower teeth.

Crown—The natural part of the tooth covered by enamel. A restorative crown is a protective shell that fits over a tooth.

Deep bite—A closed bite; a deep or excessive overbite in which the lower incisors bite too closely to or into the gum tissue or palate behind the upper teeth.

Eruption—The process of a tooth breaking through the gum tissue to grow into place in the mouth.

Impacted tooth—Any tooth that is prevented from reaching its normal position in the mouth by another tooth, bone, or soft tissue.

Incisors—The eight front teeth.

Interceptive orthodontics—Preventative orthodontics; early, simpler orthodontic treatment.

Malocclusion—The misalignment of opposing teeth in the upper and lower jaws.

Molars—The teeth behind the primary canines or the permanent premolars, with large crowns and broad chewing surfaces for grinding food.

Open bite—A malocclusion in which some teeth do not meet the opposing teeth.

Orthognatic surgery—Surgery to alter the relationships of the teeth and/or supporting bones, usually in conjunction with orthodontic treatment.

Overbite—Protrusion of the upper teeth over the lower teeth.

Plaque—A sticky film of saliva, food particles, and bacteria that attaches to the tooth surface and causes decay.

Retainer—An orthodontic appliance that is worn to stabilize teeth in a new position.

Retention treatment stage—The passive treatment period following orthodontic treatment, when retainers may be used to stabilize the teeth.

Temporomandibular joint (TMJ)—One of a pair of joints that attaches the mandible of the jaw to the temporal bone of the skull. It is a combination of a hinge and a gliding joint.

- positioners, rubber-like mouthpieces that are worn at night and bitten into for a few hours during the day
- removable retainers with a plastic plate that snaps onto the roof of the mouth and wires on the outside of the teeth
- removable, clear, plastic retainers that completely cover the sides and biting surfaces of the teeth
- semi-rigid wires that are bonded onto the inside of the incisors.

Risks

Braces may cause discomfort when they are first installed or adjusted during treatment. For the first three to five days teeth may hurt during biting. Lips, cheeks, and tongue may be irritated for one to two weeks before they toughen and adapt to the braces. Some appliances may interfere with speech for the first day or two. Damaged appliances can extend the length of treatment and negatively affect the outcome.

Food particles and plaque deposits around orthodontic appliances can cause demineralization of the tooth enamel, leading to cavities and permanent whitish scars on the teeth.

Normal results

Orthodontic treatment is usually very successful at correcting malocclusions. Even a significant size discrepancy between the upper and lower jaws often can be corrected. Sometimes, particularly in adults, corrective orthognathic surgery is required to shorten or lengthen a jawbone. The height of the lower face also can be shortened or lengthened. Sometimes surgery reduces the duration of the orthodontic treatment.

Maturational change can cause teeth to gradually shift with age—at least until one's early 20s—causing crowding. Nighttime retainers can prevent maturational movement.

Parental concerns

In general the earlier an orthodontic problem is detected, the easier and less expensive it is to correct. Parents can compare their child's **dental development** with standard charts and pictures.

When to call the doctor

Children with problems involving the width or length of the jaws should be evaluated no later than age 10 for girls and age 12 for boys. For children receiving orthodontic care, the orthodontist should be notified immediately if an appliance breaks. Indications that children may need an early orthodontic examination include:

- early or late loss of baby teeth
- crowded, misplaced, or blocked-out teeth
- upper and lower teeth that do not meet normally
- thumb- or finger-sucking
- biting of the cheek or roof of the mouth
- difficulty biting or chewing
- breathing through the mouth
- jaws that shift or make noise
- jaws and teeth that are out of proportion to the rest of the face

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Osgood-Schlatter disease *see* **Osteochondroses**

Osteochondroses

Definition

The term osteochondroses refers to a group of diseases of children and adolescents in which localized tissue death (necrosis) occurs, usually followed by full regeneration of healthy bone tissue. The singular term is osteochondrosis.

Description

During the years of rapid bone growth, blood supply to the growing ends of bones (epiphyses) may become insufficient resulting in necrotic bone, usually near joints. Since bone is normally undergoing a continuous rebuilding process, the necrotic areas are most often self-repaired over a period of weeks or months.

Osteochondrosis can affect different areas of the body and is often categorized by one of three locations: articular, non-articular, and physeal.

Physeal osteochondrosis is known as Scheuermann disease or juvenile kyphosis. It is a deformity of the thoracic spine (in the chest area, the vertebra to which ribs are attached) caused by abnormal centers of bone development at the intervertebral joints (physes). It is most common among children ages 13 to 16.

Articular disease occurs at the joints (articulations). One of the more common forms is Legg-Calvé-Perthes disease, which occurs at the hip joint. Other forms include Köhler disease (foot), Freiberg disease (second toe), and Panner disease (elbow). These diseases are most common during **adolescence**, although they have been found in individuals ranging in age from eight to 77.

Non-articular osteochondrosis occurs at any other skeletal location. For instance, Osgood-Schlatter disease of the tibia (the large inner bone of the leg between the knee and ankle) is relatively common. It is often the cause of knee and leg **pain** in active teens.

Osteochondritis dissecans (OCD) is a form of osteochondrosis in which loose bone fragments form in a joint. The knee is a common site for osteochondritis dissecans. The condition is found most often in people aged ten to 20, although it may occur at other ages. OCD is sometimes associated with some sort of past trauma to the joint. In about 30 to 40 percent of cases the same joint on both sides of the body is involved (e.g. both knees, both elbows).

Demographics

Osteochondroses are disorders of teens and young adults. Some are rare disorders, and in many cases, the number of individuals who have these disorders is not known, since cases many resolve on their own. Scheuermann disease is thought to occur in 0.4 to 8 percent of individuals. One Finnish study found that 13 percent of adolescents had Osgood-Schlatter disease. Freiberg disease is the one type of osteochondrosis that is more common in females than in males. OCD affects males about twice as often as females. All other osteochondrosis appear to affect the sexes equally.

Causes and symptoms

Many theories have been advanced to account for osteochondrosis, but none has proven fully satisfactory. Stress on the bone, ischemia (reduced blood supply), and trauma to the site are commonly mentioned factors. Athletic children are often affected when they overstress their developing limbs with a particular repetitive motion. Many cases are idiopathic, meaning that no specific cause is known.

The most common symptom for most types of osteochondrosis is pain, usually a dull, non-specific ache, at the affected joint. Pain is especially noticeable when pressure is applied. Locking of a joint or limited range of motion at a joint can also occur.

Scheuermann disease can (rarely) lead to serious kyphosis (hunchback condition) due to erosion of the vertebral bodies. Usually, however, the kyphosis is mild, causing no further symptoms and requiring no special treatment.

When to call the doctor

A doctor should be consulted whenever a child has a persistent joint pain that does not go away after resting the joint for a few days.

Diagnosis

Diagnosis is suspected based on history and symptoms. It can be confirmed by x-ray findings.

Treatment

Conservative treatment is usually attempted first. In many cases, resting the affected body part for a several days or weeks brings relief. A cast may be applied if needed to prevent movement of a joint.

Surgical intervention may be needed in some cases of osteochondritis dissecans to remove abnormal bone fragments in a joint. Rarely is spine curvature so pronounced in Scheuermann disease that the individual needs to wear a brace or have surgical intervention.

Prognosis

Accurate prediction of the outcome for individual adolescents is difficult with osteochondrosis. Some individuals heal spontaneously. Others heal with little treat-

KEY TERMS

Osteoblast—A bone-building cell.

Osteoclast—A large, multinuclear cell involved in the physiological destruction and absorption of bone.

ment other than keeping weight or stress off the affected limb. The earlier the age of onset, the better the prospects for full recovery. Surgical intervention is often successful in osteochondritis dissecans.

Prevention

No preventive measures are known.

Parental concerns

Persistent pain in the joint may keep children temporarily from playing **sports**.

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Osteogenesis imperfecta

Definition

Osteogenesis imperfecta (OI) is a group of genetic diseases in which the bones are formed improperly, making them fragile and prone to breaking.

Description

Collagen is a fibrous protein material that serves as the structural foundation of skin, bone, cartilage, and ligaments. In osteogenesis imperfecta, the collagen produced is abnormal and disorganized, which results in a number of abnormalities throughout the body, the most notable being fragile, easily broken bones.

There are four forms of OI, types I through IV. Of these, type II is the most severe and is usually fatal within a short time after birth. Types I, III, and IV have some overlapping and some distinctive symptoms, with the hallmark symptom being fragile bones.

OI is usually inherited as an autosomal dominant condition. In autosomal dominant inheritance, a single abnormal gene on one of the autosomal chromosomes (one of the first 22 non-sex chromosomes) from either parent can cause the disease. Only one parent needs to be a carrier in order for the child to inherit the disease. The parent affected by OI will have one abnormal gene and one normal gene. A child who has one parent with the disease therefore has a 50 percent chance of also inheriting the disease.

If both parents have OI caused by an autosomal dominant gene change, there is a 75 percent chance that the child will inherit one or both OI genes. In other words, there is a 25 percent chance of inheriting a faulty gene from the mother and a normal gene from the father, a 25 percent chance of inheriting a normal gene from the mother and a faulty gene from the father, a 25 percent chance of inheriting faulty genes from both parents, and a 25 percent chance of inheriting normal genes from both parents. It is difficult to predict the severity of OI in a child who has inherited two copies of the faulty gene because of its rarity.

There is no **family** history of OI in about 25 percent of children born with the disease. This occurs as a result of a spontaneous mutation of the gene in either the sperm or egg. The cause of such mutations is not known. Called new dominant mutation, the affected child subsequently has a 50 percent of passing the abnormal gene to his or her children. The risk of normal parents having a second child with OI, or of normal siblings going on to have affected children, does not appear to be greater than that of the general population.

In studies of families into which infants with OI type II were born, most of the babies had a new dominant mutation in a collagen gene. In some of these families, however, more than one infant was born with OI. Previously, researchers had seen this recurrence as evidence of recessive inheritance of OI type II. Subsequently, however, researchers concluded that the rare recurrence

of OI to a couple with a child with autosomal dominant OI is more likely due to gonadal mosaicism. Instead of mutation occurring in an individual sperm or egg, it occurs in a percentage of the cells that give rise to a parent's multiple sperm or eggs. This mutation, present in a percentage of his or her reproductive cells, can result in more than one affected child without affecting the parent with the disorder. An estimated 2 percent to 4 percent of families into which an infant with OI type II is born are at risk of having another affected child because of gonadal mosaicism.

Demographics

OI affects equal numbers of males and females. It occurs in about one of every 20,000 births.

Causes and symptoms

Evidence suggests that OI results from abnormalities in the collagen gene COL1A1 or COL1A2 and possibly abnormalities in other genes. In OI, the genetic abnormality causes one of two things to occur. It may direct cells to make an altered collagen protein and the presence of this altered collagen causes OI type II, III, or IV. Alternately, the dominant altered gene may fail to direct cells to make any collagen protein. Although some collagen is produced by instructions from the normal gene, an overall decrease in the total amount of collagen produced results in OI type I.

Type I

OI type I is the most common and mildest type. Among the common features of type I are the following:

- Bones are predisposed to fracture, with most **fractures** occurring before **puberty**; people with OI type I typically have about 20 to 40 fractures before puberty.
- Stature is normal or near-normal.
- Joints are loose and muscle tone is low.
- Sclerae (whites of the eyes) have blue, purple, or gray tint.
- Face shape is triangular.
- Tendency toward **scoliosis** (a curvature of the spine) is present.
- Bone deformity is absent or minimal.
- Dentinogenesis imperfecta may occur, causing brittle teeth.
- Hearing loss is a possible symptom, often beginning in early 20s or 30s.
- Structure of collagen is normal but the amount is lower than normal.

Type II

Sometimes called the lethal form, type II is the most severe form of OI. Among the common features of type II are the following:

- Frequently, OI type II is lethal at or shortly after birth, often as a result of respiratory problems.
- Fractures are numerous and bone deformity is severe.
- Stature is small with underdeveloped lungs.
- Collagen is formed improperly.

Type III

Among the common features of type III are the following:

- Bones fracture easily (Fractures are often present at birth, and x rays may reveal healed fractures that occurred before birth; people with OI type III may have more than 100 fractures before puberty.)
- Stature is significantly shorter than normal.
- Sclerae have blue, purple, or gray tint.
- Joints are loose and muscle development is poor in arms and legs.
- Rib cage is barrel-shaped.
- Face shape is triangular.
- Scoliosis (a curvature of the spine) is present.
- Respiratory problems are possible.
- Bones are deformed and deformity is often severe.
- Dentinogenesis imperfecta may occur.
- Hearing loss is possible.
- Collagen is formed improperly.

Type IV

OI type IV falls between type I and type III in severity. Among the common features of type IV are the following:

- Bones fracture easily, with most fractures occurring before puberty.
- Stature is shorter than average.
- Sclerae are normal in color, appearing white or near-white.
- Bone deformity is mild to moderate.
- Scoliosis (curvature of the spine) is likely.

- Rib cage is barrel-shaped.
- Face is triangular.
- Dentinogenesis imperfecta may occur.
- Hearing loss is possible.
- Collagen is formed improperly.

When to call the doctor

Parents should contact a healthcare professional if their child exhibits any of the symptoms of OI, particularly a tendency to fracture bones easily.

Diagnosis

It is often possible to diagnose OI solely on clinical features and x-ray findings. Collagen or DNA tests may help confirm a diagnosis of OI; test results may take several weeks to confirm. Approximately 10 to 15 percent of individuals with mild OI who have collagen testing, and approximately 5 percent of those who have genetic testing, test negative for OI despite having the disorder.

Diagnosis is usually suspected when a baby has bone fractures after having suffered no apparent injury. Another indication is small, irregular, isolated bones in the sutures between the bones of the skull (wormian bones). Sometimes the bluish sclerae serve as a diagnostic clue. Unfortunately, because of the unusual nature of the fractures occurring in a baby who is not yet mobile, some parents have been accused of **child abuse** before the actual diagnosis of osteogenesis imperfecta was reached.

Prenatal diagnosis

Testing is available to assist in prenatal diagnosis. Women with OI who become pregnant or women who conceive a child with a man who has OI may wish to explore prenatal diagnosis. Because of the relatively small risk (2–4%) of recurrence of OI type II in a family, families may opt for ultrasound studies to determine if a developing fetus has the disorder.

Ultrasound is the least invasive procedure for prenatal diagnosis and carries the least risk. Using ultrasound, a doctor can examine the fetus's skeleton for bowing of the leg or arm bones, fractures, shortening, or other bone abnormalities that may indicate OI. Different forms of OI may be detected by ultrasound in the second trimester. When OI occurs as a new dominant mutation and is found inadvertently on ultrasound, it may be difficult to confirm the diagnosis until after delivery since other genetic conditions can cause bowing and/or fractures prenatally.

Chorionic villus sampling is a procedure that obtains a sampling of cells from the placenta for testing. Examination of fetal collagen proteins in the tissue can reveal information about the quantitative or qualitative collagen defects that leads to OI. When a parent has OI, it is necessary for the affected parent to have the results of his or her own collagen test available. Chorionic villus sampling can be performed at ten to 12 weeks of pregnancy.

Amniocentesis is a procedure that involves inserting a thin needle into the uterus, into the amniotic sac, and withdrawing a small amount of amniotic fluid. Genetic material can be extracted from the fetal cells contained in the amniotic fluid and tested for the specific mutation known to cause OI in that family. This technique is useful only when the mutation causing OI in a particular family has been identified through previous genetic testing of affected family members, including previous pregnancies involving a baby with OI. Amniocentesis is performed at 16 to 18 weeks of pregnancy.

Treatment

There are no treatments available to cure OI, nor to prevent most of its complications. Most treatments are aimed at treating the fractures and bone deformities caused by OI. Splints, casts, braces, and rods are all used. Rodding is a surgical procedure in which a metal rod is implanted within a bone (usually the long bones of the thigh and leg). This surgery is performed when bowing or repeated fractures of these bones has interfered with a child's ability to walk.

Other treatments include hearing aids and early capping of teeth. Patients may require the use of a walker or wheelchair. **Pain** may be treated with a variety of medications. **Exercise** is encouraged as a means to promote muscle and bone strength. Swimming is a form of exercise that puts a minimal amount of strain on muscles, joints, and bones. Walking is encouraged for those who are able.

Alternative treatment

Alternative treatment such as acupuncture, naturopathic therapies, hypnosis, relaxation training, visual imagery, and biofeedback have all been used to try to decrease the constant pain of fractures.

Nutritional concerns

Smoking, excessive alcohol and **caffeine** consumption, and steroid medications may deplete bone and exacerbate bone fragility.

KEY TERMS

Chromosome—A microscopic thread-like structure found within each cell of the human body and consisting of a complex of proteins and DNA. Humans have 46 chromosomes arranged into 23 pairs. Chromosomes contain the genetic information necessary to direct the development and functioning of all cells and systems in the body. They pass on hereditary traits from parents to child (like eye color) and determine whether the child will be male or female.

Collagen—The main supportive protein of cartilage, connective tissue, tendon, skin, and bone.

Ligament—A type of tough, fibrous tissue that connects bones or cartilage and provides support and strength to joints.

Mutation—A permanent change in the genetic material that may alter a trait or characteristic of an individual, or manifest as disease. This change can be transmitted to offspring.

Sclera—The tough, fibrous, white outer protective covering of the eyeball.

Scoliosis—An abnormal, side-to-side curvature of the spine.

Prognosis

The lifespan of people with OI types I, III, and IV is not generally shortened. The prognosis for people with these types of OI is quite variable, depending on the severity of the disorder and the number and severity of the fractures and bony deformities.

Fifty percent of all babies with OI type II are stillborn. The rest of these babies usually die within a very short time after birth. In the early 2000s, some people with type II have lived into young adulthood.

Prevention

As a congenital birth defect, OI cannot be prevented. Individuals at risk of having a child with OI should be encouraged to undergo genetic counseling to more accurately determine their chances of having a child with OI. The risk of fractures can be minimized with bone- and muscle-strengthening exercises, rehabilitative therapy, and use of leg braces.

Nutritional concerns

Because the symptoms of OI are caused by collagen abnormalities and not a calcium deficiency (such as in osteoporosis), supplementation of **vitamins** or **minerals** will not cure the disease. To prevent bone loss related to calcium deficiency, which could exacerbate the fragility of bones, it is important that children with OI consume an adequate amount of calcium (generally 500 mg for children ages one to three, 800 mg for children ages four to eight, and 1,300 mg a day for preteens and teenagers).

Parental concerns

In cases in which OI is not diagnosed at birth, a child may experience numerous fractures of seemingly unexplained cause, leading healthcare providers to suspect the child is being abused. Once a child has been diagnosed, it may be helpful for parents to carry with them a letter from the child's healthcare provider detailing the diagnosis in order to facilitate care in an emergency.

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Osteopetroses

Definition

Osteopetrosis (plural osteopetroses) is a rare inherited disorder that makes bones increase in both size (mass) and fragility. It is a potentially fatal condition that can deform bone structure and distort the appearance. Osteopetrosis is also called chalk bones, ivory bones, or marble bones.

Description

In healthy individuals, bones are constantly being broken down (resorption) by cells called osteoclasts, and new bone material is constantly being formed by cells called osteoblasts. Osteopetrosis occurs when there is a failure in bone resorption. The mass of bone increases, but the new bone material that is added is porous, weak, and brittle.

There are three types of osteopetrosis. In some children with osteoporosis, bone mass begins to increase at birth, but symptoms may not become evident until adulthood. In mild cases, bone mass increases at gradual, irregular intervals until full adult height is attained. Some bones are not affected. Other forms of osteopetrosis progress at a more rapid pace and destroy bone structure, which can involve bones throughout the body, although the lower jaw is never affected.

Types of osteopetroses

Severe malignant infantile osteopetrosis (early-onset osteopetrosis) is the most severe form of osteopetrosis. It results from a child inheriting defective genes from both parents (autosomal recessive pattern of inheritance). It is most often discovered soon after birth. The ends of the long bones of the arms and legs appear widened and thickened (clubbed). Bone mass continues to increase rapidly, often filling in the hollow middle of the bone where the bone marrow, which produces red blood cells, is found. Early-onset osteopetrosis can be a fatal condition, with death occurring before the age of two. About one-third of all children with malignant infantile osteopetroses die before age ten. Although this form of osteopetrosis is called "malignant," it is not a type of **cancer**.

Intermediate osteopetrosis generally appears in children under age ten. This condition is usually less severe than early-onset or malignant infantile osteopetrosis and is not normally life-threatening.

Adult (delayed-onset) osteopetrosis may not become evident until after age 20. Albers-Schönberg disease is a mild form of this condition. People who have this disease are born with normal bone structure. Bone mass increases as they age but does not affect appearance, health, **intelligence**, or life span. Many people with adult osteopetrosis are diagnosed only when abnormalities are discovered on x rays taken for other purposes. There are two distinct types of adult osteopetrosis (types I and II). These types have different biochemical features. Individuals with type II disease have a higher risk of sustaining bone **fractures**.

Demographics

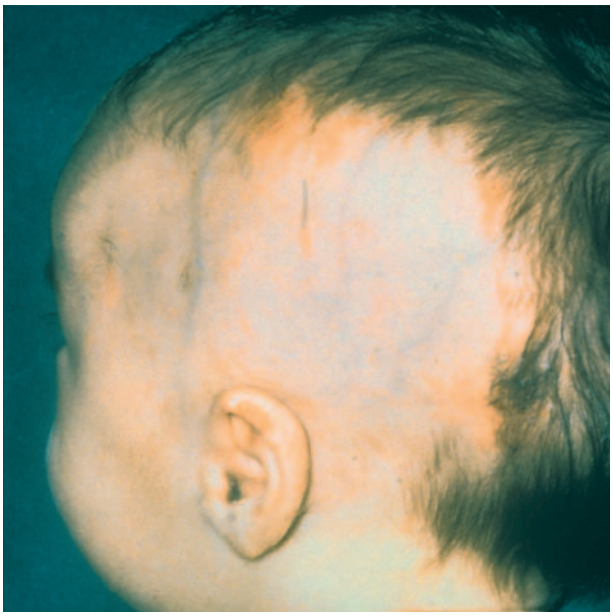
The incidence of osteopetroses is not known, although it is estimated that adult osteopetrosis occurs in about one of every 1,250 individuals. About 20,000 people in the United States have this form of the disease. Worldwide malignant infantile osteopetrosis occurs in about one in 100,000 to 500,000 births, making it exceedingly rare. Only eight to 40 children are born with this disease in the United States each year. Males and females appear to be equally affected.

Causes and symptoms

Osteopetrosis is the result of a genetic defect that causes the body to add new bone more rapidly than existing bone disintegrates. When fibrous or bony tissue invades bone marrow where red blood cells are made, the individual may develop anemia. Infection results when excess bone impairs the immune system, and hemorrhage can occur when platelet production is disrupted. When the skeleton grows so thick that nerves are unable to pass between bones, the individual may have a nerve damage, paralysis, or become blind or deaf.

Other symptoms associated with osteopetrosis include:

- bones that break easily and do not heal properly



This infant has osteopetrosis, a condition which thickens and hardens the bone. Note the unusual shape of the skull. *(Custom Medical Stock Photo Inc.)*

- bruising
- bone **pain**
- carpal tunnel syndrome
- osteoarthritis
- convulsions
- enlargement of the liver, lymph glands, or spleen
- failure to thrive (delayed growth, weight gain, and development)
- hydrocephalus (fluid on the brain)
- macrocephaly (abnormal enlargement of the head)
- paralysis or loss of control of muscles in the face or eyes

When to call the doctor

A doctor should be consulted if the child has frequent broken bones, bone pain, or failure of normal growth and development.

Diagnosis

Osteopetrosis is usually diagnosed when x rays reveal abnormalities or increases in bone density. Bone biopsy can confirm the diagnosis. Additional tests may be done to look for associated problems in vision, hearing, blood composition, and so forth.

Treatment

Interferon gamma-1b (Actimmune) is the only drug approved by the United States Food and Drug Administration (FDA) to treat osteopetrosis. Injections delay the progress of severe osteopetrosis in both children and adults.

Bone marrow transplantation (BMT) is the only therapy that can completely cure severe malignant infantile osteopetrosis. It replaces the abnormal osteoclasts with normal cells. The survival rate for children with osteopetrosis who undergo BMT is 40 to 70 percent. Because of the high risk of death, this procedure is done only with the most severely affected children where a good bone marrow match can be found.

High doses of calcitriol, an active form of vitamin D, can stimulate osteoclasts responsible for disintegration of old bone and significantly alleviate symptoms of severe disease in some individuals.

When bone overgrowth deforms the shape of the skull, surgery may be required to relieve pressure on the brain. Orthodontic treatment is sometimes necessary to correct **malocclusion**, a condition that shifts the position of the teeth and makes closing the mouth impossible.

Physical and occupational therapy can help children reach their full potential and adults to retain function and independence. Speech therapy is often needed in young children, because the heavy skeleton can cause language delays even in children of normal intelligence. Professional counseling can help children and their families cope with the emotional aspects of deformed features.

Prognosis

The severity of anemia seems to determine the course of an individual's osteopetrosis. About two-thirds of children who have severe malignant infantile osteopetrosis die before age ten unless they have a successful bone marrow transplant. Individuals who develop the adult form of the disease have normal life spans, although they experience more bone fractures and complications related to compression of cranial nerves. Nerve compression can result in facial paralysis, deafness, or blindness.

Prevention

Osteopetrosis is an inherited disease that cannot be prevented.

KEY TERMS

Cranial nerves—The set of twelve nerves found on each side of the head and neck that control the sensory and muscle functions of the eyes, nose, tongue, face, and throat.

Osteoblast—A bone-building cell.

Osteoclast—A large, multinuclear cell involved in the physiological destruction and absorption of bone.

Parental concerns

Parents with children who develop intermediate or adult forms of this disease as teens or young adults should be aware that their children are very susceptible to bone fractures and should avoid situations and **sports** where they are likely to be injured.

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Osteosarcoma *see* **Sarcomas**

Otitis externa

Definition

Otitis externa refers to an infection of the ear canal (outer ear), the tube leading from the outside opening of the ear in towards the ear drum. The infection usually develops in children and adolescents whose ears are exposed to persistent, excessive moisture.

Description

The external ear canal is a tube approximately 1 in (2.5 cm) in length that runs from the outside opening of the ear to the start of the middle ear, which is behind the tympanic membrane (ear drum). The canal is partly cartilage and partly bone. The lining of the ear canal is skin, which is attached directly to the covering of the bone. Glands within the skin of the canal produce a waxy substance called cerumen (popularly called earwax). Cerumen is designed to protect the ear canal, repel water, and keep the ear canal too acidic to allow bacteria to grow.

Continually exposing the ear canal to moisture may cause significant loss of cerumen. The delicate skin of the ear canal, unprotected by cerumen, retains moisture and becomes irritated. Without cerumen, the ear canal stops being appropriately acidic, which allows for the growth of microorganisms. Thus, the warm, moist, dark environment of the ear canal becomes a hospitable environment for development of an infection.

Otitis externa is commonly referred to as swimmer's ear.

Demographics

Although all age groups are affected by otitis externa, children, adolescents, and young adults whose ears are exposed to persistent, excessive moisture develop the infection most often. Otitis externa occurs most often in warm climates and during the summer months, when more people are participating in water activities. The ratio of occurrence in males is equal to that of females. People in some racial groups have a smaller size of the ear canal, which may predispose them to infection.

Causes and symptoms

Chidren and adolescents with otitis externa often have been diving or swimming for long periods of time, especially in polluted lakes, rivers, or ponds. Routine showering can also lead to otitis externa. Water in the ear canal can carry infectious microorganisms into the ear canal.

Bacteria, fungi, and viruses have all been implicated in causing otitis externa. However, most commonly otitis externa is caused by bacteria, especially *Pseudomonas aeruginosa*. Other bacteria that can cause otitis externa

include *Enterobacter aerogenes*, *Proteus mirabilis*, *Klebsiella pneumoniae*, *Staphylococcus epidermidis*, and bacteria of the family called Streptococci. Occasionally, fungi may cause otitis externa. These include *Candida* and *Aspergillus*. Two types of viruses, called herpesvirus hominis and varicella-zoster virus, have also been identified as causing otitis externa.

Other conditions predisposing to otitis externa include the use of cotton swabs to clean the ear canals. This pushes cerumen and normal skin debris back into the ear canal, instead of allowing the ear canal's normal cleaning mechanism of the ear to work, which would ordinarily move accumulations of cerumen and debris out of the ear. Also, putting other items into the ear can scratch the canal, making it more susceptible to infection. For example, children may insert a foreign body in their ear canal and not mention it to their parents. Hair spray or hair coloring, which can irritate the ear canal, may also lead to otitis externa. A hearing aid can trap moisture in the ear canal and should be taken out as often as possible to allow the ear an opportunity to dry out.

The first symptom of otitis externa is often **itching** of the ear canal, followed by watery discharge from the ear. Eventually, the ear begins to feel extremely painful. Any touch, movement, or pressure on the outside structure of the ear may cause severe **pain**. If the canal is excessively swollen, hearing may become muffled. The canal may appear swollen and red, and there may be evidence of foul-smelling, greenish-yellow pus.

In severe cases, otitis externa may be accompanied by **fever**. Often, this indicates that the outside ear structure has become infected as well. It will become red and swollen, and there may be enlarged and tender lymph nodes in front of, or behind, the ear.

A serious and life-threatening type of otitis externa is called malignant otitis externa. This is an infection that most commonly affects persons who have diabetes or in persons with weakened immune systems. In malignant otitis externa, a patient has usually had minor symptoms of otitis externa for some months, with pain and drainage. The causative bacteria is usually *Pseudomonas aeruginosa*. This bacteria spreads from the external canal into all of the nearby tissues, including the bones of the skull. Swelling and destruction of these tissues may lead to damage of certain nerves, resulting in spasms of the jaw muscles or paralysis of the facial muscles. Other, more severe, complications of this destructive infection include **meningitis** (swelling and infection of the coverings of the spinal cord and brain), brain infection, or brain abscess (the development of a pocket of infection with pus).

When to call the doctor

The doctor should be called if any of the following symptoms are present:

- pain in an ear with or without fever
- persistent itching of the ear or in the ear canal
- loss of hearing or decreased hearing in one or both ears
- discharge from an ear, especially if it is thick, discolored, bloody, or foul-smelling

Diagnosis

Diagnosis of uncomplicated otitis externa is usually quite simple. The symptoms alone, of ear pain worsened by any touch to the outer ear, are characteristic of otitis externa. Examination of the ear canal will usually reveal redness and swelling. It may be impossible (due to pain and swelling) to see much of the ear canal, but this inability itself is diagnostic.

If there is a need to identify the types of organisms causing otitis externa, the canal can be gently swabbed to obtain a specimen. The organisms present in the specimen can then be cultured (allowed to multiply) in a laboratory, and then viewed under a microscope to allow identification of the causative organisms.

If the rare infection malignant otitis externa is suspected, **computed tomography** scan (CT scan) or **magnetic resonance imaging** (MRI) scans will be performed to determine how widely the infection has spread within bone and tissue. A swab of the external canal will not necessarily reveal the actual causative organism, so some other tissue sample (biopsy) will need to be obtained. The CT or MRI will help the doctor decide where the most severe focus of infection is located, in order to guide the choice of a biopsy site.

Treatment

Otitis externa us usually not a dangerous condition and often clears up by itself within a few days. To aid in the healing, the infected ear canal can be washed with an over-the-counter topical antiseptic. Pain can be relieved be placing a warm heating pad or compress on the infected ear as well as through the use of an over-the-counter pain reliever such as **acetaminophen** or aspirin. During the healing process, the infected ear canal must be kept dry, even while showering, through the use of ear plugs or a shower cap.

If the pain worsens or does not improve within 24 hours, or for the fastest way to relieve pain and to prevent

the spread of infection, the doctor should be seen. The doctor will clean the ear with a suction-tipped probe or other type of suction device to relieve irritation and pain. **Antibiotics** will applied directly to the skin of the ear canal (**topical antibiotics**) to fight the infection. These antibiotics are often combined in a preparation that includes a steroid medication that reduces the itching, inflammation and swelling within the ear canal. For full treatment, eardrops are usually applied several times a day for seven to 10 days.

If the opening to the ear is narrowed by swelling, a cotton wick may be inserted into the ear canal to help carry the eardrops into the ear more effectively. The medications are applied directly to the wick, enough times per day to allow the wick to remain continuously saturated. After the wick is removed, usually after about 48 hours, the medications are then put directly into the ear canal three to four times each day.

For severe infection, oral antiobiotics may be prescribed. If the otitis externa infection is caused by the presence of a foreign body in the ear, the infection will not improve until the foreign body is removed.

In malignant otitis externa, antibiotics will almost always need to be given intravenously (IV). If the CT or MRI scan reveals that the infection has spread extensively, these IV antibiotics will need to be continued for six to eight weeks. If the infection is in an earlier stage, two weeks of IV antibiotics can be followed by six weeks of antibiotics by mouth.

Alternative treatment

Mullein (*Verbascum thapsus*) oil has anti-inflammatory properties and may be apppied to the infected ear canal (one to three drops every three hours) to help soothe and heal the ear. Garlic (*Allium sativum*) is a natural antibiotic. Garlic juice can be combined with equal parts of glycerin and a carrier oil such as olive or sweet olive and applied (one to three drops) to the infected ear every three hours.

Prognosis

The prognosis is excellent for otitis externa, for it is usually easily treated. Basic treatment measures will cure 90 percent of cases without complication. However, it may recur in certain susceptible individuals. Left untreated, malignant otitis externa may spread sufficiently to cause death.

KEY TERMS

Cerumen—The medical term for earwax.

Eardrum—A paper-thin covering stretching across the ear canal that separates the middle and outer ears.

Infectious disease—A disease caused by a virus or a bacterium. Examples of viruses causing an infectious disease are: HIV-1 virus, herpes simplex, cytomegalovirus, Epstein-Barr virus, leukemia virus. Examples of bacterial infectious diseases are: syphilis and tuberculosis.

Outer ear—Outer visible portion of the ear that collects and directs sound waves toward the tympanic membrane by way of a canal which extends inward through the temporal bone.

Prevention

Swimming in polluted water and in pools or hot tubs without good chlorine and pH control should be avoided.

Keeping the ear dry is an important aspect of prevention of otitis externa. Before swimming, a protective coating consisting of several drops of mineral oil, baby oil, or lanolin can be applied to the ear canal. After swimming, several drops of a mixture of isopropyl alcohol and white vinegar can be put into the ear canal to ensure that it dries adequately. The head should be tilted so that the solution reaches the bottom of the ear canal; then the liquid should be drained out.

Care should be taken when cleaning ears. The outer ear should be cleaned wiped with a clean washcloth. The use of pointed objects to dig into the ear canal, especially those that can scratch the skin, should be avoided.

The most serious complications of malignant otitis externa can be avoided by careful attention to early symptoms of ear pain and drainage from the ear canal. Children with conditions that put them at higher risk for this infection (diabetes or conditions that weaken the immune system) should always report new symptoms immediately to the doctor.

Parental concerns

Parents should teach their children how to clean their ears without using sharp objects and to dry their ears thoroughly after swimming, showering, or bathing.

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Judith Sims

Otitis media

Definition

Otitis media is an infection of the middle ear, which is located behind the eardrum. There are two main types of otitis media. In the first, called acute otitis media (AOM), parts of the ear are infected and swollen, and fluid and mucus are trapped inside the ear. AOM can be quite painful. In the second type, called otitis media with effusion (fluid), or OME, fluid and mucus remain trapped within the ear after the infection is over, making it more difficult for the ear to fight off new infections. This fluid may adversely affect a child's hearing.

Description

One of the most common childhood infections, Otitis media is the leading cause of visits to the doctor by children. It is also the most frequent reason children receive antibiotic prescriptions or undergo surgery.

In order to fully understand otitis media, it is helpful to have a basic knowledge of ear anatomy. Deep within the outer ear canal is the eardrum, which is a thin, transparent membrane that vibrates in response to sound. Behind the eardrum is the space called the middle ear. When the eardrum vibrates, three tiny bones within the middle ear, called ossicles, transmit these sounds to the inner ear. Nerves are stimulated in the inner ear, which then relay the sound signals to the brain. The eustachian tube, which connects the middle ear to the nose, normally equalizes pressure in the middle ear, allowing the eardrum and ossicles to vibrate correctly, so that hearing is normal.

There are certain factors particular to children that make them more at risk for otitis media. In children, the eustachian tube is shorter and less slanted than in adults. Its size and position allow bacteria and viruses to travel to the middle ear more easily. Children also have clumps of infection fighting cells, commonly called adenoids, in the area of the eustachian tube. These adenoids may enlarge with repeated respiratory tract infections and ultimately block the eustachian tubes. When these tubes are blocked, the middle ear is more likely to fill with fluid, which in turn increases the risk for infection.

Demographics

Otitis media is common. Fifty percent of children have an episode before their first birthday, and 80 percent of children have an occurrence by their third birthday. It is estimated that $3 to $4 billion are spent per year on patients with a diagnosis of acute otitis media and related complications. Ear infections are found in all age groups, but they are considerably more common in children, especially those aged six months to three years. Boys are affected more commonly than girls. Other children at higher risk include those from poor families, Native Americans, children born with **cleft palate** or other defects of the facial structures, and children with **Down syndrome**. Exposure to cigarette smoke and early entrance into daycare also increase the risk. Otitis media occurs more frequently in winter and early spring. It is less common among children who are breastfeeding. Some studies show a genetic predisposition towards developing otitis media.

Causes and symptoms

The first precondition for the development of acute otitis media is exposure to an organism capable of causing the infection. Otitis media may be caused by either viruses or bacteria. Viral infections account for approximately 15 percent of cases. The majority of other cases are caused by a variety of bacteria. The three most common bacteria are *Streptococcus pneumoniae* (responsible for 25–50% of cases), *Haemophilus influenzae* (15–30%), and *Moraxella catarrhalis* (3–20%).

Acute otitis media often occurs as an aftereffect of upper respiratory infections, in which the eustachian tube and nasal membranes become swollen and congested. This condition can lead to an impaired clearance and pressure regulation in the middle ear, which, if sustained, may be followed by viruses and bacteria traveling from the nasopharynx to the middle ear.

Otitis media with effusion may develop within weeks of an acute episode of middle ear infection, but in many cases the cause is unknown. It is often associated with an abnormal or malfunctioning eustachian tube, which causes negative pressure in the middle ear and leaking of fluid from tiny blood vessels, or capillaries, into the middle ear.

Symptoms of acute otitis media (AOM)

The following are symptoms of acute otitis media:

- **fever**
- ear pulling
- complaints of ear **pain**, ear fullness, or hearing loss by older children
- fussiness, irritability, or difficulties in hearing, feeding, or sleeping in younger children
- bloody or greenish-yellow pus draining from the ear (This seepage is the sign of a perforated the eardrum. The pain leading up to such a perforation may be severe, but it is often relieved by the rupture.)

Otitis media with effusion (OME) is the presence of middle ear fluid for six weeks or longer after the initial episode of acute otitis media. The hallmark of OME is the lack of obvious symptoms in those who most commonly have the condition. Older children often complain of muffled hearing or a sense of fullness in the ear. Younger children may turn up the television volume. Most often OME is diagnosed when someone examines the ear for another reason, such as a well-child physical. For this reason, OME is often referred to as silent otitis media.

When to call the doctor

Unresolved episodes of otitis media may lead to a variety of complications, including hearing loss and **dizziness**. Any child who reports an earache or a sense of fullness in the ear, especially if combined with a prior upper respiratory tract infection, or fever, should be evaluated by a physician.

Diagnosis

The physician will visualize the ear canal and ear drum by using a special lighted instrument called an otoscope. Normally, the light from the otoscope reflects off the eardrum in a characteristic fashion called the "cone of light." In an infection, this reflection is often shifted or absent. If fluid or pus is draining from the ear, it can be collected and sent to a laboratory to determine if any specific infectious organisms are present. Additionally, a tympanometry test will be performed. Here, the doctor inserts a probe into the ear which emits a tone with a certain amount of sound energy. The probe measures how much sound energy bounces back off the eardrum, rather than being transmitted to the middle ear. The more energy that is returned to the probe, the more blocked the middle ear is.

A diagnosis of acute otitis media is based on the following:

- recent, usually abrupt, onset of signs and symptoms of middle ear inflammation and middle ear effusion
- the presence of middle ear effusion that is indicated by any of the following: bulging of the tympanic membrane; limited or absent movement of the tympanic membrane; or discharge from the external ear
- signs or symptoms of middle ear inflammation as indicated by either distinct redness of the eardrum or ear pain that results in an interference with **sleep** or other normal activities

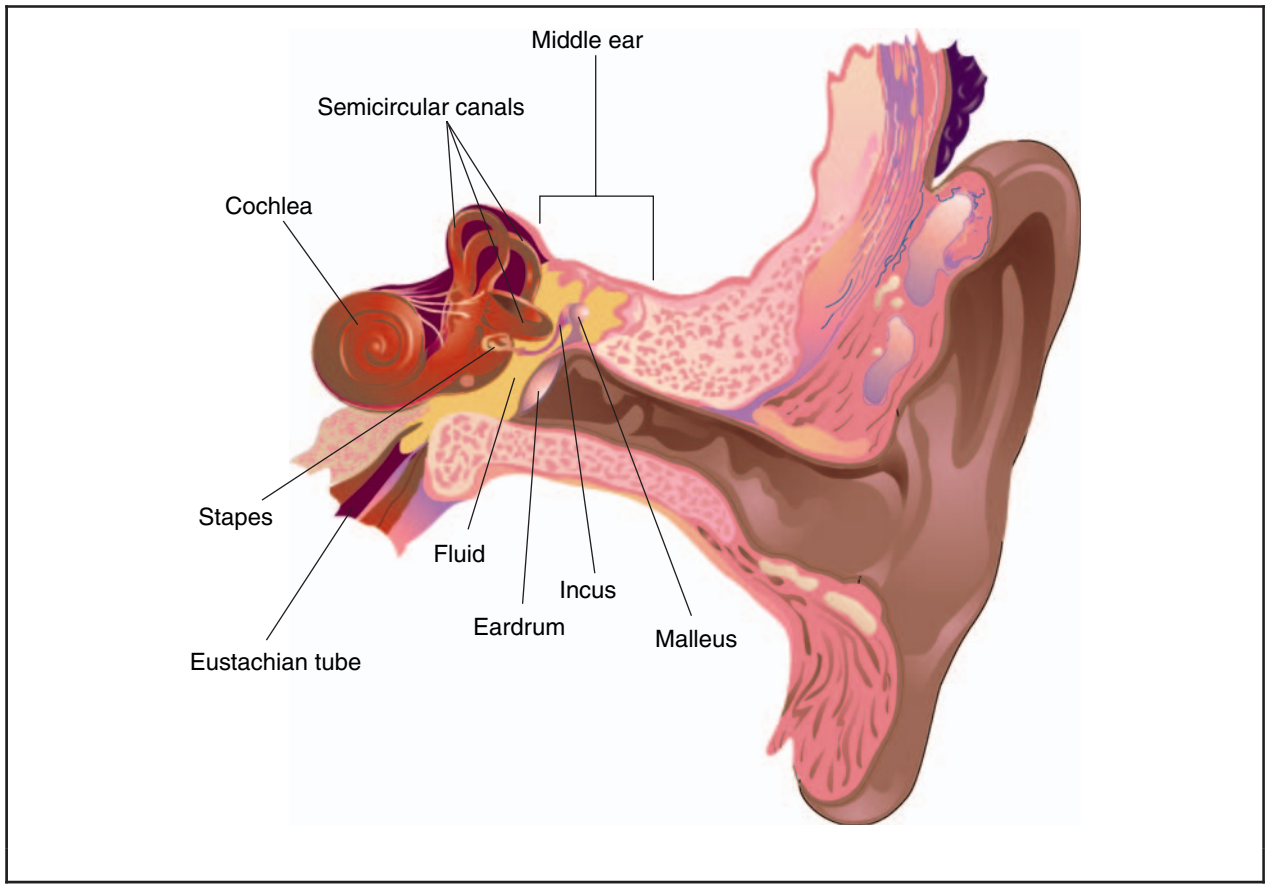
Otitis media with effusion can be more difficult to detect, since it is not painful and the child usually does not appear ill. The physician may rely on one or several tests to determine the diagnosis.

- A physical examination may reveal fluid behind the eardrum and poor movement of the eardrum. The eardrum may look clear and have no signs of redness, but may not move in response to air, as a normal eardrum would.
- A tympanometry test may reveal an impairment of eardrum mobility.
- A hearing test often shows some degree of hearing loss.

Treatment

Acute otitis media (AOM)

Treatment of AOM is focused on relieving any pain that may be present and addressing the infection itself. Usually, **acetaminophen** or ibuprofen prove adequate in



Otitis media is an ear infection in which fluid accumulates within the middle ear. A common condition occurring in childhood, it is estimated that 85 percent of all American children will develop otitis media at least once. *(Illustration by Electronic Illustrators Group.)*

relieving the pain. In cases of severe pain, narcotics may occasionally be prescribed.

Occasionally, an "observation option" will be used in a child who has uncomplicated acute otitis media. This refers to delaying antibacterial treatment of certain children for 48 to 72 hours and limiting management to symptomatic relief. The decision to observe or treat is based on the child's age, the certainty of the diagnosis, and the severity of the illness. To observe a child without initial antibacterial therapy, it is important that the parent or caregiver has a ready means of communicating with the doctor. There also must be a system in place that permits a prompt reevaluation of the child if symptoms persist or worsen. If the decision is made to use an antibiotic, the usual recommendation is for amoxicillin, preferably at a dose of 80 to 90mg/kg/day. If the initial treatment plan fails to work within 48 to 72 hours, the physician may reconsider the diagnosis of AOM. Further treatment may involve changing **antibiotics**.

Otitis media with effusion (OME)

For young children ages one to three years, most physicians prefer a conservative, or wait-and-see, approach, using antibiotics if the infection is persistent, the child is in pain, or there is evidence of hearing loss. Most cases of otitis media with effusion get better within three months without any treatment. If the child continues to have repeated episodes of OME, despite taking antibiotics, the physician may decide to try long-term, low-dose treatment with antibiotics, even after the condition has cleared. If OME persists for over three months, despite antibiotic treatment, the doctor may suggest a hearing test. If OME persists for more than four to six months, even if hearing tests are normal, the doctor may

suggest surgery to drain the eardrum and implant **ear tubes** for continuous drainage.

Surgery

In some cases, a surgical perforation to drain pus from the middle ear may be performed. This procedure is called a **myringotomy**. The hole created by the myringotomy generally heals itself in about a week. In 2002 a new minimally invasive procedure was introduced that uses a laser to perform the myringotomy. It can be performed in the doctor's office and heals more rapidly than the standard myringotomy. In some cases, the physician may decide that the placement of tubes during the myringotomy is recommended. These small tubes are placed to aid in draining the fluid from the middle ear. They fall out on their own after a few months. The decision to place these tubes is based on the following criteria:

- presence of fluid in the ears for more than three or four months following an ear infection
- fluid in the ears and more than three months of hearing loss
- changes in the structure of the eardrum as a result of ear infections
- a delay in speaking
- repeated infections that do not improve with antibiotics over several months

Another type of surgery, called an adenoidectomy, removes the adenoids. Removing the adenoids has been shown to help some children with otitis media between the ages of four to eight. It is a procedure generally reserved for those children who have recurrent otitis media after myringotomy tubes are extruded.

Alternative treatment

Treatment guidelines from the American Academy of Pediatrics and the American Academy of Family Physicians in the early 2000s state that there is insufficient evidence to either support or discourage the use of alternative medicines for acute otitis media. Increasing numbers of parents and caregivers are using various forms of nonconventional treatment for their children. Treatments that have been used for AOM include homeopathy, acupuncture, herbal remedies, chiropractic treatments, and nutritional supplements. Although most treatments are harmless, some are not. Some can have a direct and dangerous effect, whereas others may interfere with the effects of conventional treatments. Parent should inform their doctor if they are using any alternative or unconventional methods to treat their child's otitis media.

KEY TERMS

Adenoids—Common name for the pharyngeal tonsils, which are lymph masses in the wall of the air passageway (pharynx) just behind the nose.

Effusion—The escape of fluid from blood vessels or the lymphatic system and its collection in a cavity.

Eustachian tube—A thin tube between the middle ear and the pharnyx. Its purpose is to equalize pressure on either side of the ear drum.

Myringotomy—A surgical procedure in which an incision is made in the ear drum to allow fluid or pus to escape from the middle ear.

Nasopharynx—One of the three regions of the pharynx, the nasopharynx is the region behind the nasal cavity.

Ossicles—The three small bones of the middle ear: the malleus (hammer), the incus (anvil) and the stapes (stirrup). These bones help carry sound from the eardrum to the inner ear.

Prognosis

The prognosis of acute otitis media is excellent. The duration is variable. There may be improvement within 48 hours even without any treatment. Treatment with antibiotics for a week to 10 days is usually effective.

Prevention

Breastfeeding helps to pass along immunities to a child that may prevent otitis media. The position the child is in while breastfeeding is better than the usual bottle-feeding position for optimal eustachian tube function. If a child must be bottle-fed, it is best to hold the infant rather than allow him or her to lie down with the bottle. Because multiple upper respiratory infections may increase the risk for acute otitis media, reducing the exposure to large groups of children, particularly in day-care centers, may reduce the incidence. Children should also be kept away from environmental irritants such as secondhand tobacco smoke.

Parental concerns

A common concern among parents has been whether recurring episodes of otitis media will cause impairments in their child's development. Research indicates that

persistent otitis media in the first three years of life does not have an adverse effect on development.

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Otoscopic examination *see* **Ear exam with an otoscope**

Overhydration

Definition

Overhydration, also called water excess or water intoxication, is a condition in which the body contains too much water.

Description

Overhydration occurs when the body takes in more water than it excretes and its normal sodium level is diluted. This can result in digestive problems, behavioral changes, brain damage, seizures, or coma. An adult whose heart, kidneys, and pituitary gland are functioning properly would have to drink more than two gallons of water a day to develop water intoxication. This condition is most common in persons whose kidney function is impaired and may occur when doctors, nurses, or other healthcare professionals administer greater amounts of water-producing fluids and medications than the person's body can excrete.

Infants seem to be at greater risk for developing overhydration. The Centers for Disease Control and Prevention has declared that babies are especially susceptible to oral overhydration during the first month of life, when the kidneys' filtering mechanism is too immature to excrete fluid as rapidly as older infants do. Breast milk or formula provides all the fluids a healthy baby needs. Water should be given slowly, sparingly, and only during extremely hot weather. Overhydration, which has been cited as a hazard of infant swimming lessons, occurs whenever a baby drinks too much water, excretes too little fluid, or consumes and retains too much water.

Demographics

Overhydration is the most common electrolyte imbalance in hospitals, occurring in about 2 percent of all people.

Causes and symptoms

Drinking too much water rarely causes overhydration when the body's systems are working normally. People with heart, kidney, or liver disease are more likely to develop overhydration because their kidneys are unable to excrete water normally. It may be necessary for people with these disorders to restrict the amount of water they drink and/or adjust the amount of salt in their diets.

Since the brain is the organ most susceptible to overhydration, a change in behavior is usually the first symptom of water intoxication. The person may become confused, drowsy, or inattentive. Shouting and delirium are common. Symptoms of overhydration may include blurred vision, **muscle cramps** and twitching, paralysis on one side of the body, poor coordination, **nausea and vomiting**, rapid breathing, sudden weight gain, and weakness. The person's complexion is normal or flushed. Blood pressure is sometimes higher than normal, but elevations may not be noticed even when the degree of water intoxication is serious.

Overhydration can cause acidosis (a condition in which blood and body tissues have an abnormally high acid content), anemia, cyanosis (a condition that occurs when oxygen levels in the blood drop sharply), hemorrhage, and shock. The brain is the organ most vulnerable to the effects of overhydration. If excess fluid levels accumulate gradually, the brain may be able to adapt to them, and the person will have only a few symptoms. If the condition develops rapidly, confusion, seizures, and coma are likely to occur.

When to call the doctor

A doctor should be called when a person becomes confused, drowsy, or inattentive. Persons should also consider calling a doctor when a person experiences blurred vision, muscle cramps and twitching, paralysis on one side of the body, poor coordination, nausea and vomiting, rapid breathing, sudden weight gain, or weakness.

Diagnosis

Before treatment can begin, a doctor must determine whether an individual's symptoms are due to overhydration, in which excess water is found within and outside cells, or excess blood volume, in which high sodium levels prevent the body from storing excess water inside the cells. Overhydration is characterized by excess water both within and around the body's cells, while excess blood volume occurs when the body has too much sodium and cannot move water to reservoirs within the cells. In cases of overhydration, symptoms of fluid accumulation do not usually occur. On the other hand, in cases of excess blood volume, fluid tends to accumulate around cells in the lower legs, abdomen, and chest. Overhydration can occur alone or in conjunction with excess blood volume, and differentiating between these two conditions may be difficult.

KEY TERMS

Cyanosis—A bluish tinge to the skin that can occur when the blood oxygen level drops too low.

Electrolytes—Salts and minerals that produce electrically charged particles (ions) in body fluids. Common human electrolytes are sodium chloride, potassium, calcium, and sodium bicarbonate. Electrolytes control the fluid balance of the body and are important in muscle contraction, energy generation, and almost all major biochemical reactions in the body.

Shock—A medical emergency in which the organs and tissues of the body are not receiving an adequate flow of blood. This deprives the organs and tissues of oxygen and allows the build-up of waste products. Shock can be caused by certain diseases, serious injury, or blood loss.

Treatment

Mild overhydration can generally be corrected by following a doctor's instructions to limit fluid intake. In more serious cases, diuretics may be prescribed to increase urination, although these drugs tend to be most effective in the treatment of excess blood volume. Identifying and treating any underlying condition (such as impaired heart or kidney function) is a priority, and fluid restrictions are a critical component of every treatment plan.

In people with severe neurologic symptoms, fluid imbalances must be corrected without delay. A powerful diuretic and fluids to restore normal sodium concentrations are administered rapidly at first. When the person has absorbed 50 percent of the therapeutic substances, blood levels are measured. Therapy is continued at a more moderate pace in order to prevent brain damage as a result of sudden changes in blood chemistry.

Prognosis

Mild water intoxication is usually corrected by drinking less than a quart of water a day for several days. Untreated water intoxication can be fatal, but this outcome is quite rare.

Prevention

People should be careful not to drink excessive amounts of water. Persons with impaired kidney function must exert extra caution.

Parental concerns

Chronic illness, **malnutrition**, a tendency to retain water, and kidney diseases and disorders increase the likelihood of a person's becoming overhydrated. Infants and the elderly seem to be at increased risk for overhydration, as are people with certain mental disorders or **alcoholism**.

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Overweight *see* **Obesity**

P

Pacifier use

Definition

A pacifier is an artificial nipple designed for babies to suck on for comfort.

Purpose

Infants have an intense need to suck that is separate from their need to eat. Fetuses may suck their thumbs before they are born, and some newborns begin to suck immediately. Infants suck when they are tired, bored, or in need of comfort. Some babies have a stronger need to suck than others and—next to eating and being held—sucking may provide the most comfort to an infant. Babies who do not suck their thumbs or fingers often rely on pacifiers.

In Western societies 75 to 85 percent of children use pacifiers. Hospital nurseries commonly give them to newborns. Premature infants seem to grow better when they suck on pacifiers. Professionals refer to a pacifier as a transitional object that helps children adjust to new situations and relieves stress.

Most infants cry because they do not yet have methods for soothing themselves. Some newborns do not have the coordination to suck their fingers or thumb. Although breastfeeding is the most effective way to calm infants, and their hands or thumbs can be placed in their mouths, pacifiers can be very helpful for discontented babies who cannot or will not suck their thumbs or fingers.

Arguments for pacifier use

Pacifier use is controversial. Some physicians are completely opposed to pacifier use, whereas others view pacifiers as helpful if used in moderation. Pacifiers can be particularly useful for unhappy babies who are difficult to comfort. The child's energy goes into sucking on a pacifier rather than crying. Although pacifiers can give children a sense of calm and security well into their toddler years, pacifier use may be most effective during the first few months of life when fussiness, **colic**, and the need to suck are at their peaks.

Pacifiers should only be used to satisfy the need to suck. They should never be used to delay or replace nurturing or feeding. As a child grows, a pacifier can be taken away, whereas it may be harder to discourage **thumb sucking**. For babies, pacifiers can be used for the following reasons:

- to sooth a baby to **sleep**
- to help a baby to stay asleep when disturbed
- to calm a frightened baby
- to keep the baby quiet

Arguments against pacifier use

The World Health Organization recommends against pacifier use. Disadvantages of pacifier use may include:

- They may get dirty and thus contribute to poor hygiene.
- If lost during sleep, the pacifier's absence may cause the baby to wake and cry.
- The pacifier may prevent babies from using their mouths to learn about **toys** and other objects.
- The pacifier may signal to a baby that crying is unacceptable even though crying is one of a baby's few means of communication.
- The pacifier is an easy fix that may cause parents not to seek to understand what is bothering the baby.
- Pacifiers may prevent children from learning how to comfort themselves.
- Older siblings may give the baby a pacifier to quiet a baby in situations where the parents would not use it.
- Many adults dislike the sight of babies with pacifiers.

The advantage of thumb sucking is that babies can adjust sucking to the feel of their skin. Some people believe that thumb sucking is an easier habit to break than pacifier use. Thumb sucking may be preferable to pacifier use because thumbs have the following benefits:

- Unlike pacifiers, the baby can find his thumb at night.
- Thumbs are more hygienic.
- Thumbs taste better.

Description

Types of pacifiers

Pacifiers consists of a latex or silicone nipple with a firm plastic shield and handle. Latex pacifiers are softer or more flexible but wear out faster than silicone. Silicone pacifiers are firmer, hold their shapes longer, and are easier to clean. The nipple should be knotted around the back of the handle and the shield and handle should be one piece. This prevents the nipple from falling off or the plastic from breaking in two and posing a **choking** hazard. The shield should be at least 1.5 inches (6 cm) across so that it will not fit in the baby's mouth. The shield should have air holes or vents to prevent saliva from collecting behind it and causing an irritation or rash. Large circular shields can obstruct an intensely sucking baby's nasal passages. Pacifiers should have easy-to-hold handles, be dishwasher-safe, and easy to clean.

Pacifiers come in several sizes designed for premature infants, newborns, babies younger than six months, and children older than six months. Pacifier nipples come in various shapes—long, short, flattened, or with a ball-shaped end. Some are shaped like bottle nipples and others resemble a breast nipple that is being sucked on. The latter may not always fit a baby's mouth. There is no evidence that one shape is preferable to another, although the baby may prefer a certain type. Some babies gag at the texture, taste, or smell of some pacifiers. A baby bottle nipple should never be used as a pacifier since the nipple could pop out of the ring and choke the baby.

Pacifier care

Infant pacifiers should be cleaned daily by boiling or washing in a dishwasher. Once a child is six months old, the pacifier can be washed daily with warm soapy water and rinsed with clean water. Children may be taught to wash their own pacifiers. Pacifiers should never be shared with playmates. They should never be stored in plastic bags where dampness can encourage fungal growth.

Pacifier nipples should be examined regularly for deterioration, including tears, frayed edges, holes, or a change in color. Emerging teeth can tear pacifiers. A worn or damaged pacifier should be replaced immediately. Since pacifiers are lost frequently, several should be kept on hand.

Precautions

Pacifiers are sometimes attached to a baby's clothing with a clip and a short cord or ribbon to prevent them from becoming lost or dirty. However a child can become entangled in even a short cord and should never be put to bed with a pacifier attached to a cord. A pacifier that is hung on a cord around the baby's neck, tied to the baby's hand, or attached to a crib can cause strangulation. Pacifiers should never be sweetened because sweetened pacifiers constitute a leading cause of **tooth decay** in babies under age three.

Pacifiers never should be used to replace a feeding, and children should never be given a pacifier if they are hungry. A hungry baby may become upset when there is no milk in the nipple and could develop feeding problems. Thus pacifiers should only be used between or after feedings.

Terminating pacifier use

Babies need their mouths for **play** and exploration. By the time a child is **crawling** and learning to walk, pacifiers are both unhygienic and limiting. Although many experts still recommend weaning a child from pacifier use at about age two, others suggest that six to ten months is the best time to end pacifier use. Before the age of two, children have short memories and may easily forget about a pacifier that has been lost for a few days. Pacifier use should not be terminated too soon or too abruptly since a baby may substitute thumb sucking or some other behavior such as hair pulling. A two-year-old is much less likely to revert to thumb sucking.

Sometimes severe stress or emotional upset causes a child to use a pacifier for a very long time. Even children who stop because of **peer pressure** at school may continue to use a pacifier to calm down at home or to go to sleep.

Risks

Breastfeeding

Pacifiers should never be given to a breastfeeding infant unless an efficient nursing routine is well-established. Pacifiers may cause nipple confusion. Newborns must learn to breastfeed effectively, and babies suck on

breast nipples differently than on a pacifier. Pacifiers have a narrow base so that infants do not have to open their lips widely. Pacifier use may prevent infants from learning how to latch onto their mother's breast, resulting in poor feedings and sore nipples.

A number of studies have found that frequent pacifier use reduces the duration of breastfeeding and increases the likelihood that a baby will be weaned by six months. The earlier a pacifier is introduced the sooner breastfeeding ceases. The reasons for this include the following:

- Pacifier use causes babies to breastfeed less.
- Mothers may introduce a pacifier because they want to stop breastfeeding.
- Infants who are given a pacifier, with or without supplemental food, may lose interest in nursing.
- A reduction in breastfeeding decreases the mother's milk production.
- A reduction in nipple stimulation by a nursing infant decreases milk production.

Ear infections

Research has found that pacifier use increases a child's risk of ear infections—acute **otitis media**, the second most common childhood illness after colds. In one study pacifier use increased the frequency of ear infections by 50 percent. Another study found a 33 percent decrease in ear infections when pacifier use was limited to babies aged six to ten months and only used when they were falling asleep. It may be that pacifier use spreads infection or that intense sucking on pacifiers hinders proper functioning of the eustachian tube that normally keeps the middle ear open and clean. The studies suggest that pacifiers only be used with babies under ten months of age, when the need to suck is strongest and the incidence of ear infection is relatively low.

Dental development

Both dental cavities and misaligned teeth have long been associated with pacifier use. Neither pacifier use nor thumb sucking is likely to interfere with early **dental development**. However, if continued past about age three, either one can contribute to protruding front teeth and an overbite. Orthodontic pacifiers do not prevent dental abnormalities. Many dentists believe that neither pacifier use nor thumb sucking should continue once all of the baby teeth have erupted. A dentist may recommend devices that are designed to discourage pacifier use.

Other risks

Pacifiers can exacerbate any problems with developing speech and language. They have been shown to interfere with normal babbling and speech development in babies older than 12 months. A child learning to speak with a pacifier may have distorted speech. The child may replace "t" and "d" sounds—that require the front of the tongue to brush against the teeth—with "k" and "g" sounds that come from the back of the throat. Pacifiers also can interfere with children's willingness to talk and the development of their social skills.

Some research has suggested that babies exhibit fewer visually exploratory behaviors when using a pacifier; they look around less and can seem less alert. Some of these children receive less of the following:

- mental stimulation
- encouragement to explore and learn
- parental attention

Additional risks of pacifier use include the following:

- the transmission of thrush, a yeast infection, that can be difficult to eradicate in children with pacifiers
- product recall of pacifiers due to **safety** concerns
- children who are unable to give them up, even after years of use

Normal results

Many babies lose interest in their pacifiers at about four months of age, as the need to suck begins to subside. They suck on it less or spit it out. Failing to replace the pacifier after the baby cries or spits it out can be a good method for breaking the pacifier habit. It is easier to end pacifier use in a younger child. By the age of three most children have learned to communicate effectively and have other means of coping with stress. **Preschool** children may experience pressure from their peers to give up pacifiers. However, most children give up both pacifiers and thumb sucking long before they become social or dental concerns.

Parental concerns

There are numerous ways to console a crying baby other than using a pacifier. During at least the first six months of life, crying should always be responded to promptly. If infants have been well-fed within the past hour and do not need a diaper change, parents can comfort them by the following:

Toddler using a pacifier to comfort himself. *(© Lawrence Migdale/Photo Researchers, Inc.)*

- placing their face directly in front of the baby's face and talking to the child
- picking the baby up
- rocking the baby
- placing a gentle hand on the baby's stomach
- swaddling a baby in soft blankets

Parents who choose to give their baby a pacifier should take the following steps:

- not use it every time the baby's mouth is open
- try alternative methods of calming a crying baby
- encourage the child to sleep without it
- remove it as soon as the need for extra sucking is gone—usually between 12 and 15 months of age

It may take several weeks for children to give up their pacifiers. Parents can encourage their children to give them up gradually by trying the following:

- ignoring the behavior
- reassuring them with affection

KEY TERMS

Otitis media—Inflammation or infection of the middle ear space behind the eardrum. It commonly occurs in early childhood and is characterized by ear pain, fever, and hearing problems.

Overbite—Protrusion of the upper teeth over the lower teeth.

Thrush—An infection of the mouth, caused by the yeast *Candida albicans* and characterized by a whitish growth and ulcers.

Tooth eruption—The emergence of a tooth through the gum.

- limiting pacifier use to bedtime or stressful situations
- finding them a comforting alternative
- providing activities in situations where they are likely to be bored
- helping them to express their emotions in words
- gently reminding them to not to use their pacifiers
- praising them for not using a pacifier
- failing to take the pacifier to daycare or other activities
- using a calendar to mark down pacifier-free days
- rewarding pacifier-free days
- dipping the pacifier in pickle juice to make it is less appealing
- offering to trade in the pacifier for a "big kid's" toy
- not pressuring the child to give up their pacifiers
- not punishing or humiliating the child for pacifier use

See also Dental development; Orthodontics.

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Pain

Definition

Pain is an unpleasant feeling that is conveyed to the brain by nerves in the body.

Description

Pain arises from any number of situations. Injury is a major cause, but pain may also arise from an illness. It may accompany a psychological condition, such as depression, or may even occur in the absence of a recognizable trigger. The discomfort signals actual or potential injury to the body. However, pain is more than a sensation or the physical awareness of pain; it also includes perception, the subjective interpretation of the discomfort. Perception gives information on the pain's location, intensity, and something about its nature. The various conscious and unconscious responses to both sensation and perception, including the emotional response, add further definition to the overall concept of pain.

Acute pain

Acute pain often results from tissue damage, such as a skin burn or broken bone. Acute pain can also be associated with headaches or **muscle cramps**. This type of pain usually goes away as the injury heals or the cause of the pain (stimulus) is removed. To understand acute pain, it is necessary to understand the nerves that support it. Nerve cells, or neurons, perform many functions in the body. Although their general purpose, providing an interface between the brain and the body, remains constant, their capabilities vary widely. Certain types of neurons are capable of transmitting a pain signal to the brain. As a group, these pain-sensing neurons are called nociceptors, and virtually every surface and organ of the body is wired with them. The central part of these cells is located in the spine, and they send threadlike projections to every part of the body. Nociceptors are classified according to the stimulus that prompts them to transmit a pain signal. Thermoreceptive nociceptors are stimulated by temperatures that are potentially tissue damaging. Mechanoreceptive nociceptors respond to a pressure stimulus that may cause injury. Polymodal nociceptors are the most sensitive and can respond to temperature and pressure. Polymodal nociceptors also respond to chemicals released by the cells in the area from which the pain originates.

Nerve cell endings, or receptors, are responsible for pain sensation. A stimulus at this part of the nociceptor unleashes a cascade of neurotransmitters (chemicals that transmit information within the nervous system) in the spine. Each neurotransmitter has a purpose. For example, substance P relays the pain message to nerves leading to the spinal cord and brain. These neurotransmitters may also stimulate nerves leading back to the site of the injury. This response prompts cells in the injured area to release chemicals that not only trigger an immune response but also influence the intensity and duration of the pain.

Chronic and abnormal pain

Chronic pain refers to pain that persists after an injury heals, **cancer** pain, pain related to a persistent or degenerative disease, and long-term pain from an unidentifiable cause. It is estimated that one in three people

in the United States experiences chronic pain at some point in their lives. Of these people, approximately 50 million are either partially or completely disabled. Chronic pain may be caused by the body's response to acute pain. In the presence of continued stimulation of nociceptors, changes occur within the nervous system. Changes at the molecular level are dramatic and may include alterations in genetic transcription of neurotransmitters and receptors. These changes may also occur in the absence of an identifiable cause; one of the frustrating aspects of chronic pain is that the stimulus may be unknown. For example, the stimulus cannot be medically identified in as many as 85 percent of individuals suffering from lower back pain.

Other types of abnormal pain include allodynia, hyperalgesia, and phantom limb pain. These types of pain often arise from some damage to the nervous system (neuropathic). Allodynia refers to a feeling of pain in response to a normally harmless stimulus. For example, some individuals who have suffered nerve damage as a result of viral infection experience unbearable pain from just the light weight of their clothing. Hyperalgesia is somewhat related to allodynia in that the response to a painful stimulus is extreme. In this case, a mild pain stimulus, such as a pinprick, causes a maximum pain response. Phantom limb pain occurs after a limb is amputated; although an individual may be missing the limb, the nervous system continues to perceive pain originating from the area.

Demographics

Pain is experienced by all age groups, both sexes, and all races and ethnic groups.

Causes and symptoms

Pain is the most common symptom of injury and disease, and descriptions can range in intensity from a mere ache to unbearable agony. Nociceptors have the ability to convey information to the brain that indicates the location, nature, and intensity of the pain. For example, stepping on a nail sends an information-packed message to the brain: the foot has experienced a puncture wound that hurts a lot. Pain perception also varies depending on the location of the pain. The kinds of stimuli that cause a pain response on the skin include pricking, cutting, crushing, burning, and freezing. These same stimuli would not generate much of a response in the intestine. Intestinal pain arises from stimuli such as swelling, inflammation, and distension.

When to call the doctor

Parents should notify their physician or pediatrician if any of the following occurs:

- The child is in severe pain.
- The child has pain that lasts for more than three days.
- Parents have questions or concerns about their child's treatment or condition.
- The child is in the hospital and the parent thinks he or she is in pain. The sooner the pain is treated, the easier it is to control.

Diagnosis

Pain is considered in view of other symptoms and individual experiences. An observable injury, such as a broken bone, may be a clear indicator of the type of pain a person is suffering. Determining the specific cause of internal pain is more difficult. Other symptoms, such as **fever** or **nausea**, help narrow the possibilities. In some cases, such as lower back pain, a specific physiological cause may not be identified. Diagnosis of the disease causing a specific pain is further complicated by the fact that pain can be referred to (felt at) a skin site that does not seem to be connected to the site of the pain's origin. For example, pain arising from fluid accumulating at the base of the lung may be referred to the shoulder.

Since pain is a subjective experience, it may be very difficult to communicate its exact quality and intensity to other people. There are no diagnostic tests that can determine the quality or intensity of an individual's pain. Therefore, a medical examination includes a lot of questions about where the pain is located, its intensity, and its nature. Questions are also directed at what kinds of things increase or relieve the pain, how long it has lasted, and whether there are any variations in it. An individual may be asked to use a pain scale to describe the pain. One such scale assigns a number to the pain intensity; for example, 0 may indicate no pain, and 10 may indicate the worst pain the person has ever experienced. Scales are modified for infants and children to accommodate their level of comprehension.

A subsequent method of evaluating pain in children up to age four years was as of 2004 set to be implemented in 60 hospitals in the Netherlands. The Pain Observation Scale for Young Children, called POCIS, measures pain levels according to children's behavior in seven categories: facial expressions, crying, breathing, torso movements, movements in the arms and fingers and in the legs and toes, and restlessness. Physicians and nurses observe the intensity of these behaviors and calculate a pain severity score ranging from 0 to 7. Researchers

from the University of Amsterdam who developed the scale said that existing behavioral pain measures were created for premature neonates or infants and may not be appropriate for older children. Some of those measures are upsetting for children because they require restraint or physical contact by a healthcare professional.

Alternative treatment

Both physical and psychological aspects of pain can be dealt with through alternative treatment. Some of the most popular treatment options include acupressure and acupuncture, massage, chiropractic, and relaxation techniques, such as **yoga**, hypnosis, and meditation. Herbal therapies are increasingly recognized as viable options; for example, capsaicin, the component that makes cayenne peppers spicy, is used in ointments to relieve the joint pain associated with arthritis. Contrast hydrotherapy can also be very beneficial for pain relief. Lifestyles can be changed to incorporate a healthier diet and regular **exercise**. Regular exercise, aside from relieving stress, has been shown to increase endorphins, painkillers naturally produced in the body.

Prognosis

Successful pain treatment is highly dependent on successful resolution of the pain's cause. Acute pain will stop when an injury heals or when an underlying problem is treated successfully. Chronic pain and abnormal pain are more difficult to treat, and it may take longer to find a successful resolution. Some pain is intractable and requires extreme measures for relief.

Prevention

Pain is generally preventable only to the degree that the cause of the pain is preventable; diseases and injuries are often unavoidable. However, increased pain, pain from surgery and other medical procedures, and continuing pain are preventable through drug treatments and alternative therapies.

Parental concerns

If a child has a lot of pain, it is likely that more can be done to help. The first step is for parents to tell the child's doctor or nurse what their concerns are. They can ask what more can be done for the child to control pain. If parents are still concerned about their child's pain control, they can request a meeting with the doctor. Parents should list their concerns as clearly as possible. They should take a constructive approach and seek to form a partnership with the healthcare team in managing the child's pain. For parents who are still not satisfied with what is being done, some type of formal complaint to the hospital may be unavoidable. **Pain management** is the right of every child. Parents working with health providers are the best advocates for this right. The U.S. Department of Health and Human Services Agency for Health Care Policy and Research has developed guidelines for pain management. These guidelines establish a standard of care that should be followed. Parents can get a copy from the hospital library or directly from the government.

KEY TERMS

Acute pain—Pain in response to injury or another stimulus that resolves when the injury heals or the stimulus is removed.

Chronic pain—Pain that lasts over a prolonged period and threatens to disrupt daily life.

Neuron—The fundamental nerve cell of the nervous system.

Neurotransmitters—Chemicals in the brain that transmit nerve impulses.

Nociceptor—A nerve cell that is capable of sensing pain and transmitting a pain signal.

Referred pain—Pain that is experienced in one part of the body but originates in another organ or area. The pain is referred because the nerves that supply the damaged organ enter the spine in the same segment as the nerves that supply the area where the pain is felt.

Stimulus—Anything capable of eliciting a response in an organism or a part of that organism.

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Pain management

Definition

Pain management covers a number of methods to prevent, reduce, or stop pain sensations. These include the use of medications; physical methods such as ice and physical therapy; and psychological methods.

Purpose

Pain serves as an alert to potential or actual damage to the body. The definition for damage is quite broad; pain can arise from injury as well as disease. Pain that acts as a warning is called productive pain. After the message is received and interpreted, further pain offers no real benefit. Pain can have a negative impact on a person's quality of life and impede recovery from illness or injury. Unrelieved pain can become a syndrome in its own right and cause a downward spiral in a person's health and outlook. Managing pain properly facilitates recovery, prevents additional health complications, and improves a person's quality of life.

For many years it was believed that infants do not feel pain the way older children and adults do. As of the early 2000s, however, there has been a better understanding of the problems of pain, even in infancy.

Description

Before considering pain management, a review of pain definitions and mechanisms may be useful.

What is pain?

Pain is the means by which the peripheral nervous system (PNS) warns the central nervous system (CNS) of injury or potential injury to the body. The CNS comprises the brain and spinal cord, and the PNS is composed of the nerves that stem from and lead into the CNS. PNS includes all nerves throughout the body except the brain and spinal cord.

Once the brain has received and processed the pain message and coordinated an appropriate response, pain has served its purpose. The body uses natural pain killers, called endorphins, that are meant to derail further pain messages from the same source. However, these natural pain killers may not adequately dampen a continuing pain message. Pain is generally divided into two categories: acute and chronic.

Acute and chronic pain

Nociceptive pain, or the pain that is transmitted by nociceptors, is typically called acute pain. This kind of pain is associated with injury, headaches, disease, and many other conditions. It usually resolves once the condition that caused it is resolved. However, following some disorders, pain does not resolve. Even after healing or a cure has been achieved, the brain continues to perceive pain. In this situation, the pain may be considered chronic. The time limit used to define chronic pain typically ranges from three to six months, although some

healthcare professionals prefer a more flexible definition and consider pain chronic when it endures beyond a normal healing time. The pain associated with **cancer**, persistent and degenerative conditions, and neuropathy, or nerve damage, is included in the chronic category. Also, constant pain that lacks an identifiable physical cause, such as the majority of cases of low back pain, may be considered chronic.

It has been hypothesized that uninterrupted and unrelenting pain can induce changes in the spinal cord. As of 2004 evidence was accumulating that unrelenting pain or the complete lack of nerve signals increases the number of pain receptors in the spinal cord. Nerve cells in the spinal cord may also begin secreting pain-amplifying neurotransmitters independent of actual pain signals from the body. Other studies indicate that even newborn and premature infants who have constant pain will reach adulthood with greater sensitivity to pain and lower tolerance of stress.

Managing pain

Considering the different causes and types of pain, as well as its nature and intensity, management can require an interdisciplinary approach. The elements of this approach include treating the underlying cause of pain, pharmacological and nonpharmacological therapies, and some invasive (surgical) procedures.

Treating the cause of pain underpins the idea of managing it. Injuries are repaired, diseases are diagnosed, and certain encounters with pain can be anticipated and prevented. However, there are no guarantees of immediate relief from pain. Recovery can be impeded by pain, and quality of life can be damaged.

Pharmacological options

Pain-relieving drugs, otherwise called **analgesics**, include **nonsteroidal anti-inflammatory drugs** (NSAIDs), **acetaminophen**, narcotics, **antidepressants**, anticonvulsants, and others. NSAIDs and acetaminophen are available as over-the-counter and prescription medications and are frequently the initial pharmacological treatment for pain. These drugs can also be used as adjuncts to other drug therapies, which might require a doctor's prescription.

NSAIDs include aspirin, ibuprofen (Motrin, Advil, Nuprin), naproxen sodium (Aleve), and ketoprofen (Orudis KT). These drugs are used to treat pain from inflammation and work by blocking production of pain-enhancing neurotransmitters, such as prostaglandins. Acetaminophen is also effective against pain, but it is not an anti-inflammatory drug.

NSAIDs and acetaminophen are effective for most forms of mild pain, but moderate and severe pain may require stronger medication. Narcotics handle intense pain effectively and are used for cancer pain and acute pain that does not respond to NSAIDs and acetaminophen.

Narcotics may be ineffective against some forms of chronic pain, especially since changes in the spinal cord may alter the usual pain signaling pathways. Furthermore, narcotics are usually not recommended for long-term use because the body develops a tolerance to narcotics, reducing their effectiveness over time. In such situations, pain can be managed with antidepressants and anticonvulsants, which are also only available with a doctor's prescription.

Although antidepressant drugs were developed to treat depression, it has been discovered that they are also effective in combating chronic headaches, cancer pain, and pain associated with nerve damage. Antidepressants that have been shown to have analgesic (pain reducing) properties include amitriptyline (Elavil), trazodone (Desyrel), and imipramine (Tofranil). Anticonvulsant drugs share a similar background with antidepressants. Developed to treat epilepsy, anticonvulsants were found to relieve pain as well. Drugs such as phenytoin (Dilantin) and carbamazepine (Tegretol) are prescribed to treat the pain associated with nerve damage.

Other prescription drugs are used to treat specific types of pain or specific pain syndromes. For example, corticosteroids are very effective against pain caused by inflammation and swelling, and sumatriptan (Imitrex) was developed to treat migraine headaches.

Drug administration depends on the drug type and the required dose. Some drugs are not absorbed very well from the stomach and must be injected or administered intravenously. Injections and intravenous administration may also be used when high doses are needed or if an individual is nauseous. Following surgery and other medical procedures, patients may have the option of controlling the pain medication themselves. By pressing a button, they can release a set dose of medication into an intravenous solution. This procedure has also been employed in other situations requiring pain management. Another mode of administration involves implanted catheters that deliver pain medication directly to the spinal cord. Delivering drugs in this way can reduce side effects and increase the effectiveness of the drug.

Nonpharmacological options

Pain treatment options that do not use drugs are often used as adjuncts to, rather than replacements for,

drug therapy. One of the benefits of non-drug therapies is that an individual can take a more active stance against pain. Relaxation techniques, such as **yoga** and meditation, are used to decrease muscle tension and reduce stress. Tension and stress can also be reduced through biofeedback, in which an individual consciously attempts to modify skin temperature, muscle tension, blood pressure, and heart rate.

Participating in normal activities and exercising can also help control pain levels. Through physical therapy, an individual learns beneficial exercises for reducing stress, strengthening muscles, and staying fit. Regular **exercise** has been linked to production of endorphins, the body's natural pain killers.

Acupuncture involves the insertion of small needles into the skin at key points. Acupressure uses these same key points but involves applying pressure rather than inserting needles. Both of these methods may work by prompting the body to release endorphins. Applying heat or being massaged are very relaxing and help reduce stress. Transcutaneous electrical nerve stimulation (TENS) applies a small electric current to certain parts of nerves, potentially interrupt pain signals and induce the release of endorphins. To be effective, use of TENS should be medically supervised.

Invasive procedures

Three types of invasive procedures may be used to manage or treat pain: anatomic, augmentative, and ablative. These procedures involve surgery, and certain guidelines should be followed before carrying out a procedure with permanent effects. First, the cause of the pain must be clearly identified. Next, surgery should be done only if noninvasive procedures are ineffective. Third, any psychological issues should be addressed. Finally, there should be a reasonable expectation of success.

Anatomic procedures involve correcting the injury or removing the cause of pain. Relatively common anatomic procedures are decompression surgeries, such as repairing a herniated disk in the lower back or relieving the nerve compression related to carpal tunnel syndrome. Another anatomic procedure is neurolysis, also called a nerve block, which involves destroying a portion of a peripheral nerve.

Augmentative procedures include electrical stimulation or direct application of drugs to the nerves that are transmitting the pain signals. Electrical stimulation works on the same principle as TENS. In this procedure, instead of applying the current across the skin, electrodes are implanted to stimulate peripheral nerves or nerves in the spinal cord.

Ablative procedures are characterized by severing a nerve and disconnecting it from the spinal cord.

Preparation

Prior to beginning management, pain is thoroughly evaluated. Pain scales or questionnaires are used to attach an objective measure to a subjective experience. Objective measurements allow healthcare workers a better understanding of the pain being experienced by the patient. Evaluation also includes physical examinations and diagnostic tests to determine underlying causes. Some evaluations require assessments from several viewpoints, including neurology, psychiatry, psychology, and physical therapy. If pain is due to a medical procedure, management consists of anticipating the type and intensity of associated pain and managing it preemptively.

Risks

Owing to toxicity over the long term, some drugs can only be used for acute pain or as adjuncts in chronic pain management. NSAIDs have the well-known side effect of causing gastrointestinal bleeding, and long-term use of acetaminophen has been linked to kidney and liver damage. Other drugs, especially narcotics, have serious side effects, such as **constipation**, drowsiness, and **nausea**. Serious side effects can also accompany pharmacological therapies; mood swings, confusion, bone thinning, cataract formation, increased blood pressure, and other problems may discourage or prevent use of some analgesics.

Nonpharmacological therapies carry little or no risk. However, it is advised that individuals recovering from serious illness or injury consult with their healthcare providers or physical therapists before making use of adjunct therapies. Invasive procedures carry risks similar to other surgical procedures, such as infection, reaction to anesthesia, iatrogenic (injury as a result of treatment) injury, and failure.

A traditional concern about narcotics use has been the risk of promoting **addiction**. As narcotic use continues over time, the body becomes accustomed to the drug and adjusts normal functions to accommodate to its presence. Therefore, to elicit the same level of action, it is necessary to increase dosage over time. As dosage increases, an individual may become physically dependent on **narcotic drugs**.

KEY TERMS

Acute—Refers to a disease or symptom that has a sudden onset and lasts a relatively short period of time.

Central nervous system—Part of the nervous system consisting of the brain, cranial nerves, and spinal cord. The brain is the center of higher processes, such as thought and emotion and is responsible for the coordination and control of bodily activities and the interpretation of information from the senses. The cranial nerves and spinal cord link the brain to the peripheral nervous system, that is the nerves present in the rest of body.

Chronic—Refers to a disease or condition that progresses slowly but persists or recurs over time.

Iatrogenic—A condition that is caused by the diagnostic procedures or treatments administered by medical professionals. Iatrogenic conditions may be caused by any number of things including contaminated medical instruments or devices, contaminated blood or implants, or contaminated air within the medical facility.

Neuropathy—A disease or abnormality of the peripheral nerves (the nerves outside the brain and spinal cord). Major symptoms include weakness, numbness, paralysis, or pain in the affected area.

Neurotransmitter—A chemical messenger that transmits an impulse from one nerve cell to the next.

Nociceptor—A nerve cell that is capable of sensing pain and transmitting a pain signal.

Nonpharmacological—Referring to therapy that does not involve drugs.

Peripheral nervous system (PNS)—The part of the nervous system that is outside the brain and spinal cord. Sensory, motor, and autonomic nerves are included. PNS nerves link the central nervous system with sensory organs, muscles, blood vessels, and glands.

Pharmacological—Referring to therapy that relies on drugs.

Stimulus—Anything capable of eliciting a response in an organism or a part of that organism.

However, physical dependence is different from psychological addiction. Physical dependence is characterized by discomfort if drug administration suddenly stops, while psychological addiction is characterized by an overpowering craving for the drug for reasons other than pain relief. Psychological addiction is a very real and necessary concern in some instances, but it should not interfere with a genuine need for narcotic pain relief. However, caution must be taken with people with a history of addictive behavior.

Parental concerns

Infants feel pain, but do not express it in the same manner as older children or young adults. Studies indicate that the majority of parents do not know how to recognize the signs of infant pain, and pediatricians fail to teach parents what to look for. Training of parents is essential in recognizing and dealing with pain in infants and young children.

In some cases, narcotic analgesics are essential for control of childhood pain. These drugs are safe when used properly and should not be withheld for fear of addiction.

Because exposure to chronic pain by children can lead to life-long changes in their pain response, parents must learn to recognize and treat pain promptly.

Over-the-counter pain relievers may be toxic. Parents must read the labeled directions carefully and follow them exactly. For liquids, it is essential to use the proper measuring devices, such as a measuring dropper or medicinal teaspoon. Household measures are not reliable.

See also Acetaminophen; Nonsteroidal anti-inflammatory drugs.

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Pain relievers *see* **Analgesics**

Parent-child relationships

Definition

The parent-child relationship consists of a combination of behaviors, feelings, and expectations that are unique to a particular parent and a particular child. The relationship involves the full extent of a child's development.

Description

Of the many different relationships people form over the course of the life span, the relationship between parent and child is among the most important. The quality of the parent-child relationship is affected by the parent's age, experience, and self-confidence; the stability of the parents' marriage; and the unique characteristics of the child compared with those of the parent.

Characteristics of the parent

Parental self-confidence is an important indicator of parental competence. Mothers who believe that they are effective parents are more competent than mothers who feel incompetent. Also, mothers who see themselves as effective also tend to believe their infants as less difficult to handle. Parental age and previous experience are also important. Older mothers tend to be more responsive to their infants than younger mothers. In addition, parents who have had previous experience with children, whether through younger siblings, career paths, or previous children, are often times better able to cope with parenthood.

Characteristics of the child

Characteristics that may affect the parent-child relationship in a **family** include the child's physical appearance, sex, and **temperament**. At birth, the infant's physical appearance may not meet the parent's expectations, or the infant may resemble a disliked relative. As a result, the parent may subconsciously reject the child. If the parents wanted a baby of a particular sex, they may be disappointed if the baby is the opposite sex. If parents do not have the opportunity to talk about this disappointment, they may reject the infant.

Children who are loved thrive better than those who are not. Either parent or a nonparent caregiver may serve as the primary caregiver or form the primary parent-child love relationship. Loss of love from a primary caregiver can occur with the death of a parent or interruption of parental contact through prolonged hospitalizations. **Divorce** can interfere with the child's need to eat, improve, and advance. Cultural norms within the family also affect a child's likelihood to achieve particular developmental milestones.

Cultural impact

In some countries, childrearing is considered protective nurturing. Children are not rushed into new experiences like **toilet training** or being in school. In other countries, children are commonly treated in a harsh, strict manner, using shame or corporal punishment for **discipline**. In Central American nations, toilet training may begin as early as when the child can sit upright.

Childhood in the United States stretches across many years. In other countries, children are expected to enter the adult world of work when they are still quite young: girls assume domestic responsibilities, and boys do outside farm work. In addition, in Asian cultures, parents understand an infant's personality in part in terms of the child's year and time of birth.

Impact of birth order

The position of a child in the family, whether a firstborn, a middle child, the youngest, an only child, or one within a large family, has some bearing on the child's growth and development. An only child or the oldest child in a family excels in **language development** because conversations are mainly with adults. Children learn by watching other children; however, a firstborn or an only child, who has no example to watch, may not excel in other skills, such as toilet training, at an early age.

Infancy

As babies are cared for by their parents, both parties develop understandings of the other. Gradually, babies begin to expect that their parent will care for them when they cry. Gradually, parents respond to and even antici-

pate their baby's needs. This exchange and familiarity create the basis for a developing relationship.

PARENT-INFANT ATTACHMENT One of the most important aspects of infant psychosocial development is the infant's attachment to parents. Attachment is a sense of belonging to or connection with a particular other. This significant bond between infant and parent is critical to the infant's survival and development. Started immediately after birth, attachment is strengthened by mutually satisfying interaction between the parents and the infant throughout the first months of life, called bonding. By the end of the first year, most infants have formed an attachment relationship, usually with the primary caretaker.

If parents can adapt to their babies, meet their needs, and provide nurturance, the attachment is secure. Psychosocial development can continue based on a strong foundation of attachment. On the other hand, if a parent's personality and ability to cope with the infant's needs for care are minimal, the relationship is at risk and so is the infant's development.

By six to seven months, strong feelings of attachment enable the infant to distinguish between caregivers and strangers. The infant displays an obvious preference for parents over other caregivers and other unfamiliar people. **Anxiety**, demonstrated by crying, clinging, and turning away from the stranger, is revealed when separation occurs. This behavior peaks between seven and nine months and again during toddlerhood, when separation may be difficult. Although possibly stressful for the parents, **stranger anxiety** is a normal sign of healthy child attachment and occurs because of **cognitive development**. Most children develop a secure attachment when reunited with their caregiver after a temporary absence. In contrast, some children with an insecure attachment want to be held, but they are not comfortable; they kick or push away. Others seem indifferent to the parent's return and ignore them when they return.

The quality of the infant's attachment predicts later development. Youngsters who emerge from infancy with a secure attachment stand a better chance of developing happy and healthy relationships with others. The attachment relationship not only forms the emotional basis for the continued development of the parent-child relationship, but can serve as a foundation for future social connections. Secure infants have parents who sensitively read their infant's cues and respond properly to their needs.

Toddlerhood

When children move from infancy into toddlerhood, the parent-child relationship begins to change. During infancy, the primary role of the parent-child relationship is nurturing and predictability, and much of the relationship revolves around the day-to-day demands of caregiving: feeding, toileting, bathing, and going to bed.

As youngsters begin to talk and become more mobile during the second and third years of life, however, parents usually try to shape their child's social behavior. In essence, parents become teachers as well as nurturers, providers of guidance as well as affection. Socialization (preparing the youngster to live as a member of a social group) implicit during most of the first two years of life, becomes clear as the child moves toward his or her third birthday.

Socialization is an important part of the parent-child relationship. It includes various child-rearing practices, for example weaning, toilet training, and discipline.

Dimensions of the parent-child relationship are linked to the child's psychological development, specifically how responsive the parents are, and how demanding they are. Responsive parents are warm and accepting toward their children, enjoying them and trying to see things from their perspective. In contrast, nonresponsive parents are aloof, rejecting, or critical. They show little pleasure in their children and are often insensitive to their emotional needs. Some parents are demanding, while others are too tolerant. Children's healthy psychological development is facilitated when the parents are both responsive and moderately demanding.

During toddlerhood, children often begin to assert their need for autonomy by challenging their parents. Sometimes, the child's newfound assertiveness during the so-called terrible twos can put a strain on the parent-child relationship. It is important that parents recognize that this behavior is normal for the toddler, and the healthy development of independence is promoted by a parent-child relationship that provides support for the child's developing sense of autonomy. In many regards, the security of the first attachment between infant and parent provides the child with the emotional base to begin exploring the world outside the parent-child relationship.

Preschool

Various parenting styles evolve during the **preschool** years. Preschoolers with authoritative parents are curious about new experiences, focused and skilled at **play**, self-reliant, self-controlled, and cheerful.

School age

During the elementary school years, the child becomes increasingly interested in peers, but this is not be a sign of disinterest in the parent-child relationship. Rather, with the natural broadening of psychosocial and cognitive abilities, the child's social world expands to include more people and settings beyond the home environment. The parent-child relationship remains the most important influence on the child's development. Children whose parents are both responsive and demanding continue to thrive psychologically and socially during the middle childhood years.

During the school years, the parent-child relationship continues to be influenced by the child and the parents. In most families, patterns of interaction between parent and child are well established in the elementary school years.

Adolescence

As the child enters **adolescence**, biological, cognitive, and emotional changes transform the parent-child relationship. The child's urges for independence may challenge parents' authority. Many parents find early adolescence a difficult period. Adolescents fare best and their parents are happiest when parents can be both encouraging and accepting of the child's needs for more psychological independence.

Although the value of peer relations grows during adolescence, the parent-child relationship remains crucial for the child's psychological development. Authoritative parenting that combines warmth and firmness has the most positive impact on the youngster's development. Adolescents who have been reared authoritatively continue to show more success in school, better psychological development, and fewer behavior problems.

Adolescence may be a time of heightened bickering and diminished closeness in the parent-child relationship, but most disagreements between parents and young teenagers are over less important matters, and most teenagers and parents agree on the essentials. By late adolescence most children report feeling as close to their parents as they did during elementary school.

Parenting styles

Parenting has four main styles: authoritarian, authoritative, permissive (indulgent), and detached. Although no parent is consistent in all situations, parents do follow some general tendencies in their approach to childrearing, and it is possible to describe a parent-child relationship by the prevailing style of parenting. These descriptions provide guidelines for both professionals and parents interested in understanding how variations in the parent-child relationship affect the child's development.

Parenting style is shaped by the parent's developmental history, education, and personality; the child's behavior; and the immediate and broader context of the parent's life. Also, the parent's behavior is influenced by the parent's work, the parents' marriage, family finances, and other conditions likely to affect the parent's behavior and psychological well-being. In addition, parents in different cultures, from different social classes, and from different ethnic groups rear their children differently. In any event, children's behavior and psychological development are linked to the parenting style with which they are raised.

Authoritarian parents

Authoritarian parents are rigid in their rules; they expect absolute obedience from the child without any questioning. They also expect the child to accept the family beliefs and principles without questions. Authoritarian parents are strict disciplinarians, often relying on physical punishment and the withdrawal of affection to shape their child's behavior.

Children raised with this parenting style are often moody, unhappy, fearful, and irritable. They tend to be shy, withdrawn, and lack self-confidence. If affection is withheld, the child commonly is rebellious and antisocial.

Authoritative parents

Authoritative parents show respect for the opinions of each of their children by allowing them to be different. Although there are rules in the household, the parents allow discussion if the children do not understand or agree with the rules. These parents make it clear to the children that although they (the parents) have final authority, some negotiation and compromise may take place. Authoritative parents are both responsive and demanding; they are firm, but they discipline with love and affection, rather than power, and they are likely to explain rules and expectations to their children instead of simply asserting them. This style of parenting often results in children who have high **self-esteem** and are independent, inquisitive, happy, assertive, and interactive.

Permissive parents

Permissive (indulgent) parents have little or no control over the behavior of their children. If any rules exist in the home, they are followed inconsistently. Underlying reasons for rules are given, but the children decide

whether they will follow the rule and to what extent. They learn that they can get away with any behavior. Indulgent parents are responsive but not especially demanding. They have few expectations of their children and impose little or inconsistent discipline. There are empty threats of punishment without setting limits. Role reversal occurs; the children act more like the parents, and the parents behave like the children.

Children of permissive parents may be disrespectful, disobedient, aggressive, irresponsible, and defiant. They are insecure because they lack guidelines to direct their behavior. However, these children are frequently creative and spontaneous. Although low in both social responsibility and independence, they are usually more cheerful than the conflicted and irritable children of authoritarian parents.

Disengaged parents

Finally, disengaged (detached) parents are neither responsive nor demanding. They may be careless or unaware of the child's needs for affection and discipline. Children whose parents are detached have higher numbers of psychological difficulties and behavior problems than other youngsters.

Parental concerns

Child's development is affected by family conditions such as divorce, remarriage, and parental employment. The parent-child relationship has a more important influence on the child's psychological development than changes in the composition of the household. Parenting that is responsive and demanding is related to healthier child development regardless of the parent's marital or employment status. If changes in the parent's marital status or work life disrupt the parent-child relationship, short-term effects on the child's behavior may be noticeable. One goal of professionals who work with families under stress is to help them reestablish healthy patterns of parent-child interaction.

Discipline is also a concern of parents. Children's behavior offers challenges to even the most experienced and effective parents. The manner in which parents respond to a child's behavior has an effect on the child's self-esteem and future interactions with others. Children learn to view themselves in the same way the parent views them. Thus, if the parent views the child as wild, the child begins to view himself that way and soon his actions consistently reinforce his self image. This way, the child does not disappoint the parent. This pattern is a self-fulfilling prophecy. While discipline in necessary to teach a child how to live comfortably in society, it should not be confused with punishment.

KEY TERMS

Adolescence—A period of life in which the biological and psychosocial transition from childhood to adulthood occurs.

Coping—In psychology, a term that refers to a person's patterns of response to stress.

Culture—A test in which a sample of body fluid is placed on materials specially formulated to grow microorganisms. A culture is used to learn what type of bacterium is causing infection.

Discipline—In health care, a specific area of preparation or training, i.e., social work, nursing, or nutrition.

Family—Two or more emotionally involved people living in close proximity and having reciprocal obligations with a sense of commonness, caring, and commitment.

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Patau syndrome

Definition

Patau syndrome, also called trisomy 13, is a congenital (present at birth) disorder associated with the presence of an extra copy of chromosome 13. The extra chromosome 13 causes numerous physical and mental abnormalities, especially heart defects. Patau syndrome

is named for Klaus Patau, who reported the syndrome and its association with trisomy in 1960.

Description

Children normally inherit 23 chromosomes from each parent, for a total of 46 chromosomes. A typical human being has 46 chromosomes: 22 pairs of non-sex linked chromosomes and one pair of sex-linked chromosomes that determine that person's sex. Sometimes a child may end up with more than 46 chromosomes because of problems with the father's sperm or the mother's egg or because of mutations that occurred after the sperm and the egg fused to form the embryo (conception).

Normally, there are two copies of each of the 23 chromosomes: one from each parent. A condition called trisomy occurs when three, instead of two, copies of a chromosome are present in a developing human embryo. An extra copy of a particular chromosome can come either from the egg or sperm or from mutations that occur after conception.

The most well-known trisomy-related disorder is **Down syndrome** (trisomy 21), in which the developing embryo has an extra copy of chromosome 21. Patau syndrome is trisomy 13, in which the developing embryo has three copies of chromosome 13.

An extra copy of chromosome 13 is not the only cause of Patau syndrome. Other changes in chromosome 13, such as mispositioning (translocation), can also result in the characteristics classified as Patau syndrome. In these cases, an error occurs that causes a portion of chromosome 13 to be exchanged for a portion of another chromosome. There is no production of extra chromosomes, but a portion of each affected chromosome is "misplaced" (translocated) to another chromosome.

Patau syndrome causes serious physical and mental abnormalities, including heart defects; incomplete brain development; unusual facial features such as a sloping forehead, a smaller than average head (microcephaly), small or missing eyes, low set ears, and **cleft palate** or hare lip; extra fingers and toes (**polydactyly**); abnormal genitalia; spinal defects; seizures; gastrointestinal hernias, particularly at the navel (omphalocele); and **mental retardation**. Due to the severity of these conditions, fewer than 20 percent of those affected with Patau syndrome survive beyond infancy.

Demographics

Patau syndrome occurs in approximately one in 10,000 live births. In many cases, spontaneous abortion (miscarriage) occurs, which means the fetus does not survive to term. In other cases, the affected individual is stillborn. As appears to be the case in all trisomies, the risks of Patau syndrome seem to increase with the mother's age, particularly if she is over 30 when pregnant. Male and female children are equally affected, and the syndrome occurs in all races.

Causes and symptoms

The severity and symptoms of Patau syndrome vary with the type of chromosomal anomaly, from extremely serious conditions to nearly normal appearance and functioning. Full trisomy 13, which is present in the majority of the cases, results in the most severe and numerous internal and external abnormalities. Commonly, the forebrain fails to divide into lobes or hemispheres (holoprosencephaly), and the entire head is unusually small (microcephaly). The spinal cord may protrude through a defect in the vertebrae of the spinal column (myelomeningocele). Children who survive infancy have profound mental retardation and may experience seizures.

Incomplete development of the optic (sight) and olfactory (smell) nerves often accompany the brain defects described above. The eyes may be unusually small (microphthalmia) or one eye may be absent (anophthalmia). The eyes are sometimes set close together (hypotelorism) or even fused into a single structure. Incomplete development of any structures in the eye (coloboma) or failure of the retina to develop properly (retinal dysplasia) produces vision problems. Patau syndrome affected individuals may be born either partially or totally deaf, and many are subject to recurring ear infections.

The facial features of many Patau syndrome affected individuals appear flattened. The ears are generally malformed and lowset. Frequently, a child with trisomy 13 has a **cleft lip**, a cleft palate, or both. Other physical characteristics include loose folds of skin at the back of the neck, extra fingers or toes (polydactyly), permanently flexed (closed) fingers (camptodactyly), noticeably prominent heels, "rocker-bottom foot," and missing ribs. Genital malformations are common in individuals affected with Patau syndrome and include undescended testicles (cryptorchidism), an abnormally developed scrotum, and ambiguous genitalia in males, or an abnormally formed uterus (bicornuate uterus) in females.

In nearly all cases, Patau syndrome affected infants have respiratory difficulties and heart defects, including atrial and ventricular septal defects (holes between chambers of the heart); malformed ducts that cause

abnormal direction of blood flow (**patent ductus arteriosus**); holes in the valves of the lungs and the heart (pulmonary and aortic valves); and misplacement of the heart in the right, rather than the left, side of the chest (dextrocardia). The kidneys and gastrointestinal system may also be affected with cysts similar to those seen in polycystic kidney disease. These defects are frequently severe and life-threatening.

Partial trisomy of the distal segment of chromosome 13 results in generally less severe, but still serious, symptoms and a distinctive facial appearance including a short upturned nose, a longer than usual area between the nose and upper lip (philtrum), bushy eyebrows, and tumors made up of blood capillaries on the forehead (frontal capillary hemangiomata). Partial trisomy of the proximal segment of chromosome 13 is much less likely to be fatal and has been associated with a variety of facial features including a large nose, a short upper lip, and a receding jaw. Both forms of partial trisomy also result in severe mental retardation.

Beyond one month of age, other symptoms that are seen in individuals with Patau syndrome are: feeding difficulties and **constipation**, reflux disease, slow growth rates, curvature of the spine (**scoliosis**), irritability, sensitivity to sunlight, low muscle tone, high blood pressure, sinus infections, urinary tract infections, and ear and eye infections.

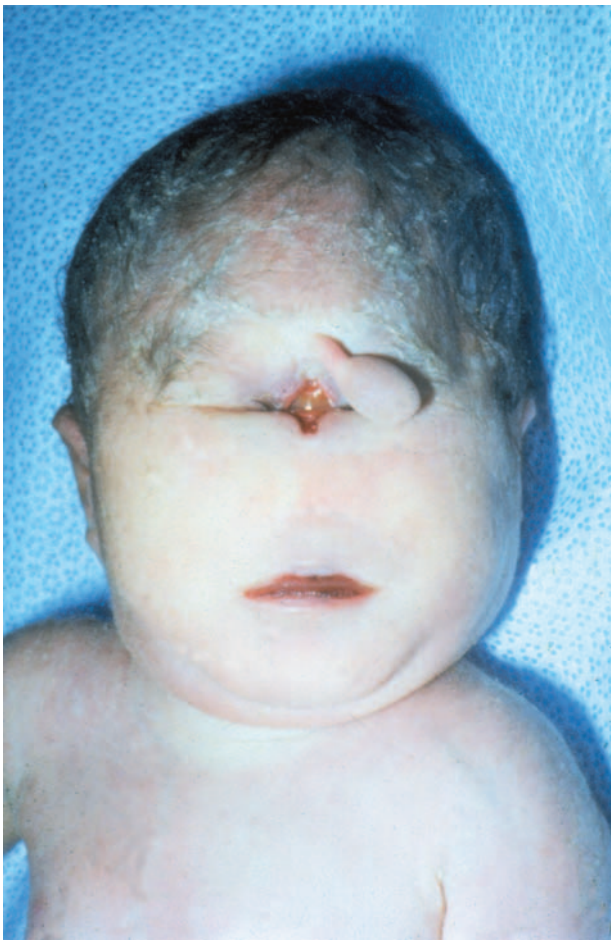
Diagnosis

Patau syndrome is detectable during pregnancy through the use of ultrasound imaging, **amniocentesis**, and chorionic villus sampling (CVS). At birth, the newborn's numerous malformations indicate a possible chromosomal abnormality. Trisomy 13 is confirmed by examining the infant's chromosomal pattern through karyotyping or another procedure. Karyotyping involves the separation and isolation of the chromosomes present in cells taken from an individual. These cells are generally extracted from cells found in a blood sample. The 22 non-sex linked chromosomes are identified by size, from largest to smallest, as chromosomes 1 through 22. The sex determining chromosomes are also identified. Patau syndrome is confirmed by the presence of three, rather than the normal two, copies of the thirteenth largest chromosome.

Treatment

Some infants born with Patau syndrome have severe and incurable birth defects. However, children with better prognoses require medical treatment to correct structural abnormalities and associated complications. For feeding problems, special formulas, positions, and techniques may be used. Tube feeding or the placement of a gastric tube (gastrostomy may be required. Structural abnormalities such as cleft lip and cleft palate can be corrected through surgery. Special diets, hearing aids, and vision aids can be used to mitigate some symptoms of Patau syndrome. Physical therapy, speech therapy, and other types of developmental therapy help the child reach his or her potential.

Since the translocation form of Patau syndrome is genetically transmitted, genetic counseling for the parents should be part of the management of the disease.



Stillborn term infant with Patau's syndrome. The baby has no eyes, no nose opening, and anelongated bulb hanging from forehead. *(© Ralph C. Eagle, M.D./Photo Researchers, Inc.)*

Prognosis

Approximately 45 percent of trisomy 13 babies die within their first month of life; up to 70 percent in the first six months; and over 70 percent by one year of age. Survival to adulthood is very rare. Only one adult is known to have survived to age 33.

KEY TERMS

Amniocentesis—A procedure performed at 16-18 weeks of pregnancy in which a needle is inserted through a woman's abdomen into her uterus to draw out a small sample of the amniotic fluid from around the baby for analysis. Either the fluid itself or cells from the fluid can be used for a variety of tests to obtain information about genetic disorders and other medical conditions in the fetus.

Chorionic villus sampling—A procedure used for prenatal diagnosis at 10–12 weeks gestation. Under ultrasound guidance a needle is inserted either through the mother's vagina or abdominal wall and a sample of the chorionic membrane. These cells are then tested for chromosome abnormalities or other genetic diseases.

Chromosome—A microscopic thread-like structure found within each cell of the human body and consisting of a complex of proteins and DNA. Humans have 46 chromosomes arranged into 23 pairs. Chromosomes contain the genetic information necessary to direct the development and functioning of all cells and systems in the body. They pass on hereditary traits from parents to child (like eye color) and determine whether the child will be male or female.

Karyotyping—A laboratory test used to study an individual's chromosome make-up. Chromosomes are separated from cells, stained, and arranged in order from largest to smallest so that their number and structure can be studied under a microscope.

Mosaicism—A genetic condition resulting from a mutation, crossing over, or nondisjunction of chromosomes during cell division, causing a variation in the number of chromosomes in the cells.

Translocation—The transfer of one part of a chromosome to another chromosome during cell division. A balanced translocation occurs when pieces from two different chromosomes exchange places without loss or gain of any chromosome material. An unbalanced translocation involves the unequal loss or gain of genetic information between two chromosomes.

Trisomy—An abnormal condition where three copies of one chromosome are present in the cells of an individual's body instead of two, the normal number.

Ultrasonography—A medical test in which sound waves are directed against internal structures in the body. As sound waves bounce off the internal structure, they create an image on a video screen. Ultrasonography is often used to diagnose fetal abnormalities, gallstones, heart defects, and tumors. Also called ultrasound imaging.

Most survivors have profound mental and physical disabilities; however, the capacity for learning in children with Patau syndrome varies from case to case. Older children may be able to walk with or without a walker. They may also be able to understand words and phrases, follow simple commands, use a few words or signs, and recognize and interact with others.

Prevention

There is no known way to prevent Patau syndrome though it can be diagnosed prenatally via amniocentesis.

Parental concerns

Parents of children born with Patau syndrome should prepare themselves for the possiblity of their child dying within days or weeks of birth, in addition to the poor survival rates past early childhood. Also, parents who have already had a child with the disease and want to have another child should discuss potential problems with their physician.

Resources

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Support Organization for Trisomy 18, 13, and Related Disorders (SOFT). 2982 South Union St., Rochester, NY 14624. Web site: <www.trisomy.org>.

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Patent ductus arteriosus

Definition

Patent ductus arteriosus (PDA) is a heart defect that occurs in infants when the ductus arteriosus (the temporary fetal blood vessel that connects the aorta and the pulmonary artery) does not close at birth.

Description

The ductus arteriosus is a temporary fetal blood vessel that connects the aorta and the pulmonary artery before birth. The ductus arteriosus should be present and open before birth while the fetus is developing in the uterus. Since oxygen and nutrients are received from the placenta and the umbilical cord instead of the lungs, the ductus arteriosus acts as a "short cut" that allows blood to bypass the deflated lungs and go straight out to the body. After birth, when the lungs are needed to add oxygen to the blood, the ductus arteriosus normally closes. The closure of the ductus arteriosus ensures that blood goes to the lungs to pick up oxygen before going out to the body. Closure of the ductus arteriosus usually occurs at birth as levels of certain chemicals, called prostagladins, change, and the lungs fill with air. If the ductus arteriosus closes correctly, the blood pumped from the heart goes to the lungs, back into the heart, and then out to the body through the aorta. The blood returning from the lungs and moving out of the aorta carries oxygen to the cells of the body. In some infants, the ductus arteriosus remains open (or patent), and the resulting heart defect is known as patent ductus arteriosus. In most cases, a small PDA does not result in physical symptoms. If the PDA is larger, health complications may occur.

In an average individual's body, the power of blood being pumped by the heart and other forces leads to a certain level of pressure between the heart and lungs. The pressure between the heart and lungs of an individual affected by PDA causes some of the oxygenated blood that should go out to the body (through the aorta) to return back through the PDA into the pulmonary artery. The pulmonary artery takes the blood immediately back to the lungs. The recycling of the already oxygenated blood forces the heart to work harder as it tries to supply enough oxygenated blood to the body. In this case, usually the left side of the heart grows larger as it works harder and must contain all of the extra blood moving back into the heart. This is known as a left-to-right or aortic-pulmonary shunt.

As noted, the size of the PDA determines how much harder the heart has to work and how much bigger the heart becomes. If the PDA is large, the bottom left side of the heart is forced to pump twice as much blood because it must supply enough blood to recycle back to the lungs and move out to the body. As the heart responds to the increased demands for more oxygenated blood by pumping harder, the pulmonary artery has to change in size and shape in order to adapt to the increased amount and force of the blood. In some cases, the increase in size and shape changes the pressure in the pulmonary artery and lungs. If the pressure in the lungs is higher than that of the heart and body, blood returning to the heart will take the short cut back into the aorta from the pulmonary artery through the PDA instead of going to the lungs. This backward flowing of blood does not carry much oxygen. If blood without much oxygen is being delivered to the body, the legs and toes will turn blue or cyanotic. This condition is called a shunt reversal.

When a PDA results in a large amount of blood being cycled in the wrong order, either through a left-to-right shunt or shunt reversal, the overworked, enlarged heart may stop working (congestive heart failure) and the lungs can become filled with too much fluid (pulmonary edema). At this time, there is also an increased risk for bacterial infection that can inflame the lining of the heart (endocarditis). These three complications are very serious.

Demographics

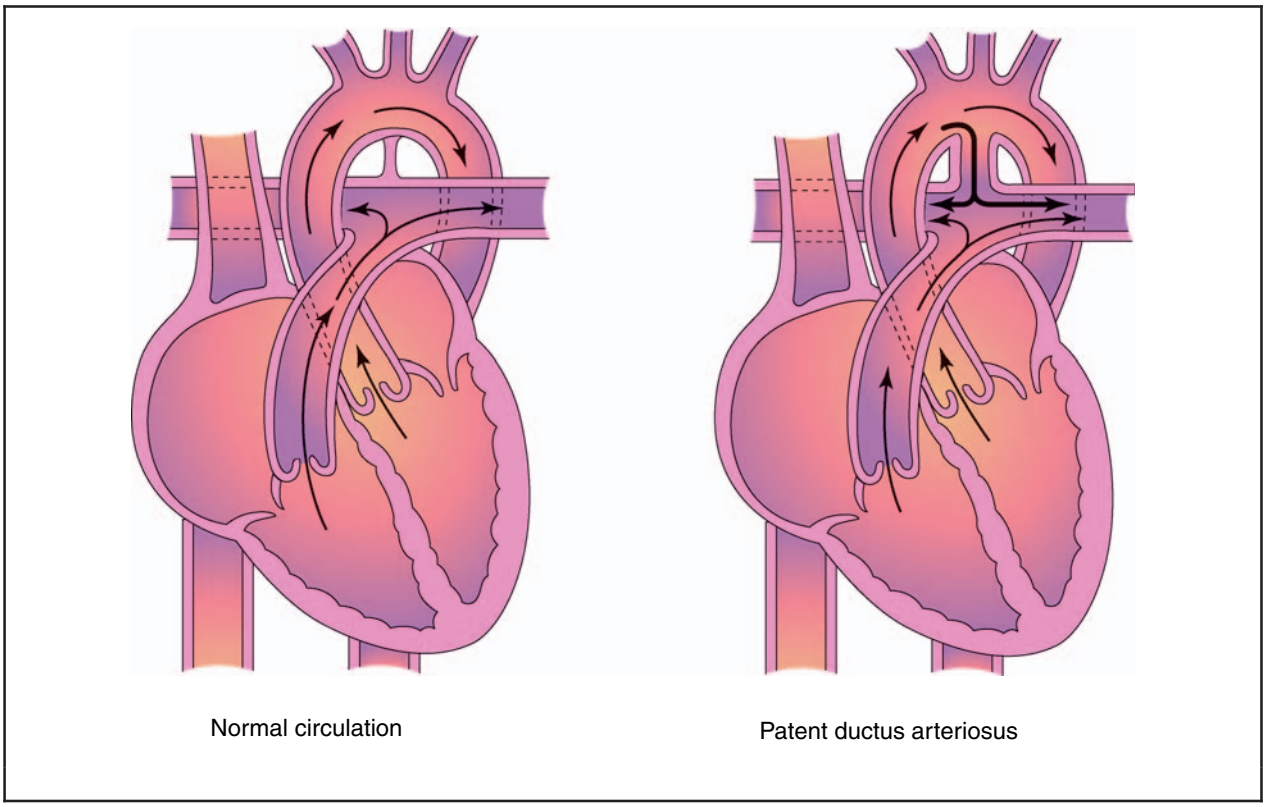
PDA is a very common heart defect, accounting for 5 to 10 percent of all types of **congenital heart disease**. Though an exact incidence of PDA is difficult to determine, researchers estimate that eight or nine in every 1,000 children are affected. PDA can occur in full-term infants, but it is seen most often in preterm infants, infants born at a high altitude, and babies whose mothers had a **rubella** infection during pregnancy. PDA occurs in individuals of every ethnic origin and does not occur more often in any one country or ethnic population; however, it is two to three times more common in females than males.

Causes and symptoms

PDA can be caused by environmental exposure before birth or the inheritance of a specific changed or mutated gene or genes. It can be a symptom of a genetic syndrome or may be caused by a combination of genetic and environmental factors (multifactorial).

Environmental exposures that can increase the chance for a baby to be affected by PDA include fetal exposure to rubella before birth, preterm delivery, and birth at a high altitude location.

PDA can be an inherited condition in families with isolated PDA or part of a genetic syndrome. In either case, there are specific gene changes or mutations which



Patent ductus arteriosus (PDA) is the failure of the ductus arteriosus to close after birth, allowing blood to inappropriately flow from the aorta into the pulmonary artery. *(Illustration by Electronic Illustrators Group.)*

lead to a defect in the elastic tissue forming the walls of the ductus arteriosus. As of 2004 the genes causing isolated PDA have not been identified, but it is known that PDA can be inherited through a **family** in an autosomal dominant pattern or an autosomal recessive pattern. Every person has approximately 30,000 genes, which tell the body how to grow and develop correctly. Each gene is present in pairs since one is inherited from the mother and one is inherited from the father. In an autosomal dominant condition, only one specific changed or mutated copy of the gene for PDA is necessary for a person to have PDA. If a parent has an autosomal dominant form of PDA, there is a 50 percent chance for each child to have the same or similar condition.

PDA can also be inherited in an autosomal recessive manner. A recessive condition occurs when a child receives two changed or mutated copies of the gene for a particular condition, such as PDA (one copy from each parent). Individuals with a single changed or mutated copy of a gene for a recessive condition are known as carriers and have no health problems related to the condition. However, when two people who each carry a changed or mutated copy of the same gene for a recessive condition have children, there is a chance with each pregnancy for the child to inherit the two changed or mutated copies from each parent. In this case, the child would have PDA. For two known carriers, there is a 25 percent risk with each child to have a child with PDA, a 50 percent chance to have a child who is a carrier, and a 25 percent chance to have a child who is neither affected nor a carrier.

Most cases of PDA occur as the result of multifactorial inheritance which is caused by the combination of genetic factors and environmental factors. The combined factors lead to isolated defects in the elastic tissue forming the walls of the ductus arteriosus.

The main sign of PDA is a constant heart murmur that sounds like the hum of a refrigerator or other machinery. This murmur is usually heard by the doctor using a stethoscope.

Other signs and symptoms of PDA include:

- shortness of breath after exertion such as crying, eating, or activity
- labored or fast breathing at rest

- cyanosis, or blue lips or fingernails especially after eating, crying, or activity
- problems with feeding and poor weight gain
- frequent colds and problems with the lungs

Diagnosis

Diagnosis is most often made by detecting the characteristic "machinery" heart murmur heard by a doctor through a stethoscope. Tests such as a chest x ray, echocardiograph, and ECG are used to support the initial diagnosis and to determine the severity of the PDA.

Treatment

The treatment and management of PDA depends upon the size of the PDA, the presence of other heart defects, and the symptoms experienced by the affected individual. In some cases, the PDA will close spontaneously and no further treatment will be required. In individuals for whom the PDA remains open, there are three treatment options: medical management, device closure, and surgical repair.

Medical management with medications such as indomethocin is often the first course of treatment in premature infants with PDA. Indomethocin causes the muscles within the heart to tighten, closing the PDA. In infants for whom indomethocin does not close the PDA, full-term infants, and older children, device closure or surgery may be necessary.

Device closure of PDA is a medical procedure in which a device such as a coil, very small rings of wire, or an occluder, a tiny wire mesh patch, is placed over the PDA causing the blood to clot and thus closing the open ductus. The device is inserted into the heart in a process called cardiac catheterization. A small thin flexible tube is inserted into a blood vessel, usually in the groin area, and guided into the heart. The device is then passed through the catheter to the PDA. For most patients, this procedure is highly effective, and no further treatment is required. In children for whom this procedure is unsuccessful, children with very large PDA, or those with multiple heart defects, surgery may be necessary.

In surgical closure of PDA, called a ligation, the chest cavity is opened, the blood vessels are separated, and the PDA is sewn closed.

Prognosis

Individuals can survive with a small opening remaining in the ductus arteriosus. Treatment, including surgery, of a larger PDA is usually successful and frequently occurs without complications. Proper treatment allows children to lead normal lives.

KEY TERMS

Aorta—The main artery located above the heart that pumps oxygenated blood out into the body. The aorta is the largest artery in the body.

Cyanosis—A bluish tinge to the skin that can occur when the blood oxygen level drops too low.

Ductus arteriosus—The temporary channel or blood vessel between the aorta and pulmonary artery in the fetus.

Echocardiogram—A record of the internal structures of the heart obtained from beams of ultrasonic waves directed through the wall of the chest.

Electrocardiagram (ECG, EKG)—A record of the electrical activity of the heart, with each wave being labeled as P, Q, R, S, and T waves. It is often used in the diagnosis of cases of abnormal cardiac rhythm and myocardial damage.

Endocarditis—Inflammation of the inner membrane lining heart and/or of the heart valves caused by infection.

Oxygenated blood—Blood carrying oxygen through the body.

Pulmonary artery—An artery that carries blood from the heart to the lungs.

Pulmonary edema—An accumulation of fluid in the tissue of the lungs.

Prevention

While there is no known prevention for PDA, appropriate prenatal care for expectant women is important and may prevent premature delivery, a major risk factor for PDA.

Parental concerns

Slow weight gain is common in children with congenital heart defects and may be alarming for parents. The pediatrician closely monitors the child's growth rate. There are many ways to increase the caloric intake of infants and children with PDA. A pediatrician, nurse, or dietitian can assist parents in ways to insure the child is getting proper **nutrition**. Infants with PDA may not be able to tolerate a large volume of breast milk or formula and therefore may need to be fed more frequently.

In addition to slow growth, children with PDA may be more susceptible to infections such as colds,

pneumonia, and a rare but potentially life threatening infection of the heart called endocarditis. Children with PDA may need to receive preventative **antibiotics** prior to dental work. Annual flu vaccines are recommended, and parents should watch for signs of infection such as **fever**, **cough**, chills, and any difficulty breathing.

When to call the doctor

Parents of children with PDA should watch for the following symptoms and contact the doctor if any of these occurs:

- problems feeding or slow weight gain
- difficulty breathing
- shortness of breath after crying, eating, or activity
- blue color in the lips and fingernails with activity
- fever of 100.4°F (38°C) or higher
- chills, cough, or lethargy
- itchy rash or swollen skin

Resources

ORGANIZATIONS

Congenital Heart Anomalies Support, Education, and Resources (CHASER). 2112 North Wilkins Rd., Swanton, OH 43558. Web site: <www.csun.edu/~hfmth006/chaser>.

Kids with Heart. 1578 Careful Dr., Green Bay, WI 54304. Web site: <www.execpc.com/~kdswhrt>.

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Peer acceptance

Definition

Peer acceptance is the degree to which a child or adolescent is socially accepted by peers. It includes the level of peer popularity and the ease with which a child or adolescent can initiate and maintain satisfactory peer relationships.

Description

Peer acceptance and relationships are important to children's social and emotional development. Peer acceptance and friendship provide a wide range of learning and development opportunities for children. These include companionship, recreation, building social skills, participating in group problem solving, and managing competition and conflict. They also allow for self-exploration, emotional growth, and moral and ethical development. Parents, teachers, and other adults are a good source of social support for children, but it is among other children that kids learn how to interact with each other.

When examining peer acceptance among children, researchers usually look at two areas that are related to a child's psychological and social development. The first area is the child's social standing in the peer group as a whole and is indicated by the child's level of social acceptance by other members in the group, usually classmates. The second area is the child's individual friendships, characterized by both the quantity and quality of these friendships.

Although genes may be a factor in a child's **social competence** and level of peer acceptance, environmental factors are also extremely important. Some of the factors contributing to peer acceptance include:

- the quality of attachment between mother or primary caregiver and child during infancy
- during childhood, the quantity and quality of opportunities for interaction with different types of peers in different environments, such as in the **family**, at school, church, camp, activity centers, in **sports**, or in the neighborhood
- parenting style (A highly nurturing but moderately controlling authoritative parenting style is associated with the highest levels of social competence. By contrast, a low nurturing, highly controlling authoritarian parenting style is associated with children's aggressiveness, while the high nurturing but low-controlling permissive style is associated with failure to take responsibility for behavior.)

Regarding having friends, the academic benefits show up very early in a child's school career. Research suggests that those who start kindergarten from **preschool** with a friend in their class make a better adjustment to school than those who do not start with a friend. Furthermore, children who maintain their friendships as the school year progresses like school better, and children who make new friends make greater gains in school performance.

Infancy and toddlerhood

The first step in childhood affiliations is the categorization of people into groups. Although some researchers believe that the ability to categorize is an achievement of toddlerhood, others suggest it is present in infancy. In children, the top three categories of peer affiliation are age, sex, and race. Children do not appear to make racial distinctions before they are of preschool age but age and sex discriminations are made earlier. There is evidence that infants make categorical distinctions between males and females and between adults and children before they are a year old. Signs of a preferential attraction to others like the self also appear at an early age. Year-old infants are interested in and attracted to other infants—including those they have never met before—at an age when they are wary of strange adults. By the age of two, they begin to show a preference for children of their own sex. There is also research that suggests the quality of attachment between mothers or primary caregivers during infancy can contribute to peer acceptance later in childhood.

Customs of child rearing and patterns of parent-infant interaction vary widely from culture to culture, but the children's playgroup is universal. If the number of children in a given locality is small, the playgroup will consist of children of both sexes and a range of ages; if the number is larger, the children generally divide up into age- and sex-segregated groups. Girls' groups tend to be split up into subgroups. It is the social category, or psychological group that is important here. Children can categorize themselves as members of a social category even if it does not assemble in one place.

Preschool

It is important to recognize the role of the peer group in maintaining a preschool-age child's level of social acceptance. Once a child has established a reputation among peers either as someone with whom it is fun to **play** or as someone with whom joint play is unpleasant or dissatisfying, this reputation may influence the way other children perceive the child's later behavior. If a negative reputation is developed, helping the child become accepted may require more than a change in the child's behavior; it may also be necessary to point out to the other children when the child's behavior changes and to guide them to respond to the child in positive ways.

Research on imaginary companions suggests that young children who create them do so to compensate for poor social relationships, according to a study published in the May 2004 issue of the *International Journal of Behavioral Development*. As a result, there is less peer acceptance of children with imaginary companions. Several other studies have shown that fantasy play is also related to peer acceptance in children in preschool. Using a scoring system that included the reality and unfamiliarity levels of fantasy play, researchers found players who scored high had higher self-ratings of peer acceptance than did average scoring fantasy players. However, the high scoring fantasy players had lower teacher ratings of peer acceptance than the average scoring fantasy players. Researchers suggest the difference may occur because the high scoring fantasy players were unable to distinguish imagined popularity from actual peer acceptance.

School age

In school-aged children, factors such as physical attractiveness, cultural traits, and disabilities greatly affect the level of peer acceptance, with a child's degree of social competence being the best predictor of peer acceptance. The peer groups of **adolescence**, especially teens, are often based on athletic, social, or academic interests and abilities; on distinctions of race, ethnicity, and social class; and on proclivities such as drug use and delinquency. Children who are peer-accepted or popular have fewer problems in middle and high school, and teens who are peer-accepted have fewer emotional and social adjustment problems as adults. Peer-accepted children may be shy or assertive, but they often have well-developed **communication skills**.

Peer-accepted children tend to be able to function in the following ways:

- Correctly interpret other children's body language and tone of voice. Well-liked children can distinguish subtleties in emotions. For example, they can distinguish between anger directed toward them and anger directed toward a parent.
- Directly respond to the statements and gestures of other children. Well-liked children will say other children's names, establish eye contact, and use touch to get attention.
- Give reasons for their own statements, gestures, and actions. For example, well-liked children will explain why they want to do something the other child does not want to do.
- Cooperate with, show tact towards, and compromise with other children, demonstrating the willingness to subordinate the self by modifying behavior and opinions in the interests of others. For example, when joining a new group where a conversation is already in progress, well-liked children will listen first, establishing a tentative presence in the group before speaking, even if it is to change the subject.

These skills are crucial in initiating and maintaining relationships and in resolving conflicts. By contrast, rejected children tend either towards aggressive, **antisocial behavior** or withdrawn, depressive behavior. They also do not listen well, tend not to offer reasons for their behavior, do not positively reinforce their peers, and have trouble cooperating. Antisocial children interrupt people, dominate other children, and either verbally or physically attack them. Depressive or withdrawn children may be excessively reserved, submissive, anxious, and inhibited. Competitiveness or dominance by itself is not necessarily indicative of low peer acceptance. In fact, popular children tend to have characteristics associated with both competitiveness and friendliness.

The need to be "one of the gang" is stronger as children approach the teen years than at any other age. Children of all ages need to feel that they fit in—that they belong.

Common problems

Children learn to relate to peers by engaging in peer relationships. Some children have problems making friends or "fitting in." Often a vicious circle develops where a rejected child is given fewer and fewer opportunities by his peers to relate and thereby learn new skills. Lack of opportunity to participate normally in peer interaction is especially a problem for children who differ in some obvious way, either culturally, racially, or through some mental or physical disability. Parents and teachers should address issues of peer acceptance as early as possible in order to prevent loss of self-confidence and **self-esteem**.

In addition to providing direct social skills training or counseling for the child with peer acceptance problems, parents and teachers can create opportunities for non-threatening social interaction to occur. Though children should never be forced to play together since this can create the rejection it is intended to remedy, popular and less-popular preschoolers can be encouraged to interact with one another. For example, a less sociable child may be encouraged to answer and ask questions of others. Older children should be provided opportunities to interact in smaller groups and in one-on-one situations, where it may be easier to try out new behaviors and make up for social mistakes. Shy or withdrawn children can be encouraged to develop outside interests that will place them in structured contact with others. In school, peer helping programs and collaborative learning provide opportunities for popular and less-popular children to work together. Ideally, collaboration should highlight the less-popular students' strengths, such as special interests and talents, rather than weaknesses. At any age, the small, positive changes in behavior should be reinforced with attention and praise.

KEY TERMS

Fantasy play—Play activities in which children act out their fantasies.

Gene—A building block of inheritance, which contains the instructions for the production of a particular protein, and is made up of a molecular sequence found on a section of DNA. Each gene is found on a precise location on a chromosome.

Primary caregiver—A person who is responsible for the primary care and upbringing of a child.

Parental concerns

Peer rejection in childhood often brings with it serious emotional difficulties. Rejected children are frequently discontent with themselves and with their relationships with other children. Many of these children experience strong feelings of loneliness and social dissatisfaction. Rejected children also report lower self-esteem and may be more depressed than other children. Peer rejection is also predictive of later life problems, such as dropping out of school, juvenile delinquency, and mental health problems. Dropping out of school seems to be a particularly frequent outcome. Results from research indicate that, on average, about 25 percent of low-accepted children drop out of school compared to 8 percent of other children, according to the National Network for Child Care at Iowa State University.

There are various reasons why children are disliked by their peers. When trying to find ways to help these children, it is easy to fall into the trap of thinking about what they do that bothers others. This focuses only on reducing these behavior problems but most rejected children also lack important social skills. They may not cooperate or be responsive to others, or they may not know how to respond in certain social situations. Teaching a child the missing skills is often more effective in improving peer relationships than working only on reducing negative behavior.

When to call the doctor

Parents may need to seek professional psychological help for children who suffer from peer rejection, especially when the child is depressed or shows overly **aggressive behavior**. Help may also be needed for adolescents whose acceptance by peers relates to common

negative behaviors, such as gang affiliation, bullying, **smoking**, and drug and alcohol abuse.

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Ken R. Wells

Peer pressure

Definition

Peer pressure is the influence of a social group on an individual.

Description

Children and teenagers feel social pressure to conform to the group of peers with whom they socialize. This peer pressure can influence how children dress, what kind of music they listen to, and what types of behavior they engage in, including risky behaviors such as using drugs, cigarettes, and alcohol, and engaging in sex. The intensity of peer pressure differs from situation to situation.

Peer groups are usually cliques of friends who are about the same age. Peer pressure can begin in early childhood with children trying to get other kids to **play** the games they want. It generally increases through childhood and reaches its intensity in the preteen and teen years. Virtually all adolescents in middle and high school deal with peer pressure, often on a daily basis. It is how children and teens learn to get along with others of their own age group and eventually learn how to become independent. Depending on the group trying to apply the influence, peer pressure can be negative or positive.

Starting in middle school, children begin to spend more time with their friends and less time with their parents and **family**. Although some children remain loners and not part of any group, most preteens tend to be part of a small group of friends called a clique. In children ages eleven to fourteen, it is most common for members of these cliques to be of the same sex. Children will spend a lot of time with friends in their clique, interacting by going to the movies or the mall, talking on the

telephone, or chatting online with instant messaging. They know which kids belong to particular cliques and who the loners are. Within the cliques, talk about the opposite sex is popular as is making arrangements for out of school activities.

Children also generally belong to a crowd, which is a larger group of kids from several cliques. While members of the cliques are close friends, members of the crowd outside a clique are casual acquaintances. Crowds are often large groups with common interests such as athletes (jocks), kids who like school (preppies), kids lacking good looks or social skills but who excel at particular intellectual interests (nerds), and drug users (druggies).

Some kids give in to peer pressure because they want to be liked, to fit in, or because they worry that other kids may make fun of them if they do not go along with the group. Others may go along because they are curious to try something new that others are doing. The idea that "everyone is doing it" may influence some kids to ignore their better judgment or their common sense. Peer pressure can be extremely strong and seductive. Experiments have shown how peer pressure can influence children to change their minds from what they know for sure is acceptable behavior to unacceptable behavior just because everyone else in their peer group is doing it. These studies have also shown that all it takes for individuals to stand their ground on what they know is right is for one other peer to join them. That principle holds true for youth of any age in peer pressure situations, according to the Online organization KidsHealth (<www.kidshealth.org>).

Children and adolescents cannot always avoid negative peer pressure. It may continue to be a fact of life through childhood, **adolescence**, and into adulthood. Quoted from an article in the September 2002 issue of *Current Health 2, A Weekly Reader Publication*, the following are strategies young people can use to deal with negative peer pressure effectively:

- Avoid putting yourself in situations that make you feel uncomfortable. For example, if you don't want to start **smoking**, stay away from areas where you know kids go to smoke.
- Choose your friends wisely. If you hang around with people who share your values, chances are you'll never be asked to do something you don't want to do.
- Think about the consequences whenever you are asked to do something you are not sure about. Stop for a moment and ask: Will this activity get me in trouble? Will it be harmful to my health?
- Be true to yourself. Think about the reasons why you are considering doing something you are uncomfortable with. Is it to gain popularity? Although there is nothing wrong with wanting to be popular, there are right ways and wrong ways to achieve it. If you change your behavior just to fit in with a particular group, you are not being true to yourself.
- Learn how to say no. This is perhaps the most difficult thing in the world for many people to do, but it is an essential skill if you are to successfully fend off negative peer pressure. There are many ways to say no, some of them subtle and some of them a little more "in your face." Several examples are: "You see it your way. I see it my way." "If you are really a friend, then back off." "You must think I'm pretty dumb to fall for that one."

Toddlerhood

Peer pressure can be found in groups as young as age two, when children will do things simply because other kids are doing it or tell them to. This can effect the child's behavior, social and emotional development, eating habits, play time, and sleeping patterns.

Preschool

Preschoolers will go out of their way to think and act like their friends, even though they know it may go against what they have been taught by their parents. At the ages of three and four, children start to see there are other values, opinions, and rules besides those set by their parents. They may demand to do things that their parents do not allow, such as watching television beyond a certain time or time limit, eating junk food, and playing with **toys** their parents do not deem appropriate, such as toy guns, simply because their friends do so.

At this age, it is normal for children to start challenging their parents, testing the limits and rules to see how far they can bend or break them. Many pediatricians suggest parents should remain firm, not overreact, and then move on. Peer pressure can have positive benefits for preschoolers, such as taking a nap or eating vegetables when they see their friends doing it.

School age

At ages five to eight, children make a concerted effort to please their friends, classmates, and playmates. Peer pressure can be a positive influence if friends encourage each other to strive to do better in school, **sports**, and creative activities. For example, a child may try harder at soccer if he or she has a friend who does well or may read more if that is what a friend does.

Peer pressure can also have a negative influence on children ages five to eight when a friend or friends encourage them to act in a way that is not natural for the child. Many pediatricians and child psychologists say it is best not to prohibit the child from hanging out with these friends but to make sure the child is aware of the consequences of unacceptable behavior. Focus on specific negative behaviors and explain why they are bad. Most children will not respond well if a parent or primary caregiver forbids them to associate with a friend or group of friends.

The effects of peer pressure usually begin to be seen heavily by middle school and through high school. As children turn into adolescents, involvement with their peers and the attraction of peer identification increases. Teens begin to experience rapid physical, emotional, and social changes, and they begin to question adult standards and the need for parental guidance. It is reassuring for teens to turn for advice to friends who understand and sympathize with them.

Adolescents expand their peer relationships to occupy a central role in their lives, often replacing their parents and family as their main source of advice, socializing, and entertainment activities. The peer group is a source of affection, sympathy, understanding, and experimentation. It is also a supportive setting for achieving the two primary developmental tasks of teens: finding answers to questions about their identity and discovering their autonomous self that is separate and independent from their parents.

At adolescence, peer relations expand to occupy a particularly central role in young people's lives. New types (opposite sex, romantic ties) and levels (crowds) of peer relationships emerge. Peers typically replace the family as the center of a young person's socializing and leisure activities. Teenagers have multiple peer relationships, and they confront multiple peer cultures that have remarkably different norms and value systems. The perception many adults have that peer pressure is one culture or a unified front of dangerous influence is inaccurate. More often than not, peers reinforce family values, but they have the potential to encourage problem behaviors as well. Although the negative peer influence is overemphasized, more can be done to help teenagers experience the family and the peer group as mutually constructive environments. The following are facts about parent, adolescent and peer relations.

- During adolescence, parents and adolescents become more physically and psychologically distant from each other. This normal distancing is seen in decreases in emotional closeness and warmth, increases in parent-adolescent conflict and disagreement, and an increase in time adolescents spend with peers. Unfortunately, this tendency sometimes is encouraged by parents who are emotionally unavailable to their teenaged children.
- Increases in family strains such as economic pressures or **divorce** may prompt teenagers to depend more on peers for emotional support. By the high school years, most teenagers report feeling closer to friends than parents. Stress caused by work, marital dissatisfaction, family break-up caused by divorce, entering a step-family relationship, lower family income or increasing expenses, all produce increased individual and family stress.
- In 10 to 20 percent of families, parents and adolescents are in distressed relationships characterized by emotional coldness and frequent outbursts of anger and conflict. Unresolved conflicts produce discouragement and withdrawal from family life. Adolescents in these families are at high risk for various psychological and behavioral problems.
- Youth **gangs**, commonly associated with inner-city neighborhoods, are a recognizable peer group among youth in smaller cities, suburbs, and even rural areas. Gangs are particularly visible in communities with a significant portion of economically disadvantaged families and when parents are conflicted, distant, or unavailable.
- Formal dating patterns of the 1980s have been replaced in the early 2000s with informal socializing patterns in mixed-sex groups. This may encourage casual sexual relationships that heighten the risk of exposure to human **immunodeficiency** virus (HIV) and other sexually transmitted diseases.
- There has been an increase in part-time employment among youth, but it has had little impact on peer relations. To find time for work, teenagers drop **extracurricular activities**, reduce time spent on homework, and withdraw from family interactions, but they protect time spent with friends.

Common problems

Negative peer pressure occurs when a child's or teen's friends or other people their age try to convince them to do something that is either harmful to their body or is against the law. Examples include drinking alcohol, taking drugs, smoking cigarettes, cutting classes, vandalizing, and **stealing**. Although teens usually know when something is bad for them, they often choose to do it because they want to be liked, to fit in, to be accepted, or because they're afraid they'll be looked down upon or made fun of.

KEY TERMS

Clique—A close group of friends having similar interests and goals and whom outsiders regard as excluding them.

Human immunodeficiency virus (HIV)—A transmissible retrovirus that causes AIDS in humans. Two forms of HIV are now recognized: HIV-1, which causes most cases of AIDS in Europe, North and South America, and most parts of Africa; and HIV-2, which is chiefly found in West African patients. HIV-2, discovered in 1986, appears to be less virulent than HIV-1 and may also have a longer latency period.

Primary caregiver—A person who is responsible for the primary care and upbringing of a child.

Bruce A. Epstein in "How to combat negative peer pressure," in the September 2002 issue of *Current Health 2, A Weekly Reader Publication*, is quoted as saying, The "desire to be accepted by their peers is perhaps the strongest motivating force during dolescence." Many studies reinforce his theory. One study showed, for example, that a student who knew the correct answer to a question gave the wrong answer just because everyone else in the class gave the wrong answer.

There are various reasons why children are disliked by their peers. When trying to find ways to help these children, it is easy to fall into the trap of thinking about what they do that bothers others. This focuses only on reducing these behavior problems but most rejected children also lack important social skills. They may not cooperate or be responsive to others, or they may not know how to respond in certain social situations. Teaching a child the missing skills is often more effective in improving peer relationships than working only on reducing negative behavior.

Parental concerns

Peer rejection in childhood often brings with it serious emotional difficulties. Rejected children are frequently discontent with themselves and with their relationships with other children. Many of these children experience strong feelings of loneliness and social dissatisfaction. Rejected children also report lower **self-esteem** and may be more depressed than other children. Peer rejection is also predictive of later life problems, such as dropping out of school, juvenile delinquency, and mental health problems. Dropping out of school seems to be a particularly frequent outcome. Results from research indicate that, on average, about 25 percent of low-accepted children drop out of school compared to 8 percent of other children, according to the National Network for Child Care at Iowa State University.

When to call the doctor

Parents may need to seek professional psychological help for children suffering from peer rejection, especially when the child is depressed or shows overly **aggressive behavior**. Help may also be needed for adolescents whose acceptance by peers relates to common negative behaviors, such as criminal activities, gang affiliation, bullying, smoking, and drug and alcohol abuse. Professional psychological help may also be needed if the child is depressed. If the child talks about or threatens **suicide**, professional help should be sought immediately.

Resources

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National Academy of Child & Adolescent Psychiatry. 3615 Wisconsin Ave. NW, Washington, DC 20016. Web site: <www.aacap.org>.

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Ken R. Wells

Penicillins

Definition

Penicillins are a group of closely related **antibiotics** that kill bacteria.

Description

There are several types of penicillins, each used to treat different kinds of infections, such as skin infections, dental infections, ear infections, respiratory tract infections, urinary tract infections, gonorrhea, and other infections caused by bacteria. These drugs will not work for olds, flu, and other infections caused by viruses.

Examples of penicillins are penicillin V (Beepen-VK, Pen-Vee K, V-cillin K, Veetids) and amoxicillin (Amoxil, Polymox, Trimox, Wymox). Penicillins are sometimes combined with other ingredients called beta-lactamase inhibitors, which protect the penicillin from bacterial enzymes that may destroy it before it can do its work. The drug Augmentin, for example, contains a combination of amoxicillin and a beta-lactamase inhibitor, clavulanic acid. Penicillins are available only with a prescription.

The original form of penicillin is called penicillin G. It is a narrow-spectrum antibiotic, which can be destroyed by stomach acid, but it is still useful against anaerobic bacteria (bacteria that can live in the absence of air). Newer penicillins are resistant to stomach acid, such as penicillin V, or have a broader spectrum, such as ampicillin and amoxicillin.

General use

Penicillins are useful against infections in many parts of the body, including the mouth and throat, skin and soft tissue, tonsils, heart, lungs, and ears. However, since many bacteria are resistant to penicillin, it is often wise to do a culture and sensitivity test before using penicillins. In some cases, there are only a few types of bacteria that are likely to be a problem, and so it is appropriate to use a penicillin without testing. For example, dentists often prescribe penicillin to prevent infections after dental surgery.

Precautions

Penicillins are usually very safe. The greatest risk is an allergic reaction, which can be severe. People who have been allergic to cephalosporins are likely to be allergic to penicillins. Moreover, people with certain medical conditions or who are taking certain other medicines can have problems if they take penicillins. Before taking these drugs, patients should be sure to let the physician know about any of the following conditions.

Low-sodium diet

Some penicillin medicines contain large enough amounts of sodium to cause problems for people on low-sodium diets. Parents of children on on such a diet should make sure that the physician treating the infection knows about the special diet.

Diabetes

Penicillins may cause false positive results on urine sugar tests for diabetes. People with diabetes should check with their physicians to see if they need to change their diet or the doses of their diabetes medicine.

Phenylketonuria

Some formulations of Augmentin contain phenylalanine. People with **phenylketonuria** (PKU) should consult a physician before taking this medicine.

Side effects

The most common side effect of penicillin is **diarrhea**. **Nausea**, **vomiting**, and upset stomach are also

common. With some penicillins, particularly the broad spectrum products, there is a risk of increased growth of organisms that are not affected by penicillin. This situation can lead to candidal infections of the mouth and vagina.

Most side effects of penicillin cannot be prevented. Amoxicillin has a lower incidence of diarrhea than ampicillin and is the preferred drug in most cases.

Interactions

Birth control pills may not work properly when taken at the same time as penicillin. Penicillins may also interact with many other medicines. When this happens, the effects of one or both of the drugs may change or the risk of side effects may be greater. People who take penicillin should let their physician know all other medicines they are taking. Among the drugs that may interact with penicillins are the following:

- acetaminophen (Tylenol) and other medicines that relieve **pain** and inflammation
- medicine for overactive thyroid
- other antibiotics
- blood thinners
- antiseizure medicines such as Depakote and Depakene
- blood pressure drugs such as Capoten, Monopril, and Lotensin

The list above does not include every drug that may interact with penicillins. A physician or pharmacist should be consulted before a patient combines penicillins with any other prescription or nonprescription (over-the-counter) medicine.

Parental concerns

Parents should verify that their children have an infection requiring antibiotic therapy. Unnecessary use of antibiotics leads to development of bacterial resistance, while it subjects the child to some needless risk of adverse effects and wastes money.

Liquid forms of penicillin should be refrigerated after reconstitution. These preparations must be shaken well before use and measured with a medicinal teaspoon, not a household teaspoon.

Any adverse effects should be discussed with the prescriber. Penicillin should not be used in patients allergic to the drug; however, an incorrect report of an allergy to penicillin may cause prescribers to select a different drug which may cause even more severe side effects.

KEY TERMS

Anaerobic—An organism that grows and thrives in an oxygen-free environment.

Beta-lactamase—An enzyme produced by some bacteria that destroys penicillins.

Broad spectrum—A term applied to antibiotics to indicate that they are effective against many different types of bacteria.

Enzyme—A protein that catalyzes a biochemical reaction without changing its own structure or function.

Microorganism—An organism that is too small to be seen with the naked eye, such as a bacterium, virus, or fungus.

Mononucleosis—An infection, caused by the Epstein-Barr virus, that causes swelling of lymph nodes, spleen, and liver, usually accompanied by extremely sore throat, fever, headache, and intense long-lasting fatigue. Also called infectious mononucleosis.

Penicillins should be administered exactly as directed. Users should never give larger, smaller, more frequent, or less frequent doses. To make sure the infection clears up completely, patients should take the medicine for as long as it has been prescribed. They should not stop taking the drug just because symptoms begin to improve. This point is important with all types of infections, but it is especially important with strep infections, which can lead to serious heart problems if they are not cleared up completely.

This medicine should be used only for the infection for which it was prescribed. Different kinds of penicillins cannot be substituted for one another. Do not save some of the medicine to use on future infections. It may not be the right treatment for other kinds of infections, even if the symptoms are the same.

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Perforated eardrum

Definition

A perforated eardrum (tympanum perforation) is an opening or rupture in the eardrum (tympanic membrane), the thin membrane that separates the outer ear canal from the middle ear. A perforated eardrum may be caused by infection, trauma, or negative pressure from underwater diving or an airplane flight. The hole or rupture can cause temporary hearing loss, **pain**, and occasional discharge.

Description

The eardrum (tympanic membrane) is a thin, semi-transparent membranous wall that stretches across the ear canal and separates the outer ear from the middle ear. The side that faces outward into the ear canal is covered with skin and the inside is covered with mucous membrane. The eardrum vibrates when sound waves travel into the ear canal and strike it. One of the bones of the middle ear (the malleus) attaches to the center of the membrane and receives vibrations, transmitting them to other bones (the incus and stapes) and the inner ear fluid, and finally to nerves in the brain where sound is perceived. The middle ear is connected to the nose by the eustachian tube, a narrow channel that runs from the ear drum to the back of the throat.

In addition to conducting sound, the eardrum also protects the middle ear from bacteria and possible infection. When perforation occurs, bacteria can pass more easily into the middle ear, potentially causing ear infections.

Perforation is most commonly caused by either chronic or acute infection of the middle ear, usually related to infection of the nose and throat (nasopharynx). It may also be the result of trauma from direct injury, pressure, or loud noise. In general, the larger the opening in the eardrum, the greater the potential for temporary hearing loss. The location of the perforation also affects the degree of hearing loss. Severe hearing loss may follow a skull fracture that disrupts the bones in the middle ear. Eardrum perforation caused by a loud noise may result in disturbing ear noise (tinnitus) as well as a temporary hearing loss. Tinnitus usually fades in a few days and, over time, hearing loss improves.

Demographics

Perforated eardrum occurs commonly in people of all ages; it is especially common in early childhood when children are exposed regularly to colds and upper respiratory infections in their contact with other children. Middle ear infection, the most frequent cause of perforated eardrum, is the most prevalent reported illness in children between six months and 20 months of age.

Causes and symptoms

Middle ear infection (**otitis media**) is the most common cause of perforation of the eardrum. Infection usually stems from an upper respiratory infection in which swelling (edema) in the eustachian tube causes fluid and mucous to gather behind the eardrum. Bacteria that find their way from the nasopharynx into the built up fluid may cause a middle ear infection. The resulting congestion builds up pressure behind the eardrum, causing severe pain and spontaneous rupture, which reduces the pain immediately. Infected or bloody mucus may then drain from the ear. In some cases, the doctor may decide to rupture the membrane on purpose, making an incision (**myringotomy**) that relives pressure, reduces pain, and allows the infection to drain. Chronic middle ear infections can also erode a hole in the eardrum, which can eventually destroy the tiny bones of the middle ear and increase the likelihood of infected material passing from the nasopharynx into the middle ear, causing more infections.

The eardrum can also become damaged by direct injury from a foreign object or sudden noise. It is possible to perforate the eardrum by injury from the following:

- cotton-tipped swabs
- trauma, such as a hard bang or hitting the ear with an open hand
- fracture of the skull
- a nearby explosion or other extremely loud noise

Other causes of tympanum perforation include pressure trauma (barotrauma) injury such as the following:

- sudden change of air pressure during diving into water
- sudden change of air pressure during air travel
- middle ear tumor, which puts pressure against the inner ear drum

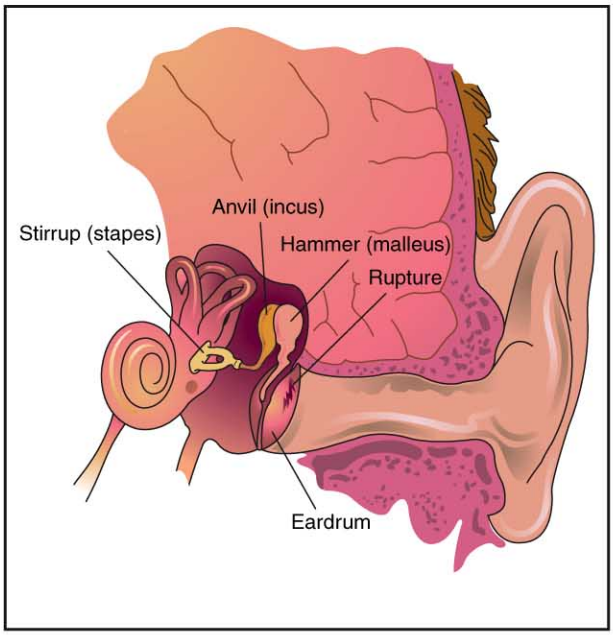
Symptoms may include an earache or severe pain in the ear or a sudden decrease in ear pain followed by drainage of clear, bloody, or pus-filled fluid. Hearing loss may be the first symptom experienced, either immediate or delayed, but will usually be restored. Tinnitus may occur as buzzing, swishing, or ringing, which will typically subside after a few days. Rarely, a small hole may remain in the eardrum after a pressure-equalizing tube (tympanostomy tube) falls out or is removed by a doctor. Tympanostomy tube insertion can also cause perforation.

When to call the doctor

If a child with a cold or upper respiratory infection cries constantly or complains of pain in the ear, it is wise to have the ears checked by a physician. Likewise, if the child seems to have pain or difficulty hearing after any type of trauma to the ear (injury with a foreign object, a bang or slap to the head, exposure to extremely loud noise, or after recent air travel), the doctor should be consulted. Early treatment of ear infection may help avoid perforation or hearing loss.

Diagnosis

The doctor may examine the ear with an otoscope, a microscope-type device with a light source for direct inspection of the ear. This examination makes possible the diagnosis of eardrum perforation by allowing the doctor to see an opening in the eardrum or damage to bones in the middle ear. Hearing tests with an audiogram may be done to measure the extent of any hearing loss. If drainage from infection is present, the doctor may have the material cultured in the laboratory to identify the organism causing infection. The nose and throat may also be cultured to see if the same organism is responsible for an upper respiratory infection. MRI or CT imaging studies may be done to rule out skull fracture, middle ear tumor, or acoustic neuroma, which may cause pain and hearing loss and be confused with a possible perforated eardrum.



A perforated eardrum is a hole or rupture in the eardrum, the thin membrane that separates the outer ear canal from the middle ear. It may result in temporary hearing loss and occasional discharge. *(Illustration by Electronic Illustrators Group.)*

Treatment

Treatment of a perforated eardrum is aimed at relieving pain or pressure behind the eardrum, treating any infection of the ear itself or of the upper respiratory tract, and restoring hearing. **Antibiotics** are usually the treatment of choice for existing ear infection or to prevent infection and reduce the likelihood of recurrence. Painkillers can relieve associated ear pain. Sometimes the doctor may lance a bulging eardrum and place a tympanostomy tube in it to relieve pain and pressure and allow the ear infection to drain before spontaneous rupture occurs.

A perforated eardrum usually heals by itself within two months. Sometimes, a paper patch is placed over the eardrum until the membrane heals. Three or four patches may be needed before the perforation closes completely. If the eardrum does not heal on its own, surgical repair (tympanoplasty) may be necessary to resolve the infection and restore hearing.

KEY TERMS

Edema—The presence of abnormally large amounts of fluid in the intercellular tissue spaces of the body.

Eustachian tube—A thin tube between the middle ear and the pharnyx. Its purpose is to equalize pressure on either side of the ear drum.

Otitis media—Inflammation or infection of the middle ear space behind the eardrum. It commonly occurs in early childhood and is characterized by ear pain, fever, and hearing problems.

Otoscope—A hand-held instrument with a tiny light and a funnel-shaped attachment called an ear speculum, which is used to examine the ear canal and eardrum.

Tinnitus—A noise, ranging from faint ringing or thumping to roaring, that originates in the ear not in the environment.

The ear should be kept clean and dry while the eardrum heals; it may help to insert ear plugs into the ears when showering or shampooing to block any water from getting in. (Cotton balls are not advised because they actually help moisture get into the ear through a wicking effect.) Pain in the ear may be eased by applying warm compresses.

Prognosis

While a perforated eardrum may be uncomfortable, it usually heals on its own within two months. Any hearing loss or ear noise that accompanies the perforation is usually temporary.

Prevention

A perforated eardrum can be prevented by avoiding insertion of any object into the ear to clean it or to remove ear wax (cerumen). Excess cerumen should only be removed by a doctor. If a foreign object becomes lodged in the ear, only a doctor should try to remove it.

Preventing ear infection is the primary way to prevent a perforated eardrum. Because infection-causing bacteria are found in the ears, nose, and throat in most cases (65-75%) of middle ear infection, avoiding contact as much as possible with children or adults who have colds or upper respiratory infections is one way to reduce the occurrence of infections in children that may lead to middle ear infection. School-age children are especially susceptible to repeat infections. Promptly treating all nose and throat infections will help avoid ear infections. Early treatment of ear infection is another way to guard against a ruptured eardrum and associated hearing loss.

Parental concerns

Parents will likely be concerned about the possibility of ear infection, especially in children who have already had ear infections. It may help to be especially watchful for symptoms of colds, sinus infections, and upper respiratory symptoms that may lead to ear infection. Physicians may recommend immune system boosters to help prevent infection. Parents can also make sure the child is getting proper rest and **nutrition** to increase resistance to the **common cold** and infections present in other children with whom their child plays or attends school.

See also Otitis media (middle ear infection).

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Perinatal infection

Definition

An infection caused by a bacteria or virus that can be passed from a mother to her baby during pregnancy or delivery is called a perinatal infection.

Description

Perinatal infections include bacterial or viral illnesses that can be passed from a mother to her baby either while the baby is still in the uterus or during the delivery process. Maternal infection can, in some cases, cause complications at birth. The mother may or may not experience active symptoms of the infection during the pregnancy. Some perinatal infections are sexually transmitted.

Transmission

Transmission of many perinatal infections occurs during **childbirth**, particularly in cases when invasive techniques such as episiotomy or artificial rupture of membranes are employed. In other cases, transmission may occur during pregnancy, if the infectious agent can cross the placental barrier, and it may occur during breastfeeding, if the infectious agent can be found in breast milk.

Demographics

The incidence of perinatal infection depends on the causative agent of infection. For example, perinatal transmission of cytomegalovirus occurs in two to 24 out of every 1,000 live births. The rate of transmission of genital herpes during pregnancy is one to two out of every 2,000 pregnancies; the rate of transmission during childbirth changes to one out of every 2,000 to 5,000 live births. Perinatal transmission of group beta streptococcus causes neonatal infection in one to five out of every 1,000 live births, and **rubella** (German **measles**), 0.02 out of every 1,000 live births. HIV is transmitted from untreated mother to child in 25 to 40 percent of cases, but in only 1 percent of cases if mother receives treatment and the infant receives prophylaxis.

Causes and symptoms

The following represent some of the more common infections that can be transmitted perinatally.

Chlamydia

The bacterium *Chlamydia trachomatis* is the cause of the most common bacterial sexually transmitted disease in the United States, causing more than 4 million infections each year. The majority of women with chlamydial infection experience no obvious symptoms. The infection affects the reproductive tract and causes pelvic inflammatory disease, infertility, and ectopic pregnancy (when the fertilized egg implants somewhere other than in the uterus). This infection can cause premature rupture of the membranes and early labor. It can be passed to the infant during delivery and can cause ophthalmia neonatorum (an eye infection) within the first month of life and **pneumonia** within one to three months of age. Symptoms of chlamydial pneumonia are a repetitive **cough** and rapid breathing. Wheezing is rare and the infant is usually without a **fever**.

Cytomegalovirus

Cytomegalovirus (CMV) is a common virus in the herpes virus family. It is found in saliva, urine, and other body fluids and can be spread through sexual contact or other more casual forms of physical contact such as kissing. In adults, CMV may cause mild symptoms of swollen lymph glands, fever, and fatigue. Many people who carry the virus experience no symptoms at all. Infants can become infected with CMV while still in the uterus if the mother becomes infected or develops a recurrence of the infection during pregnancy. Although most infants exposed to CMV before birth develop normally and do not show any symptoms, as many as 6,000 infants who were exposed to CMV before birth are born with serious complications each year. CMV interferes with normal fetal development and can cause **mental retardation**, blindness, deafness, or epilepsy in these infants.

Genital herpes

Genital herpes, which is usually caused by **herpes simplex** virus type 2 (HSV-2), is a sexually transmitted disease that causes painful sores on the genitals. Women who have their first outbreak of genital herpes during pregnancy are at high risk of miscarriage or delivering a low birth weight baby. The infection can be passed to the infant at the time of delivery if the mother has an active sore. The most serious risk to the infant is the possibility of developing HSV-2 **encephalitis**, an inflammation of the brain, with symptoms of irritability and poor feeding.

Hepatitis B

Hepatitis B is a contagious virus that causes liver damage and is a leading cause of chronic liver disease and cirrhosis. Approximately 20,000 infants are born

each year to mothers who test positive for the hepatitis B virus. These infants are at high risk for developing hepatitis B infection through exposure to their mothers blood during delivery.

Human immunodeficiency virus (HIV)

Human **immunodeficiency** virus (HIV) is a serious, contagious virus that causes acquired immunodeficiency syndrome (**AIDS**). About 25 to 40 percent of untreated pregnant women pass the infection on to their newborn infants, while only 1 percent of treated pregnant women transmit the virus. There are often no symptoms of HIV in infants, but within a few months most infants who are infected show signs of opportunistic infections such as **failure to thrive**, chronic thrush, and persistent **diarrhea**.

Human papillomavirus

Human papillomavirus (HPV) is a sexually transmitted disease that causes genital **warts** and can increase the risk of developing some cancers. HPV appears to be transferred from the mother to the infant during the birth process and can cause tracheal narrowing due to lesions (warts) from the virus.

Rubella (German measles)

Rubella is a virus that causes German measles, an illness that includes rash, fever, and symptoms of an upper respiratory tract infection. Most people are exposed to rubella during childhood and develop antibodies to the virus so they never get it again. Rubella infection during early pregnancy can pass through the placenta to the developing infant and cause serious birth defects, including heart abnormalities, mental retardation, blindness, and deafness.

Group beta streptococcus

Group beta streptococcus (GBS) infection is the most common bacterial cause of infection and death in newborn infants. Although rates have declined in the United States since the introduction of **antibiotics** to at-risk women during labor in the 1980s, about 1,600 cases and 80 newborn deaths still occur each year. In women, GBS can cause vaginitis and urinary tract infections. Both infections can cause premature birth, and the bacteria can be transferred to the infant in the uterus or during delivery. GBS causes pneumonia, **meningitis**, and other serious infections in infants.

Syphilis

Syphilis is a sexually transmitted bacterial infection that can be transferred from a mother to an infant through the placenta before birth. Up to 50 percent of infants born to mothers with syphilis are premature or stillborn or die shortly after birth. Infected infants may have severe birth defects. Those infants who survive infancy may develop symptoms of syphilis up to two years later.

When to call the doctor

Pregnant women who exhibit symptoms of infection should contact their healthcare provider to determine if the infection can be passed vertically to the child during pregnancy, childbirth, and/or breastfeeding. In some cases, early detection and treatment of infection can minimize the risk of perinatal transmission.

Diagnosis

How a bacterial or viral infection is diagnosed depends on the causative agent. Examples include the following:

- Chlamydia can be diagnosed by taking a cotton swab sample of the cervix and vagina during the third trimester of the pregnancy. Chlamydial cell cultures take three to seven days to grow. DNA probes are available for more rapid diagnosis.
- Past or recent infection with cytomegalovirus (CMV) can be identified by antibody tests and CMV can be grown from body fluids.
- Genital herpes is suspected with the outbreak of a particular kind of genital sore. The sore can be cultured and tested to confirm that HSV-2 is present.
- Hepatitis B can be identified through a blood test for the hepatitis B surface antigen (HBsAg) in pregnant women. The test is part of prenatal health programs.
- Human immunodeficiency virus (HIV) can be detected using a blood test and is part of most prenatal screening programs.
- Human papillomavirus (HPV) causes the growth of warts in the genital area. The wart tissue can be removed with a scalpel and tested to determine what type of HPV virus caused the infection.
- Pregnant women are usually tested for antibodies to rubella, which would indicate that they have been previously exposed to the virus and, therefore, would not develop infection during pregnancy if exposed.
- Group beta streptococcus (GBS) can be detected by a vaginal or rectal swab culture and sometimes from a

urine culture. Blood tests can be used to confirm GBS infection in infants who exhibit symptoms.

- Pregnant women are usually tested for syphilis as part of the prenatal screening, generally with a blood test.

Treatment

Methods of treating some of the more common causes of perinatal infection include:

- Chlamydia: Pregnant women can be treated during the third trimester with oral erythromycin, for seven to 14 days depending on the dose used. Newborn infants can be treated with erythromycin liquid for ten to 14 days at a dosage determined by their body weight.
- Cytomegalovirus (CMV): No drugs or vaccines were as of 2004 available for prevention or treatment of CMV except in immunocompromised persons.
- Genital herpes: The **antiviral drugs** acyclovir or famciclovir can be administered to the mother during pregnancy. Little is known about the risks of these drugs to the fetus; however, the risk of birth defects does not seem to be any higher than for women who do not take these medications. Infants with suspected HSV-2 can be treated with acyclovir. Delivery of the infant by **cesarean section** is recommended if the mother has an active case of genital herpes.
- Hepatitis B: Infants born to mothers who test positive to the HBsAg test should be treated with hepatitis B immune globulin at birth to give them immediate protection against developing hepatitis B. All infants should also receive a series of three **hepatitis B vaccine** injections as part of their routine immunizations.
- Human immunodeficiency virus (HIV): Recent studies have shown that prenatal care and HIV testing before delivery are major opportunities for preventing perinatal **HIV infection**. Pregnant women with HIV should be treated as early in the pregnancy as possible with zidovudine (AZT). Other newer drugs designed to treat HIV/AIDS also may be used during pregnancy with the knowledge that these drugs may have unknown effects on the infant. Infants born with HIV should receive aggressive drug treatment to prevent development of AIDS. Most of the drugs designed to treat HIV are routinely used during pregnancy because of the mother's health needs and because transmission rate is directly related to the mother's viral load. Teratogenicity is not fully established for some of the subsequent HIV medications.
- Human papillomavirus: Genital warts are very difficult to treat and frequently recur even after treatment. They can be removed by cryotherapy (freezing), laser or electrocauterization (burning), or surgical excision (cutting). Some medications (imiquimod 5% cream, podophyllin, trichloroacetic acid, or topical 5-fluorouracil) can be applied to help dissolve genital warts. Cesarean delivery rather than vaginal delivery reduces the risk of transmission of HPV from mothers to infants.
- Rubella (German measles): No treatment is available. Some healthcare providers may recommend giving the mother an injection of immune globulin (to boost the immune system to fight off the virus) if she is exposed to rubella early in the pregnancy. However, no evidence to support the use of these injections existed as of 2004. Exposure to rubella early in pregnancy poses a high risk that the infant will have serious birth defects. Termination of the pregnancy may be considered. Women who have not been previously exposed to rubella are usually vaccinated immediately after the first pregnancy to protect infants of future pregnancies.
- Group beta streptococcus (GBS): Pregnant women diagnosed with GBS late in the pregnancy should be treated with antibiotics injected intravenously to prevent premature labor. In 2003, the Centers for Disease Control and Prevention (CDC) issued revised guidelines for preventing perinatal GBS disease. They began recommending that women not only be tested as soon as they learn of their pregnancy, but again at 35 to 37 weeks of gestation. The CDC also recommended updated prophylaxis regimens for women with penicillin **allergies**, as well as other guidelines for patients with threatened preterm deliveries and other recommendations. If transmission of GBS to the newborn infant already is suspected or if the baby develops symptoms of infection, infants often are treated with antibiotics.
- Syphilis: Antibiotic therapy, usually penicillin, given early in the pregnancy can be used to treat the infection and may prevent transmission to the infant.

Prognosis

The prognosis of a neonate who has contracted an infection perinatally depends on the specific infection. Examples include the following:

- Chlamydia: Without treatment, the most serious consequences of chlamydial infection are related to complications of premature delivery. Treatment of the mother with antibiotics during the third trimester can prevent premature delivery and the transfer of the infection to the baby. Infants treated with antibiotics for eye infection or pneumonia generally recover.
- Cytomegalovirus: The chance for recovery after exposure to CMV is very good for both the mother and the

infant. Exposure to CMV can be serious and even life threatening for mothers and infants whose immune systems are compromised, for example, those receiving **chemotherapy** or who have HIV/AIDS. Those infants who develop birth defects after CMV exposure may have serious, lifelong complications.

- Genital herpes: Once a woman or infant is infected, outbreaks of genital herpes sores can recur at any point during their lifetimes.
- Hepatitis B: Infants treated at birth with immune globulin and the series of vaccinations are protected from development of hepatitis B infection. Infants infected with hepatitis B develop a chronic, mild form of hepatitis and are at increased risk for developing liver disease.
- Human immunodeficiency virus (HIV): A combination of treatment with highly active antiretroviral therapy during pregnancy, zidovudine (AZT) during delivery, and AZT to the baby for six weeks after birth significantly reduces the chance that the infant will be infected with HIV from the mother.
- Human papillomavirus: Once infected with HPV, there is a lifelong risk of developing warts and an increased risk of some cancers.
- Rubella (German measles): Infants exposed to rubella virus in the uterus are at high risk for severe birth defects, including heart defects, blindness, and deafness.
- Streptococcus: Infection of the urinary tract or genital tract of pregnant women can cause premature birth. Infants infected with GBS can develop serious, life-threatening infections.
- Syphilis: Premature birth, birth defects, or the development of serious syphilis symptoms is likely to occur in untreated pregnant women.

Prevention

Use of a barrier method of contraceptive (e.g. **condom**) can prevent transmission of some sexually transmitted infections during intercourse. Intravenous drug use and sexual intercourse with infected partners increase the risks of exposure to most of these infections. Pregnant women can be tested for many of the bacterial or viral infections described; however, effective treatment may not be available to protect the infant.

In some cases, the method of childbirth may impact the chance of passing an infection from mother to child. For instance, research has shown that delivering a baby by caesarian section over vaginal delivery reduces the risk of transmitting HIV from mother to child.

KEY TERMS

Cesarean section—Delivery of a baby through an incision in the mother's abdomen instead of through the vagina; also called a c-section, cesarean birth, or cesarean delivery.

Ectopic pregnancy—A pregnancy that develops outside of the mother's uterus, such as in the fallopian tube. Ectopic pregnancies often cause severe pain in the lower abdomen and are potentially life-threatening because of the massive blood loss that may occur as the developing embryo/fetus ruptures and damages the tissues in which it has implanted.

Encephalitis—Inflammation of the brain, usually caused by a virus. The inflammation may interfere with normal brain function and may cause seizures, sleepiness, confusion, personality changes, weakness in one or more parts of the body, and even coma.

Episiotomy—An incision made in the perineum (the area between the vulva and the anus) during labor to assist in delivery and to avoid abnormal tearing of the perineum.

Perinatal—Referring to the period of time surrounding an infant's birth, from the last two months of pregnancy through the first 28 days of life.

Pneumonia—An infection in which the lungs become inflamed. It can be caused by nearly any class of organism known to cause human infections, including bacteria, viruses, fungi, and parasites.

Nutritional concerns

A woman's nutritional status may contribute to her ability to fight off infections, particularly in cases of **malnutrition**. A well-balanced diet rich in nutrients such as **folic acid**, calcium, iron, zinc, vitamin D, and the B **vitamins** is recommended for pregnant women. Mothers are recommended to eat approximately 300 additional calories day (above and beyond a normal non-pregnancy diet) to support the fetus's growth and development.

Parental concerns

Minimizing the risk of transmitting a maternal infection to a fetus is often a major concern for parents. The first step is identifying possible maternal infections. Proper prenatal care in many cases allows for early

diagnosis and thus early treatment of certain infections, thus improving the newborn's prognosis.

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Periodontal disease

Definition

Periodontal diseases are a group of diseases that affect the tissues that support and anchor the teeth. Left untreated, periodontal disease results in the destruction of the gums, alveolar bone (the part of the jaws where the teeth arise), and the outer layer of the tooth root.

Description

Periodontal (meaning "around the tooth") disease is usually seen as a chronic (long-term) inflammatory disease. An acute (sudden) infection of the tissue surrounding the teeth (periodontal tissue) may occur, but acute inflammation usually resolves on its own and is not treated by a dentist.

Periodontal diseases affect the gums, which consist of the gingiva, periodontal ligament, cementum, and alveolar bone. The gingiva is a pink-colored mucous membrane that covers part of the teeth and the alveolar bone. The periodontal ligament, also called the periodontal membrane, is the tough, fibrous tissue that holds the teeth in the gums. The cementum is a bony layer that covers the lower parts of the teeth. The alveolar bone is a set of ridges along the jaw bones (maxillary and mandible) from which the teeth arise.

Periodontal disease most often develops when a pocket or space is formed between the teeth and the gums. This pocket is called the gingival sulcus. A number of distinct forms of periodontal disease are known, including gingivitis, acute necrotizing ulcerative gingivitis, adult periodontitis, and localized juvenile periodontitis. Although many people have some form of periodontal disease, serious cases are not common.

Gingivitis is an inflammation of the outermost soft tissue of the gums. The gums become red and inflamed, lose their normal shape, and bleed easily. Gingivitis may remain a chronic disease for years without affecting other periodontal tissues. Chronic gingivitis may lead to a deepening of the pockets between the gum and tooth. In some children, gingivitis and bleeding gums are among the early signs of leukemia.

Acute necrotizing ulcerative gingivitis is seen mainly in young adults. This form of gingivitis is characterized by painful, bleeding gums, and death (necrosis) and erosion of gums between the teeth.

Localized juvenile periodontitis is a less common form of periodontal disease and is seen mainly in young people. Localized juvenile periodontitis usually affects

the molars (back grinding teeth) and incisors. Among the distinctions that separate this form of periodontitis are the low incidence of bacteria in the periodontal pocket, minimal plaque formation, and mild inflammation.

Pericoronitis is a condition found in children whose molars are in the process of erupting through the gum. The disease is seen more frequently in the lower molar teeth. As the molar emerges, a flap of gum still covers the tooth. The flap of gum traps bacteria and food, leading to mild irritation. If the upper molar fully emerges before the lower one, it may bite down on the flap during chewing and increase the irritation of the flap, leading to infection. In severe cases, the infection can spread to the neck and cheeks.

Periodontitis, also called pyorrhea, is a condition in which gingivitis has extended down around the tooth and into the supporting bone structure. Plaque and tarter build-up lead to the formation of large pockets between the gums and teeth. When this happens, anaerobic bacteria (bacteria that do not need oxygen) grow in the pockets. The pockets eventually extend down around the roots of the teeth where the bacteria cause damage to the bone structure supporting the teeth.

Herpetic gingivostomatitis, which is relatively common in children, is an inflammation of the gums and mouth caused by the **herpes simplex** virus. This disease is contagious, but tends to heal without medical intervention in about two weeks.

Desquamative gingivitis occurs mainly in postmenopausal women and is not well understood.

Trench mouth, also called Vincent's disease, is a suddenly developing (acute) complication of gingivitis. It causes tissue death and open sores on the gums and is often accompanied by **fever**, fatigue, and painful bleeding gums. Trench mouth usually develops because of poor **oral hygiene**, stress, fatigue, and **smoking**. It requires immediate treatment by a dentist, since **pain** can increase to the point where eating and swallowing become difficult, and the inflammation can spread to nearby tissues of the face and neck.

Demographics

Periodontal disease is common. It is estimated that 9–17 percent of children between the ages of three and 11 years have gingivitis. The number increases sharply at **puberty**, with 70–90 percent of teens developing the disease. More boys than girls have gingivitis, probably because girls have better oral hygiene habits than boys, rather than because of any physiological differences.

Some medical conditions are associated with an increased likelihood of developing periodontitis. These diseases include diabetes, **Down syndrome**, **AIDS**, and any disease or condition that compromises the immune system and reduces the number of white blood cells in the body for extended periods.

Causes and symptoms

Bacteria present on the gingival tissues cause periodontal diseases. The mechanisms by which bacteria in the periodontal pocket cause tissue destruction in the surrounding region are not fully understood. However, removal of bacteria through good oral hygiene practices and regular dental care helps reduce or eliminate these diseases. There are indications that a tendency toward developing periodontal disease is genetic, with up to 30 percent of the population being highly susceptible despite aggressive oral hygiene habits.

Other factors that put individuals at higher risk for developing periodontal diseases include smoking, stress, poor diet, and taking certain medications such as antidepressants, some heart medicines, and **oral contraceptives**. Gingivitis can be aggravated by hormones and may temporarily worsen during puberty and pregnancy. Individuals with diabetes and diseases that depress the immune system are more likely to develop periodontal disease.

The main symptoms of periodontal disease include:

- bleeding gums
- red, sore, or swollen gums
- gums that have receded from the base of the teeth
- chronic bad breath
- loose permanent teeth
- open sores on the gums

When to call the dentist

Beginning as toddlers, all children need regular checks-up by a dentist. Children who have chronically bleeding gums, open sores on the gums, or who complain of gum or tooth pain, should see a dentist promptly. Those with bleeding gums should see their pediatrician urgently, as this is also a symptom of leukemia in some children.

Diagnosis

Diagnosis of periodontal disease is made by observation of infected gums. Usually a dentist diagnoses and characterizes the various types of periodontal disease.

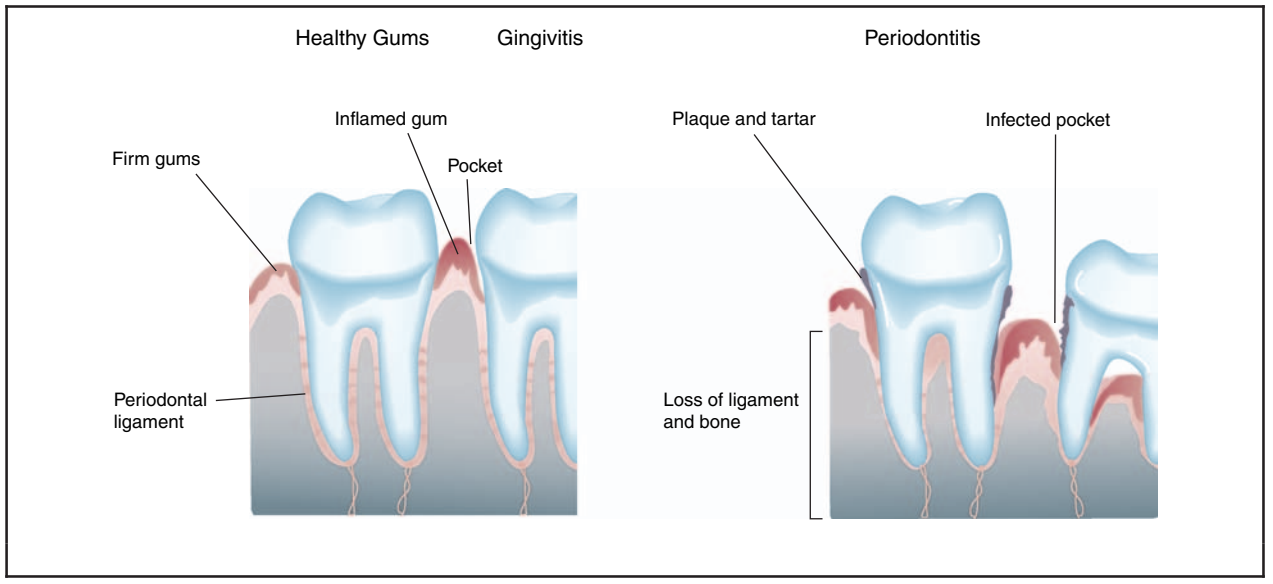

Diagram showing healthy gums (far left) with no pockets or redness; gingivitis (left) with inflamed gum and pocket; and periodontitis (right) with infected pockets, plaque, and tartar. *(Illustration by Argosy, Inc.)*

Many periodontal diseases are distinguished based on the severity of the infection and the number and type of tissues involved.

Diagnosis of periodontitis includes measuring the size of the pockets formed between the gums and teeth. Normal gingival pockets are shallow. If periodontal disease is severe, jawbone loss will be detected in **x rays** of the teeth. If too much bone is lost, the teeth become loose and can change position. This will also be seen in x-ray images.

Treatment

Tartar can be removed only by professional dental treatment. Following treatment, periodontal tissues usually heal quickly. Administering the needed **vitamins** and improving diet treats gingivitis caused by poor **nutrition** or vitamin deficiencies. Removing debris under the flap of gum covering the molar treats pericoronitis.

Treatment of periodontitis requires professional dental care. The pockets around the teeth are cleaned, and all tartar and plaque removed. In periodontitis, tartar and plaque can extend far down the tooth root. Normal dental hygiene, brushing and flossing, cannot reach deep enough to effectively treat periodontitis. In cases where pockets are very deep (more than 0.25 in, or 0.64 cm, deep), surgery is required to clean the pocket. This is performed in a dental office. Sections of gum that are not likely to reattach to the teeth may be removed to promote healing by healthy sections of gum. Abscesses are treated with a combination of **antibiotics** and surgery. If antibiotics are needed for gum disease, they are usually given orally. The antibiotics may be delivered directly to the infected gum and bone tissues to ensure that high concentrations reach the infected area. Abscess infections, especially of bone, are difficult to treat and require long term antibiotic therapy to prevent a reoccurrence of infection.

There are no useful drugs to treat herpetic gingivostomatitis, but acyclovir is used in high-risk patients or those with a compromised immune system. Herpes lesions heal by themselves without treatment. After the herpetic lesions have disappeared, the gums usually return to normal if good oral hygiene is resumed.

Prognosis

Most cases of periodontal disease are mild and can be cleared up with improved oral hygiene, as well as tooth and gum cleaning by a trained professional. Serious cases of periodontal disease may be persistent, but they can usually be controlled. Untreated periodontal disease may cause teeth to loosen and fall out, and infection may spread to surrounding tissues.

Prevention

Good oral hygiene, a well-balanced nutritious diet, and regular dental visits for tooth cleaning all help prevent periodontal disease. Prompt attention to gingivitis can prevent it from progressing to more serious periodontal diseases.

KEY TERMS

Alveolar bone—A set of ridges from the jawbones.

Cementum—A bony substance that covers the root of the tooth.

Gingiva—The gum tissue surrounding the teeth.

Gingival sulcus—The space between the tooth and the gum that often traps food and bacteria, leading to periodontal disease.

Periodontal ligament—Also called the periodontal membrane, this tough fibrous tissue holds the teeth in place in the gums.

Plaque—A deposit, usually of fatty material, on the inside wall of a blood vessel. Also refers to a small, round demyelinated area that develops in the brain and spinal cord of an individual with multiple sclerosis.

Tartar—A hardened yellow or brown mineral deposit from unremoved plaque. Also called calculus.

Parental concerns

Sometimes parents are less concerned about their child's first (baby) teeth than their permanent teeth. However, poor oral hygiene and lack of care of the first set of teeth are apt to be reflected in problems with the gums and the permanent teeth.

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Peroneal muscular atrophy *see* **Charcot-Marie-Tooth disease**

Peroxisomal disorders

Definition

Peroxisomal disorders are a group of congenital diseases characterized by the absence of normal peroxisomes in the cells of the body.

Description

Peroxisomes are organelles within a cell that contain enzymes responsible for critical cellular processes. A cell can contain several hundred peroxisomes, round or oval bodies with diameters of about 0.5 micron that contain proteins that function as enzymes in metabolic processes. By definition, a peroxisome must contain catalase, which is an enzyme that breaks down hydrogen peroxide.

Peroxisomal disorders are subdivided into two major categories: those disorders resulting from a failure to form intact, normal peroxisomes, resulting in multiple metabolical abnormalities, which are referred to as peroxisome biogenesis disorders (PBD) or generalized peroxisomal disorders; and those disorders resulting from the deficiency of a single peroxisomal enzyme. There are about 25 known peroxisomal disorders, although the number of diseases that are considered to be separate, distinct peroxisomal disorders varies among researchers and healthcare practitioners.

Approximately 50 different biochemical reactions occur entirely or partially within a peroxisome. Some of the processes are anabolic (constructive), resulting in the synthesis of essential biochemical compounds, including bile acids, cholesterol, plasmalogens, and docosahexanoic acid (DHA), which is a long chain fatty acid that is a component of complex lipids, including the membranes of the central nervous system. Other reactions are catabolic (destructive) and lead to the

destruction of some fatty acids, including very long chain fatty acids (VLCFAs, fatty acids with more than 22 carbon atoms in their chains), phytanic acid, pipecolic acid, and the prostoglandins. The peroxisome is involved in breaking down VLCFAs to lengths that the body can use or get rid of.

When VLCFAs accumulate due to abnormal functioning of the peroxisomes, they are disruptive to the structure and stability of certain cells, especially those associated with the central nervous system and the myelin sheath, which is the fatty covering of nerve fibers. The peroxisomal disorders that include effects on the growth of the myelin sheath are considered to be part of a group of genetic disorders referred to as leukodystrophies.

Peroxisomal disorders form a heterogeneous disease group, with different degrees of severity. The differences among these disorders are continuous, with overlap between abnormalities. Examples of peroxisomal disorders are:

- X-linked adrenoleukodystrophy (X-ALD), a sex-linked disorder characterized by progressive symptoms that begin as behavioral changes, muscle weakness, and speech difficulties.
- Zellweger syndrome (ZS), which is usually fatal within the first year of life.
- Neonatal adrenoleukodystrophy (NALD), which is usually fatal within the first ten years.
- Infantile Refsum disease (IRD), which is not as devastating as ZS and NALD, as the children with this disorder with time and patience can develop some degree of motor, cognitive, and **communication skills**, although death generally occurs during the second decade of life.
- Rhizomelic chondrodysplasia punctata (RCDP), which in its most severe form is fatal within the first year or two of life; however, survival into the teens has been known to occur. It is characterized by shortening of the proximal limbs (i.e., the legs from knee to foot and the arms from elbow to hand).
- Zellweger-like syndrome, which is fatal in infancy and known to be a defect of three particular enzymes.

Transmission

Most peroxisomal disorders are inherited autosomal recessive diseases. This means that both parents need to be carriers of the defective gene in order for a child to develop the disease. If both parents are carriers but do not show signs of disease, each child has a 25 percent chance of having the disease. If one parent has the disease and the other is a carrier, each child has a 50 percent chance of having the disease. As a sex-linked genetic disorder, the daughters of males affected with X-ALD become carriers and the sons are not affected. The children of female carriers have a 50 percent chance of having the genetic mutation, which means that sons who inherit the mutation have the disease, and daughters who inherit the mutation are carriers.

Demographics

Peroxisomal disorders occur in all countries, among all races and ethnic groups. They are extremely rare, with frequencies reported at one in 30,000 to one in 150,000, although these numbers are only estimates. X-ALD is the most common of the peroxisomal disorders, affecting about one in 20,000 males. It is estimated that there are about 1,400 people in the United States with the disorder. ZS is estimated to affect one in 50,000 to 100,000 live births.

Causes and symptoms

The range of disease abnormalities may be a result of a corresponding range of peroxisome failure. For example, in severe cases of ZS, the failure is nearly complete, while in IRD, there is some degree of peroxisome activity. In peroxisomal single-enzyme disorders, the peroxisome is intact and functioning, but there is a defect in only one enzymatic process, with only one corresponding biochemical abnormality. These disorders, however, can be as severe as those in which peroxisomal activity is nearly or completely absent.

In general, **developmental delay**, **mental retardation**, and vision and **hearing impairment** are common in those who have these disorders. Acquisition of speech appears to be especially difficult, and because of the reduced communication abilities, **autism** is common in those who live longer. Peroxisomal disorder patients have decreased muscle tone (**hypotonia**), which in the most severe cases is generalized, while in less severe cases, is usually restricted to the neck and trunk muscles. Sometimes this lack of control is only noticeable by a curved back in the sitting position. Head control and independent sitting is delayed, with most patients unable to walk independently.

Failure to thrive is a common characteristic of patients with peroxisomal disorder, along with an enlarged liver, abnormalities in liver enzyme function, and loss of fats in stools (steatorrhea). Peroxisomal disorders are also associated with facial abnormalities, including high forehead, frontal bossing (swelling), small face, low set ears, and slanted eyes. These characteristics may

not be prominent in some children and are especially difficult to identify in an infant.

In X-ALD there is a deficiency in the enzyme that breaks down VLCFAs, which then accumulate in myelin and the adrenal glands. Onset of X-ALD-related neurological symptoms occurs at about five to 12 years of age, with death occurring within one to ten years after onset of symptoms. In addition to physical abnormalities seen in other types of peroxisomal disorders, common symptoms of X-ALD also include behavioral changes such as abnormal withdrawal or aggression, poor memory, dementia, and poor academic performance. Other symptoms are muscle weakness and difficulties with hearing, speech, and vision. As the disease progresses, muscle tone deteriorates, swallowing becomes difficult, and the patient becomes comatose. Unless treated with a diet that includes a mixture of oils called Lorenzo's oil, the disease will result in paralysis, hearing loss, blindness, vegetative state, and death. There are also milder forms of X-ALD, an adult onset ALD that typically begins between the ages of 21 and 35, and a form that is occasionally seen in women who are carriers of the disorder. In addition to X-ALD, there are at least ten other single-enzyme peroxisomal disorders, each with its own specific abnormalities.

When to call the doctor

A healthcare provider should be contacted if a child develops symptoms suggestive of peroxisomal disorder or if a child already diagnosed with a peroxisomal disorder shows signs of worsening disease.

Diagnosis

Since hearing and vision deficiencies may be difficult to identify in infants, peroxisomal disorders are usually detected by observations of failure to thrive, hypotonia, mental retardation, widely open fontanel, abnormalities in liver enzymes, and an enlarged liver. If peroxisomal disorders are suspected, blood plasma assays for VLCFAs, phytanic acid, and pipecolic acid are conducted. Additional tests include plasmalogen biosynthesis potential.

It is possible to diagnose peroxisomal disorders in utero. For example, for X-ALD, diagnosis can be made from cultured skin fibroblasts or amniotic fluid cells. This allows prenatal diagnosis and carrier identification in 90 percent of those affected. As of the early 2000s it has been shown that biochemical diagnosis can be performed through chorionic villus testing, a procedure performed very early in the first trimester of pregnancy.

Treatment

For many of the peroxisomal disorders, there is no standard course of treatment, with supportive treatment strategies focusing on alleviation of complications and symptoms. Bone marrow transplants may be effective for children with X-ALD if administered early in the course of the childhood form of the disease. Physical and psychological therapies are important for all types of peroxisomal disorders.

Alternative treatment

Patients with peroxisomal disorders, and particularly X-ALD, have been treated with a mixture of glycerol trioleate-glycerol trieucate (4:1 by volume), prepared from olive and rapeseed oils, and referred to as Lorenzo's oil (developed by the parents of a son, Lorenzo, who had X-ALD, whose story was documented in the 1992 movie, *Lorenzo's Oil*), to decrease the levels of VLCFA. Other diets that have been tried with varying success include dietary supplementation with plasmalogen precursors to increase plasmalogen levels and with cholic acid to normalize bile acids.

Nutritional concerns

In general, most treatments that are attempted for peroxisomal disorders are dietary, whereby attempts are made to artificially correct biochemical abnormalities associated with the disorders. Therapies include supplementation of the diet with antioxidant **vitamins** or limitation of intake of fatty acids, especially VLCFAs.

Another area of dietary therapy that is being investigated is the supplementation of the diet with pure DHA, given as early in life as possible, in conjunction with a normal well-balanced diet. Some results have indicated that if given soon enough during development, DHA therapy may prevent some of the devastating consequences of peroxisomal disorders, including the loss of vision and brain damage.

Other treatment strategies include addition of important missing chemicals. For example, in disorders where there is faulty adrenal function, replacement adrenal hormone therapy is used.

Any dietary changes should be monitored biochemically to determine if the supplements are having their desired effects and are not causing additional adverse effects.

Prognosis

Peroxisomal disorders range from life-threatening to cases in which people may function with some degree of

KEY TERMS

Adrenal glands—A pair of endocrine glands (glands that secrete hormones directly into the bloodstream) that are located on top of the kidneys. The outer tissue of the glands (cortex) produces several steroid hormones, while the inner tissue (medulla) produces the hormones epinephrine (adrenaline) and norepinephrine.

Autosomal recessive mutation—A pattern of genetic inheritance where two abnormal genes are needed to display the trait or disease.

Autosome—A chromosome not involved in sex determination.

Enzyme—A protein that catalyzes a biochemical reaction without changing its own structure or function.

Fontanelle—One of several "soft spots" on the skull where the developing bones of the skull have yet to fuse.

Organelle—A specialized structure within a cell, which is separated from the rest of the cell by a membrane composed of lipids and proteins, where chemical and metabolic functions take place.

mental and motor delays. As of 2004, there was not yet a cure for peroxisomal disorders. Enzyme replacement therapies, including enzyme infusion, transplantation, and gene therapy, may hold promise for future advances in the treatment of these disorders. As of the early 2000s research is conducted in order to increase scientific understanding of these disorders and find ways to prevent, treat, and cure them.

Prevention

It is not possible to prevent the transmission of an abnormal peroxisomal gene from parent to child or spontaneous mutations that may arise.

Parental concerns

Numerous professional and parent-led organizations exist to support parents as they first learn of a peroxisomal disorder diagnosis and as they provide care for their child. Genetic counseling is recommended for known or suspected carriers. As genes are identified that result in the disorders, genetic testing is being developed to identify carriers, who then can manage their reproduction to avoid the possibility of children being born with these deficiencies.

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Personality development

Definition

Personality development is the development of the organized pattern of behaviors and attitudes that makes a person distinctive. Personality development occurs by the ongoing interaction of **temperament**, character, and environment.

Description

Personality is what makes a person a unique person, and it is recognizable soon after birth. A child's personality has several components: temperament, environment, and character. Temperament is the set of genetically determined traits that determine the child's approach to the world and how the child learns about the world. There are no genes that specify personality traits, but some genes do control the development of the nervous system, which in turn controls behavior.

A second component of personality comes from adaptive patterns related to a child's specific environment. Most psychologists agree that these two factors—temperament and environment—influence the development of a person's personality the most. Temperament, with its dependence on genetic factors, is sometimes referred to as "nature," while the environmental factors are called "nurture."

While there is still controversy as to which factor ranks higher in affecting personality development, all experts agree that high-quality parenting plays a critical role in the development of a child's personality. When parents understand how their child responds to certain situations, they can anticipate issues that might be problematic for their child. They can prepare the child for the situation or in some cases they may avoid a potentially difficult situation altogether. Parents who know how to adapt their parenting approach to the particular temperament of their child can best provide guidance and ensure the successful development of their child's personality.

Finally, the third component of personality is character—the set of emotional, cognitive, and behavioral patterns learned from experience that determines how a person thinks, feels, and behaves. A person's character continues to evolve throughout life, although much depends on inborn traits and early experiences. Character is also dependent on a person's **moral development**.

In 1956, psychiatrist Erik Erikson provided an insightful description as to how personality develops based on his extensive experience in psychotherapy with children and adolescents from low, upper, and middle-class backgrounds. According to Erikson, the socialization process of an individual consists of eight phases, each one accompanied by a "psychosocial crisis" that must be solved if the person is to manage the next and subsequent phases satisfactorily. The stages significantly influence personality development, with five of them occurring during infancy, childhood, and **adolescence**.

Infancy

During the first two years of life, an infant goes through the first stage: *Learning Basic Trust or Mistrust (Hope)*. Well-nurtured and loved, the infant develops trust and security and a basic optimism. Badly handled, the infant becomes insecure and learns "basic mistrust."

Toddlerhood

The second stage occurs during early childhood, between about 18 months to two years and three to four years of age. It deals with *Learning Autonomy or Shame (Will)*. Well-parented, the child emerges from this stage with self-confidence, elated with his or her newly found control. The early part of this stage can also include stormy **tantrums**, stubbornness, and negativism, depending on the child's temperament.

Preschool

The third stage occurs during the "play age," or the later **preschool** years from about three to entry into formal school. The developing child goes through *Learning Initiative or Guilt (Purpose)*. The child learns to use imagination; to broaden skills through active **play** and fantasy; to cooperate with others; and to lead as well as to follow. If unsuccessful, the child becomes fearful, is unable to join groups, and harbors guilty feelings. The child depends excessively on adults and is restricted both in the development of play skills and in imagination.

School age

The fourth stage, *Learning Industry or Inferiority (Competence)*, occurs during school age, up to and possibly including junior high school. The child learns to master more formal skills:

- relating with peers according to rules
- progressing from free play to play that is structured by rules and requires teamwork (team sports)
- learning basic intellectual skills (reading, arithmetic)

At this stage, the need for self-discipline increases every year. The child who, because of his or her successful passage through earlier stages, is trusting, autonomous, and full of initiative, will quickly learn to be industrious. However, the mistrusting child will doubt the future and will feel inferior.

Adolescence

The fifth stage, *Learning Identity or Identity Diffusion (Fidelity)*, occurs during adolescence from age 13 or 14. Maturity starts developing during this time; the

young person acquires self-certainty as opposed to self-doubt and experiments with different constructive roles rather than adopting a negative identity, such as delinquency. The well-adjusted adolescent actually looks forward to achievement, and, in later adolescence, clear sexual identity is established. The adolescent seeks leadership (someone to inspire him or her), and gradually develops a set of ideals to live by.

The Child Development Institute (CDI) rightfully points out that very little knowledge is available on the type of specific environment that will result, for example, in traits of trust being more developed in a person's personality. Helping the child through the various stages of emotional and personality development is a complex and difficult task. Searching for the best ways of accomplishing this task accounts for most of the research carried out in the field of child development today.

Renowned psychologist Carl Rogers emphasized how childhood experiences affect personality development. Many psychologists believe that there are certain critical periods in personality development—periods when the child will be more sensitive to certain environmental factors. Most experts believe that a child's experiences in the **family** are important for his or her personality development, although not exactly as described by Erikson's stages, but in good agreement with the importance of how a child's needs should to be met in the family environment. For example, children who are toilet trained too early or have their **toilet training** carried out too strictly may become rebellious. Another example is shown by children who learn appropriate behavior to their sex lives when there is a good relationship with their same-sex parent.

Another environmental factor of importance is culture. Researchers comparing cultural groups for specific personality types have found some important differences. For example, Northern European countries and the United States have individualistic cultures that put more emphasis on individual needs and accomplishments. In contrast, Asian, African, Central American, and South American countries are characterized more by community-centered cultures that focus on belonging to a larger group, such as a family, or nation. In these cultures, cooperation is considered a more important value than competitiveness, which will necessarily affect personality development.

Common problems

Infants who are just a few weeks old display differences between each other in how active they are, how responsive they are to change, and how irritable they are. Some infants cry constantly while others seem happy and stay fairly quiet. Child development research conducted by the CDI has identified nine temperamental traits that may contribute to a child's personality development being challenging or difficult:

- activity level (how active the child is generally)
- distractibility (degree of concentration and paying attention when the child is not particularly interested)
- intensity (how loud the child is)
- regularity (the predictability of biological functions like appetite and sleep)
- sensory threshold (how sensitive the child is to physical stimuli: touch, taste, smell, sound, light)
- approach/withdrawal (characteristic responses of a child to a new situation or to strangers)
- adaptability (how easily the child adapts to transitions and changes such as switching to a new activity)
- persistence (stubbornness, inability to give up)
- mood (tendency to react to the world primarily in a positive or negative way)

Temperamental traits are enduring personality characteristics that are neither "good" nor "bad." Early on, parents can work with the child's temperamental traits rather than oppose them. Later, as the child grows up, parents can help the child to adapt to his or her own world in spite of inborn temperament.

Parental concerns

Most children experience healthy personality development. However, some parents worry as to whether their infant, child, or teenager has a personality disorder. Parents are usually the first to recognize that their child has a problem with emotions or behaviors that may point to a personality disorder.

Children with **personality disorders** have great difficulty dealing with other people. They tend to be inflexible, rigid, and unable to respond to the changes and normal stresses of life and find it very difficult to participate in social activities. When these characteristics are present in a child to an extreme, when they are persistent and when they interfere with healthy development, a diagnostic evaluation with a licensed physician or mental health professional is recommended.

When to call the doctor

Parents who suspect that their child has a personality disorder should seek professional help. It is a very

KEY TERMS

Behavior—A stereotyped motor response to an internal or external stimulus.

Character—An individual's set of emotional, cognitive, and behavioral patterns learned and accumulated over time.

Cognition—The act or process of knowing or perceiving.

Cognitive—The ability (or lack of) to think, learn, and memorize.

Gene—A building block of inheritance, which contains the instructions for the production of a particular protein, and is made up of a molecular sequence found on a section of DNA. Each gene is found on a precise location on a chromosome.

Identity—The condition of being the same with, or possessing, a character that is well described, asserted, or defined.

Maturity—A state of full development or completed growth.

Personality—The organized pattern of behaviors and attitudes that makes a human being distinctive. Personality is formed by the ongoing interaction of temperament, character, and environment.

Socialization—The process by which new members of a social group are integrated in the group.

Temperament—A person's natural disposition or inborn combination of mental and emotional traits.

important first step in knowing for sure whether there is a disorder, and if so, what treatment can best help the child. Child and adolescent psychiatrists are trained to help parents sort out whether their child's personality development is normal.

See also Bonding; Cognitive development; Temperament.

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Personality disorders

Definition

Personality disorders (PD) are a group of psychiatric conditions characterized by experience and behavior patterns that cause serious problems with respect to any two of the following: thinking, mood, personal relations, and the control of impulses.

Description

Most personality disorders are associated with problems in personal development and character which peak during **adolescence** and are then defined as personality disorders. Children and adolescents with a personality disorder have great difficulty dealing with others. They tend to be inflexible, rigid, with inadequate response to the changes and demands of life. They have a narrow view of the world and find it hard to participate in social activities. There are many formally identified personality disorders, each with its own types of associated behaviors. Most PDs, however, fall into three distinct categories or clusters, namely: cluster A, which includes disorders characterized by odd or eccentric behavior; cluster B, which includes disorders marked by dramatic, emotional or erratic behavior; and cluster C, which includes disorders accompanied by anxious and fearful behavior. The most common disorders in each cluster are given below.

Cluster A disorders

These disorders include the following:

- Schizoid personality disorder. Schizoid personalities are introverted, withdrawn, solitary, emotionally cold, and distant. Often absorbed with their own thoughts and feelings, they **fear** closeness and intimacy with others. People suffering from schizoid personality tend to be more daydreamers than practical action takers, often living "in a world of their own."
- Paranoid personality disorder. Paranoid personalities interpret the actions of others as deliberately threatening or demeaning. People with paranoid personality disorder are untrusting, unforgiving, and often resort to angry or aggressive outbursts without justification because they see others as unfaithful, disloyal, or dishonest. Paranoid personalities are often jealous, guarded, secretive, and scheming, and may appear to be emotionally "cold" or excessively serious.
- Schizotypal personality disorder. Schizotypal personalities tend to have odd or eccentric manners of speaking or dressing. They often have strange, outlandish, or paranoid beliefs and thoughts. People with schizotypal personality disorder have difficulties **bonding** with others and experience extreme **anxiety** in social situations. They tend to react inappropriately or not react at all during a conversation, or they may talk to themselves. They also have delusions characterized by "magical thinking," for example, by saying that they can foretell the future or read other people's minds.

Cluster B disorders

Cluster B disorders include the following:

- **Antisocial personality disorder**. Antisocial personalities typically ignore the normal rules of social behavior. These individuals are impulsive, irresponsible, and callous. They often have a history of violent and irresponsible behavior, aggressive and even violent relationships. They have no respect for other people and feel no remorse about the effects of their behavior on others. Antisocial personalities are at high risk for substance abuse, since it helps them to relieve tension, irritability, and boredom.
- Borderline personality disorder. Borderline personalities are unstable in interpersonal relationships, beha-

vior, mood, and self-image. They are prone to sudden and extreme mood changes, stormy relationships, unpredictable and often self-destructive behavior. These personalities have great difficulty with their own sense of identity and often experience the world in extremes, viewing experiences and others as either "black" or "white." They often form intense personal attachments only to quickly dissolve them over a perceived offense. Fears of **abandonment** and rejection often lead to an excessive dependency on others. **Self-mutilation** or suicidal threats may be used to get attention or manipulate others. Impulsive actions, persistent feelings of boredom or emptiness, and intense anger outbursts are other traits of this disorder.

- Narcissistic personality disorder. Narcissistic personalities tend to have an exaggerated sense of self-importance, and are absorbed by fantasies of unlimited success. They also seek constant attention, and are oversensitive to failure, often complaining about multiple physical disorders. They also tend to be prone to extreme mood swings between self-admiration and insecurity, and tend to exploit interpersonal relationships.

Cluster C disorders

Cluster C disorders include the following:

- Avoidant personality disorder. Avoidant personalities are often fearful of rejection and unwilling to become involved with others. They are characterized by excessive social discomfort, **shyness**, fear of criticism, and avoidance of social activities that involve interpersonal contact. They are afraid of saying something considered foolish by others and are deeply hurt by any disapproval from others. They tend to have no close relationships outside the **family** circle and are upset at their inability to form meaningful relationships.
- **Dependent personality disorder**. As the name implies, dependent personalities exhibit a pattern of dependent and submissive behavior, relying on others to make decisions for them. They fear rejection, need constant reassurance and advice, and are oversensitive to criticism or disapproval. They feel uncomfortable and helpless if they are alone and can be devastated when a close relationship ends. Typically lacking in self-confidence, the dependent personality rarely initiates projects or does things independently.
- Compulsive personality disorder. Compulsive personalities are conscientious, reliable, dependable, orderly, and methodical, but with an inflexibility that often makes them incapable of adapting to changing circumstances. They have such high standards of achievement that they constantly strive for perfection. Never satisfied with their performance or with that of others, they take on more and more responsibilities. They also pay excessive attention to detail, which makes it very hard for them to make decisions and complete tasks. When their feelings are not under strict control, when events are unpredictable, or when they must rely on others, compulsive personalities often feel a sense of isolation and helplessness.

Demographics

In 2001 to 2002, fully 16.4 million Americans (7.9% of all adults) had obsessive-compulsive personality disorder; 9.2 million (4.4%) had paranoid personality disorder; 7.6 million (3.6%) had antisocial personality disorder; 6.5 million (3.1%) had schizoid personality disorder; 4.9 million (2.4%) had avoidant personality disorder; and 1.0 million (0.5%) had dependent personality disorder. According to the National Institutes of Health, nearly 31 million Americans meet criteria for at least one personality disorder. A 2004 survey showed that nearly 14.8 percent of adult Americans met diagnostic criteria for personality disorders as defined by the American Psychiatric Association's Diagnostic and Statistical Manual of Mental Disorders. The risk of having avoidant, dependent, and paranoid personality disorders is greater for females than males, whereas risk of having antisocial personality disorder is greater for males than females. There are no gender differences in the risk of having compulsive or schizoid personality disorders. In general, other risk factors contributing to the emergence of personality disorders include being Native American or African American; being a young adult; having a low socioeconomic status; and having any other status than married.

Causes and symptoms

The exact cause of personality disorders is unknown. However, evidence points to genetic and environmental factors such as a history of personality disorders in the family. Some experts believe that traumatic events occurring in early childhood exert a crucial influence upon behavior later in life. Others propose that people are genetically predisposed to personality disorders or that they have an underlying biological disturbance (anatomical, electrical, or neurochemical).

Symptoms vary widely depending on the specific type of PD, but according to the American Psychiatric Association, individuals with personality disorders have most of the following symptoms in common:

- self-centeredness that manifests itself through a "me-first," self-preoccupied attitude

- lack of individual accountability that results in a "victim mentality" and blaming others for their problems
- lack of empathy and caring
- manipulative and exploitative behavior
- unhappiness, suffering from depression, and other mood and anxiety disorders
- vulnerability to other mental disorders
- distorted or superficial understanding of self and others' perceptions that results in being unable to see how objectionable, unacceptable, and disagreeable their behavior is
- self-destructive behavior
- socially maladaptive, changing the "rules of the game," or otherwise influencing the external world to conform to their own needs

When to call the doctor

An appointment should be made with a healthcare provider or a mental health professional if a child has persistent symptoms of a personality disorder. Parents are often concerned about their child's emotional health or behavior, but they do not know where to start to get help. The mental health system can also be complicated and difficult for parents to understand. When worried about their child's behavior, parents can start by talking to the child's pediatrician or family physician about their concerns. Personality disorders require treatment and parents should try to find a mental health professional with advanced training and experience with children, adolescents, and families. Parents should always ask about the professional's training and experience. It is also very important to find a good match between child, family, and the mental health professional.

Diagnosis

The character of a person is shown through his or her personality, by the way the person thinks, feels, and behaves. When the behavior is inflexible, maladaptive, and antisocial, then that individual is diagnosed with a personality disorder. Personality disorders are diagnosed following a psychological evaluation that records the history and severity of the symptoms. A personality disorder must fulfill several criteria. A deeply ingrained, inflexible pattern of relating, perceiving, and thinking that is serious enough to cause distress or impaired functioning defines a personality disorder. Personality disorders are usually recognizable by adolescence or earlier, continue throughout adulthood, and become less obvious in middle age.

Treatment

There are many types of help available for the different personality disorders. Treatment may include individual, group, or family psychotherapy. Medications, prescribed by a patient's physician, may also be helpful in relieving some of the symptoms of personality disorders, such as problems with anxiety and delusions. Psychotherapy is a form of treatment designed to help children and families understand and resolve the problems due to PD and modify the inappropriate behavior. In some cases a combination of medication with psychotherapy may be more effective. PD psychotherapy focuses on helping patients see the unconscious conflicts that are causing their disorder. It also helps them become more flexible and is aimed at reducing the behavior patterns that interfere with everyday living. In psychotherapy, patients have the opportunity to learn to recognize the effects of their behavior on others. The different types of psychotherapies available to children and adolescents include the following:

- Cognitive behavior therapy (CBT). CBT is focused on improving a child's moods and behavior by examining confused or distorted patterns of thinking. With CBT, the child learns that thoughts cause feelings and moods that can influence behavior. For example, if a child has problematic behavior patterns, the therapist seeks to identify the underlying thinking that is causing them. The therapist then helps the child replace this thinking with thoughts that result in more appropriate feelings and behaviors.
- Dialectical behavior therapy (DBT). DBT is used to treat older adolescents with suicidal thoughts or who intentionally engage in self-destructive behavior or who have borderline personality disorder. DBT teaches how to take responsibility for one's problems and how to deal with conflict and negative feelings. DBT often involves a combination of group and individual sessions.
- **Family therapy**. This therapy approach is designed to help the family unit function in more positive and constructive ways by exploring patterns of communication and providing support and education. Family therapy sessions can include the child or adolescent along with parents and siblings.
- Group therapy (GT). GT uses group dynamics and peer interactions to increase understanding, communication, and improve social skills.
- **Play** therapy. This type of therapy is directed at helping younger children. It involves the use of **toys**, blocks, dolls, puppets, **drawings**, and games to help the child recognize, identify, and verbalize feelings. The psychotherapist observes how the child uses play

materials and identifies themes or patterns to understand the child's problems. Through a combination of talk and play the child has an opportunity to better understand conflicts, feelings, and behavior.

Alternative treatment

Alternative treatments are available for personality disorders and most are complementary to conventional psychotherapy. They include the following:

- Coloring therapy. CT uses the activity of coloring as a self-help medium. While a person colors (with felt tipped markers, colored pens, pencils, etc.) a state of consciousness similar to meditation occurs. The approach is based on how people speak to themselves on the "inside." During a coloring session, people are asked to listen to the thoughts going on in their minds so as to become aware of where their thoughts, feelings, and opinions come from.
- Creative arts therapies. These therapies include art therapy, dance/movement therapy, drama therapy, music therapy, poetry therapy, and psychodrama. They use arts and creative processes to promote health, communication, and expression; they encourage the integration of physical, emotional, cognitive, and social functioning while enhancing self-awareness and facilitating change.
- Neurolinguistic programming. NLP is a method of examining the way a person thinks and acts through language and using this knowledge to effect change.

Nutritional concerns

The notion that foods and nutrients influence brain function and behavior generated in the early 2000s widespread interest in the general public and in the scientific community. However, the evaluation data are still ambiguous when it comes to establishing a direct link between personality disorders and diet, aside from recommending the avoidance of alcoholic and stimulant beverages.

Prognosis

The PD outlook varies. Some personality disorders diminish during middle age without any treatment, while others persist throughout life despite treatment.

Prevention

The prevention of personality disorders is an area surrounded with pessimism and controversy. Many mental health specialists believe that these disorders are untreatable, that individuals with personality disorder have little capacity for change; therefore not surprisingly, they remain skeptical about prevention prospects. However, even though the innate **temperament** of a person cannot be modified, understanding the factors that influence the development of personality disorders (such as genetic risks and environmental factors) may help prevention. Accordingly, some mental health professionals advocate primary prevention steps, which should include education of parents and primary healthcare workers, as well as early psychotherapy and protection of traumatized children, which can be carried out by child developing services. Some evidence suggests that traditional doctor-patient relationships are of much less value than programs which enable parents to see their own role as crucial and their own actions as able to bring changes for the better in their child's behavior. High quality parenting plays a critical role in child development and, thus, in the prevention of personality disorders.

Parental concerns

Understanding personality disorders can be challenging for parents as well as for children. During the last third of the twentieth century, great advances were made in the areas of diagnosis and treatment of personality disorders. Parents can help children understand that these are real illnesses that can be treated. In order for parents to talk with a child about a personality disorder, they must be knowledgeable of the subject. Parents may have to do some homework to become better informed. They should have a basic understanding and answers to questions such as what are personality disorders, who gets them, what causes them, how are diagnoses made, and what treatments are available. When explaining to a child about how personality disorders affect a person, it may be helpful to explain that feelings of anxiety, worry, and irritability are common for most people. However, when these feelings get very intense, last for a long period of time, and begin to interfere with school and relationships, it may be a sign of a personality disorder that can, however, be treated.

A child's personality disorder often causes disruption to both the parents' and the child's world. Parents may have difficulty being objective. They may blame themselves or worry that others such as teachers or family members will blame them. Recognizing these feelings and seeking the help of professional care providers and support groups is the best way to cope with this issue.

Medication can also be an effective part of the treatment for several personality disorders in childhood and adolescence. A doctor's recommendation to use

KEY TERMS

Anxiety—Worry or tension in response to real or imagined stress, danger, or dreaded situations. Physical reactions, such as fast pulse, sweating, trembling, fatigue, and weakness, may accompany anxiety.

Caring—The demonstration of an awareness of and a concern for the good of others.

Character—An individual's set of emotional, cognitive, and behavioral patterns learned and accumulated over time.

Delusion—A belief that is resistant to reason or contrary to actual fact. Common delusions include delusions of persecution, delusions about one's importance (sometimes called delusions of grandeur), or delusions of being controlled by others.

Eccentric—Deviating from the center; conduct and behavior departing from accepted norms and conventions.

Empathy—A quality of the client-centered therapist, characterized by the therapist s conveying appreciation and understanding of the client's point of view.

Erratic—Having no fixed course; behavior that deviates from common and accepted opinions.

Introversion—A personal preference for solitary, non-social activities and settings.

Maladaptive—Unsuitable or counterproductive; for example, maladaptive behavior is behavior that is inappropriate to a given situation.

Personality—The organized pattern of behaviors and attitudes that makes a human being distinctive. Personality is formed by the ongoing interaction of temperament, character, and environment.

Substance abuse—Maladaptive pattern of drug or alcohol use that may lead to social, occupational, psychological, or physical problems.

Temperament— A person's natural disposition or inborn combination of mental and emotional traits.

medication often raises many concerns and questions in both the parents and the child. The physician who recommends medication should be experienced in treating psychiatric illnesses in children and adolescents. He or she should fully explain the reasons for medication use, what benefits the medication should provide, as well as the possible negative side-effects or dangers and other treatment alternatives.

See also Antisocial behavior; Antisocial personality disorder; Anxiety.

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Pertussis *see* **Whooping cough**

Pervasive developmental disorders

Definition

Pervasive developmental disorders are a group of neurological disorders that include autistic disorder (**autism**), Asperger's syndrome, childhood disintegrative disorder, Rett's syndrome, and pervasive developmental disorder not otherwise specified (PDDNOS). These disorders are characterized by delayed development in functional, socialization, and **communication skills**.

Description

The term pervasive developmental disorders was first used in the 1980s to describe a class of neurological disorders that involved impaired social and communication skills and repetitive behaviors.

Due to difficulties in accurately describing these disorders using the term pervasive developmental disorders, some neurological and psychiatric specialists have proposed new terminology to describe this class of disorders, including autistic spectrum disorders and multi-system neurological disorders.

Asperger's syndrome

Asperger's syndrome is characterized by difficulties with social relationships and skills and with poor coordination and restricted range of interests. Children with Asperger's syndrome generally have a normal to above average **intelligence** level and adequate knowledge of vocabulary and grammar but poor concentration and ability to understand language subtleties, such as humor. Asperger's syndrome is often incorrectly referred to as "high-functioning autism."

Autistic disorder

Autistic disorder, also referred to as autism, is characterized by moderate to severe communication, socialization, and behavioral problems, and in some children, **mental retardation**.

Childhood disintegrative disorder

Childhood disintegrative disorder is extremely rare, relative to the other pervasive developmental disorders. Children with this disorder develop normally until at least two years of age, after which an obvious regression in multiple functional skills occurs, including bladder and bowel control, ability to move, and language skills.

Pervasive developmental disorder not otherwise specified (PDDNOS)

Children are diagnosed with PDDNOS if their symptoms do not fit any of the other four types and/or they do not have the degree of impairment of the other four types. PDDNOS involves developmental impairments, such as communication and social skills, and repetitive behaviors that cannot be attributed to a specific developmental disorder or personality disorder. Usually, children with PDDNOS do not exhibit symptoms until age three or four.

Rett's syndrome

Rett's syndrome occurs primarily in female children and is characterized by normal development for the first six to 18 months, followed by a noticeable change in behavior and loss of some abilities, especially motor skills. As the child ages, significant loss of speech, hand movement, and reasoning develops. Children with Rett's syndrome usually repeat certain movements and gestures, in particular, hand wringing or hand washing. Rett's syndrome is the rarest of the pervasive developmental disorders.

Demographics

About one in 1,000 children born in the United States is diagnosed with autistic disorder, and it is four to five times more common in boys. Rett's syndrome has been diagnosed primarily in girls. Although autism is the most well-known of these disorders, PDDNOS is at least twice as common in children.

Causes and symptoms

As of 2004, the causes of these disorders were unknown. While genetics is believed to play a primary role, some children in families with a history of pervasive developmental disorders do not have a disorder. Medical researchers believe that genetic susceptibility plus additional factors contribute to the development of one of these disorders. Factors under investigation as a cause of these disorders include immune system problems, **allergies**, drugs, environmental pollution, and infections. Autopsy studies of individuals with pervasive developmental disorders have shown that brain cell structure is different, particularly in the brain stem area. In addition, because many individuals with pervasive developmental disorders are also affected by seizures, "electrical miswiring" of the brain may also contribute to these disorders. Researchers have used **magnetic resonance imaging** (MRI) and positron emission tomography (PET) to find

subtle differences in the brain structure and function of children with these disorders.

Symptoms of pervasive developmental disorders may be visible as early as infancy; however, the typical age of onset is age three. Although each of the five types has some distinctive symptoms, in general, early symptoms of a pervasive developmental disorder include the following:

- impaired language skills
- difficulties relating to people, objects, or activities
- unusual play
- repetitive body movements or behavior patterns
- difficulties handling changes in routine or surroundings
- unusual responses to sensory stimuli, like loud noises and lights

When to call the doctor

Parents should see a physician as soon as they notice developmental problems or delays in their infant or child.

Diagnosis

Pervasive developmental disorders are diagnosed using the *Diagnostic and Statistical Manual of Mental Disorders* (DSM), which provides criteria for physicians to diagnose the specific type. Diagnosis of a pervasive developmental disorder is difficult because there is no specific medical test, like a blood test or imaging test that can confirm the diagnosis. Some physicians may hesitate to diagnose very young children with a specific type of pervasive developmental disorder.

Diagnosis of these disorders usually requires consultation and **assessment** by a specialist in childhood developmental disorders, such as a child psychiatrist, pediatric neurologist, neuropsychologist, or developmental child psychologist. These specialists evaluate laboratory medical tests, neurological tests, and **psychological tests**; interview parents and children; and observe and assess behaviors. Educational skill testing, communication assessment, and motor skill assessment may also be conducted. Medical tests that may be performed to rule out other medical conditions include electroencephalography, MRI, and blood tests.

Once a pervasive developmental disorder is diagnosed, the diagnosis must be narrowed to one of the five types, which is achieved by using pre-established DSM criteria that outline the key differences among the types. For example, for childhood disintegrative disorder to be diagnosed, symptoms must be preceded by at least two years of normal development and onset of decline and regression must occur prior to age 10 years.

Treatment

As of 2004, no cure existed for these disorders, and no specific therapy works for all individuals. Treatment depends on the severity of the disorder and consists of specialized therapy, **special education**, and medication to address specific behavioral problems. Medications that may be prescribed to treat specific symptoms include anti-depressants, anti-anxiety medications, anti-spasmodic and anti-seizure medications, and stimulants. Therapeutic interventions include applied behavior analysis (the Lovaas method), auditory integration training, behavior modification programs, play therapy, occupational and physical therapy, animal-assisted therapy, art/music/dance therapy, sensory integration, and speech therapy.

Alternative treatment

Alternative treatments for pervasive developmental disorders focus on **nutrition**. Some evidence has shown that vitamin therapy with vitamin B6 and magnesium supplementation can help children with autism and PDDNOS. Because some children with pervasive developmental disorders have **food sensitivities** or **food allergies**, allergy testing and subsequent dietary modification may help. In food-allergic children, certain foods have been shown to increase hyperactivity and autistic behavior. Anti-yeast therapy has also been proposed because children with autism and PDDNOS sometimes have higher yeast levels in their bodies. Administering anti-yeast medications has decreased negative behaviors in some children. Before parents turn to alternative therapy, they should consult a physician to make sure it does not interfere or interact with any other medications.

Prognosis

Pervasive developmental disorders are not life-threatening and do not affect normal life expectancy. Prognosis depends on the severity and type of disorder and the effectiveness of early interventions. Early intervention with specialized educational and behavioral support programs improves the quality of life and level of functioning of children with these disorders. However, because of their impaired communication and social skills, about 70 percent of individuals with a pervasive developmental disorder are never able to live on their own.

KEY TERMS

Electroencephalography—The recording of electrical impulses produced by the brain's activity via electrodes attached to a patient's scalp.

Magnetic resonance imaging (MRI)—An imaging technique that uses a large circular magnet and radio waves to generate signals from atoms in the body. These signals are used to construct detailed images of internal body structures and organs, including the brain.

Positron emission tomography (PET)—A computerized diagnostic technique that uses radioactive substances to examine structures of the body. When used to assess the brain, it produces a three-dimensional image that shows anatomy and function, including such information as blood flow, oxygen consumption, glucose metabolism, and concentrations of various molecules in brain tissue.

Prevention

Pervasive developmental disorders are caused by a complex interaction of genetics, neurological factors, and environmental factors. As of 2004, there was no genetic test to detect these disorders, and there is no way to prevent their development.

Parental concerns

The majority of children with a pervasive developmental disorder will require special education services. By law, public schools must evaluate children at no cost and provide special education services to eligible children with disabilities. Some private or alternative schools may be dedicated to serving children with such disorders and offer more comprehensive education and therapeutic options, but at an additional cost to parents.

Parenting children with pervasive developmental disorders is difficult and emotionally demanding. Parents and families can benefit from joining a support group. Benefits of parent support groups include information sharing, emotional support, and educational assistance.

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Jennifer E. Sisk, MA

Pharyngitis *see* **Sore throat**

Phenylketonuria

Definition

Phenylketonuria (PKU) is a rare metabolic disorder caused by a deficiency in the production of the hepatic (liver) enzyme phenylalanine hydroxylase (PAH).

Description

PKU is the most serious form of a class of diseases referred to as hyperphenylalaninemia, all of which involve above normal (elevated) levels of phenylalanine in the blood. The primary symptom of untreated PKU, **mental retardation**, is the result of consuming foods that contain the amino acid phenylalanine, which is toxic to brain tissue.

PKU is an inherited, autosomal recessive disorder. It is the most common genetic disease involving amino acid metabolism. As of 2004, PKU was incurable, but early, effective treatment can prevent the development of serious mental incapacity.

PKU is caused by the liver's inability to produce a particular type of PAH enzyme. This enzyme converts (metabolizes) the amino acid called phenylalanine into another amino acid, tyrosine. This is the only role of PAH in the body. A lack of PAH results in the buildup of abnormally high phenylalanine concentrations (or levels) in the blood and brain. Above normal levels of phenylalanine are toxic to the cells that make up the nervous system and cause irreversible abnormalities in brain structure and function in PKU patients. Phenylalanine is a type of teratogen (any substance or organism that can cause birth defects in a developing fetus).

The liver is the body's chief protein-processing center. Proteins are one of the major food nutrients. They are generally very large molecules composed of strings of smaller building blocks or molecules called amino acids. About twenty amino acids exist in nature. The body breaks down proteins from food into individual amino acids and then reassembles them into human proteins. Proteins are needed for growth and repair of cells and tissues and are the key components of enzymes, antibodies, and other essential substances.

PKU effects on the human nervous system

The extensive network of nerves in the brain and the rest of the nervous system are made up of nerve cells. Nerve cells have specialized extensions called dendrites and axons. Stimulating a nerve cell triggers nerve impulses (signals) that speed down the axon. These nerve impulses then stimulate the end of an axon to release chemicals called neurotransmitters that spread out and communicate with the dendrites of neighboring nerve cells.

Many nerve cells have long, wire-like axons that are covered by an insulating layer called the myelin sheath. This covering helps speed nerve impulses along the axon. In untreated PKU patients, abnormally high phenylalanine levels in the blood and brain can produce nerve cells with deformed axons and dendrites and cause imperfections in the myelin sheath referred to as hypomyelination and demylenation. This loss of myelin can short circuit nerve impulses (messages) and interrupt cell communication. A number of brain scan studies also indicate a degeneration of the white matter in the brains of older patients who have not maintained adequate dietary control.

PKU can also affect the production of one of the major neurotransmitters in the brain, called dopamine. The brain makes dopamine from the amino acid tyrosine. PKU patients who do not consume enough tyrosine in their diets cannot produce sufficient amounts of dopamine. Low dopamine levels in the brain disrupt normal communication between nerve cells, which results in impaired cognitive (mental) function.

Some research suggests that nerve cells of PKU patients also have difficulty absorbing tyrosine. This abnormality may explain why many PKU patients who receive sufficient dietary tyrosine still experience some form of learning disability.

Behavior and academic performance

IQ (**intelligence** quotient) tests provide a measure of cognitive function. The IQ of PKU patients is generally lower than the IQ of their healthy peers. Students with PKU often find academic tasks difficult and must struggle harder to succeed than their non-PKU peers. They may require special tutoring and need to repeat some of their courses. Even patients undergoing treatment programs may experience problems with typical academic tasks as math, reading, and spelling. Visual perception, visual-motor skills, and critical thinking skills can also be affected. Ten years of age seems to be an important milestone for PKU patients. After these individuals reach age 10, variations in their diets seem to have less influence on their IQ development.

People with PKU tend to avoid contact with others, appear anxious, and show signs of depression. However, some patients may be much more expressive and tend to have hyperactive, talkative, and impulsive personalities. It is also interesting to note that people with PKU are less likely to display such antisocial behaviors as **lying**,

teasing, and active disobedience. It should be emphasized that, as of 2004, research findings were still quite preliminary and more extensive research is needed to clearly show how abnormal phenylalanine levels in the blood and brain might affect behavior and academic performance.

Demographics

One in 50 individuals in the United States has inherited a gene for PKU. About 5 million Americans are PKU carriers. About one in 15,000 babies tests positive for PKU in the United States. Studies indicate that the incidence of this disease in Caucasian and Native American populations is higher than in African-American, Hispanic, and Asian populations.

Causes and symptoms

PKU symptoms are caused by alterations or mutations in the genetic code for the PAH enzyme. Mutations in the PAH gene prevent the liver from producing adequate levels of the PAH enzyme needed to break down phenylalanine. The PAH gene and its PKU mutations are found on chromosome 12 in the human genome. In more detail, PKU mutations can involve many different types of changes, such as deletions and insertions, in the DNA of the gene that codes for the PAH enzyme.

PKU is described as an inherited, autosomal recessive disorder. The term autosomal means that the gene for PKU is not located on either the X or Y sex chromosome. The normal PAH gene is dominant to recessive PKU mutations. A recessive genetic trait, such as PKU, is one that is expressed—or shows up—only when two copies are inherited (one from each parent).

A person with one normal and one PKU gene is called a carrier. A carrier does not display any symptoms of the disease because the carrier's liver produces normal quantities of the PAH enzyme. However, PKU carriers can pass the PKU genetic mutation on to their children. Two carrier parents have a 25 percent chance of producing a baby with PKU symptoms, and a 50 percent chance having a baby that is a carrier for the disease. Although PKU conforms to these basic genetic patterns of inheritance, the actual expression, or phenotype, of the disease is not strictly an either/or situation. This is because there are at least 400 different types of PKU mutations. Although some PKU mutations cause rather mild forms of the disease, others can initiate much more severe symptoms in untreated individuals. The more severe the PKU mutation, the greater the effect on **cognitive development** and performance (mental ability).

Untreated PKU patients develop a broad range of symptoms related to severely impaired cognitive function, sometimes referred to as mental retardation. Other symptoms can include extreme patterns of behavior, delayed speech development, seizures, a characteristic body odor, and light body pigmentation. The light pigmentation is due to a lack of melanin, which normally colors the hair, skin, and eyes. Melanin is made from the amino acid tyrosine, which is lacking in untreated cases of PKU. Physiologically, PKU patients show high levels of phenylalanine and low levels of tyrosine in the blood. Babies do not show any visible symptoms of the disease for the first few months of life. However, typical PKU symptoms usually do show up by a baby's first birthday.

Diagnosis

The primary diagnostic test for PKU is the measurement of phenylalanine levels in a drop of blood taken from the heel of a newborn baby's foot. This screening procedure is referred to as the Guthrie test (Guthrie bacterial inhibition assay). In this test, PKU is confirmed by the appearance of bacteria growing around high concentrations of phenylalanine in the blood spot. PKU testing was introduced in the early 1960s and is the largest genetic screening program in the United States. It is required by law in all 50 states. Early diagnosis is critical. It ensures early the treatment PKU babies need to develop normally and avoid the ravages of PKU.

The American Academy of Pediatrics recommends that this test be performed on infants between 24 hours and seven days after birth. The preferred time for testing is after the baby's first feeding. If the initial PKU test produces a positive result, then follow-up tests are performed to confirm the diagnosis and to determine if the elevated phenylalanine levels may be caused by some medical condition other than PKU. Treatment for PKU is recommended for babies that show a blood phenylalanine level of 7 to 10 mg/dL or higher for more than a few consecutive days. Another, more accurate test procedure for PKU measures the ratio (comparison) of the amount of phenylalanine to the amount of tyrosine in the blood.

Subsequent diagnostic procedures (called mutation analysis and genotype determination) can actually identify the specific types of PAH gene mutations inherited by PKU infants. Large-scale studies have helped to clarify how various mutations affect the ability of patients to process phenylalanine. This information can help doctors develop more effective customized treatment plans for each of their PKU patients.

Treatment

The severity of the PKU symptoms experienced by people with this disease is determined by both lifestyle and genetic factors. In the early 1950s, researchers first demonstrated that phenylalanine-restricted diets could eliminate most of the typical PKU symptoms—except for mental retardation. As of 2004, dietary therapy (also called **nutrition** therapy) is the most common form of treatment for PKU patients. PKU patients who receive early and consistent dietary therapy can develop fairly normal mental capacity to within about five IQ points of their healthy peers. By comparison, untreated PKU patients generally have IQ scores below 50.

Infants with PKU should be put on a specialized diet as soon as they are diagnosed to avoid progressive brain damage and other problems caused by an accumulation of phenylalanine in the body. A PKU diet helps patients maintain very low blood levels of phenylalanine by restricting the intake of natural foods that contain this amino acid. Even breast milk is a problem for PKU babies. Special PKU dietary mixtures or formulas are usually obtained from medical clinics or pharmacies.

Phenylalanine is actually an essential amino acid. This means that it has to be obtained from food because the body cannot produce this substance on its own. Typical diets prescribed for PKU patients provide very small amounts of phenylalanine and higher quantities of other amino acids, including tyrosine. The amount of allowable phenylalanine can be increased slightly as a child grows older.

In addition, PKU diets include all the nutrients normally required for good health and normal growth, such as carbohydrates, fats, **vitamins**, and **minerals**. High protein foods such as meat, fish, chicken, eggs, nuts, beans, milk, and other dairy products are banned from PKU diets. Small amounts of moderate protein foods (such as grains and potatoes) and low protein foods (some fruits and vegetables and low protein breads and pastas) are allowed. Sugar-free foods, such as diet soda, which contain the artificial sweetener aspartame, are also prohibited foods for PKU patients because aspartame contains the amino acid phenylalanine.

Ideally, school-age children with PKU should be taught to assume responsibility for managing their diets, recording food intake, and for performing simple blood tests to monitor their phenylalanine levels. Blood tests should be done in the early morning when phenylalanine levels are highest. Infants and young children require more frequent blood tests than older children and adults. The amount of natural foods allowed in a diet can be adjusted to ensure that the level of phenylalanine in the blood is kept within a safe range—2 to 6 mg/dL before 12 years of age and 2 to 15 mg/dL for PKU patients over 12 years old.

Blood taken from a newborn's heel is tested for phenylketornuia (PKU). *(© Custom Medical Stock Photo, Inc.)*

A specialized PKU diet can cause abnormal fluctuations in tyrosine levels throughout the day. Thus, some health professionals recommend adding time-released tyrosine that can provide a more constant supply of this amino acid to the body. It should be noted that some PKU patients show signs of learning disabilities even with a special diet containing extra tyrosine. Research studies suggests that these PKU patients may not be able to process tyrosine normally.

For PKU caregivers, providing a diet that is appealing as well as healthy and nutritious is a constant challenge. Many PKU patients, especially teenagers, find it difficult to stick to the relatively bland PKU diet for extended periods of time. Some older patients decide to go off their diet plan simply because they feel healthy. However, many patients who abandon careful nutritional

KEY TERMS

Amino acid—An organic compound composed of both an amino group and an acidic carboxyl group. Amino acids are the basic building blocks of proteins. There are 20 types of amino acids (eight are "essential amino acids" which the body cannot make and must therefore be obtained from food).

Axon—A long, threadlike projection that is part of a neuron (nerve cell).

Enzyme—A protein that catalyzes a biochemical reaction without changing its own structure or function.

Gene—A building block of inheritance, which contains the instructions for the production of a particular protein, and is made up of a molecular sequence found on a section of DNA. Each gene is found on a precise location on a chromosome.

Genetic disease—A disease that is (partly or completely) the result of the abnormal function or expression of a gene; a disease caused by the inheritance and expression of a genetic mutation.

Intelligence quotient (IQ)—A measure of somebody's intelligence, obtained through a series of aptitude tests concentrating on different aspects of intellectual functioning.

Metabolism—The sum of all chemical reactions that occur in the body resulting in growth, transformation of foodstuffs into energy, waste elimination, and other bodily functions. These include processes that break down substances to yield energy and processes that build up other substances necessary for life.

Mutation—A permanent change in the genetic material that may alter a trait or characteristic of an individual, or manifest as disease. This change can be transmitted to offspring.

Myelin—A fatty sheath surrounding nerves throughout the body that helps them conduct impulses more quickly.

Nervous system—The system that transmits information, in the form of electrochemical impulses, throughout the body for the purpose of activation, coordination, and control of bodily functions. It is comprised of the brain, spinal cord, and nerves.

Phenylalanine—An essential amino acid that must be obtained from food since the human body cannot manufacture it. It is necessary for normal growth and development and for normal protein metabolism.

Protein—An important building block of the body, a protein is a large, complex organic molecule composed of amino acids. It is involved in the formation of body structures and in controlling the basic functions of the human body.

Recessive—Refers to an inherited trait that is outwardly obvious only when two copies of the gene for that trait are present. An individual displaying a recessive trait must have inherited one copy of the defective gene from each parent.

management develop cognitive problems, such as difficulties remembering, maintaining focus, and paying attention. Many PKU health professionals contend that all PKU patients should adhere to a strictly controlled diet for life.

One promising line of PKU research involves the synthesis (manufacturing) of a new type of enzyme that can break down phenylalanine in food consumed by the patient. This medication would be taken orally and could prevent the absorption of digested phenylalanine into the patient's bloodstream.

In general, medical researchers express concern about the great variation in treatment programs available in the early 2000s to PKU patients around the world. They have highlighted the urgent need for consistent international standards for proper management of PKU patients, which should emphasize comprehensive psychological as well as physiological monitoring and **assessment**.

PKU and pregnancy

Women with PKU must be especially careful with their diets if they want to have children. They should ensure that phenylalanine blood levels are under control before conception and throughout pregnancy. Mothers with elevated (higher than normal) phenylalanine levels are high risk for having babies with significant birth defects, such as microencephaly (smaller than normal head size), **congenital heart disease** (abnormal heart structure and function), stunted growth, mental retardation, and psychomotor (coordination) difficulties. This condition is referred to as maternal PKU and can even affect babies who do not have the PKU disease.

Prognosis

Early newborn screening, careful monitoring, and life-long strict dietary management can help PKU patients to live normal, healthy, and long lives.

Parental concerns

Every state in the United States has mandatory newborn screening programs in place for phenylketonuria, as well as other diseases. Parents who suspect that other genetic diseases may run in their families should speak to their healthcare providers before their baby's birth to ascertain what other screening tests should be run.

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Children's PKU Network. 1520 State St., Suite 111, San Diego, CA 92101–2930. Web site: <www.pkunetwork.org>.

March of Dimes Birth Defects Foundation. 1275 Mamaroneck Ave., White Plains, NY 10605. Web site: <www.modimes.org>.

National PKU News. 6869 Woodlawn Avenue, NE, #116, Seattle, WA 98115–5469. Web site: <www.pkunews.org>.

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Phenylpropanolamine *see* **Decongestants**

Phobias

Definition

A phobia is an intense and unrealistic **fear** brought on by an object, event, or situation, which can interfere with the ability to socialize, work, or go about everyday life.

Description

Almost all children develop specific fears at some age. Sometimes the fear is a result of a particular event, but some fears arise on their own. Many fears are associated with certain age groups. Very young children (through age two) tend to fear loud noises, strangers, large objects, and being away from their parents. Preschoolers often have imaginary fears, such as monsters who might eat them, strange noises, being alone in the dark, or thunder. School-age children have concrete fears, such being hurt, doing badly in school, dying, or natural disasters. When the child is afraid of something past the age at which it is normal, when the fear interferes with the child's ability to function normally, then the fear ranks as a phobia.

Phobias belong to a large group of mental problems known as **anxiety** disorders that include **obsessive-compulsive disorder** (OCD), panic disorder, and post-traumatic stress disorder. Phobias themselves can be divided into three specific types:

- specific phobias (formerly called simple phobias, most common in children)
- social phobia
- agoraphobia (not common in children)

Specific phobias

As its name suggests, a specific phobia is the fear of a particular situation or object, for example, flying on an airplane or going to the dentist. Found in one out of every 10 Americans, specific phobias seem to run in families and are roughly twice as likely to appear in women. If the person rarely encounters the feared object, the phobia does not cause much harm. However, if the feared object or situation is common, it can seriously disrupt the person's everyday life. Common examples of specific phobias, which can begin at any age, include fear of insects, snakes, and dogs; escalators, elevators, and bridges; high places; and open spaces. Children often have specific phobias that they outgrow over time, and they can learn specific fears from adults or other children around them, or even from television.

Social phobia

People with social phobia have deep fears of being watched or judged by others and being embarrassed in public. This may extend to a general fear of social situations. They may be more specific or circumscribed, such as a fear of giving speeches or of performing (stage fright). More rarely, people with social phobia may have trouble using a public restroom, eating in a restaurant, or signing their name in front of others. Young children

often have a fear of strangers that is quite normal; social phobia is not usually diagnosed until a child reaches adolescence and has crippling fears that interfere with normal function.

Social phobia is not the same as **shyness**. Shy people may feel uncomfortable with others, but they do not experience severe anxiety, they do not worry excessively about social situations beforehand, and they do not avoid events that make them feel self-conscious. On the other hand, people with social phobia may not be shy; they may feel perfectly comfortable with people except in specific situations. Social phobias may be only mildly irritating, or they may significantly interfere with daily life. It is not unusual for people with social phobia to turn down job offers or avoid relationships because of their fears.

Agoraphobia

Agoraphobia is the intense fear of being trapped and having a panic attack in a public place. It usually begins between ages 15 and 35 and affects three times as many women as men or approximately 3 percent of the population.

An episode of spontaneous panic is usually the initial trigger for the development of agoraphobia. After an initial panic attack, the person becomes afraid of experiencing a second one. People are literally fearful of fear. They worry incessantly about when and where the next attack may occur. As they begin to avoid the places or situations in which the panic attack occurred, their fear generalizes. Eventually the person completely avoids public places. In severe cases, people with agoraphobia can no longer leave their homes for fear of experiencing a panic attack.

Demographics

Approximately one person in five (18 percent) of all Americans experience phobias that interfere with their daily lives. Almost all children experience some specific fears at some point, but not many rise to the level of phobia or require professional treatment.

Causes and symptoms

Experts do not really know why phobias develop, although research suggests the tendency to develop phobias may be a complex interaction between heredity and environment. Some hypersensitive people have unique chemical reactions in the brain that cause them to respond much more strongly to stress. These people also may be especially sensitive to **caffeine**, which triggers certain brain chemical responses.

Advances in neuroimaging have also led researchers to identify certain parts of the brain and specific neural pathways that are associated with phobias. One part of the brain that was as of 2004 being studied is the amygdala, an almond-shaped body of nerve cells involved in normal fear conditioning. Another area of the brain that appears to be linked to phobias is the posterior cerebellum.

While experts believe the tendency to develop phobias runs in families and may be hereditary, a specific stressful event usually triggers the development of a specific phobia or agoraphobia. For example, someone predisposed to develop phobias who experiences severe turbulence during a flight might go on to develop a phobia about flying. What scientists do not understand is why some people who experience a frightening or stressful event develop a phobia and others do not.

Social phobia typically appears in childhood or **adolescence**, sometimes following an upsetting or humiliating experience. Certain vulnerable children who have had unpleasant social experiences (such as being rejected) or who have poor social skills may develop social phobias. The condition also may be related to low **self-esteem**, unassertive personality, and feelings of inferiority.

A person with agoraphobia may have a panic attack at any time, for no apparent reason. While the attack may last only a minute or so, the person remembers the feelings of panic so strongly that the possibility of another attack becomes terrifying. For this reason, people with agoraphobia avoid places where they might not be able to escape if a panic attack occurs. As the fear of an attack escalates, the person's world narrows.

While the specific trigger may differ, the symptoms of different phobias are remarkably similar: feelings of terror and impending doom, rapid heartbeat and breathing, sweaty palms, and other features of a panic attack. People may experience severe anxiety symptoms in anticipating a phobic trigger. For example, someone who is afraid to fly may begin having episodes of pounding heart and sweating palms at the mere thought of getting on a plane in two weeks.

When to call the doctor

A doctor, mental health professional, or counselor should be consulted when irrational fears interfere with a child's normal functioning.

Diagnosis

A mental health professional can diagnose phobias after a detailed interview and discussion of both mental and physical symptoms. Children are often less able to accurately describe their symptoms or discuss their fears, and so should be encouraged to talk about them with parents. Social phobia is often associated with other anxiety disorders, depression, or substance abuse.

Treatment

People who have a specific phobia that is easy to avoid (such as snakes) and that does not interfere with their lives may not need to get help. When phobias do interfere with a person's daily life, a combination of psychotherapy and medication can be quite effective. Medication is used less often in young children, but more frequently in older children or adolescents with severe phobias and associated depression. While most health insurance covers some form of mental health care, most do not cover outpatient care completely, and most have a yearly or lifetime maximum.

Medication can block the feelings of panic and, when combined with cognitive-behavioral therapy, can be quite effective in reducing specific phobias and agoraphobia.

Cognitive-behavioral therapy adds a cognitive approach to more traditional behavioral therapy. It teaches individuals how to change their thoughts, behaviors, and attitudes, while providing techniques to lessen anxiety, such as deep breathing, muscle relaxation, and refocusing.

One cognitive-behavioral therapy is desensitization (also known as exposure therapy), in which people are gradually exposed to the frightening object or event until they become used to it and their physical symptoms decrease. For example, someone who is afraid of snakes might first be shown a photo of a snake. Once the person can look at a photo without anxiety, he might then be shown a video of a snake. Each step is repeated until the symptoms of fear (such as pounding heart and sweating palms) disappear. Eventually, the person might reach the point where he can actually touch a live snake. Three-fourths of affected people are significantly improved with this type of treatment.

Another, more dramatic, cognitive-behavioral approach is called flooding. It exposes the person immediately to the feared object or situation. The person remains in the situation until the anxiety lessens.

Several drugs are used to treat specific phobias by controlling symptoms and helping to prevent panic attacks. These include anti-anxiety drugs (benzodiazepines) such as alprazolam (Xanax) or diazepam (Valium). Blood pressure medications called beta blockers, such as propranolol (Inderal) and atenolol (Tenormin), appear to work well in the treatment of circumscribed social phobia, when anxiety gets in the way of performance, such as public speaking. These drugs reduce over-stimulation, thereby controlling the physical symptoms of anxiety.

In addition, some **antidepressants** may be effective when used together with cognitive-behavioral therapy. These include the monoamine oxidase inhibitors (MAO inhibitors) phenelzine (Nardil) and tranylcypromine (Parnate), as well as selective serotonin reuptake inhibitors (SSRIs) like fluoxetine (Prozac), paroxetine (Paxil), sertraline (Zoloft) and fluvoxamine (Luvox).

In all types of phobias, symptoms may be eased by lifestyle changes, such as the following:

- eliminating caffeine
- cutting down on alcohol
- eating a good diet
- getting plenty of exercise
- reducing stress

Treating agoraphobia is more difficult than other phobias because there are often so many fears involved, such as fear of open spaces, traffic, elevators, and escalators. Treatment includes cognitive-behavioral therapy with antidepressants or anti-anxiety drugs. Paxil and Zoloft are used to treat panic disorders with or without agoraphobia.

Prognosis

Phobias are among the most treatable mental health problems; depending on the severity of the condition and the type of phobia, most properly treated people can go on to lead normal lives. Research suggests that once a person overcomes the phobia, the problem may not return for many years, if it returns at all. Children most often outgrow their specific phobias, with or without treatment.

Untreated phobias are another matter. In adults, only about 20 percent of specific phobias go away without treatment, and agoraphobia gets worse with time if untreated. Social phobias tend to be chronic and are not likely go away without treatment. Moreover, untreated phobias can lead to other problems, including depression, **alcoholism**, and feelings of shame and low self-esteem. Therefore, specific phobias that persist into adolescence should receive professional treatment.

KEY TERMS

Agoraphobia—Abnormal anxiety regarding public places or situations from which the person may wish to flee or in which he or she would be helpless in the event of a panic attack.

Benzodiazepine—One of a class of drugs that has hypnotic and sedative action, used mainly as tranquilizers to control symptoms of anxiety. Diazepam (Valium), alprazolam (Xanax), and chlordiazepoxide (Librium) are all benzodiazepines.

Beta blockers—The popular name for a group of drugs that are usually prescribed to treat heart conditions, but that also are used to reduce the physical symptoms of anxiety and phobias, such as sweating and palpitations. These drugs, including nadolol (Corgard) and digoxin (Lanoxin), block the action of beta receptors that control the speed and strength of heart muscle contractions and blood vessel dilation. Beta blockers are also called beta-adrenergic blocking agents and antiadrenergics.

Monoamine oxidase (MAO) inhibitors—A type of antidepressant that works by blocking the action of a chemical substance known as monoamine oxidase in the nervous system.

Neuroimaging—The use of x-ray studies and magnetic resonance imaging (MRI) to detect abnormalities or trace pathways of nerve activity in the central nervous system.

Selective serotonin reuptake inhibitors (SSRIs)—A class of antidepressants that works by blocking the reabsorption of serotonin in the brain, thus raising the levels of serotonin. SSRIs include fluoxetine (Prozac), sertraline (Zoloft), and paroxetine (Paxil).

Serotonin—A widely distributed neurotransmitter that is found in blood platelets, the lining of the digestive tract, and the brain, and that works in combination with norepinephrine. It causes very powerful contractions of smooth muscle and is associated with mood, attention, emotions, and sleep. Low levels of serotonin are associated with depression.

Social phobia—An anxiety disorder characterized by a strong and persistent fear of social or performance situations in which the individual might feel embarrassment or humiliation.

A group of researchers in Boston reported in 2003 that phobic anxiety appears to be a risk factor for Parkinson's disease (PD) in males, although as of 2004 it is not known whether phobias cause PD or simply share an underlying biological cause.

While most specific phobias appear in childhood and subsequently fade away, those that remain in adulthood often need to be treated. Unfortunately, most people never get the help they need; only about 25 percent of people with phobias ever seek help for their condition.

Prevention

There was, as of 2004, no known way to prevent the development of phobias. Medication and cognitive-behavioral therapy may help prevent the recurrence of symptoms once they have been diagnosed.

Nutritional concerns

Unless a phobia involves fear of eating a needed food, there are no nutritional concerns associated with phobias.

Parental concerns

Parents should be observant to ensure that unusual fears or phobias do not interfere in the lives of their children. Parents should recognize that a child's fears are real, and encourage the child to talk about his or her feelings, without trivializing the fear. Parents should be sympathetic, but not allow the child to avoid situations in which the child must encounter the feared object or events. If a school-age child has fears that interfere with the child's education, ability to make friends, or participate in other normal activities, a professional should be consulted.

Resources

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ABIL Incorporated. 400 West 32nd Street, Richmond, Virginia 23225. Web site: <www.anxietysupport.org/b001menu.htm>.

Agoraphobics in Motion. 1719 Crooks, Royal Oak, MI 48067. Web site: <www.aim-hq.org/>.

American Academy of Family Physicians. 11400 Tomahawk Creek Parkway, Leawood, KS 66211–2672. Web site: <www.aafp.org/>.

American Academy of Pediatrics. 141 Northwest Point Boulevard, Elk Grove Village, IL 60007–1098. Web site: <www.aap.org/>.

American Psychiatric Association. 1400 K Street NW, Washington, DC 20005. Web site: <www.psych.org/>.

American Psychological Association. 750 First Street NW, Washington, DC, 20002–4242. Web site: <www.apa.org/>.

Anxiety Disorders Association of America.8730 Georgia Avenue, Suite 600, Silver Spring, MD 20910. Web site: <www.adaa.org/>.

National Anxiety Foundation.3135 Custer Dr., Lexington, KY 40517. Web site: <www.lexington-on-line.com/naf.html>.

National Institute of Mental Health. 6001 Executive Boulevard, Rm. 8184, MSC 9663, Bethesda, MD 20892–9663. Web site: <www.nimh.nih.gov/home.cfm>.

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L. Fleming Fallon, Jr., MD, DrPH

Pica

Definition

Pica is the persistent craving and compulsive eating of non-food substances.

Description

The puzzling phenomenon of pica has been recognized and described since ancient times. Pica has been observed in ethnic groups worldwide, in both primitive and modernized cultures, in both sexes, and in all age groups. The word pica comes from the Latin name for magpie, a bird known for its unusual and indiscriminate eating habits. In addition to humans, pica has been observed in other animals, including the chimpanzee.

Demographics

True pica affects people of all ages, although it is more common in children. There are some regional variations concerning specific substances. For example, eating clay is more prevalent among women in the American southeast than in other areas of the country. Adolescents may chew ice due to **peer pressure** or because they are deficient in iron. Without a blood test for serum iron, there is no way to differentiate these causes.

Causes and symptoms

Pica in humans has many different subgroups, defined by the substance that is ingested. Some of the most commonly described types of pica are eating earth, soil, or clay (geophagia); ice (pagophagia); and starch (amylophagia). However, pica involving dozens of other substances, including cigarette butts and ashes, hair, paint chips, and paper have also been reported.

Although pica can occur in individuals of any background, a higher incidence of pica is associated with:

- pregnancy
- developmental disabilities
- **mental retardation**
- psychiatric disease and autism
- early childhood (under age three)
- poor **nutrition** or low blood levels of iron and other **minerals**
- certain cultural or religious traditions

When to call the doctor

A health care professional should be consulted whenever a child over the age of three repeatedly ingests non-food substances for a period over one month. The behavior might be merely habitual, but it can become a compulsion that needs treatment.

Diagnosis

In order for the diagnosis of pica to be made, there must be a history of persistent consumption of a non-food substance continuing for a minimum period of one month. Infants and toddlers are typically excluded from this diagnosis since mouthing objects is a normal developmental behavior at that age. Individuals with mental retardation who function at or below an approximate cognitive level of 18 months may also be exempt from this diagnosis.

Pica is most often diagnosed when a report of such behaviors can be provided by an individual or documented by another person. In other cases, pica is diagnosed after studies have been performed to assess the presenting symptoms. For example, imaging studies ordered to assess severe gastrointestinal complaints may reveal intestinal blockage with an opaque substance; such a finding is suggestive of pica. Biopsy of intestinal contents can also reveal findings, such as parasitic infection, consistent with pica. Pica may also be suspected if abnormal levels of certain minerals or chemicals are detected in the blood.

Treatment

Treatment of pica will often depend on the cause and type of pica. Conventional medical treatment may be appropriate in certain situations. For example, supplementation with iron-containing **vitamins** has been shown to cause the unusual cravings to subside in some iron-deficient people.

Medical complications and health threats, including high lead levels, bowel perforation or intestinal obstruction, will require additional medical management, beyond addressing the underlying issue of pica.

Because most cases of pica do not have an obvious medical cause, treatment with counseling, education, and nutritional management is often more successful and more appropriate than treatment with medication. Some therapists specializing in eating disorders may have expertise in treating pica.

Prognosis

The prognosis for individuals with pica varies greatly, according to the type and amount of substance ingested, the extent of presenting side effects, and the success of treatment. Many of the side effects and complications of pica can be reversed once the behavior is stopped, while other complications, including infection and bowel perforation, pose significant health threats and if not successfully treated may result in death.

When seen in children, pica behavior tends to lessen with age. However, individuals with a history of pica are more likely to experience it again. Counseling and nutritional education can reduce the risk of recurrence.

Prevention

There are no known methods of preventing pica. However, once pica is known or suspected, measures can be taken to reduce further ingestion of non-food substances. Removing the particular substance from readily accessible areas can be helpful. Close observation of the individual with pica may limit inappropriate eating behaviors.

Nutritional concerns

Pica may be a symptom of an underlying nutritional deficiency. Correcting the deficiency usually stops the pica.

KEY TERMS

Amylophagia—The compulsive eating of purified starch, typically cornstarch or laundry starch.

Geophagia—The compulsive eating of earth substances, including sand, soil, and clay.

Pagophagia—The compulsive eating of ice.

Parental concerns

Parents should monitor the food and other substances that their children eat. Repeated ingestion of non-food substances may be cause for concern. An evaluation by a pediatrician is recommended in such circumstances. Parents should be especially careful of children who eat paint chips, because this can cause lead poisoning if the paint is from an older home in which lead paint was used.

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L. Fleming Fallon, Jr., MD, DrPH

Piercing and tattoos

Definition

Body piercing and tattoos are forms of body art that have been practiced throughout history by various cultures.

Purpose

Tattoos and body piercing are done as expressions of independence, for religious or cultural reasons, or to adorn one's body. Tattooing is accomplished by injecting pigment into the deeper layers of the skin, usually by way of needles or air pressure.

Piercing is performed quickly and without anesthesia by either a spring-loaded ear-piercing gun or piercing needles, with the needle diameter varying from six to 18 gauge. The skin is cleaned, and then the needle and jewelry are inserted through the tissue in one swift motion. Piercing is typically completed in tattoo or beauty parlors.

Description

Various cultures have embraced adorning the human body with piercings and tattoos throughout history. In 1992, the 4,000-year-old body of a tattooed man was discovered in a glacier on the Austrian border, and historical research has shown that Egyptians identified tattooing with fertility and nobility in the period from

Teenage girl with tattoos. *(© PictureNet/Corbis.)*

4000 to 2000 B.C. Similar to tattooing, body piercing also has a long history, which includes being used as a symbol of royalty and courage. In some hunting and gathering societies, body piercing and tattoos have long been used in initiation rites and as symbols of socialization and enculturation.

In industrialized cultures in the early 2000s, tattoos and piercing have become a popular art form enjoyed by people of all ages. They also are indicative of a psychology of **self-mutilation**, defiance, independence, and belonging, as for example in prison or gang cultures.

Popular piercing sites include the ear, nasal septum, eyebrow, tongue, cheek, nipple, navel, labia, and penis. Tattoos permanently mark various areas on the body.

Originating from the Tahitian word *tattau*, meaning "to mark," tattoos are relatively permanent marks or designs on the skin. An electric needle injects colored pigment into small, deep holes made in the skin to form the tattoo. Prison tattoo techniques are usually very crude, in marked contrast to the highly skilled art practiced in Japan and also performed in the United States and in Europe. In the early 2000s, the ancient art of Mehndi, or temporary tattooing of the skin with a paste made of henna has become popular in the United States and around the world. Henna is a stain normally made for hair and, therefore, exempt from U.S. Food and Drug Administration regulation. Although seemingly safe because it does not pierce the skin, henna tattoos using black henna, a paste that contains parahenylenediamine, can actually be dangerous when absorbed into the skin of some people.

Risks

While piercing and tattooing are popular, both present distinct health risks. Tattoos can lead to the transmission of infectious diseases, such as **hepatitis B** and C and theoretically HIV, when proper sterilization and **safety** procedures are not followed. Black henna tattoos can cause significant **allergies** and **rashes**, leading to renal (kidney) failure and even death in those who are sensitive to their ingredients. These types of tattoos have appeared particularly dangerous to young children. Body piercing also presents the risk of chronic infection, scarring, hepatitis B and C, **tetanus**, and skin allergies to the

jewelry that is used. One study reported that 17 percent of college students with piercings suffered a medical complication such as infection or tearing. Use of piercing guns and preferences for upper ear piercing have led to increased infections. The force of the gun's delivery further complicates matters around the delicate cartilage of the upper ear and some people require surgical intervention.

Body piercing and tattooing are unregulated in most parts of the United States, and illegal in some. The American Dental Association opposes oral (tongue, lip, or cheek) piercing, and the American Academy of Dermatology is against all forms of body piercing except ear lobe piercing.

Some of the signs of an infection from either piercing or tattoos are obvious, such as inflammation of the pierced or tattooed area, while the symptoms of hepatitis C, the most common blood-borne infection in the United States, may not be so obvious. Allergic responses to tattoos may occur due to the pigment compounds used, such as oxides of iron, mercury, chromium, cadmium, and cobalt and synthetic organic dyes. Symptoms of an allergic reaction include swelling, redness, and severe **itching**. The symptom of henna tattoo reaction is an eczema-like rash around the tattoo site. The patch should be tested for reaction severity before it proceeds to anaphylactic shock or severe allergic reaction.

Most infections from piercing are due to the use of non-sterile techniques. The skin pathogens streptococcus and staphylococcus are most frequently involved in skin infections from piercing. The fleshy tissue around the pierced area may weaken and tear, leading for example, to a badly disfigured earlobe. Other common complications include **contact dermatitis** and scars. Piercing can result in endocarditis (inflammation of the heart), urethral rupture (when the labia is pierced), and a serious infection of the penis foreskin (when the foreskin is pierced) leading to severe disability or even, on rare occasions, death.

Normal results

Though painful initially, many tattoos and piercings heal well, do not become infected, and are satisfying to the wearer. Those interested in getting a tattoo or piercing should look for an established business with clean facilities. Equipment should be sterilized between uses, and the person applying the tattoo or piercing should wear clean latex gloves. The skin should be cleaned and an antiseptic applied to minimize the risk of infection. Common problems are infection, which can range in severity from mild to severe, and deciding that a particular tattoo or piercing is unsatisfactory. Piercings can close on their own if allowed to heal naturally shortly after the piercing is done. If done too late, the hole may close incompletely or not at all. Tongue piercings are among the most likely to become infected and often cause tooth damage.

Common healing times for body piercings

Ear lobe: six to eight weeks

Ear cartilage: four months to one year

Eyebrow: six to eight weeks

Nostril: two to four months

Nasal septum: six to eight months

Nasal bridge: eight to ten weeks

Tongue: four weeks (Can cause partial paralysis if jewelry pierces a nerve.)

Lip: two to three months

Nipple: three to six months

Navel: four months to one year

Female genitalia: four to ten weeks

Male genitalia: four weeks to six months

SOURCE: Akron Children's Hospital, "Tips to Grow By," 2003.

(Table by GGS Information Services.)

Treatment of a local infection from piercing includes warm compresses and antibacterial ointment for local infections to a five-day course of oral antibiotic therapy. If hepatitis B or C is confirmed, a series of diet and lifestyle changes, such as the elimination of alcohol, is recommended to control the disease.

There are five methods to remove tattoos when desired, including using a laser to break up tattoo pigments; surgical removal that involves cutting the tattoo away; sanding the skin with a wire brush to remove the epidermis and dermis layers in a process called dermabrasion; using a salt solution to soak the tattooed skin (salabrasion); and scarification, removing the tattoo with an acid solution to form a scar in its place. Topical steroids can often treat reactions to henna tattoos, but improvement may take several weeks.

Depending on the type of infection resulting from either piercing or tattoos, the treatment and prognosis vary. Minor infections respond well to antibiotic therapy, while blood-borne diseases such as hepatitis B and C cause life-altering results. Disfigurement may or may not be fully correctable by later plastic surgery. People particularly sensitive or allergic to the ingredients in

KEY TERMS

Endocarditis—Inflammation of the inner membrane lining of the heart and/or of the heart valves caused by infection.

Hepatitis—An inflammation of the liver, with accompanying liver cell damage or cell death, caused most frequently by viral infection, but also by certain drugs, chemicals, or poisons. May be either acute (of limited duration) or chronic (continuing). Symptoms include jaundice, nausea, vomiting, loss of appetite, tenderness in the right upper abdomen, aching muscles, and joint pain. In severe cases, liver failure may result.

Socialization—The process by which new members of a social group are integrated in the group.

black henna may suffer serious consequences, even death, if their reaction progresses. Others may be left with scarring or altered pigmentation along the tattoo design.

Parental concerns

Obviously, the best way to prevent infection from piercing or tattoos is not to get one in the first place. However, the risks can be minimized. Procedures should be performed in a sterile environment by an experienced professional. The person performing the procedure should remove a new needle from the plastic in front of the person to be tattooed and should put on a new pair of sterile gloves. Anyone considering a henna tattoo should require proof from the artist that he or she is using pure, safe brown henna, not the unsafe black henna.

Piercing should be completed with smoothly polished jewelry made of 14 or 18 carat gold, titanium, surgical steel, or niobium. An allergic reaction can result with the use of jewelry made of brass plate or containing a nickel alloy. Healing time from a piercing ranges from six months to two years. A piercing should be completed in a sterile environment that uses every precaution to reduce the risk of infection. Excessive force, such as exerting a strong pull, should never be applied to jewelry inserted into pierced body parts to avoid tearing and injuring tissues.

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L. Fleming Fallon, Jr., MD, DrPH

Pimples *see* **Acne**

Pinkeye *see* **Conjuctivitis**

Pinta

Definition

Pinta is a bacterial infection of the skin that causes lesions, red to bluish-black colored spots and splotches, and discoloration of the skin.

Description

Pinta is a skin infection caused by the bacterium *Treponema carateum*, a relative of the bacterium that causes syphilis. The word "pinta" comes from Spanish and means "painted." Pinta is also known as "azula" (blue), and "mal de pinto" (pinto sickness). It is one of several infections caused by different *Treponema* bacteria, which are called "endemic" or "non-venereal" treponematoses.

Transmission

Pinta is spread from one person to another by direct skin-to-skin contact. The bacteria enter the skin through a small cut, scratch, or other skin damage. Once inside the body, warmth and moisture allow the bacteria to multiply. The bacterial infection causes red, scaly lesions on the skin.

Demographics

Pinta is primarily found in rural, poverty-stricken areas of northern South America, Mexico, and the Caribbean. The disease is usually acquired during late childhood and **adolescence**. It is very rare in the United States. In the 1950s, it is estimated that there were about one million cases of Pinta in South and Central America. That number has been reduced drastically, and recently there are believed to be only a few hundred cases a year in those areas.

Causes and symptoms

Pinta is caused by an infection with the bacterium *Treponema carateum*. Symptoms occur two to four weeks after exposure to the bacteria. The first sign of infection is a red, scaly, slowly enlarging bump on the skin. This is called the primary lesion. The primary lesion usually appears at the site where the bacteria entered the skin, most often on the arms, legs, or face. Smaller lesions then form around the primary lesion. These are called satellite lesions. Lymph nodes located near the infected area may become enlarged, but are painless.

The second stage of pinta occurs between one and 12 months after the primary lesion stage. Many flat, red, scaly, itchy lesions called pintids occur either near the primary lesion, or scattered around the body. Pintid lesions progress through a range of color changes, from red to bluish-black. The skin of older lesions will become depigmented (lose normal color).

When to call the doctor

If the parent notices red scaly lumps, strange patterns of discoloration, or lesions on a child's skin the doctor should be contacted.

Diagnosis

Pinta can be diagnosed by dermatologists (doctors who specialize in skin diseases) and infectious disease specialists. The appearance of the lesions helps in the diagnosis. A blood sample will be taken from the patient's arm to test for antibodies to *Treponema carateum*. A scraping of a lesion will be examined under the microscope to look for *Treponema* bacteria.

Treatment

Pinta is usually treated with a single injection of benzathine penicillin G (Bicillin). This is very effective and is the treatment of choice. However if the child is allergic to penicillin, alternate **antibiotics** can be prescribed.

Prognosis

Approximately 24 hours after the treatment the skin lesions are no longer infectious and the child can no longer transmit the disease to others. Treatment results in a complete cure, but will not undo any skin damage caused by the late stages of disease. The lesions heal slowly over many months. If pinta spreads to the eyes, irreversible eyelid deformities may persist.

Prevention

Good personal hygiene may help prevent pinta. In general, children should avoid physical contact with persons who have skin lesions.

KEY TERMS

Depigmented—Characterized by a loss of normal color; discolored.

Dermatologist—A physician that specializes in diseases and disorders of the skin.

Parental concerns

Pinta only affects the skin and does not affect life expectancy, even if not treated, and it can usually be cured completely. The most significant concern is that if pinta is not diagnosed and treated promptly, the pigmentation changes in the skin may be permanent. This can cause the child to have a negative self-image and possibly lead to rejection by other children.

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National Organization for Rare Disorders, Inc. 55 Kenosia Ave, PO Box 1968, Danbury, CT 06813-1968. (203) 744-0100. <www.rarediseases.org>

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Pinworms

Definition

Pinworms, *Enterobius vermicularis*, are small, white worms that can live in the intestines, are common in young children, and are easily treated.

Description

Adult pinworms live in the large intestines. Males and females are about 5 mm and 10 mm long, respectively, with the diameter of a strand of thread. After copulation, the males die. When the female is ready to lay eggs, she crawls out of the anus, and violently expels the eggs on the skin around the anus. Some of the eggs become airborne and land elsewhere, but the majority stay on the skin of the buttocks. A single female can produce more than 10,000 eggs. After laying her eggs, the female also dies. At body temperature, the eggs develop quickly and are infective in about six hours. When ingested by another person, the eggs hatch in the small intestine. Juvenile worms grow into adult, sexually mature worms in about a month. These tiny worms are quite complex in that they have mouths, throats, gastrointestinal tracts, and a nervous system. The males and females have a complex reproductive tract and reproduce sexually. There is evidence that a protozoan parasite (*Dientamoeba fragilis*) is transmitted among humans in the eggs of pinworms. Thus, they may occur simultaneously.

Transmission

Pinworms are extremely contagious, and the eggs are infective within a few hours of being laid. They are usually spread from child to child by contaminated fingers. When children scratch their itchy bottoms, the tiny eggs get under their fingernails. As they move around the house or classroom, the eggs can be spread. Eggs can stay on a child's skin for several hours. They can survive for two weeks on clothes, bedding, and **toys**. Children who touch the contaminated materials and then place their fingers in their mouths have provided a route for the tiny eggs to enter their bodies. The eggs stay in the upper part of the intestine until they hatch, then move down the length of the intestine and out the anus to lay eggs, and the cycle continues. The entire life cycle lasts four to six weeks. Sometimes adults breathe in the eggs when the bed covers are shaken; however, this is very uncommon. Transmission easily occurs by children not washing their hands thoroughly and spreading the infection to others. It is for this reason that if one member of a **family** is infected with pinworms, the whole family is treated.

Demographics

The pinworm *Enterobius vermicularis* is one of the most common nematode parasitic infections of humans in North America and Europe. It is estimated that pinworms infect more than 400,000,000 people throughout the world or approximately 10 percent of humans. There are no differences in pinworm infections on the basis of race or socioeconomic class. Neither is pinworm infection an indication of poor hygiene. This is a very easily

transmissible infection that is quite widespread in children. Since the majority of children experience no ill effects whatsoever, extreme measures to treat pinworms are not indicated.

Causes and symptoms

Pinworm infections can be asymptomatic or result in mild gastrointestinal upsets. A common symptom associated with pinworm infections is perianal (around the anus) **itching**. Scratching of the perianal skin to relieve the itching can lead to bacterial infections that result in more itching, etc. Eventually, this cycle produces a great deal of discomfort. Children who are infected with pinworms often show symptoms that include restlessness, irritability, and insomnia. In females, the adult pinworms can enter the vagina and cause additional irritation. Since the pinworm almost always stays in the gastrointestinal tract or vagina, there is usually no systemic illness. A few children do develop intense nighttime itching of the skin around the anus. Girls who develop vaginal pinworm may experience vaginal itching or a vaginal discharge.

When to call the doctor

If the child seems restless at night and complains of itching in the morning, parents should call their health-care provider to obtain a pinworm lab kit. The kit consists of a tongue depressor with a piece of clear tape on the end. To use the kit, a parent should press the end of the tongue depressor, with the tape on it, against the child's anal skin. The tape is then placed, sticky-side down on a glass slide. The health-care provider will be able to see the eggs with a microscope, and the parent may even see them around the anus during the tape test.

Diagnosis

Stool and blood tests are not helpful in diagnosing pinworms. Seeing a worm is what determines the diagnosis. The parent must check the child's skin with a flashlight during the night and the first thing in the morning and look for white, wiggling threads. Occasionally a wiggling worm may be seen on the surface of a stool. Since pinworms are so common, children with nighttime anal itching are often treated without any lab test. The classic diagnostic tool is to apply a piece of transparent tape to the skin near the anus first thing in the morning. The health-care provider can attach it to a glass slide and then examine it under a microscope for the presence of eggs. A pinworm lab kit can usually be supplied by a provider's office if necessary.

Pinworm (*Enterobius vermicularis*) is a common parasite acquired through fecal-oral transmission. *(© J. Seibert/Custom Medical Stock Photo, Inc.)*

Treatment

Treatment is with a single dose of an anti-pinworm drug such as albendazole (Albenza) or mebendazole (Vermox). Vermox comes as a chewable tablet and most children, as well as adults, experience no side effects with the medication. Allergic reactions have been rarely reported, and very rare cases of convulsions have occurred. The medication kills the worms about 95 percent of the time, but it does not kill the eggs. Therefore, retreatment in two weeks is recommended. Girls with vaginal itching alone do not necessarily need treatment, since the problem will often disappear on its own. Many healthcare providers disagree as to whether to treat the whole family, while others believe it is essential to treat the entire household. It is possible that a girl may be an asymptomatic carrier, which results in numerous reinfections. If everyone is treated, however, this problem will be alleviated. If the child is over two years of age, Pin-X (pyrantel pamoate) is an over-the-counter alternative to Vermox that is available as a liquid.

Prognosis

Treatment is usually very successful if followed with the prevention guidelines to prevent reinfection and doing a retreatment within two weeks after the first.

Prevention

Pinworm infections and reinfections can be diminished by the following:

- Make certain children wash their hands before meals and after using the restroom.

KEY TERMS

Systemic—Relating to an entire body system or the body in general.

- Keep children's fingernails trimmed.
- Discourage nail-biting and scratching the anal area.
- Have children change into a clean pair of underwear each day.
- Have children bathe in the morning to reduce egg contamination.
- Open bedroom blinds and curtains during the day as eggs are sensitive to sunlight.
- After each treatment, change night clothes, underwear, and bedding and wash them.

Parental concerns

Since pinworms are so common and usually occur in children age 12 and under, there is no reason for concern unless the infection keeps reoccurring. In that case, meticulous cleaning and treatment with retreatment needs to be enforced.

Resources

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Pituitary dwarfism

Definition

Dwarfism is a condition in which the growth of the individual is very slow or delayed, resulting in less than normal adult stature. The word pituitary refers to the pituitary gland, which regulates the production of certain chemicals called hormones. Therefore, pituitary dwarfism is decreased bodily growth due primarily to hormonal problems. The end result is a proportionate little person, because the height and the growth of all other structures of the individual are decreased.

Description

Pituitary dwarfism is caused by problems arising from the pituitary gland. The pituitary gland, also called the hypophysis, is a gland at the base of the brain that produces many different hormones. This gland is divided into the anterior (front) and posterior (back) halves. The anterior pituitary produces six hormones: growth hormone, adrenocorticotropin (corticotropin), thyroid stimulating hormone (thyrotropin), prolactin, follicle stimulating hormone, and lutenizing hormone. The posterior pituitary gland only produces two hormones: antidiuretic hormone (vasopressin) and oxytocin.

The growth process begins in the lower part of the forebrain in a small organ called the hypothalamus. The hypothalamus releases hormones that regulate the production of other hormones. When the hypothalamus releases growth hormone-releasing hormone (GHRH), the anterior pituitary is stimulated to release growth hormone (GH). Growth hormone then acts on the liver and other tissues and stimulates them to secrete insulin-like growth factor-1 (IGF-1). IGF-1 directly promotes the development of bone and muscle, causing bones to grow in length, and muscles to increase protein synthesis (make more protein).

Since growth is a complex phenomenon, it may be slowed down or stopped by abnormalities arising at any point in the process. Thus, dwarfism can result if there is a deficiency in any of these hormones, if there is a failure in the receptor cells receiving the hormonal stimuli, or if the target cells are unable to respond.

At its most basic, pituitary dwarfism results from decreased production of hormones by the anterior pituitary. When none of the hormones of the anterior pituitary are adequately produced, this is called panhypopituitarism. A common form of pituitary dwarfism is due to deficiencies in the production of growth hormone (GH). When less GH than normal is produced during childhood, an

individual's arms, legs, and other structures continue to develop in normal proportions, but at a decreased rate.

Demographics

It is estimated that between one in 14,000 and one in 27,000 babies born each year have some form of dwarfism. In 2004, more than 20,000 children in United States were receiving supplemental GH therapy. It is estimated that about one quarter of them had organic causes of GH deficiencies. There appears to be no racial or ethnic component to pituitary dwarfism, but males seem to be afflicted more than females.

Causes and symptoms

Investigations are underway to determine the specific genetic mutations that can cause dwarfism. Pituitary dwarfism can be caused by:

- genetics
- accident-related trauma to the pituitary gland
- surgical injury of the pituitary
- central nervous system tumor
- central nervous system trauma
- central nervous system radiation
- leukemia

In most cases, the cause of dwarfism is not known (idiopathic).

A child with a growth hormone deficiency is often small with an immature face and chubby body build. The child's growth does not follow the normal growth curve patterns. In cases of tumor, most commonly craniopharyngioma (a tumor near the pituitary gland), children and adolescents may have neurological symptoms such as headaches, **vomiting**, and problems with vision. The child may also have symptoms of double vision. The symptom, however, that all children with pituitary dwarfism share is that they do not grow at the same rate as their peers.

When to call the doctor

If a child appears to be smaller than children two or more years younger than he or she is, the doctor should be consulted.

Diagnosis

Growth hormone deficiency is present at birth, but since the primary symptoms of pituitary dwarfism are height and growth at a reduced rate, the condition is not diagnosed until later in childhood. Charting a child's growth in comparison to age norms will help lead to a diagnosis. Another diagnostic technique uses an x ray of the child's hand to determine the child's bone age by comparing this to the child's actual chronological age. The bone age in affected children is usually two or more years behind the chronological age. This means that if a child is 10 years old, his or her bones will look like they are those of an eight-year-old. The levels of growth hormone and IGF-1 may also be measured with blood tests.

The doctor will do a complete examination to make sure that delayed growth is not caused by other underlying problems, such as tumor. **X rays** of the area where the pituitary gland is located, or more advanced imaging such as **magnetic resonance imaging** (MRI) or **computed tomography** (CT), may help the doctor make a diagnosis and may show whether there have been any changes to the pituitary gland itself.

Treatment

Growth hormone replacement therapy can be administered if the child is lacking growth hormone. A pediatric endocrinologist, a doctor specializing in the hormones of children, administers this type of therapy before a child's bone growth plates have fused or joined. Once the growth plates have fused, GH replacement therapy is rarely effective.

Until 1985, growth hormone was obtained from the pituitary glands of human cadavers. However, some disease complications resulted, and the United States Food and Drug Administration (FDA) banned this source of GH. In 1985, recombinant DNA techniques produced a safe and unlimited supply of GH in the lab. Now, the only growth hormone used for treatment is that which is made in a laboratory.

If growth hormone is not the only hormone deficiency, the doctor must prescribe ways to raise the levels of the other deficient hormones, if these options are available. A careful balancing of all of the hormones produced by the pituitary gland is necessary for patients with panhypopituitarism, making this form of dwarfism complex and difficult to manage.

Prognosis

The prognosis depends on the cause of the dwarfism. A panhypopituitarism dwarf does not pass through the initial onset of adult sexual development (**puberty**) and never produces enough gonadotropic (sex) hormones to develop adult sexual function. These individuals also have other medical conditions that may prove fatal.

KEY TERMS

Adrenocorticotropic hormone (ACTH)—Also called adrenocorticotropin or corticotropin, this hormone is produced by the pituitary gland to stimulate the adrenal cortex to release various corticosteroid hormones.

Antidiuretic hormone (ADH)—Also called vasopressin, a hormone that acts on the kidneys to regulate water balance.

Craniopharyngioma—A tumor near the pituitary gland in the craniopharyngeal canal that often results in intracranial pressure.

Deprivational dwarfism—A condition where emotional disturbances are associated with growth failure and abnormalities of pituitary function.

Follicle-stimulating hormone (FSH)—A pituitary hormone that in females stimulates the ovary to mature egg capsules (follicles) and in males stimulates sperm production.

Growth hormone—A hormone that eventually stimulates growth. Also called somatotropin.

Hormone—A chemical messenger secreted by a gland or organ and released into the bloodstream. It travels via the bloodstream to distant cells where it exerts an effect.

Luteinizing hormone—A hormone secreted by the pituitary gland that regulates the menstrual cycle and triggers ovulation in females. In males it stimulates the testes to produce testosterone.

Oxytocin—A hormone that stimulates the uterus to contract during child birth and the breasts to release milk.

Panhypopituitarism—Generalized decrease of all of the anterior pituitary hormones.

Prolactin—A hormone that helps the breast prepare for milk production during pregnancy.

Puberty—The point in development when the ability to reproduce begins. The gonads begin to function and secondary sexual characteristics begin to appear.

Thyroid-stimulating hormone (TSH)—A hormone produce by the pituitary gland that stimulates the thyroid gland to produce the hormones that regulate metabolism. Also called thyrotropin.

Dwarfism due only to growth hormone deficiency has a much better prognosis if treated early with replacement GH. These individuals do pass through puberty and mature sexually; however, without treatment, they remain proportionately small in stature.

The success of treatment with GH varies. An increase in height of 4–6 in. (10–15 cm) can occur in the first year of treatment. Following this first year, the response to the hormone replacement therapy is less pronounced. Even after this first year, however, the child usually continues to grow at a faster rate than he or she would without GH therapy. Long-term use is considered successful if the individual grows at least 0.75 in. (2 cm) per year more than he or she would without the hormone. However, if GH therapy is not given before the growth plates of the long bones—such as the legs and arms—fuse, the individual will not grow. Prognosis is generally better the earlier a child starts GH replacement therapy.

Improvement for individuals with other causes of dwarfism, such as a tumor, varies greatly. The prognosis usually depends on successful resolution of the underlying problem, whether there is any permanent damage, and the age of the child.

Prevention

There is no known way to prevent pituitary dwarfism, although in some cases it may be caused by traumatic injury to the pituitary gland. Engaging in safe behaviors may reduce the risk of injury-induced pituitary deficiencies.

Parental concerns

Children with pituitary dwarfism are smaller than other children, but they are just as smart and can lead long, healthy lives. It is important for parents not to expect less of their child with pituitary dwarfism simply because the child looks younger than he or she actually is. Chores and conversations should be appropriate to the actual age of the child. Children with pituitary dwarfism may face thoughtless comments from others on occasion, and the parents' reaction to such comments can strongly determine how the child feels about himself or herself.

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ORGANIZATIONS

Human Growth Foundation. 997 Glen Cove Avenue, Suite 5 Glen Head, NY 11545. (800) 451-6434 Fax: (516) 671-4055 <www.hgfound.org>

Little People of America, Inc. 5289 NE Elam Young Parkway, Suite F-700 Hillsboro, OR 97124. (888) LPA-2001 or (530) 846-1562 Fax: (503) 846-1590. <www.plaonline.org.org>

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Pityriasis rosea

Definition

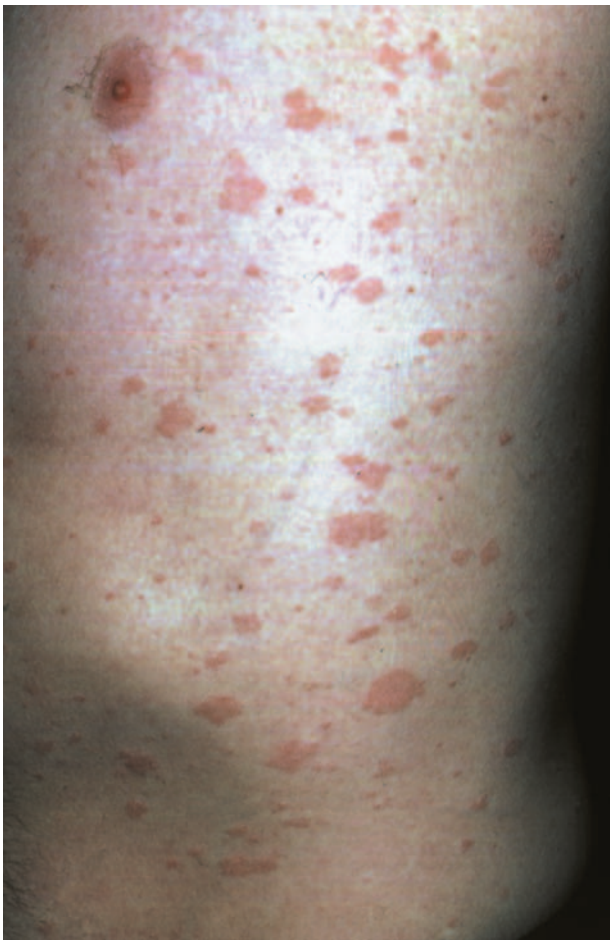
Pityriasis rosea is a mild skin disorder common among children and young adults, manifesting initially as a single round spot on the body and followed later by a rash of colored spots on the body and upper arms.

Demographics

Pityriasis rosea is most common in young adults and appears up to 50 percent more often in women. The condition occurs most often in spring and fall and can occur in epidemics within dormitories, army barracks, or other locations where young people live in close proximity to each other.

Causes and symptoms

It is unclear whether pityriasis rosea is contagious. Although some experts suspect the rash may be triggered by a virus, no infectious agent had, as of 2004, been found. Some scientists believe that the rash is an immune response to some type of infection in the body.



Torso covered with pityriasis rosea. It often appears on the torso and upper parts of the limbs of young people and may be contagious. *(Photograph by Dr. P. Marazzi. Photo Researchers, Inc.)*

Sometimes, before the symptoms appear, people experience preliminary symptoms, including **fever**, malaise, **sore throat**, or **headache**. Symptoms begin with a single, large round spot called a herald patch on the body, followed days or weeks later by slightly raised, scaly-edged round or oval pink-copper colored spots on the trunk and upper arms. The distribution of the spots, which have a wrinkled center and a sharp border, sometimes resemble a Christmas tree. They may be mild to severely itchy, and they can spread to other parts of the body.

Diagnosis

Although the diagnosis is usually obvious, if there is any confusion, other conditions (such as a fungal condition or syphilis) can be ruled out through examination of skin scrapings or blood tests.

KEY TERMS

Antihistamine—A drug used to treat allergic conditions that blocks the effects of histamine, a substance in the body that causes itching, vascular changes, and mucus secretion when released by cells.

Steroids—Hormones, including aldosterone, cortisol, and androgens, that are derived from cholesterol and that share a four-ring structural characteristic.

Treatment

The rash usually clears up on its own, over the course of about 12 weeks. During that time, external and internal medications may be given for **itching** and inflammation. Mild inflammation and itching can be relieved with antihistamine drugs or calamine lotion, zinc oxide, or other mild lubricants or anti-itching creams. Gentle, soothing strokes should be used to apply the ointments, since vigorous rubbing may cause the lesions to spread. More severe itching and inflammation is treated with topical steroids. Moderate exposure to sun or ultraviolet light may help heal the lesions, but patients should avoid being sunburned.

Soap makes the rash more uncomfortable; patients should bathe or shower with plain lukewarm water and apply a thin coating of bath oil to freshly-dried skin afterwards.

Prognosis

These spots, which may be itchy, last for three to 12 weeks. Symptoms rarely recur.

Parental concerns

After the rash has cleared up, parents often notice that areas where there were spots may appear lighter (hypopigmented) or darker (hyperpigmented) in color than the surrounding skin. Hypopigmentation can be particularly obvious in darker skinned patients. These skin changes will resolve within weeks to months after the rash has cleared.

Resources

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ORGANIZATIONS

American Academy of Dermatology. 930 N. Meacham Road, PO Box 4014, Schaumburg, IL 60168–4014. Web site: <www.aad.org>.

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PKU *see* **Phenylketonuria**

Platelet count

Definition

A platelet count is a diagnostic test that determines the number of platelets in the patient's blood. Platelets, which are also called thrombocytes, are small disk-shaped blood cells produced in the bone marrow and involved in the process of blood clotting. There are normally between 150,000–450,000 platelets in each microliter of blood. Low platelet counts or abnormally shaped platelets are associated with bleeding disorders. High platelet counts or low platelet counts sometimes indicate disorders of the bone marrow.

Purpose

The primary functions of a platelet count are to assist in the diagnosis of bleeding disorders and to monitor patients who are being treated for any disease involving bone marrow failure. Patients who have leukemia, polycythemia vera, or aplastic anemia are given periodic platelet count tests to monitor their health.

Description

Blood collection and storage

Platelet counts use a freshly collected blood specimen to which a chemical called EDTA has been added to prevent clotting before the test begins. About 5 mL of blood are drawn from a vein in the patient's inner elbow region, or other area. Blood drawn from a vein helps to produce a more accurate count than blood drawn from a fingertip. Collection of the sample takes only a few minutes. After collection, the mean platelet volume of EDTA-blood will increase over time. This increase is

caused by a change in the shape of the platelets after removal from the body. The changing volume is relatively stable for a period of one to three hours after collection. This period is the best time to count the sample when using electronic instruments, because the platelets will be within a standard size range.

Counting methods

Platelets can be observed in a direct blood smear for approximate quantity and shape. A direct smear is made by placing a drop of blood onto a microscope slide and spreading it into a thin layer. After staining to make the various blood cells easier to see and distinguish, a laboratory technician views the smear through a light microscope. Accurate assessment of the number of platelets requires other methods of counting. There are three methods used to count platelets: hemacytometer, voltage-pulse counting, and electro-optical counting.

Hemacytometer counting: The microscopic method uses a phase contrast microscope to view blood on a hemacytometer slide. A sample of the diluted blood mixture is placed in a hemacytometer, which is an instrument with a grid etched into its surface to guide the counting. For a proper count, the platelets should be evenly distributed in the hemacytometer. Errors in platelet counting are more common when blood is collected from capillaries than from veins.

Electronic counting: Electronic counting of platelets is the most common method. There are two types of electronic counting, voltage-pulse and electro-optical counting systems. In both systems, the collected blood is diluted and counted by passing the blood through an electronic counter. For these instruments to work properly, the sample must not contain other material that might mistakenly be counted as platelets. Electronic counting instruments sometimes produce artificially low platelet counts. If a platelet and another blood cell pass through the counter at the same time, the instrument will not count the larger cell, which will cause the instrument to accidentally miss the platelet. Clumps of platelets will also not be counted. In addition, if the patient has a high white blood cell count, electronic counting may yield an unusually low platelet count because white blood cells may filter out some of the platelets before the sample is counted. On the other hand, if the red blood cells in the sample have burst, their fragments will be falsely counted as platelets.

Aftercare

Because platelet counts are sometimes ordered to diagnose or monitor bleeding disorders, patients with these disorders should be cautioned to watch the puncture site for signs of additional bleeding.

KEY TERMS

Capillaries—The tiniest blood vessels with the smallest diameter. These vessels receive blood from the arterioles and deliver blood to the venules. In the lungs, capillaries are located next to the alveoli so that they can pick up oxygen from inhaled air.

EDTA—A colorless compound used to keep blood samples from clotting before tests are run.

Hemocytometer—An instrument used to count platelets or other blood cells.

Leukemia—A cancer of the blood-forming organs (bone marrow and lymph system) characterized by an abnormal increase in the number of white blood cells in the tissues. There are many types of leukemias and they are classified according to the type of white blood cell involved.

Phase contrast microscope—A light microscope in which light is focused on the sample at an angle to produce a clearer image.

Thrombocyte—Another name for platelet.

Thrombocytopenia—A persistent decrease in the number of blood platelets usually associated with hemorrhaging.

Thrombocytosis—An abnormally high platelet count. It occurs in polycythemia vera and other disorders in which the bone marrow produces too many platelets.

Risks

Risks for a platelet count test are minimal in normal individuals. Children with bleeding disorders, however, may have prolonged bleeding from the puncture wound or the formation of a bruise (hematoma) under the skin where the blood was withdrawn. Rarely an infection may occur at the needle puncture site.

Normal results

The normal range for a platelet count is 150,000–450,000 platelets per microliter of blood.

Abnormal results

An abnormally low platelet level (thrombocytopenia) is a condition that may result from increased destruction of platelets, decreased production, or increased usage of platelets. In **idiopathic thrombocytopenic purpura** (ITP), platelets are destroyed at abnormally high rates. Another cause of a low platelet count is an enlarged spleen. Hypersplenism is characterized by the collection (sequestration) of platelets in the spleen. Disseminated intravascular coagulation (DIC) is a condition in which blood clots occur within blood vessels in a number of tissues. Leukemia and aplastic anemia can result in a low platelet count because of decreased production of platelets in the bone marrow. All of these diseases produce reduced platelet counts. Abnormally high platelet levels (thrombocytosis) may indicate either a benign reaction to an infection, surgery, or certain medications; or a disease like polycythemia vera, in which the bone marrow produces too many platelets too quickly.

Parental concerns

The **pain** from the needle puncture only lasts a moment. The parent should comfort a child as needed. Older children can be prepared for the test ahead of time, and the reason why the test is being given should also be explained if the child is old enough to understand.

When to call a doctor

If the bleeding does not stop at the needle puncture site, or hours to days later, there are signs of infection (redness and swelling), then parents should contact a doctor.

Resources

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Mark A. Best

Play

Definition

Play is the work of children. It consists of those activities performed for self-amusement that have behavioral, social, and psychomotor rewards. It is child-directed, and the rewards come from within the individual child; it is enjoyable and spontaneous.

Description

Play is an important part of the childhood development. Through play children learn about shapes, colors, cause and effect, and themselves. Besides cognitive thinking, play helps the child learn social and psychomotor skills. It is a way of communicating joy, **fear**, sorrow, and **anxiety**.

In the early 2000s, children of all ages and from every socioeconomic background often prefer television, computers, and battery-operated **toys** to self-directed, imaginative, and creative play. This tendency leaves children developmentally deprived, because imaginative and fantasy play allows children to explore their world and express their innermost thoughts and feelings, hopes and fears, likes and dislikes. Through play, decisions are made without penalty or fear of failure. Play allows children to gain control of their thoughts, feelings, actions, and helps them achieve self-confidence.

Play takes different forms for different children, and its definition entails many aspects. Play is the direct opposite of work; it is frivolous. It provides freedom and invites the impulse to engage in foolishness. Yet it provides a means for ego development and a process by which social skills and physical skills develop as well.

Play with imagination and fantasy is the child's natural medium of self-expression and one that gives cues about the child's conscious and unconscious states. In play therapy, clinicians employ various techniques designed to reveal the child's psychological and social development. Clinician-directed play therapy is, therefore, not naturally self-directed play, but play designed by a professional to facilitate understanding of the child and the child's healing process.

Categories of play

Categories of play are not mutually exclusive; different forms or categories of play may overlap. Having choices is important since an action that appeals to one child may be of no interest to another, and the child's interest is likely to change throughout the play period. An understanding of play in many forms can help parents understand its importance for children of all ages. Some specific categories of play are as follows.

- Physical play. When children run, jump, and play games such as chase, hide-and-seek, and tag, they engage in physical play. This play has a social nature

because it involves other children. It also provides **exercise**, which is essential for normal development.

- Expressive play. Certain forms of play give children opportunities to express feelings by engaging with materials. Materials used in expressive play include tempera paints, fingerpaints, watercolors, crayons, colored pencils and markers, and drawing paper; clay, water, and sponges; beanbags, pounding benches, punching bags, and rhythm instruments; and shaving cream, pudding, and gelatin. Parents can take an active role in expressive play by using the materials alongside the child.
- Manipulative play. Children control or master their environment through manipulative play. They manipulate the environment and other people as much as possible. Manipulative play starts in infancy. Infants play with their parents; for example, they drop a toy, wait for the parent to pick it up, clean it, and return it, and then they drop it again. This interaction brings the infant and parent together in a game. Children move objects such as puzzle pieces and gadgets to better understand how they work.
- Symbolic play. Certain games can symbolically express a child's problems. Because there are no rules in symbolic play, the child can use this play to reinforce, learn about, and imaginatively alter painful experiences. The child who is in an abusive **family** may pretend to be a mother who loves and cuddles her child rather than one who verbally or physically abuses her child. Or in play this same child might act out abusive experience by hitting or screaming at a doll that symbolizes the child. Parents can be surprised by their child's perception of family issues. Children mimic their parents in certain play; in other games they may pretend they are the heroes they read about in books or see on television. At certain developmental stages children believe they can fly or disappear. Symbolic play may be used by children to cope with fear of separation when they go to school or to the hospital.
- Dramatic play. Children act out situations they suspect may happen to them, that they are fearful will happen, or that they have witnessed. Dramatic play can be either spontaneous or guided and may be therapeutic for children in the hospital.
- Familiarization play. Children handle materials and explore experiences in reassuring, enjoyable ways. Familiarization prepares children for potentially fearful and painful experiences, such as surgery or parental separation.
- Games. Some video and card games are played by one child alone. Games with rules are rarely played by children younger than four years of age. Board games, card games, and **sports** are enjoyed typically by school-age children. In these games children learn to play by the rules and to take turns. Older children enjoy games with specific rules; however, younger children tend to like games that allow them to change the rules.
- Surrogate play. For children who are too ill or incapacitated to play, another child or a parent may serve as surrogate. Watching the surrogate who plays on behalf of the sick child is stimulating to the sick child. When parents engage in expressive art by painting or redecorating a room while the physically challenged child watches, they stimulate the child.

Functions of play

Play reinforces the child's growth and development. Some of the more common functions of play are to facilitate physical, emotional, cognitive, social, and **moral development**.

PHYSICAL DEVELOPMENT Play aids in developing both fine and **gross motor skills**. Children repeat certain body movements purely for pleasure, and these movements develop body control. For example, an infant will first hit at a toy, then will try to grasp it, and eventually will be able to pick it up. Next, the infant will shake the rattle or perhaps bring it to the mouth. In these ways, the infant moves from simple to more complex gestures.

EMOTIONAL DEVELOPMENT Children who are anxious may be helped by role playing. Role playing is a way of coping with emotional conflict. Children may escape through play into a fantasy world in order to make sense out of the real one. Also, a child's self-awareness deepens as he explores an event through role-playing or symbolic play.

When a parent or sibling plays a board game with a child, shares a bike ride, plays baseball, or reads a story, the child learns self-importance. The child's **self-esteem** gets a boost. Parents send positive messages to their child when they communicate pleasure in providing him or her with daily care. From these early interactions, children develop a vision of the world and gain a sense of their place in it.

COGNITIVE DEVELOPMENT Children gain knowledge through their play. They exercise their abilities to think, remember, and solve problems. They develop cognitively as they have a chance to test their beliefs about the world.

Children increase their problem-solving abilities through games and puzzles. Children involved in make-believe play can stimulate several types of learning. Language is strengthened as the children model others and organize their thoughts to communicate. Children play-

ing house create elaborate narratives concerning their roles and the nature of daily living.

Children also increase their understanding of size, shape, and texture through play. They begin to understand relationships as they try to put a square object in a round opening or a large object in a small space. Books, videos, and educational toys that show pictures and matching words also increase a child's vocabulary while increasing the child's concept of the world.

SOCIAL DEVELOPMENT A newborn cannot distinguish itself from others and is completely self-absorbed. As the infant begins to play with others and with objects, a realization of self as separate from others begins to develop. The infant begins to experience joy from contact with others and engages in behavior that involves others. The infant discovers that when he coos or laughs, mother coos back. The child soon expects this response and repeats it for fun, playing with his mother.

As children grow, they enjoy playful interaction with other children. Children learn about boundaries, taking turns, teamwork, and competition. Children also learn to negotiate with different personalities and the feelings associated with winning and losing. They learn to share, wait, and be patient.

MORAL DEVELOPMENT When children engage in play with their peers and families, they begin to learn some behaviors are acceptable while others are unacceptable. Parents start these lessons early in the child's life by teaching the child to control **aggressive behavior**. Parents can develop morals while reading to children by stressing the moral implications in stories. Children can identify with the moral fictional characters without assuming their roles. With peers they quickly learn that taking turns is rewarding and cheating is not. Group play helps the child appreciate teamwork and share and respect others' feelings. The child learns how to be kind and charitable to others.

Age-related play

As children develop, their play evolves, too. Certain types of play are associated with, but not restricted to, specific age groups.

- Solitary play is independent. The child plays alone with toys that are different from those chosen by other children in the area. Solitary play begins in infancy and is common in toddlers because of their limited social, cognitive, and physical skills. However, it is important for all age groups to have some time to play by themselves.
- Parallel play is usually associated with toddlers, although it happens in any age group. Children play side by side with similar toys, but there is a lack of group involvement.
- Associative play involves a group of children who have similar goals. Children in associate play do not set rules, and although they all want to be playing with the same types of toys and may even trade toys, there is no formal organization. Associative play begins during toddlerhood and extends though **preschool** age.
- Cooperative play begins in the late preschool period. The play is organized by group goals. There is at least one leader, and children are definitely in or out of the group.
- Onlooker play is present when the child watches others playing. Although the child may ask questions of the players, there is no effort to join the play. This type of play usually starts during toddler years but can take place at any age.

Common problems

Promoting play for a sick child is a challenge when the child cannot voluntarily engage in play. Parents need to realize the importance of play to the well being of a sick child. Children can bring favorite books, games, and stuffed animals to the hospital. In hospitals young children need toys that they can manipulate independently, so that parents are free sometimes to focus on medical issues and the healthcare team.

Play activities vary depending on cultural and socioeconomic circumstances. When children do not speak the group's language, games such as stacking blocks or building with tinker toys are appealing. Playing tapes of well-loved children's songs can be effective too. The child does not need to be able to understand the words to enjoy the music or clap with the rhythm.

Assessing child health through play

Acutely ill children do not have the strength, the attention span, or the interest in play. They may enjoy being read to and the comfort of holding a favorite stuffed animal. Once the acute phase of an illness is over, the child's interest in playing returns. Spontaneous interest in play is a good index of health. The toys selected for play are good indicators of the child's recovery progress.

Play in a medical setting

When a child goes to see the doctor, the waiting room is likely to have other children in it. The arriving child may hear other children cry as they leave the examining room. The child may dread the examination.

Parents should pack a favorite toy or book with which to distract the child. Having a parent sit with them is comforting, and they may venture a few feet away to examine toys in the toy box. Older children who go with the parent and the sick sibling to see the doctor should have toys and games for their entertainment, too, so the parent can focus on the sick child.

Hospitalized children can release fear, anger, or tension through effective play. Children in the hospital for a week or longer may enjoy playing school or socializing in the playroom with other children of their age. However, physical play for sick children must be supervised by a parent or healthcare provider.

Therapeutic play

When a child is ill or traumatized the care plan may include therapeutic play. Unlike normal play in design and intent, therapeutic play is guided by the health professional to meet the physical and psychological needs of the child. Because play is the language of children, children who have difficulty putting their thoughts in words can often speak clearly through play therapy. There are three divisions of therapeutic play, including:

- Energy release. Children release anxiety by pounding, hitting, running, punching, or shouting. Toddlers pound pegs with a plastic hammer or pretend to cut wood with a toy saw. An anxious preschooler pounds a ball of modeling clay flat; a relaxed child may build the clay into shapes. Balloons tied over the bed of a school-age child or adolescent can be punched.
- Dramatic play. Children act out or dramatize real-life situations. They act out anxiety and emotional stress from abuse, neglect, **abandonment**, and various painful physical experiences. Imaginative preschool children enjoy dramatic play. An abused or wounded child might not communicate the experience verbally but may be able to use an anatomically correct doll to show what happened. Therapeutic play can teach children about medical procedures or help them work through their feelings about what has happened to them in the medical setting.
- Creative play. Some children are too angry or fearful to act out their feelings through dramatic play. However, they may be able to draw a picture that expresses their emotions or communicates what they know. To encourage this expression children can be given blank paper and crayons or markers and asked to draw a picture about how they feel. Some children are so concerned about a particular body part that instead of drawing a self portrait, they will draw only the body part that worries them.

Many children draw pictures that reflect punitive images to explain unhappy experiences. They need reassurance that they are not being punished. Health-care providers need to make sure that these children are not being abused. Other children may draw pictures that are symbolic of death (an airplane crashing, boats sinking, burning buildings, or children in graves). These children need assurances that they are not going to die. Some **drawings** express the child's fear of abandonment and loss of independence. Pictures may suggest the parent cannot find the little child who is in the hospital. The child needs to be reassured that their parents know where they are. They need to know when the parents will visit and the parents should appear when they say they will be there.

Older school-age children and adolescents may not be interested in drawing, but they can make a list of experiences they like and dislike.

Parental concerns

Parents express interest in age-related play that prepares children for group exercises in preschool. They want to know the right kind of play for an only child or sick child who may not be able to play with other children in their age group. The following age-related play and toys serve as a guide to parents with these concerns.

- Infant. The infant enjoys watching other members of the family; the infant enjoys rocking, strolling, time spent in a swing, supervised time on a blanket on the floor, **crawling**, walking with help, and being sung and read to. Play is self-absorbed; it is difficult, if not impossible to direct play. Infants are engaged in the vigorous process of self-discovery, learning their world by looking, listening, chewing, smelling, and grasping. Most of their learning comes through play. They need safe toys that appeal to all of their senses and stimulate their interest and curiosity. Infants need toys and play that include oral movements. They like peek-a-boo; playing with the parent's fingers, hair, face, and the infant's own body parts; playing in water. Soft stuffed animals, crib mobiles, squeeze toys rattles, busy boxes, mirrors, and musical toys. Parents can give them water toys for the bath, safe kitchen utensils, and push toys (after they begin to walk), and large print books.
- Toddler. Toddlers fill and empty containers and begin dramatic play. As they increase their motor skills, they enjoy feeling different textures, exploring the home environment, and mimicking others. They like to be read to and to look at books and television. Toddlers enjoy manipulating small objects such as toy people, cars, and animals. Favorite toys are mechanical; objects of different textures such as clay, sand, finger

paints, and bubbles; push-pull toys; large balls; sand and water play; blocks; painting or coloring with large crayons; nesting toys; large puzzles; and trucks and dolls. Toddlers explore their bodies and those of others. Therapeutic play can begin at this age.

- Preschooler. Dramatic play is prominent. This age group likes to run, jump, hop, and in general increase motor skills. The children like to build and create whether it is sand castles or mud pies. Play is simple and imaginative. Simple collections begin. Preschoolers enjoy riding toys, building materials such as sand and blocks, dolls, drawing materials, cars, puzzles, books, appropriate television and videos, nonsense rhymes, and singing games. Preschoolers love pretending to be something or somebody and playing dress up They enjoy finger paints, clay, cutting, pasting, and simple board and card games.
- School-age child. Play becomes organized and has a direction. The early school-age child continues dramatic play with increased **creativity** but loses some spontaneity. The child gains awareness of rules when playing games and begins to compete in sports. Children in this age group enjoy collections (comic books, baseball cards, and stamps), dolls, pets, guessing games, board games, riddles, physical games, competitive play, reading, bike riding, hobbies, sewing, listening to the radio, television, and videos, and cooking.
- Adolescent. Athletic sports are the most common form of play. Strict rules are in place, and competition is important. Adolescents also enjoy movies; telephone conversations and parties; listening to music; and experimenting with makeup, hairstyles, and fashion. They also begin developing an interest in peers of the opposite sex.

Play for the sick child

Children who are confined to a bed need to have play periods built into their day. The length of play and the toys will depend the individual child's age and physical and emotional states. Short-term school projects appeal to school-age children because these activities help the children feel industrious and think about their future wellness. Parents can help children with their baths; encourage them to drink enough fluids; and prompt them to do deep breathing and muscle strengthening exercises.

Safety issues

Toys and games should be screened for **safety**, especially those used by a sick child. The toys should be washable with no sharp edges and no small parts that could be swallowed or aspirated. Cylinder-shaped toys of 1-inch (2.5-cm) diameter (the size of a regular hot dog) are the most dangerous size because they can occlude the trachea (windpipe) if they are aspirated. As a rule, if a toy can fit through the center of a toilet tissue tube, it is too small.

Toddler playing by himself with toys. *(© Villareal/Photo Researchers, Inc.)*

Parents should be certain that toys do not lead children into danger. Tossing a ball to a toddler on bed rest may be safe, but if a child in a cast leans to catch the ball, he may fall. Chasing a ball may lead to falls and collisions. If children are bored with a toy because it is not stimulating enough or they have played with it too long, they may begin to use the toy in an unsafe way. For example, the child may throw blocks across the room for fun instead of stacking them.

Indoor toys

For home care of the sick child, parents may need to buy new toys suitable for indoor use. The ill child may need soft toys for bed play and sit-down toys such as magic markers, puzzles, books, or board games, for quiet out-of-bed play.

KEY TERMS

Accommodation—The process in which a schema changes to accomodate new knowledge.

Assimilation—The process of taking in new information by incorporating it into an existing schema.

Associative play—Preschoolers play together in a similar activity with little organization or responsibility.

Cooperative play—School-age children play in an organized structure or compete for goal or outcome.

Experimental play therapy—Play therapy based on the belief that a child has the ability to solve his or her own problems within the context of a warm and caring therapeutic environment.

Observation—Infants and children watch an object, although not actively engaged in it, as in watching a mobile.

Parallel play—Toddlers play side by side but seldom try to interact with each other, playing separately with a similar toy.

Play therapy or therapeutic play—A type of psychotherapy for young children involving the use of toys and games to build a therapeutic relationship and encourage the child's self-expression.

Play-based assessment—A form of developmental assessment that involves observation of how a child plays alone, with peers, or with parents or other familiar caregivers, in free play or in special games.

When to call the doctor

Parents and teachers who spend time observing and understanding childhood behaviors may want to report to the child's therapist what they see the child do.

Skin care is essential for children who are bedridden or in a cast or restraints. Children lose interest in playing if they are uncomfortable or in **pain**. Parents should look for pressure over the buttocks, elbows, heels, and other parts of the child's body. The skin should be inspected often and massaged with a moisturizing lotion to increase circulation. Redness, irritation, and sores should be reported immediately to the healthcare provider.

When children are ill, the rate of bladder and bowel elimination may slow down because of reduced physical action. School-age children and adolescents may hesitate to drink or eat a normal diet because toileting is uncomfortable or performed without privacy. Parents may need to seek medical advice about digestive and elimination aids and about adjusting the child's diet and fluid intake to promote normal elimination.

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Aliene S. Linwood, RN, DPA, FACHE

PMS *see* **Premenstrual syndrome**

Pneumonia

Definition

Pneumonia is an infection of the lungs that can be caused by nearly any class of organism known to cause human infections, including bacteria, viruses, fungi, and parasites. It results in an inflammatory response within the small air spaces of the lung (alveoli).

Description

Pneumonia can develop gradually in children after exposure to the causative organism, or it can develop quickly after another illness, reducing the lungs' ability

to receive and distribute oxygen. It can be mild and easily cured with **antibiotics** and rest, or it can be severe and require **hospitalization**. The onset, duration, and severity of pneumonia depend upon the type of infective organism invading the body and the response of the child's immune system in fighting the infection. Respiratory distress represents 20 percent of all admissions of children to hospitals, and pneumonia is the underlying cause of most of these admissions.

To understand pneumonia, it is important to understand the basic anatomic features of the respiratory system. The human respiratory system begins at the nose and mouth, where air is breathed in (inspired) and out (expired). The nasopharynx is the air tube extending from the nose that directs air into the lungs. Air breathed in through the mouth travels through the oropharynx, which also carries swallowed food, water, and salivary secretions through the food tube (esophagus) and then into the stomach. The nasopharynx and oropharynx merge into the larynx, which is protected by a trap door called the epiglottis. The epiglottis normally prevents substances that have been swallowed, as well as substances that have been regurgitated (vomited), from heading down through the larynx into the lungs.

The larynx flows into the trachea, which is the broadest part of the respiratory tract. The trachea divides into the right and left bronchi, each branching off into multiple smaller bronchi that course throughout the lung tissue. Each bronchus divides into tubes of smaller and smaller diameter, finally ending in the terminal bronchioles. The alveoli, in which oxygen and carbon dioxide are exchanged, are clustered at the ends of the bronchioles. Lung stroma, the tissue of the lung, serves a supportive role for the bronchi, bronchioles, and alveoli.

The main function of the respiratory system is to help distribute oxygen, the most important energy source for the body's cells. Oxygen enters the body as inspired air and travels through the respiratory system to the alveoli. The oxygen is then picked up by hemoglobin, the oxygen-carrying protein in red blood cells, and delivered throughout the body through the circulatory system. Oxygen in the inspired air is exchanged within the alveoli of the lungs for carbon dioxide, a waste product of human metabolism. Carbon dioxide leaves the lungs during expiration.

The healthy human lung is sterile, with no normally resident bacteria or viruses, unlike the upper respiratory system and parts of the gastrointestinal system, where bacteria dwell even in a healthy state. Multiple safeguards along the path of the respiratory system are designed to keep invading organisms from causing infection. The first line of defense includes tiny hairs in the nostrils that filter out large particles. The epiglottis helps prevent food and other swallowed substances from entering the larynx and the trachea. Sneezing and coughing, both provoked by the presence of irritants within the respiratory system, help to clear such irritants from the respiratory tract. Mucus produced through the respiratory system also serves to trap dust and infectious organisms. Tiny hair like projections (cilia) from cells lining the respiratory tract beat constantly to move debris trapped by mucus upwards and out of the respiratory tract. This mechanism of protection is referred to as the mucociliary escalator. Finally, cells lining the respiratory tract produce several types of immune substances that protect against various organisms. Other cells (macrophages) along the respiratory tract surround and kill invading organisms.

Organisms that cause pneumonia, then, are usually prevented from entering the lungs by virtue of these host defenses. However, when a large number of organisms are encountered at once or when the immune system is weakened, the usual defenses may be overwhelmed and infection may occur. This can happen either by inhaling contaminated air droplets or by the aspiration of organisms inhabiting the upper airways. Aspiration pneumonia is a type of pneumonia in which something is aspirated from the upper airway into the lungs. This can be food from the mouth, a foreign object or substance that has entered the mouth, or regurgitated stomach contents (vomitus) aspirated into the lungs as it travels to the mouth.

The invading organism causing pneumonia provokes an immune response in the lungs that causes inflammation of the lung tissue (pneumonitis), a condition that actually makes the lung environment more ideal for infection. Small blood vessels in the lungs (capillaries) begin to empty protein-rich fluid into the alveoli, a condition that results in a less functional area for oxygen-carbon dioxide exchange. The individual becomes relatively oxygen deprived, while retaining potentially damaging carbon dioxide. This results in rapid respiration (tachypnea or faster and faster breathing) in an effort to bring in more oxygen and blow off more carbon dioxide.

Consolidation, a feature of bacterial pneumonia, occurs when the alveoli, which are normally hollow air spaces within the lung, instead become solid due to quantities of fluid and debris. Viral pneumonias and mycoplasma pneumonias do not result in consolidation. These types of pneumonia primarily infect the walls of the alveoli and the stroma of the lung. Bacterial and viral pneumonia occur mostly in winter months, while mycoplasma pneumonia is more common in summer and fall.

Bacterial pneumonia develops after the child inhales or aspirates pathogens. Viral pneumonia stems primarily from inhaling infected droplets from the upper airway into the lungs. In neonates, pneumonia may result from colonization of the infant's nasopharynx by organisms that were in the birth canal at the time of delivery.

In addition to exposure to sufficient quantities of causative organisms, certain other conditions can increase the risk of pneumonia. These include the following:

- abnormal anatomical structure, particularly of the chest or lungs
- cigarette smoke, inhaled directly by a smoker or second-hand
- immune system deficiencies (**common variable immunodeficiency**, **immunoglobulin deficiency syndromes**, **HIV infection**, and others)
- swallowing difficulties as a result of **stroke** or seizures
- intoxication by alcohol and drugs that may interfere with normal **cough** reflex and decrease the chance of clearing unwanted debris from the respiratory tract
- viruses that may interfere with ciliary function, allowing themselves or other invading microorganisms such as bacteria access to the lower respiratory tract
- various chronic conditions such as **asthma**, **cystic fibrosis**, diabetes, emphysema, and neuromuscular diseases that may interfere with the seal of the epiglottis
- advanced age and associated immune system weakness
- esophageal disorders that may result in stomach contents passing upwards
- genetic factors and associated changes in DNA
- post-operative complications including the use of certain therapeutic drugs, suppressed cough reflex, breathing difficulties, and **pain** at the surgical site that affects breathing
- malnutrition
- radiation treatment for breast **cancer**, which may weaken lung tissue

The epidemic of immmunodeficiency virus (HIV), the virus that causes acquired **immunodeficiency** syndrome (**AIDS**), has resulted in a huge increase in the incidence of pneumonia. Because AIDS results in immune system suppression, individuals with AIDS are highly susceptible to all kinds of pneumonia, including some previously rare parasitic types that would not cause illness in someone with a normal immune system.

Pneumonia is also the most common fatal infection acquired by already hospitalized patients. Even in nonfatal cases, pneumonia is a significant economic burden on the healthcare system. One study estimates that U.S. workers who develop pneumonia cost employers five times as much in health care as the average worker.

Transmission

Pneumonia is not usually passed from one person to another. The bacterial and viral organisms that cause pneumonia, however, can be transmitted through airborne or direct contact.

Demographics

Every year in the United States, two million people of all ages develop pneumonia, including 4 percent of all the children in the country. It is the sixth most common disease leading to death and the fourth leading cause of death in the elderly; 40,000 to 70,000 people die from pneumonia each year. The incidence of pneumonia in children younger than one year of age is 35 to 40 per 1,000; 30 to 35 per 1,000 children ages two to four; and 15 per 1,000 children between ages five and nine. Fewer than 10 children in 1,000 over age nine are reported to develop pneumonia. The Centers for Disease Control and Prevention (CDC) reports that the number of deaths from pneumonia in the United States declined between 2001 and 2004.

Causes and symptoms

The list of organisms that can cause pneumonia is lengthy and includes nearly every class of infecting organism: viruses, bacteria, bacteria-like organisms, fungi, and parasites (including certain worms). Different organisms are more frequently encountered by different age groups, and other individual characteristics may increase risk for infection by particular types of organisms:

- Viruses cause the majority of pneumonias in young children, especially respiratory syncytial virus, parainfluenza and **influenza** viruses, and adenovirus.
- Adults are more frequently infected with bacteria such as *Streptococcus pneumoniae*, *Haemophilus influenzae*, and *Staphylococcus aureus*.
- Pneumonia in older children and young adults is often caused by the bacteria-like *Mycoplasma pneumoniae*, the cause of pneumonia that is often called "walking" pneumonia.
- *Pneumocystis carinii* causes pneumonia in immunosuppressed individuals such as patients being treated with **chemotherapy** or people with AIDS. Classically

considered a parasite, it appears to be more related to fungi.

- *Chlamydia psittaci* can be infective in some individuals, such as poultry farm workers, who have direct contact with bird droppings.

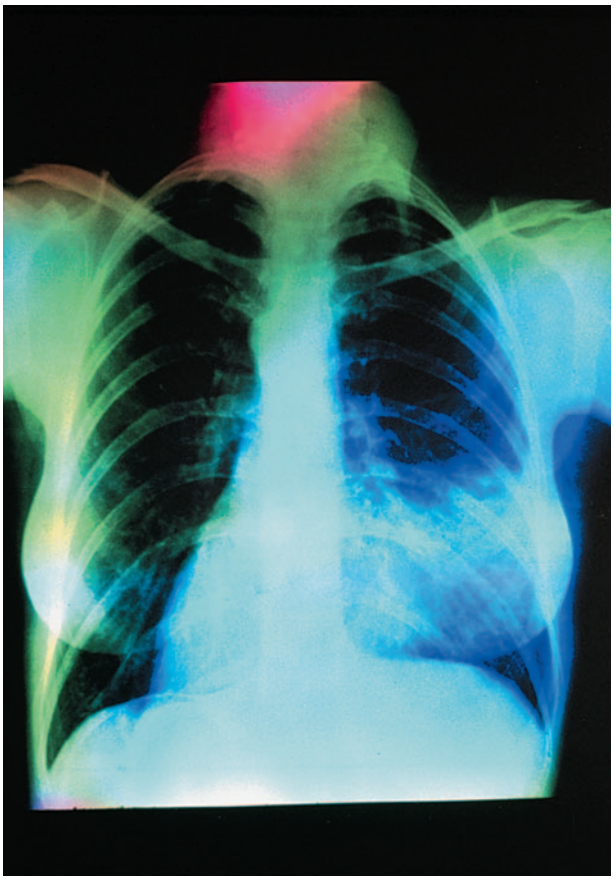
Pneumonia is suspected in a child who has symptoms such as **fever**, cough, chest pain, difficulty breathing (shortness of breath or dyspnea), and an increased number of breaths per minute (respiration). Fever with a shaking chill is even more suspicious. Mucus production is typically increased and leaky capillaries in the lungs may tinge the mucus with blood. The alveoli fill further with fluid and debris from the large number of white blood cells being produced to fight the infection. Children may cough up clumps of sputum or phlegm, secretions produced in the alveoli during the infection or inflammatory condition. These clumps may appear streaked with pus or blood. In severe pneumonia, mucus plugs and the accumulation of fluid together decrease the efficiency of gas exchange in the lung, resulting in signs of oxygen deprivation. Reduced oxygen levels in the blood may produce a blue appearance of the nail beds or lips (cyanosis).

Diagnosis

Diagnosis is based on the parents' report of the onset of illness and the symptoms that have developed, combined with examination of the chest. Physical examination may indicate labored breathing. Listening with a stethoscope may reveal abnormal crackling sounds (rales), and tapping on the back, which normally yields a resonant sound due to air filling the alveoli, may yield a dull thump if the alveoli are filled with fluid and debris.

Laboratory diagnostic tests may include staining sputum samples on a glass slide and looking at the stained specimen under a microscope to determine if white cells, red cells, or bacteria are present. Identification of the specific type of bacteria may require culturing the sputum, a microbiological technique that identifies disease-causing bacterial organisms in infected material. A small sample of sputum will be streaked on a special plate filled with medium that allows the specific organism to be grown in the laboratory under certain conditions. The bacteria can then be identified and, by performing antibiotic sensitivity tests on the bacteria, appropriate treatment can usually be prescribed. In addition, oxygen and carbon dioxide levels may be measured (blood gases) and the exchange evaluated (oximetry).

If pneumonia is present, a rapid rate of respiration may be noted; tachypnea is defined as a respiratory rate over 50 respirations per minute in infants younger than one year. Older children will have tachypnea if the respiratory rate is greater than 40 per minute.



A chest x ray showing pneumonia in the lower lobe of a patient's right lung. The alveoli (air sacs) of the lung become blocked with pus, which forces air out and causes the lung to become solidified. *(National Audubon Society Collection/Photo Researchers, Inc.)*

X-ray examination of the chest may reveal certain abnormal changes associated with pneumonia. Localized shadows obscuring areas of the lung may indicate a bacterial pneumonia, while streaky or patchy changes in the x-ray film may indicate viral or mycoplasma pneumonia. These changes on x ray, however, are known to lag in time behind actual symptoms.

Treatment

Prior to the discovery of penicillin and other antibiotics, bacterial pneumonia was almost always fatal. In the early 2000s, especially given early in the course of the disease, antibiotics are very effective against bacterial causes of pneumonia. Penicillin was, as of 2004, still the first choice for treating children with pneumonia unless the child is known to be penicillin-resistant. Oral

KEY TERMS

Alveoli—The tiny air sacs clustered at the ends of the bronchioles in the lungs in which oxygen-carbon dioxide exchange takes place.

Aspiration—The process of removing fluids or gases from the body by suction. Also refers to the inhalation of food or liquids into the lungs.

Cilia—Tiny hairlike projections on certain cells within the body. Cilia produce lashing or whipping movements to direct or cause motion of substances or fluids within the body. Within the respiratory tract, the cilia act to move mucus along, in an effort to continually flush out and clean the respiratory tract.

Consolidation—A condition in which lung tissue becomes firm and solid rather than elastic and air-filled, arising because of accumulated fluids and tissue debris.

Culture—A test in which a sample of body fluid is placed on materials specially formulated to grow microorganisms. A culture is used to learn what type of bacterium is causing infection.

Cyanosis—A bluish tinge to the skin that can occur when the blood oxygen level drops too low.

Pneumocystis carinii—An organism that causes pneumonia in immunodeficient individuals, such as people with AIDS.

Respiratory system—The organs that are involved in breathing: the nose, the throat, the larynx, the trachea, the bronchi and the lungs. Also called the respiratory tract.

Sputum—The substance that is coughed up from the lungs and spit out through the mouth. It is usually a mixture of saliva and mucus, but may contain blood or pus in patients with lung abscess or other diseases of the lungs.

Stroma—A term used to describe the supportive tissue surrounding a particular structure. An example is the tissue that surrounds and supports the actually functional lung tissue.

Tachypnea—Rapid breathing.

amoxicillin or cephalosporins are often administered first in treating milder cases of pneumococcal pneumonia in children younger than age five, though they are not used in newborns. Erythromycin and tetracycline are broad-spectrum antibiotics that are known to improve recovery time for symptoms of mycoplasma pneumonia. They do not, however, eradicate the organisms. If the results of culture and sensitivity positively identify the causative bacteria, an antibiotic is prescribed for that demonstrated sensitivity. Viruses do not usually respond to antibiotics. Amantadine and acyclovir may be helpful against certain viral pneumonias.

Linezolid (Zyvox), the first of a new line of antibiotics known as oxazolidinones, is used to treat penicillin-resistant organisms that cause pneumonia. Another newer drug known as ertapenem (Invanz) is reported to be effective in treating bacterial pneumonia.

The child is also be given fluids and possibly drug therapy to thin mucus secretions (mucolytic agents) or medication to open the airways of the lung (brochodilators). **Cough suppressants** may be given as well as pain medication and fever-reducing medication. Hospitalized children may receive extra oxygen, respiratory therapy, and intravenous antibiotics and fluids.

Alternative treatment

Vitamin C is known to improve immune response and to help reduce inflammation. Grape seed extract enhances immune system functioning and helps protect lung tissue. These are adjunctive measures that do not destroy the causative organism as antibiotics do. Although garlic and certain herbs such as yerba mansa may have antibiotic properties, they cannot replace specific antibiotics used to treat pneumonia.

Prognosis

Prognosis varies according to the type of organism causing the infection, the status of the immune system, and the overall health of the affected child. Generally, there are lower mortality rates from pneumonia in the United States than elsewhere in the world. *Streptococcus pneumoniae*, the most common organism causing pneumonia, has a significantly lower death rate of about 5 percent. More complications occur in the very young or very old with multiple areas of the lung infected simultaneously. The presence of chronic illnesses such as diabetes, cirrhosis, and congestive heart failure may increase the chance of complications. Individuals with immunodeficiency disorders, various types of cancer, or AIDS are also more prone to complications. In children, cystic fibrosis, aspiration problems, immunodeficiencies, and congenital or acquired lung malformation may increase the risk of pneumonia from *S. pneumoniae*.

Recovery following pneumonia with *Mycoplasma pneumoniae* is nearly 100 percent. However, in the very young or very old or immunodeficient, *Staphylococcus aureus* has a death rate of 30 to 40 percent. Similarly,

infections with a number of gram negative bacteria (such as those in the gastrointestinal tract that can cause infection following aspiration) have a death rate of 25 to 50 percent.

Prevention

Because many bacterial pneumonias occur in people who were first infected with the influenza virus (the flu), yearly flu vaccinations can decrease the risk of pneumonia for the elderly and children or adults with chronic diseases such as asthma, cystic fibrosis, other lung or heart diseases, **sickle cell anemia**, diabetes, kidney disease, and cancer.

A specific vaccine against *Streptococcus pneumoniae* can be protective for people with chronic illnesses.

Immunodeficient individuals are at higher risk for infection with *Pneumocystis carinii* and are frequently put on a regular preventive drug regimen of trimethoprim sulfa and/or inhaled pentamidine to avoid pneumocystis pneumonia.

Parental concerns

Pneumonia in a child can produce severe symptoms that can be frightening to both the child and parents, particularly when breathing is compromised or cyanosis is noted. When symptoms seem to suggest pneumonia, immediate attention allows early treatment so that breathing difficulties can be corrected quickly and drug therapy begun in order to destroy the causative organism. Parents can try to reassure young children and keep them as calm as possible, knowing that **anxiety** also increases breathing difficulties.

See also Common variable immunodeficiency.

Resources

BOOKS

"Pneumonia." Section 6, Chapter 73 in *The Merck Manual of Diagnosis and Therapy*, edited by Mark H. Beers and Robert Berkow. Whitehouse Station, NJ: Merck Research Laboratories, 2003.

ORGANIZATIONS

American Lung Association. 1740 Broadway, New York, NY 10019. Web site: <www.lungusa.org>.

Centers for Disease Control and Prevention. 1600 Clifton Rd., NE, Atlanta, GA 30333. Web site: <www.cdc.gov>.

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Poison ivy, oak, and sumac

Definition

Poison ivy, oak, and sumac are allergic skin **rashes** (or *Rhus dermatitis*) caused by the plants of the same name. All three plants secrete a potent, irritating oil known as urushiol that causes blistering and intense **itching** once it penetrates the skin.

Description

The allergic rash of poison ivy, oak, and sumac is characterized by red, weeping blisters and severe itching. The rash usually appears within one to two days of initial contact with the plant oil, although it may take longer to appear in areas where the skin is thicker, and lasts from one to three weeks (longer in severe cases). It starts as itchy, inflamed red patches or streaks, and as the oil penetrates into the skin, blisters and small papules form.

Poison plant rash cannot be spread from person to person by contact with the rash itself or fluid from the blisters, and scratching does not spread the rash (although it can cause scarring and potential infection). Only urushiol oil can cause the rash.

Transmission

Urushiol oil or resin is found in the leaves, roots, and woody parts (i.e., vines and stems) of the poison ivy, oak, and sumac plants. It is a clear substance that is released by the plant when it is cut or bruised. Leaves are bruised easily, especially in the spring, so even a gentle brush against a plant can cause the urushiol to seep out and onto the skin.

Urushiol can remain active for years. For that reason, even dead poison ivy, oak, or sumac plants must be handled with care. Plants should never be burned or shredded, as airborne particles can spread the oil to sensitive areas like the face and eyes and may potentially cause damage to lungs.

The three main sources of poison plant rash—poison ivy, poison oak, and poison sumac—are members of the *Anacardiaceae*, or cashew, family. While they are usually concentrated most heavily in a few specific regions of the country, all three have been found in locations throughout the United States. Identifying the plant, particularly if people live in a wooded area or have a lot of vegetation in their yards or neighborhood, is essential to preventing the rash.

Poison ivy, known as *Rhus radicans* or *Toxicondendron radicans*, is found throughout the United States. The plant grows in vines (typical in the Midwest, East coast, and South) or small bushes (in the North, West, and Great Lakes region), and has clusters of three leaves. (Hence the popular saying: "Leaves of three, let them be.") The leaves are red in the spring; green throughout the summer; and yellow, red, and orange in the fall when they also produce white berries.

Poison oak is a small shrub. The plant, which is also known as *Rhus diversiloba* or *Toxicondendron diversilobum*, is found in the western United States. Like poison ivy, poison oak leaves change color with the seasons. The plant also produces white berries in the fall.

The small, woody shrubs that are poison sumac are most common in the Eastern United States. Also known as *Rhus vernix* or *Toxicondendron vernix*, poison sumac differs in appearance from the three-leaf clusters of poison ivy and oak. It is feather-like in appearance, with two rows of leaves arranged on either side of a long stem, topped off by a long leaf at the tip. It can be distinguished from regular, non-poisonous sumac by its berries, which are green to white as opposed to the bright red berries of regular sumac.

Demographics

According to the American Academy of Dermatology, an estimated 85 percent of the population is allergic to the urushiol oil found in poison ivy, oak, and sumac. Every year up to 50 million Americans develop a poison ivy, oak, or sumac rash. The chance of developing an allergic sensitivity to these poison plants decreases with age, and adults who have never been exposed to urushiol only have a 50 percent chance of developing **contact dermatitis** when exposed to poison ivy, oak, or sumac.

In addition, allergic sensitivity to poison plants tends to lessen with age. It is possible for children who are highly reactive to urushiol to grow into adults who are barely sensitive to poison ivy, oak, or sumac, regardless of how many times they have been exposed to the plant oil.

Causes and symptoms

While direct skin-to-plant contact with poison ivy, oak, or sumac is probably the most frequent cause of the rash, the irritants from the plants can also be passed on indirectly. Urushiol oil can be transmitted on clothing, pets, garden tools, shoes, or virtually anything that touches a plant.

Most children will not get a rash the very first time they are exposed to poison ivy, oak, or sumac, although this is when the sensitivity, or immune response, to urushiol develops. Not everyone acquires an allergic sensitivity to urushiol, but in those that do, the next time they are exposed to the plant and urushiol penetrates the skin, a rash is inevitable.

The first and most annoying symptom of a poisonous plant rash is severe itching. This may precede the rash or start at the same time as the rash appears. The rash, which is red and inflamed, usually begins to appear within two days after the initial exposure and is usually in a pattern of streaks or patches that approximates where the plant made contact with the skin. Blisters and/or red papules may form soon after the rash appears.

When to call the doctor

Mild cases of poison plant rash can usually be treated at home with over-the-counter creams and itch-relief measures, such as ice packs. A child who is not getting adequate relief from these treatments should see a doctor. Prescription cortisone cream or prednisone treatment may be necessary to relieve the itching.

Anyone who is experiencing symptoms of anaphylactic shock (such as difficulty breathing, **dizziness**, **nausea**, rash, swelling, itchy eyes, loss of consciousness) after exposure to poison ivy, oak, or sumac should be taken to the nearest hospital or emergency care facility for immediate treatment. Poison plant rashes that spread to the eyes and affect vision should also be treated by a doctor as soon as possible. If rash blisters are broken while scratching and begin to show signs of infection (for example, **pain**, swelling, puss, systemic **fever**), a doctor should examine them as soon as possible in case **antibiotics** are necessary.

Diagnosis

Poison plant rashes are diagnosed through an examination of the rash. A physician can distinguish poison ivy, oak, or sumac from other allergic contact **dermatitis** through a brief patient interview. If the contact with the plant was direct, the diagnosis may be obvious. If it was indirect (for example, from dog fur or garden tools), the

doctor may need to rule out other **allergies**, especially if there were other new potential allergens in the child's environment (for example, a new pet, food, soap, or medication).

Treatment

Treatments for the itching of poison ivy, oak, or sumac rashes range from calamine lotion and oatmeal baths to over-the-counter **antihistamines** and topical creams. Mild rashes may be relieved with a tub soak in baking soda solution, an oatmeal bath, or aluminum acetate (Domeboro solution). Calamine lotion and menthol ointments lessen the itching and dry out weeping blisters. Over-the-counter hydrocortisone creams and ointments and numbing sprays and lotions containing benzocaine and other anesthetic agents can relieve itching as well.

Benadryl and other oral antihistamines are also effective in soothing the discomfort and itch of poison plant rashes, but they can also cause drowsiness and are best used before bedtime.

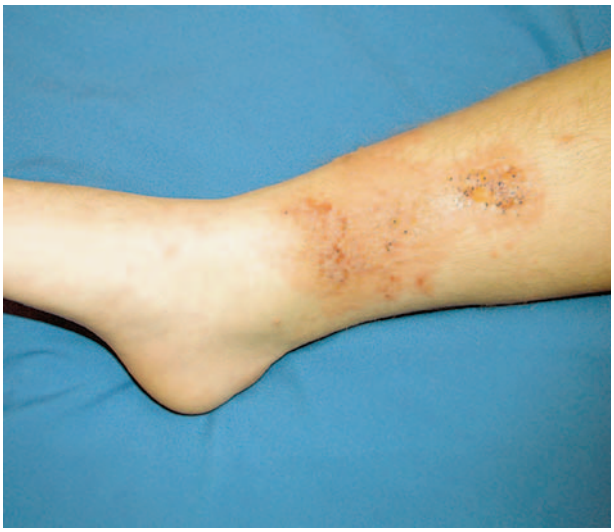
In severe cases of poison plant rash, a prescription-strength cortisone cream or corticosteroid treatment (either oral or injections) may be required to relieve swelling and itching. These medications should be taken under a doctor's supervision according to the directions for use only for the period of time prescribed, as overuse of corticosteroid creams has the potential of interfering with a child's normal growth and development. Corticosteroid treatment may not be a preferred treatment in children with diabetes, as the drug has the potential of increasing blood glucose levels.

There are several lotions and creams on the market that remove urushiol oil from the skin and can prevent further spreading of the rash if oil remains, or even prevent the rash entirely if applied early enough following exposure. Rubbing (isopropyl) alcohol can also remove urushiol on both skin and household objects.

Alternative treatment

The sap of the jewelweed plant (*Impatiens capensis*) is thought to be helpful in binding to and removing urushiol from skin. Either the plant itself (which grows wild in the Eastern United States, particularly in damp environments) can be rubbed on exposed skin, or a soap product made from Jewelweed (e.g., Burt's Bees Poison Ivy Soap) can be used to wash away urushiol. The plant must be used shortly after exposure to poison ivy, oak, or sumac to work.

A soak in tea tree oil (*Melaleuca alternaifolia*) or the application of gel from the aloe vera plant can also be useful in alleviating itching and in drying the blisters of poison plant rash. Tea tree oil also has antiseptic properties and may be useful in warding off infection when poison plant rash blisters break. Jewelweed, tea tree oil, and aloe vera are not recognized by the U.S. Food and Drug Administration as treatments for poison plant rash.



Poison ivy rash. *(© Scott Camazine/Photo Researchers, Inc.)*

Prognosis

The rash of poison ivy, oak, or sumac may last anywhere from a week to three weeks. In severe cases, it may linger up to a month. Usually there is no long-term effects or skin damage, but scratching that breaks the skin could potentially lead to permanent scarring. Infections can occur if blisters break and bacteria enters the open wound. Keeping the rash clean and any open blisters bandaged can lessen the chance of infection.

If urushiol enters the respiratory tract, which typically happens when the plant is burned and the smoke is breathed in, it can be life threatening. Anyone who encounters this type of exposure to poison ivy, oak, or sumac should seek emergency medical care immediately.

Prevention

Children should be advised to stay out of areas where poison ivy, oak, or sumac is known to grow. When people are hiking or camping, exposed skin should be covered with long sleeves and pants. There are several topical skin creams on the market that contain bentoquatum, which forms a protective barrier designed to repel urushiol oil (e.g., Ivy Block, Stokoguard). These may be

a useful preventative tool against poison plant rash as well.

If exposure does occur, washing with soap and cool water within the first 30 minutes of contact can sometimes prevent a rash. If soap is not available, rinse with water alone. A full body shower is best to eliminate all traces of the urushiol and prevent re-exposure from undetected oil remaining on other parts of the body. Again, water should be cool, as warm water will open pores and allow urushiol to penetrate the skin more quickly.

Other over-the-counter skin cleansers formulated to remove urushiol oil (e.g. Tecnu, IvyStat, IvyCleanse) can also stop or lessen the severity of a rash if they are applied early enough following exposure (i.e., before the urushiol begins penetrating the skin). These products can also be used to decontaminate garden tools and other items that have come in contact with the plant oil. Rubbing alcohol (isopropyl alcohol) is also helpful in decontaminating objects and skin.

Any clothing that has been exposed to poisonous plants should be handled carefully and laundered immediately. The same goes for shoes and garden gloves, which are common culprits of harboring urushiol oil. If possible, use latex or other disposable gloves to handle contaminated items and throw them away immediately afterwards.

Pet fur can also carry urushiol oil into the home. People should make sure outdoor pet areas are free of poisonous plants and never let a dog run unleashed in the woods or other areas with dense vegetation. Pets are typically not sensitive to urushiol, but a dog or cat that seems to be experiencing symptoms of poison plant rash following exposure should be taken to the veterinarian for **assessment**.

Eliminating known poison ivy, oak, or sumac growth in the yard or garden is also an important preventative step, but eradicating the weeds can be difficult. Glyphosate-based herbicides like Roundup and triclopyr-based herbicides like Ortho Brush-B-Gon will kill poison plants, but they can also take out any other surrounding foliage they come into contact with. If herbicides are used they should be applied carefully and may have to be rubbed directly on to the leaves and stems to avoid damaging other plantings.

Another option for eliminating poison plants is to pull them by hand. Proper protection of all exposed skin is important to prevent a reaction. The entire plant, including the root system, must be pulled. As an alternative, landscaping fabric or another barrier can be placed over poison ivy, oak, or sumac to kill the plants and prevent future growth. Dead plants still contain urushiol and must be handled carefully during removal. All plants should be disposed of according to local waste regulations. Never compost or burn poison plants because of the potential of spreading the oil through the garden or air. Mowing over the vines or plants can also send urushiol into the air and has the potential to cause a serious allergic reaction.

KEY TERMS

Corticosteroids—A group of hormones produced naturally by the adrenal gland or manufactured synthetically. They are often used to treat inflammation. Examples include cortisone and prednisone.

Papule—A solid, raised bump on the skin.

Urushiol—The oil from poison ivy, oak, and sumac that causes severe itching, blistering, and rash.

Parental concerns

The itching and discomfort of poison ivy, oak, and sumac rashes can disrupt **sleep**, make a child irritable and anxious, and pose a major distraction to schoolwork and other tasks that require concentration.

Soothing the itching is the best way to help a child get through the misery of a poisonous plant rash. Covering the affected areas with bandages may be useful in curtailing scratching and preventing potential scarring. A child's fingernails should be kept clean and trimmed short to lessen the chance of bursting and infecting blisters if scratching does occur.

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Paula Ford-Martin

Poisoning

Definition

Poisoning occurs when any substance interferes with normal body functions after it is swallowed, inhaled, injected, or absorbed. The branch of medicine that deals with the detection and treatment of poisons is known as toxicology.

Description

Children are the most common victims of poisoning in the United States. Curiosity, inability to read warning labels, a desire to imitate adults, and inadequate supervision lead to most childhood poisonings.

The elderly are the second most likely group to be poisoned. Mental confusion, poor eyesight, and the use of multiple drugs are the leading reasons this group has a high rate of accidental poisoning. A substantial number of poisonings also occur as **suicide** attempts or drug overdoses.

Poisons are common in the home and workplace, yet there are basically two major types. One group consists of products that were never meant to be ingested or inhaled, such as shampoo, paint thinner, pesticides, houseplant leaves, and carbon monoxide. The other group contains products that can be ingested in small quantities, but which are harmful if taken in large amounts, such as pharmaceuticals, medicinal herbs, or alcohol. Other types of poisons include the bacterial toxins that cause **food poisoning**, such as *Escherichia coli*; heavy metals, such as the lead found in the paint on older houses; and the venom found in the **bites and stings** of some animals and insects. The staff at a poison control center and emergency room doctors have the most experience diagnosing and treating poisoning cases.

Demographics

Poisonings are a common occurrence. About 10 million cases of poisoning occur in the United States each year. In 80 percent of the cases, the victim is a child under the age of five. About 50 children die each year from poisonings.

Causes and symptoms

The effects of poisons are as varied as the poisons themselves; however, the exact mechanisms of only a few are understood. Some poisons interfere with the metabolism. Others destroy the liver or kidneys, such as heavy metals and some **pain** relief medications, including **acetaminophen** and **nonsteroidal anti-inflammatory drugs** (ibuprofen). A poison may severely depress the central nervous system, leading to coma and eventual respiratory and circulatory failure. Potential poisons in this category include anesthetics (e.g. ether and chloroform), opiates (e.g., morphine and codeine), and barbiturates. Some poisons directly affect the respiratory and circulatory system. Carbon monoxide causes death by binding with hemoglobin that would normally transport oxygen throughout the body. Certain corrosive vapors trigger the body to flood the lungs with fluids, effectively drowning the person. Cyanide interferes with respiration at the cellular level. Another group of poisons interferes with the electrochemical impulses that travel between neurons in the nervous system. Yet another group, including cocaine, ergot, strychnine, and some snake venoms, causes potentially fatal seizures.

Severity of symptoms can range from **headache** and **nausea** to convulsions and death. The type of poison, the amount and time of exposure, and the age, size, and health of the victim are all factors which taken together determine the severity of symptoms and the chances for recovery.

Plant poisoning

There are more than 700 species of poisonous plants in the United States. Plants are second only to medicines

in causing serious poisoning in children under age five. There is no way to tell by looking at a plant if it is poisonous. Some plants, such as the yew shrub, are almost entirely toxic: needles, bark, seeds, and berries. In other plants, only certain parts are poisonous. The bulb of the hyacinth and daffodil are toxic, but the flowers are not; while the flowers of the jasmine plant are the poisonous part. Moreover, some plants are confusing because portions of them are eaten as food while other parts are poisonous. For example, the fleshy stem (tuber) of the potato plant is nutritious; however, its roots, sprouts, and vines are poisonous. The leaves of tomatoes are poisonous, while the fruit is not. Rhubarb stalks are good to eat, but the leaves are poisonous. Apricots, cherries, peaches, and apples all produce healthful fruit, but their seeds contain a form of cyanide that can kill a child if chewed in sufficient quantities. One hundred milligrams (mg) of moist, crushed apricot seeds can produce 217 mg of cyanide.

Common houseplants that contain some poisonous parts include the following:

- aloe
- amaryllis
- cyclamen
- dumb cane (also called Dieffenbachia)
- philodendron

Common outdoor plants that contain some poisonous part include the following:

- bird of paradise flower
- buttercup
- castor bean
- chinaberry tree
- daffodil
- English ivy
- eucalyptus
- foxglove
- holly
- horse chestnut
- iris
- jack-in-the-pulpit
- jimsonweed (also called thornapple)
- larkspur
- lily-of-the-valley
- morning glory
- nightshade (several varieties)
- oleander
- potato
- rhododendron
- rhubarb
- sweet pea
- tomato
- wisteria
- yew

Symptoms of plant poisoning range from irritation of the skin or mucous membranes of the mouth and throat to nausea, **vomiting**, convulsions, irregular heartbeat, and even death. It is often difficult to tell if a person has eaten a poisonous plant because there are no tell-tale empty containers and no unusual lesions or odors around the mouth.

Many cases of plant poisoning involve plants that contain hallucinogens, such as peyote cactus buttons, certain types of mushrooms, and marijuana. Poisoning has occurred with *Datura*, or moonflower, a plant that has become popular with young people trying to imitate Native American **puberty** rites.

Other cases of plant poisoning result from the use of herbal dietary supplements that have been contaminated by toxic substances. The Food and Drug Administration (FDA) has the authority to monitor herbal products on the market and issue warnings about accidental poisoning or other adverse affects associated with these products. For example, in 2002 a manufacturer of nettle capsules found to contain lead recalled the product following a warning from the FDA. Other dietary supplements have been found to contain small quantities of prescription medications or even toxic plants.

Household chemicals

Many products used daily in the home are poisonous if swallowed. These products often contain strong acids or strong bases (alkalis). Toxic household cleaning products include the following:

- ammonia
- bleach
- dishwashing liquids
- drain openers
- floor waxes and furniture polishes
- laundry detergents, spot cleaners, and fabric softeners
- mildew removers
- oven cleaners

- toilet bowl cleaners

Personal care products found in the home can also be poisonous. These include:

- deodorant
- hairspray
- hair straighteners
- nail polish and polish remover
- perfume
- shampoo

Signs that a person has swallowed one of these substances include evidence of an empty container nearby, nausea or vomiting, and **burns** on the lips and skin around the mouth if the substance is a strong acid or alkali. The chemicals in some of these products may leave a distinctive odor on the breath.

Pharmaceuticals

Both over-the-counter and prescription medicines can help the body heal if taken as directed. However, when taken in large quantities, or with other drugs with which there may be an adverse interaction, they can act as poisons. Drug overdoses, both accidental and intentional, are the leading cause of poisoning in adults. Medicinal herbs should be treated like pharmaceuticals and taken only in designated quantities under the supervision of a knowledgeable person. Herbs that have healing qualities when taken in small doses can be toxic in larger doses or may interact with prescription medications in unpredictable ways.

Drug overdoses cause a range of symptoms, including excitability, sleepiness, confusion, unconsciousness, rapid heartbeat, convulsions, nausea, and changes in blood pressure. The best initial evidence of a drug overdose is the presence of an empty container near the victim.

Other causes of poisonings

People can be poisoned by fumes they inhale. Carbon monoxide is the most common form of inhaled poison. Other toxic substances that can be inhaled include:

- farm and garden insecticides and herbicides
- gasoline fumes
- insect repellent
- paint thinner fumes

When to call the doctor

A doctor or poison control center should be called if any form of poisoning is suspected or if children or other persons behave in an odd manner.

Diagnosis

Initially, poisoning is suspected if the victim shows changes in behavior and signs or symptoms previously described. Hallucinations or other psychiatric symptoms may indicate poisoning by a hallucinogenic plant. Evidence of an empty container or information from the victim is helpful in determining exactly what substance has caused the poisoning. Some acids and alkalis leave burns on the mouth. Petroleum products, such as lighter fluid or kerosene, leave a distinctive odor on the breath. The vomit may be tested to determine the exact composition of the poison. Once hospitalized, the person may be given blood and urine tests to determine his or her metabolic condition.

Treatment

Treatment for poisoning depends on the poison swallowed or inhaled. Contacting the poison control center or hospital emergency room is the first step in getting proper treatment. The poison control center's telephone number is often listed with emergency numbers on the inside cover of the telephone book, or it can be reached by dialing the operator. The poison control center will ask for specific information about the victim and the poison then give appropriate first aid instructions. If the person is to be taken to a hospital, a sample of vomit and the poison container should be taken along, if they are available.

For acid, alkali, or petroleum product poisonings, the person should not vomit. Acids and alkalis can burn the esophagus if they are vomited, and petroleum products can be inhaled into the lungs during vomiting, resulting in **pneumonia**.

Once the victim is under medical care, doctors have the option of treating the person with a specific remedy to counteract the poison (antidote) or with activated charcoal to absorb the substance inside the individual's digestive system. In some instances, pumping the stomach may be required. This technique, which is known as gastric lavage, involves introducing 20 to 30 mL of tap water or 9 percent saline solution into the person's digestive tract and removing the stomach contents with a siphon or syringe. The process is repeated until the washings are free of poison. Medical personnel will also

provide supportive care as needed, such as intravenous fluids or mechanical ventilation.

If the doctor suspects that the poisoning was not accidental, he or she is required to notify law enforcement authorities. Most cases of malicious poisoning concern **family** members or acquaintances of the victim, but the number of intentional random poisonings of the general public has increased in the late 1990s and early 2000s. A case reported in 2003 involved the use of nicotine to poison 1,700 pounds of ground beef in a Michigan supermarket. Over 100 persons fell ill after eating the poisoned beef.

Prognosis

The outcome of poisoning varies from complete recovery to death and depends on the type and amount of the poison, the health of the victim, and the speed with which medical care is obtained.

Prevention

Most accidental poisonings are preventable. The number of deaths of children from poisoning has declined from about 450 per year in the 1960s to about 50 each year in the 1990s. This decline has occurred mainly because of better packaging of toxic materials and better public education.

Actions to prevent poisonings include:

- removing plants that are poisonous
- keeping medicines and household chemicals locked and in a place inaccessible to children
- keeping medications in child-resistant containers
- never referring to medicine as candy
- keeping cleaners and other poisons in their original containers
- disposing of outdated prescription medicines
- not purchasing over-the-counter medications with damaged protective seals or packaging
- avoiding the use of herbal preparations not made by a reputable manufacturer

Parental concerns

Parents should monitor the activities and substances to which their children are exposed. The number of the nearest Poison Control Center should be posted next to every telephone in the house. The number can be found on the first page of any telephone book.

KEY TERMS

Antidote—A remedy to counteract a poison or injury. Also refers to a substance which cancels the effect of homeopathic remedies

Emetic—A medication intended to cause vomiting. Emetics are sometimes used in aversion therapy in place of electric shock. Their most common use in mainstream medicine is in treating accidental poisoning.

Gastric lavage—Also called a stomach pump. For this procedure, a flexible tube is inserted through the nose, down the throat, and into the stomach and the contents of the stomach are suctioned out. The inside of the stomach is rinsed with a saline (salt water) solution.

Toxicology—The branch of medical pharmacology dealing with the detection, effects, and antidotes of poisons.

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American Academy of Emergency Medicine. 611 East Wells St., Milwaukee, WI 53202. Web site: <www.aaem.org/>.

American Academy of Family Physicians. 11400 Tomahawk Creek Parkway, Leawood, KS 66211–2672. Web site: <www.aafp.org/>.

American Academy of Pediatrics. 141 Northwest Point Blvd., Elk Grove Village, IL 60007–1098. Web site: <www.aap.org/default.htm>.

American Association of Poison Control Centers. 3201 New Mexico Ave., NW, Washington, DC 20016. Web site: <www.aapcc.org/>.

American College of Emergency Physicians. PO Box 619911, Dallas, TX 75261–9911. Web site: <www.acep.org/>.

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Polio

Definition

Poliomyelitis, also called polio or infantile paralysis, is a highly infectious viral disease that may attack the central nervous system and is characterized by symptoms that range from a mild nonparalytic infection to total paralysis in a matter of hours.

Description

There are three known types of polioviruses (called 1, 2, and 3), each causing a different strain of the disease and all being members of the viral family of enteroviruses (viruses that infect the gastrointestinal tract). Type 1 is the cause of epidemics, and many cases of paralysis, which is the most severe manifestation of the infection. The virus is usually a harmless parasite of human beings. Some statistics quote one in 200 infections as leading to paralysis, while others state that one in 1,000 cases reach the central nervous system (CNS). When it does reach the CNS, inflammation and destruction of the spinal cord motor cells (anterior horn cells) occurs, which prevents them from sending out impulses to muscles. This causes the muscles to become limp or soft, and they cannot contract, a condition called flaccid paralysis and is the type found in polio. The extent of the paralysis depends on where the virus strikes and the number of cells that it destroys. Usually, some of the limb muscles are paralyzed; the abdominal muscles or muscles of the back may be paralyzed, affecting posture. The neck muscles may become too weak for the head to be lifted. Paralysis of the face muscles may cause the mouth to twist or

the eyelids to droop. Life may be threatened if paralysis of the throat or of the breathing muscles occurs.

Humans are the only natural host for polioviruses, and it most commonly infects younger children, although older children and adults can be infected. Crowded living conditions and poor hygiene encourage the spread of poliovirus. Risk factors for this paralytic illness include older age, pregnancy, abnormalities of the immune system, and a recent episode of excessively strenuous **exercise** concurrent with the onset of the CNS phase. As of 2004, the last naturally occurring polio case in the United States was diagnosed in 1979.

Causes and symptoms

Poliovirus can be spread by direct exposure to an infected individual, and more rarely, by eating foods contaminated with waste products from the intestines (feces) and/or droplets of moisture (saliva) from an infected person. Thus, the major route of transmission is fecal-oral, which occurs primarily with poor sanitary conditions. The virus is believed to enter the body through the mouth with primary multiplication occurring in the lymphoid tissues in the throat, where it can persist for about one week. During this time, it is absorbed into the blood and lymphatics from the gastrointestinal tract where it can reside and multiply, sometimes for as long as 17 weeks. Once absorbed, it is widely distributed throughout the body until it ultimately reaches the CNS (the brain and spinal cord). The infection is passed on to others when poor hand washing allows the virus to remain on the hands after eating or using the bathroom. Transmission remains possible while the virus is being excreted and it can be transmitted for as long as the virus remains in the throat or feces. The incubation period ranges from three to 21 days, but cases are most infectious from seven to ten days before and after the onset of symptoms.

There are two basic patterns to the virus: the minor illness (abortive type) and the major illness (which may be paralytic or nonparalytic). The minor illness accounts for 80 to 90 percent of clinical infections and is found mostly in young children. It is mild and does not involve the CNS. Symptoms include a slight **fever**, fatigue, **headache**, **sore throat**, and **vomiting**, which generally develop three to five days after exposure. Recovery from the minor illness occurs within 24 to 72 hours. Symptoms of the major illness usually appear without a previous minor illness and generally affect older children and adults.

About 10 percent of people infected with poliovirus develop severe headache and **pain** and stiffness of the neck and back. This is due to an inflammation of the meninges (tissues which cover the spinal cord and brain). This syndrome is called aseptic **meningitis**. The term aseptic is used to differentiate this type of meningitis from those caused by bacteria. The patient usually recovers completely from this illness within several days.

About 1 percent of people infected with poliovirus develop the most severe form. Some of these patients may have two to three symptom-free days between the minor illness and the major illness, but the symptoms often appear without any previous minor illness. Symptoms again include headache and back and neck pain. The major symptoms, however, are due to invasion of the motor nerves, which are responsible for movement of the muscles. This viral invasion causes inflammation and then destruction of these nerves. The muscles, therefore, no longer receive any messages from the brain or spinal cord. The muscles become weak, floppy, and then totally paralyzed. All muscle tone is lost in the affected limb and the muscle becomes soft (flaccid). Within a few days, the muscle begins to decrease in size (atrophy). The affected muscles may be on both sides of the body (symmetric paralysis) but are often on unbalanced parts of the body (asymmetric paralysis). Sensation or the ability to feel is not affected in these paralyzed limbs.

When poliovirus invades the brainstem (the stalk of brain which connects the two cerebral hemispheres with the spinal cord, called bulbar polio), a person may begin to have trouble breathing and swallowing. If the brainstem is severely affected, the brain's control of such vital functions as heart rate and blood pressure may be disturbed, a condition that can lead to death.

The maximum state of paralysis is usually reached within just a few days. The remaining, unaffected nerves then begin the process of attempting to grow branches, which can compensate for the destroyed nerves. Fortunately, the nerve cells are not always completely destroyed. By the end of a month, the nerve impulses start to return to the apparently paralyzed muscle and by the end of six months, recovery is almost complete. If the nerve cells are completely destroyed; however, paralysis is permanent.

Diagnosis

Fever and asymmetric flaccid paralysis without sensory loss in a child or young adult almost always indicate poliomyelitis. Using a long, thin needle inserted into the lower back to withdraw spinal fluid (lumbar puncture)

will reveal increased white blood cells and no bacteria (aseptic meningitis). Nonparalytic poliomyelitis cannot be distinguished clinically from aseptic meningitis due to other agents. Virus isolated from a throat swab and/or feces or blood tests demonstrating the rise in a specific antibody is required to confirm the diagnosis.

Treatment

There is no specific treatment for polio except symptomatic. Therapy is designed to make the patient more comfortable (pain medications and hot packs to soothe the muscles), and intervention if the muscles responsible for breathing fail (for instance, a ventilator to take over the work of breathing). During active infection, rest on a firm bed is indicated. Physical therapy is the most important part of management of paralytic polio during recovery.

Prognosis

When poliovirus causes only the minor illness or simple aseptic meningitis, the patient can be expected to recover completely. Among patients with the major illness, about 50 percent recover completely. About 25 percent of such patients have slight disability, and about 25 percent have permanent and serious disability. Approximately 1 percent of all patients with major illness die. The greatest return of muscle function occurs in the first six months, but improvements may continue for two years.

Post-polio syndrome (PPS) is a condition that can strike polio survivors anywhere from 10 to 40 years after their recovery from polio. It is caused by the death of individual nerve terminals in the motor units that remain after the initial polio attack. Symptoms include fatigue, slowly progressive muscle weakness, muscle and joint pain, and muscular atrophy. The severity of PPS depends upon how seriously the survivors were affected by the first polio attack.

Prevention

There are two types of polio immunizations available in the United States, but since the year 2000, one is rarely used. A vaccine takes advantage of the fact that infection with polio leads to an immune reaction, which will give the person permanent, lifelong immunity from reinfection with the form of poliovirus for which the person was vaccinated.

The Salk vaccine (also called the killed **polio vaccine** or inactivated polio vaccine, IPV) consists of a series of three shots that are given just under the skin to children at the ages of two months, four months, and any time between six and 18 months. A fourth injection is given between the ages of four to six years as a booster. This immunization contains no live virus, just the components of the virus that provoke the recipient's immune system to react as if the recipient were actually infected with the poliovirus. The recipient thus becomes immune to infection with the poliovirus.

Since the year 2000, the Sabin vaccine (also called the oral polio vaccine or OPV) has been discontinued in the United States, although it is still being used in other countries. It contains the live, but weakened, poliovirus and because OPV uses the live virus, it has the potential to cause infection in individuals with weak immune defenses (both in the person who receives the vaccine and in close contacts). Approximately nine cases a year of vaccine related polio was associated with OPV in the United States. Although this is a rare complication,

KEY TERMS

Aseptic—Sterile; containing no microorganisms, especially no bacteria.

Asymmetric—Not occurring equally on both sides of the body.

Atrophy—The progressive wasting and loss of function of any part of the body.

Brainstem—The stalk of the brain which connects the two cerebral hemispheres with the spinal cord. It is involved in controlling vital functions, movement, sensation, and nerves supplying the head and neck.

Epidemic—Refers to a situation in which a particular disease rapidly spreads among many people in the same geographical region in a relatively short period of time.

Flaccid—Flabby, limp, weak, or floppy.

Gastrointestinal—Pertaining to the digestive organs and structures, including the stomach and intestines.

Lymph—Clear, slightly yellow fluid carried by a network of thin tubes to every part of the body. Cells that fight infection are carried in the lymph.

Paralysis—Loss of the ability to move one or more parts of the body voluntarily due to muscle or nerve damage.

Symmetric—Occurring on both sides of the body, in a mirror-image fashion.

occurring in only one in 6.8 million doses administered and one in every 6.4 million doses from having close contact with someone who received the vaccine, the risk of having polio from OPV was greater than it was of naturally acquiring it.

Following the launching of the Global Polio Eradication Initiative, the number of cases fell 99 percent from an estimated 350,000 cases to less than 3,500 cases worldwide in 2000. At the end of 2000, the number of polio-infected countries was approximately 20, down from 125. The goal of the World Health Organization (WHO) is to have polio eliminated from the planet by the year 2005. The virus has still been identified in Africa and parts of Asia, so travelers to those areas may want to check with their physicians concerning booster vaccinations.

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Polio vaccine

Definition

The poliomyelitis (**polio**) vaccine protects against poliovirus infections. The vaccine helps the body produce antibodies (protective substances) that will prevent an individual from contracting polio. There are two forms of the vaccine that can be given; the one preferred is the inactivated poliovirus vaccine (IPV). IPV is preferred because it contains the inactivated or dead virus, which is considered safer for administration. The Sabin oral polio vaccine was made with a live but weakened virus, which gives the advantage of passive immunity for large groups (i.e. because it is easily passed on through the oral fecal route in households, schoolrooms, etc., even if only a portion of the community is immunized, everyone eventually develops immunity). The Sabin oral polio vaccine has the disadvantage of causing polio-like symptoms in some immune compromised hosts. Since 2000, the live virus vaccine is rarely used in the United States, but it is still being used in other countries.

Description

The purpose of any vaccine is to prevent disease. Mass immunizations in the United States have served to eradicate polio in the Americas. In 1988, the World Health Organization embarked upon a mission to eradicate polio by the year 2000. The intent was to immunize the world's children by methodically establishing sites where the masses could be reached. The mandate was to eradicate the virus by 2005. Most of the remaining virus can be found on the Indian subcontinent and Nigeria. Attempts to immunize children in Indian have met with good results, but Nigeria halted their immunization sites due to rumors that Western donors had tampered with the vaccine to spread HIV and cause sterility in Muslim males. Following a ban on the vaccine that lasted nearly one year, the virus spread across Nigeria to 10 African countries that were previously polio-free. The Muslim leaders in Nigeria lifted the ban in summer 2004. Immunization from the IPV triggers an excellent immune response and long-lasting immunity to all three poliovirus types.

General use

The inactivated poliovirus vaccine is injected into a muscle or under the skin and is usually given by a health care professional in a hospital, clinic, or provider's office. The use of this vaccine must be officially recorded. Federal law requires that the vaccine manufacturer's name, the lot number of the vaccine, the name, address, and phone number of the person giving the vaccine, and the date of vaccine administration be recorded in a permanent medical record. For children, the vaccine is usually started at two months of age and given again at four months of age. The next dose should be given between six and 18 months of age with a final booster dose at age four to six years, for a total of four doses. Serious reactions to the inactivated poliovirus vaccine are

rare in small children. It is necessary to receive all doses of the vaccine and there is no generic vaccine available.

Precautions

Individuals with an immune deficiency disease need to be counseled before taking the vaccine, and anyone with allergic reactions to prior vaccines and preservatives should be cautious. A provider may want to delay giving a child a dose of IPV or may not give it at all if the child has a known severe allergy to the **antibiotics** neomycin, streptomycin, or polymyxin B. A child who had a life-threatening reaction to a previous IPV should not receive another one.

There is no preparation necessary for the vaccine; however, if an individual is ill on the scheduled date, it is essential to make arrangements for a follow-up appointment as no dose can be missed.

Side effects

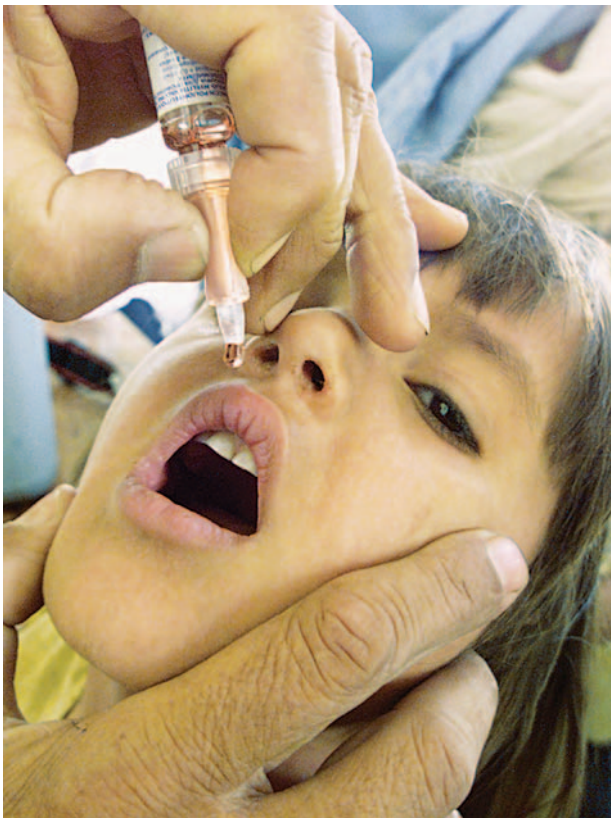
Children receiving the inactivated poliovirus vaccine should be carefully observed for 24–72 hours after receiving the injection. If any serious side effects occur, the healthcare provider or an emergency service provider should be called immediately. For problems that may occur following the vaccine, parents are asked to call the vaccine adverse event reporting system toll-free at (800) 822–7967 to report them. The health care professional may administer a dose of a non-aspirin pain/fever reliever at the time of the vaccine and advise giving the medicine every four to six hours for 24 hours after the vaccine. This may serve to reduce **pain** and **fever** associated with the vaccine.

Side effects that usually do not require immediate medical attention, unless they persist and are bothersome, include:

- fussiness
- decreased appetite
- low-grade fever (102°F [39° C] or less)
- pain, tenderness, redness, swelling, or a ‘‘knot’’ at the injection site
- fatigue
- vomiting

Side effects that should be reported as soon as possible are:

- limp, pale, or less alert child
- difficulty breathing, shortness of breath, or wheezing
- difficulty swallowing



Oral polio vaccines are no longer recommended for use in the United States, but are still used in many parts of the world. *(© Reuters/Corbis)*

- high fever (103°F [39.4°C] or more)
- inconsolable crying for three hours or more
- seizures (convulsions)
- severe skin rash, **hives**, or itching
- swelling of eyes or face
- unusual sleepiness

Interactions

Before administering the vaccine, the healthcare provider should be informed as to whether the recipient has any of the following conditions:

- an immune deficiency (natural or due to **cancer chemotherapy** radiation, steroid therapy, or HIV infection)
- fever or infection
- an unusual reaction to this poliovirus vaccine, oral poliovirus vaccine, other medicines, foods, dyes, or preservatives

KEY TERMS

World Health Organization (WHO)—An international organization within the United Nations system that is concerned with world health and welfare.

- pregnant or trying to get pregnant
- breastfeeding

Prevention

While it is important to mention these items to the physician, they are not necessarily contraindications for the vaccine. The provider also needs to know what medicines an individual is taking, including non-prescription medicines, nutritional supplements, herbal products, tobacco, and whether or not he or she is a user of illegal drugs or a frequent user of drinks with **caffeine** or alcohol. Any of these may affect the way the vaccine works.

Risks

There have been no adverse effects from IPV reported to date. However, IPV induces only little immunity in the intestinal tract. If an individual is infected with the wild-type poliovirus, the virus can multiply in the intestines and be shed in stools, ultimately heightening the risk of viral circulation within the community. This scenario is unlikely in the United States.

Parental concerns

Parents need to be aware of any existing **allergies** in their families that might cause a reaction from vaccines and their preservatives, and they need to be observant of a child for the first 24–72 hours after receiving the vaccine. Traveling to other parts of the world may necessitate a booster vaccine if the polio virus is known to be present in that vicinity.

See also Polio.

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Linda K. Bennington, MSN, CNS

Polydactyly and syndactyly

Definition

Polydactyly and syndactyly are congenital malformations of the fingers and/or toes. Polydactyly is the presence of extra fingers or toes, and syndactyly is the fusing together or webbing of two or more fingers or toes.

Description

Polydactyly and syndactyly can vary in the degree of severity. Polydactyly may range from small skin tags on the side of the hands to fully formed extra fingers with bone, blood vessels, and muscle tissue. Polydactyly is classified as postaxial if the extra digit is located beside the little finger or preaxial if the extra digit is located near the thumb. Syndactyly may be either a complete fusion of fingers or toes or a looser webbing of skin between them. Syndactyly and polydactyly may occur simultaneously when extra digits are fused in a condition known as polysyndactyly. Polydactyly and syndactyly are usually isolated conditions, meaning the child will have no other birth defects; however, both are also found in many complex and sometimes lethal groups of anomalies or syndromes.

Demographics

Syndactyly of the fingers is the most common malformation affecting the hand. It occurs in approximately one in 2,000 live births. Postaxial polydactyly, or an extra pinky finger, is the second most common malfor-

mation of the hand, occurring in approximately one in 3,000 births. Polydactyly of the toes, especially of the little toe, is also a common malformation, occurring in approximately two in 1,000 births. Thirty percent of all children with polydactyly have **family** members with some type of polydactyly, and it is more common in African Americans. Syndactyly is an equally common malformation and runs in families as well.

Causes and symptoms

Polydactyly and syndactyly are conditions that occur in the developing fetus. Most often these conditions are caused by genetic factors. Both polydactyly and syndactyly can be caused by the presence of an autosomal dominant trait. An autosomal dominant trait is a gene that is not related to the chromosome that determines gender; therefore, it affects boys and girls equally. Because the gene is dominant, when one parent has the gene, each of his or her children has a 50 percent chance of having polydactyly or syndactyly.

The primary symptom of polydactyly is the presence of extra digits on the hands or feet. Polydactyly rarely causes any difficulties for the child. The extra fingers and/or toes are usually removed for cosmetic reasons. In children with syndactyly of the hand, finger function may be impaired and, in cases where fingers of different lengths are connected by tissue, finger growth may be limited unless surgery to separate the fingers is performed.

Diagnosis

Diagnosis is made during the initial physical examination at birth. Some children with polydactyly will need radiographs or x rays to determine if there is bone present in the extra digit. This will indicate what type of surgery is necessary to remove the extra finger or toe. When polydactyly is more severe and involves digits with bone, a pediatric orthopedic surgeon will perform the repair. In children with syndactyly, the surgeon must determine if the fusion involves muscle tissue and blood vessels, and in children with severe polydactyly of the fingers, a surgeon specializing in hands may evaluate the child as well. Some children with syndactyly may also have cardiac or heart problems; therefore, an electrocardiogram (EKG) may be ordered to evaluate heart function.

Treatment

For children with minor cases of polydactyly, the extra finger or toe may be tied at its base to restrict blood flow into it. Eventually the extra digit will fall off. In more significant cases, a pediatric orthopedic surgeon will remove the extra finger or toe and reconstruct the part of the hand or foot that was affected. The surgeon will save the digit that best fits with the others. This surgery usually occurs when the child is between one and two years old.

Polydactyly is the occurrence of extra or partial fingers or toes. *(Photograph by Joseph R. Siebert. Custom Medical Stock Photo, Inc.)*

Prognosis

For children with isolated polydactyly and syndactyly, the prognosis is excellent. After surgery most children will have full use of their fingers and toes. In children with syndromes and additional birth defects, the prognosis may be poor; however, the problems these children may experience will not be related to polydactyly or syndactyly, but rather to the sometimes lethal conditions that accompany the other defects present.

Prevention

There is no known prevention for polydactyly and syndactyly.

Parental concerns

Parents may be concerned about their child's ability to use his hands and feet. For most children, surgery corrects the condition and allows normal use of the hands and feet. Parents may also be concerned about the appearance of the hands and feet. In most cases, following surgery, they will have a normal appearance.

Children with more severe polydactyly and syndactyly will have surgery to correct the malformation. These children will need to be prepared for the experience.

KEY TERMS

Autosomal dominant trait—One of the non-X or non-Y chromosomes that will always express itself when present.

Congenital—Present at birth.

Electrocardiagram (ECG, EKG)—A record of the electrical activity of the heart, with each wave being labeled as P, Q, R, S, and T waves. It is often used in the diagnosis of cases of abnormal cardiac rhythm and myocardial damage.

Gene—A building block of inheritance, which contains the instructions for the production of a particular protein, and is made up of a molecular sequence found on a section of DNA. Each gene is found on a precise location on a chromosome.

Malformation—An irregular or abnormal formation or structure.

Postaxial—Situated behind or away from the axis or midline of the body.

Preaxial—Situated in front of the axis or midline of the body.

Radiograph—The actual picture or film produced by an x-ray study.

Syndrome—A group of signs and symptoms that collectively characterize a disease or disorder.

Webbing—A tissue or membrane that connects two digits at their base or for the greater part of their length.

Polydactyly and syndactyly correction surgery is usually performed when the child is between the ages of one and two years. When talking to children of this age, tone of voice is as important as the words used. Using a calm and comforting voice and simple language, parents can tell the child what will happen. Young children should be told the day before surgery that they will be going to the hospital. Parents may want to use dolls or stuffed animals to show their child where the doctor will work. If a child must spend the night, it is helpful to bring a toy or familiar object to make the child feel more comfortable in the hospital. Afterwards, the child may reenact the experience during **play** or in **drawings**. Parents should be prepared to talk and play with their child in this way.

When to call the doctor

Following surgery, parents should call the doctor if any of these symptoms occur:

- Unexpected bleeding from the wound
- An oral temperature of over 101 degrees
- **Pain** that is nor relieved by pain medication
- A persistent dry cough
- Nausea or **vomiting** that does not improve
- Redness or swelling around the incision site

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Porphyrias

Definition

The porphyrias are disorders in which the body produces too much porphyrin and insufficient heme (an iron-containing nonprotein portion of the hemoglobin molecule). Porphyrin is a foundation structure for heme and certain enzymes. Excess porphyrins are excreted as waste in urine and stool. Overproduction and overexcretion of porphyrins causes low, unhealthy levels of heme and certain important enzymes, creating various physical symptoms.

Description

Biosynthesis of heme is a multistep process that begins with simple molecules and ends with a large,

complex heme molecule. Each step of the chemical pathway is directed by its own task-specific protein, called an enzyme. As a heme precursor molecule moves through each step, an enzyme modifies the precursor in some way. If a precursor molecule is not modified, it cannot proceed to the next step, causing a buildup of that specific precursor.

This situation is the main characteristic of the porphyrias. Owing to a defect in one of the enzymes of the heme biosynthesis pathway, protoporphyrins or porphyrins (heme precursors) are prevented from proceeding further along the pathway. These precursors accumulate at the stage of the enzyme defect, causing an array of physical symptoms in an affected child. Specific symptoms depend on the point at which heme biosynthesis is blocked and which precursors accumulate. In general, the porphyrias primarily affect the skin and the nervous system. Symptoms can be debilitating or life threatening in some cases. Porphyria is most commonly an inherited condition. It can also, however, be acquired after exposure to poisonous substances.

Heme

Heme is produced in several tissues in the body, but its primary biosynthesis sites are the liver and the bone marrow. Heme synthesis for immature red blood cells, namely the erythroblasts and the reticulocytes, occurs in the bone marrow.

Although production is concentrated in the liver and bone marrow, heme is utilized in various capacities in virtually every tissue in the body. In most cells, heme is a key building block in the construction of factors that oversee metabolism and transport of oxygen and energy. In the liver, heme is a component of several vital enzymes, particularly cytochrome P450. Cytochrome P450 is involved in the metabolism of chemicals, **vitamins**, fatty acids, and hormones; it is very important in transforming toxic substances into easily excretable materials. In immature red blood cells, heme is the featured component of hemoglobin. Hemoglobin is the red pigment that gives red blood cells their characteristic color and their essential ability to transport oxygen.

Heme biosynthesis

The heme molecule is composed of porphyrin and an iron atom. Much of the heme biosynthesis pathway is dedicated to constructing the porphyrin molecule. Porphyrin is a large molecule shaped like a four-leaf clover. An iron atom is placed at its center point in the last step of heme biosynthesis.

The production of heme may be compared to a factory assembly line. At the start of the line, raw materials are fed into the process. At specific points along the line, an addition or adjustment is made to further development. Once additions and adjustments are complete, the final product rolls off the end of the line.

The heme "assembly line" is an eight-step process, requiring eight different and properly functioning enzymes:

1. delta-aminolevulinic acid synthase
2. delta-aminolevulinic acid dehydratase
3. porphobilogen deaminase
4. uroporphyrinogen III cosynthase
5. uroporphyrinogen decarboxylase
6. coproporphyrinogen oxidase
7. protoporphyrinogen oxidase
8. ferrochelatase

The control of heme biosynthesis is complex. Various chemical signals can trigger increased or decreased production. These signals can affect the enzymes themselves or the production of these enzymes, starting at the genetic level. For example, one point at which heme biosynthesis may be controlled is at the first step. When heme levels are low, greater quantities of delta-aminolevulinic acid (ALA) synthase are produced. As a result, larger quantities of heme precursors are fed into the biosynthesis pathway to step up heme production.

Porphyrias

Under normal circumstances, when heme concentrations are at an appropriate level, precursor production decreases. However, a glitch in the biosynthesis pathway—represented by a defective enzyme—means that heme biosynthesis does not reach completion. Because heme levels remain low, the synthesis pathway continues to churn out precursor molecules in an attempt to correct the heme deficit.

The net effect of this continued production is an abnormal accumulation of precursor molecules and development of some type of porphyria. Each type of porphyria corresponds with a specific enzyme defect and an accumulation of the associated precursor. Although there are eight steps in heme biosynthesis, there are only seven types of porphyrias; a defect in ALA synthase activity does not have a corresponding porphyria.

Enzymes involved in heme biosynthesis display subtle, tissue-specific variations; therefore, heme biosynthesis may be impeded in the liver, but normal in the immature red blood cells, or vice versa. Incidence of porphyria

varies widely between types and occasionally by geographic location. Although certain porphyrias are more common than others, their greater frequency is only relative to other types. All porphyrias are considered to be rare disorders.

In the past, the porphyrias were divided into two general categories based on the location of the porphyrin production. Porphyrias affecting heme biosynthesis in the liver were referred to as hepatic porphyrias. Porphyrias that affect heme biosynthesis in immature red blood cells were referred to as erythropoietic porphyries. (Erythropoiesis is the process through which red blood cells are produced.) As of 2001, porphyrias are usually grouped into acute and non-acute types. Acute porphyrias produce severe attacks of **pain** and neurological effects. Non-acute porphyrias present as chronic diseases.

The acute porphyrias, and the heme biosynthesis steps at which enzyme defects occur, are:

- ALA dehydratase deficiency porphyria (step 2). This porphyria type is very rare. The inheritance pattern appears to be autosomal recessive. In autosomal recessively inherited disorders, a child must inherit two defective genes, one from each parent. A parent with only one gene for an autosomal recessive disorder does not display symptoms of the disease.
- Acute intermittent porphyria (step 3). Acute intermittent porphyria (AIP) is also known as Swedish porphyria, pyrroloporphyria, and intermittent acute porphyria. AIP is inherited as an autosomal dominant trait, which means that only one copy of the defective gene needs to be present for the disorder to occur. Simply inheriting this gene, however, does not necessarily mean that a child will develop the disease. Approximately five to 10 per 100,000 children in the United States carry a gene for AIP, but only 10 percent of these people, mostly teenage or older, ever develop symptoms of the disease.
- Hereditary coproporphyria (step 6). Hereditary coproporphyria (HCP) is inherited in an autosomal dominant manner. As with all porphyrias, it is an uncommon ailment. By 1977, only 111 cases of HCP were recorded; in Denmark, the estimated incidence is two in one million people.
- Variegate porphyria (step 7). Variegate porphyria (VP) is also known as porphyria variegata, protocoproporphyria, South African genetic porphyria, and Royal malady (supposedly King George III of England and Mary, Queen of Scots, suffered from VP). VP is inherited in an autosomal dominant manner and is especially prominent in South Africans of Dutch descent. Among that population, the incidence is approximately three in 1,000 persons. It is estimated that there are 10,000 cases of VP in South Africa. Interestingly, it appears that the affected South Africans are descendants of two Dutch settlers who came to South Africa in 1680. Among other populations, the incidence of VP is estimated to be one to two cases per 100,000 persons.

The non-acute porphyrias, and the steps of heme biosynthesis at which they occur, are:

- Congenital erythropoietic porphyria (step 4). Congenital erythropoietic porphyria (CEP) is also called Gunther's disease, erythropoietic porphyria, congenital porphyria, congenital hematoporphyria, and erythropoietic uroporphyria. CEP is inherited in an autosomal recessive manner. It is a rare disease, estimated to affect fewer than one in one million people. Onset of dramatic symptoms usually occurs in infancy, but may hold off until adulthood.
- Porphyria cutanea tarda (step 5). Porphyria cutanea tarda (PCT) is also called symptomatic porphyria, porphyria cutanea symptomatica, and idiosyncratic porphyria. PCT may be acquired, typically as a result of disease (especially hepatitis C), drug or alcohol use, or exposure to certain poisons. PCT may also be inherited as an autosomal dominant disorder, however most people remain latent—that is, symptoms never develop. PCT is the most common of the porphyrias, but the incidence of PCT is not well defined. However, PCT does not typically develop in children.
- Hepatoerythopoietic porphyria (step 5). Hepatoerythopoietic porphyria (HEP) affects heme biosynthesis in both the liver and the bone marrow. HEP results from a defect in uroporphyrinogen decarboxylase activity (step 5), and is caused by defects in the same gene as PCT. Disease symptoms, however, strongly resemble congenital erythropoietic porphyria. HEP seems to be inherited in an autosomal recessive manner.
- Erythropoietic protoporphyria (step 8). Also known as protoporphyria and erythrohepatic protoporphyria, erythropoietic protoporphyria (EPP) is more common than CEP; more than 300 cases have been reported. In these cases, onset of symptoms typically occurred in childhood.

Causes and symptoms

General characteristics

The underlying cause of all porphyrias is a defective enzyme important to the heme biosynthesis pathway.

Porphyrias are inheritable conditions. In virtually all cases of porphyria, an inherited factor causes the enzyme's defect. An environmental trigger—such as diet, drugs, or sun exposure—may be necessary before any symptoms develop. In many cases, symptoms do not develop. These asymptomatic individuals may be completely unaware that they have a gene for porphyria.

All of the hepatic porphyrias—except porphyria cutanea tarda—follow a pattern of acute attacks separated by periods during which no symptoms are present. For this reason, this group is often referred to as the acute porphyrias. The erythropoietic porphyrias and porphyria cutanea tarda do not follow this pattern and are considered to be chronic conditions.

The specific symptoms of each porphyria vary based on which enzyme is affected and whether that enzyme occurs in the liver or in the bone marrow. The severity of symptoms can vary widely, even within the same type of porphyria. If the porphyria becomes symptomatic, the common factor between all types is an abnormal accumulation of protoporphyrins or porphyrin.

ALA dehydratase porphyria (ADP)

ADP is characterized by a deficiency of ALA dehydratase. ADP is caused by mutations in the delta-aminolevulinate dehydratase gene (ALAD) at 9q34. Being located at 9q34 means that it is on the long arm (q) of chromosome 9 in the 34 region. Of the few cases on record, the prominent symptoms are **vomiting**, pain in the abdomen, arms, and legs, and neuropathy. (Neuropathy refers to nerve damage that can cause pain, **numbness**, or paralysis.) The nerve damage associated with ADP could cause breathing impairment or lead to weakness or paralysis of the arms and legs.

Acute intermittent porphyria (AIP)

AIP is caused by a deficiency of porphobilinogen deaminase, which occurs due to mutations in the hydroxymethylbilane synthase gene (HMBS) located at 11q23.3. Symptoms of AIP usually do not occur unless a person with the deficiency encounters a trigger substance. Trigger substances can include hormones (for example **oral contraceptives**, **menstruation**, pregnancy), drugs, and dietary factors. Most people with this deficiency never develop symptoms.

Attacks occur after **puberty** and commonly feature severe abdominal pain, **nausea**, vomiting, and **constipation**. Muscle weakness and pain in the back, arms, and legs are also typical symptoms. During an attack, the urine is a deep reddish color. The central nervous system may also be involved. Possible psychological symptoms include hallucinations, confusion, seizures, and mood changes.

Congenital erythropoietic porphyria (CEP)

CEP is caused by a deficiency of uroporphyrinogen III cosynthase due to mutations in the uroporphyrinogen III cosynthase gene (UROS) located at 10q25.2-q26.3. Symptoms are often apparent in infancy and include reddish urine and possibly an enlarged spleen. The skin is unusually sensitive to light and blisters easily if exposed to sunlight. (Sunlight induces protoporphyrin changes in the plasma and skin. These altered protoporphyrin molecules can cause skin damage.) Increased hair growth is common. Damage from recurrent blistering and associated skin infections can be severe. In some cases facial features and fingers may be lost to recurrent damage and infection. Deposits of protoporphyrins can sometimes lead to red staining of the teeth and bones.

Porphyria cutanea tarda (PCT)

PCT is caused by deficient uroporphyrinogen decarboxylase. PCT is caused by mutations in the uroporphyrinogen decarboxylase gene (UROD) located at 1p34. PCT may occur as an acquired or an inherited condition. The acquired form usually does not appear until adulthood. The inherited form may appear in childhood, but often demonstrates no symptoms. Early symptoms include blistering on the hands, face, and arms following minor injuries or exposure to sunlight. Lightening or darkening of the skin may occur along with increased hair growth or loss of hair. Liver function is abnormal but the signs are mild.

Hepatoerythopoietic porphyria (HEP)

HEP is linked to a deficiency of uroporphyrinogen decarboxylase in both the liver and the bone marrow. HEP is an autosomal recessive disease caused by mutations in the gene responsible for PCT, the uroporphyrinogen decarboxylase gene (UROD), located at 1p34. The gene is shared, but the mutations, inheritance, and specific symptoms of these two diseases are different. The symptoms of HEP resemble those of CEP.

Hereditary coproporphyria (HCP)

HCP is similar to AIP, but the symptoms are typically milder. HCP is caused by a deficiency of coproporphyrinogen oxidase due to mutations in a gene by the same name at 3q12. The greatest difference between HCP and AIP is that people with HCP may have some skin sensitivity to sunlight. However, extensive damage to the skin is rarely seen.

Variegate porphyria (VP)

VP is caused by a deficiency of protoporphyrinogen oxidase. There is scientific evidence that VP is caused by mutation in the gene for protoporphyrinogen oxidase located at 1q22. Like AIP, symptoms of VP occur only during attacks. Major symptoms of this type of porphyria include neurological problems and sensitivity to light. Areas of the skin that are exposed to sunlight are susceptible to burning, blistering, and scarring.

Erythropoietic protoporphyria (EPP)

Owing to deficient ferrochelatase, the last step in the heme biosynthesis pathway—the insertion of an iron atom into a porphyrin molecule—cannot be completed. This enzyme deficiency is caused by mutations in the ferrochelatase gene (FECH) located at 18q21.3. The major symptoms of this disorder are related to sensitivity to light—including both artificial and natural light sources. Following exposure to light, a child with EPP experiences burning, **itching**, swelling, and reddening of the skin. Blistering and scarring may occur but are neither common nor severe. EPP is associated with increased risks for gallstones and liver complications. Symptoms can appear in childhood and tend to be more severe during the summer when exposure to sunlight is more likely.

Diagnosis

Depending on the array of symptoms a child may exhibit, the possibility of porphyria may not immediately come to a physician's mind. In the absence of a **family** history of porphyria, non-specific symptoms, such as abdominal pain and vomiting, may be attributed to other disorders. Neurological symptoms, including confusion and hallucinations, can lead to an initial suspicion of psychiatric disease. Diagnosis is more easily accomplished in cases in which non-specific symptoms appear in combination with symptoms more specific to porphyria, like neuropathy, sensitivity to sunlight, or certain other manifestations. Certain symptoms, such as urine the color of port wine, are hallmark signs very specific to porphyria. DNA analysis is not yet of routine diagnostic value.

A common initial test measures protoporphyrins in the urine. However, if skin sensitivity to light is a symptom, a blood plasma test is indicated. If these tests reveal abnormal levels of protoporphyrins, further tests are done to measure heme precursor levels in red blood cells and the stool. The presence and estimated quantity of porphyrin and protoporphyrins in biological samples are easily detected using spectrofluorometric testing. Spectrofluorometric testing uses a spectrofluorometer that directs light of a specific strength at a fluid sample. The porphyrins and protoporphyrins in the sample absorb the light energy and fluoresce, or glow. The spectrofluorometer detects and measures fluorescence, which indicates the amount of porphyrins and protoporphyrins in the sample.

Whether heme precursors occur in the blood, urine, or stool gives some indication of the type of porphyria, but more detailed biochemical testing is required to determine their exact identity. Making this determination yields a strong indicator of which enzyme in the heme biosynthesis pathway is defective; which, in turn, allows a diagnosis of the particular type of porphyria.

Biochemical tests rely on the color, chemical properties, and other unique features of each heme precursor. For example, a screening test for acute intermittent porphyria (AIP) is the Watson-Schwartz test. In this test, a special dye is added to a urine sample. If one of two heme precursors—porphobilinogen or urobilinogen—is present, the sample turns pink or red. Further testing is necessary to determine whether the precursor present is porphobilinogen or urobilinogen—only porphobilinogen is indicative of AIP.

Other biochemical tests rely on the fact that heme precursors become less soluble in water (able to be dissolved in water) as they progress further through the heme biosynthesis pathway. For example, to determine whether the Watson-Schwartz urine test is positive for porphobilinogen or urobilinogen, chloroform is added to the test tube. Chloroform is a water-insoluble substance. Even after vigorous mixing, the water and chloroform separate into two distinct layers. Urobilinogen is slightly insoluble in water, while porphobilinogen tends to be water-soluble. The porphobilinogen mixes more readily in water than chloroform, so if the water layer is pink (from the dye added to the urine sample), that indicates the presence of porphobilinogen, and a diagnosis of AIP is probable.

As a final test, measuring specific enzymes and their activities may be done for some types of porphyrias; however, such tests are not done as a screening method. Certain enzymes, such as porphobilinogen deaminase (the defective enzyme in AIP), can be easily extracted from red blood cells; other enzymes, however, are less readily collected or tested. Basically, an enzyme test involves adding a certain amount of the enzyme to a test tube that contains the precursor it is supposed to modify. Both the production of modified precursor and the rate at which it appears can be measured using laboratory equipment. If a modified precursor is produced, the test indicates that the enzyme is doing its job. The rate at which the modified precursor is produced can be

compared to a standard to measure the efficiency of the enzyme.

Treatment

Treatment for porphyria revolves around avoiding acute attacks, limiting potential effects, and treating symptoms. Treatment options vary depending on the specific type of porphyria diagnosed. Gene therapy has been successful for both CEP and EPP. In the future, scientists expect development of gene therapy for the remaining porphyrias. Given the rarity of ALA dehydratase porphyria, definitive treatment guidelines for this rare type have not been developed.

Acute intermittent porphyria, hereditary coproporphyria, and variegate porphyria

Treatment for acute intermittent porphyria, hereditary coproporphyria, and variegate porphyria follows the same basic regime. A child who has been diagnosed with one of these porphyrias can prevent most attacks by avoiding precipitating factors, such as certain drugs that have been identified as triggers for acute porphyria attacks. Individuals must maintain adequate **nutrition**, particularly with respect to carbohydrates. In some cases, an attack can be stopped by increasing carbohydrate consumption or by receiving carbohydrates intravenously.

When attacks occur prompt medical attention is necessary. Pain is usually severe, and narcotic **analgesics** are the best option for relief. Phenothiazines can be used to counter nausea, vomiting, and **anxiety**, and chloral hydrate or diazepam is useful for sedation or to induce **sleep**. Hematin, a drug administered intravenously, may be used to halt an attack. Hematin seems to work by signaling the pathway of heme biosynthesis to slow production of precursors. Older girls, who tend to develop symptoms more frequently than boys owing to hormonal fluctuations, may find ovulation-inhibiting hormone therapy to be helpful.

Gene therapy is a possible future treatment for these porphyrias. An experimental animal model of AIP has been developed and research is in progress.

Congenital erythropoietic porphyria

The key points of congenital erythropoietic porphyria treatment are avoiding exposure to sunlight and prevention of skin trauma or skin infection. Liberal use of **sunscreens** and consumption of beta-carotene supplements can provide some protection from sun-induced damage. Medical treatments such as removing the spleen or administering transfusions of red blood cells can create short-term benefits, but these treatments do not offer a cure. Remission can sometimes be achieved after treatment with oral doses of activated charcoal. Severely affected patients may be offered bone marrow transplantation which appears to confer long-term benefit.

Porphyria cutanea tarda

As with other porphyrias, the first line of defense is avoidance of factors, especially alcohol, that could bring about symptoms. Regular blood withdrawal is a proven therapy for pushing symptoms into remission. If an individual is anemic or cannot have blood drawn for other reasons, chloroquine therapy may be used.

Erythropoietic protoporphyria

Avoiding sunlight, using sunscreens, and taking beta-carotene supplements are typical treatment options for erythropoietic protoporphyria. The drug cholestyramine may reduce the skin's sensitivity to sunlight as well as the accumulated heme precursors in the liver. Liver transplantation has been used in cases of liver failure, but it has not effected a long-term cure of the porphyria.

Alternative treatment

Acute porphyria attacks can be life-threatening events, so attempts at self-treatment can be dangerous. Alternative treatments can be useful adjuncts to conventional therapy. For example, some people may find relief for the pain associated with acute intermittent porphyria, hereditary coproporphyria, or variegate porphyria through acupuncture or hypnosis. Relaxation techniques, such as **yoga** or meditation, may also prove helpful in **pain management**.

Prognosis

Even when porphyria is inherited, symptom development depends on a variety of factors. In the majority of cases, a person remains asymptomatic throughout life. About 1 percent of acute attacks can be fatal. Other symptoms may be associated with temporarily debilitating or permanently disfiguring consequences. Measures to avoid these consequences are not always successful, regardless of how diligently they are pursued. Although pregnancy has been known to trigger porphyria attacks, dangers associated with pregnancy as not as great as was once thought.

Prevention

For the most part, the porphyrias are attributable to inherited genes; such inheritance cannot be prevented.

KEY TERMS

Autosomal dominant—A pattern of inheritance in which only one of the two copies of an autosomal gene must be abnormal for a genetic condition or disease to occur. An autosomal gene is a gene that is located on one of the autosomes or non-sex chromosomes. A person with an autosomal dominant disorder has a 50 percent chance of passing it to each of their offspring.

Autosomal recessive—A pattern of inheritance in which both copies of an autosomal gene must be abnormal for a genetic condition or disease to occur. An autosomal gene is a gene that is located on one of the autosomes or non-sex chromosomes. When both parents have one abnormal copy of the same gene, they have a 25 percent chance with each pregnancy that their offspring will have the disorder.

Biosynthesis—The manufacture of materials in a biological system.

Bone marrow—The spongy tissue inside the large bones in the body that is responsible for making the red blood cells, most white blood cells, and platelets.

Chromosome—A microscopic thread-like structure found within each cell of the human body and consisting of a complex of proteins and DNA. Humans have 46 chromosomes arranged into 23 pairs. Chromosomes contain the genetic information necessary to direct the development and functioning of all cells and systems in the body. They pass on hereditary traits from parents to child (like eye color) and determine whether the child will be male or female.

Enzyme—A protein that catalyzes a biochemical reaction without changing its own structure or function.

Erythropoiesis—The process through which new red blood cells are created; it begins in the bone marrow.

Erythropoietic—Referring to the creation of new red blood cells.

Gene—A building block of inheritance, which contains the instructions for the production of a particular protein, and is made up of a molecular sequence found on a section of DNA. Each gene is found on a precise location on a chromosome.

Hematin—A drug administered intravenously to halt an acute porphyria attack. It causes heme biosynthesis to decrease, preventing the further accumulation of heme precursors.

Heme—The iron-containing molecule in hemoglobin that serves as the site for oxygen binding.

Hemoglobin—An iron-containing pigment of red blood cells composed of four amino acid chains (alpha, beta, gamma, delta) that delivers oxygen from the lungs to the cells of the body and carries carbon dioxide from the cells to the lungs.

Hepatic—Refers to the liver.

Neuropathy—A disease or abnormality of the peripheral nerves (the nerves outside the brain and spinal cord). Major symptoms include weakness, numbness, paralysis, or pain in the affected area.

Porphyrin—An organic compound found in living things that founds the foundation structure for hemoglobin, chlorophyll, and other respiratory pigments. In humans, porphyrins combine with iron to form hemes.

Protoporphyrin—A kind of porphyrin that links with iron to form the heme of hemoglobin.

However, symptoms can be limited or prevented by avoiding factors that trigger symptom development.

Children with a family history of an acute porphyria should be screened for the disease. Even if symptoms are absent, it is useful to know about the presence of the gene to assess the risks of developing the associated porphyria. This knowledge also reveals whether a person's offspring may be at risk. Prenatal testing for certain porphyrias is possible. Prenatal diagnosis of congenital erythropoietic porphyria has been successfully accomplished. Any prenatal tests, however, would not indicate whether a child would develop porphyria symptoms; only that the potential is there.

Parental concerns

Many children with porphyria do not have symptoms. Many acute attacks can be prevented by knowing what causes the attacks, and avoiding those things in the diet or environment that result in acute attacks.

When to call a doctor

Notify a doctor if the child appears to have an acute attack. Some signs and symptoms of an acute attack are: pain, red, burning or blistering skin, red urine, neurological changes, or psychological changes.

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American Porphyria Foundation. PO Box 22712, Houston, TX 77227. (713) 266-9617. <www.porphyriafoundation.com/>.

OTHER

Gene Clinics. Available online at <www.geneclinics.org>.

National Institute of Diabetes & Digestive & Kidney Diseases. Available online at <www.niddk.nih.gov>.

Online Mendelian Inheritance in Man (OMIM). Available online at <www3.ncbi.nlm.nih.gov/Omim>.

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Port-wine stain *see* **Birthmarks**

Post-concussion syndrome

Definition

Post-concussion syndrome (PCS) is a common but controversial disorder with a variety of symptoms including, but not limited to, **headache**, **dizziness**, fatigue, and personality changes.

Description

Post-concussion syndrome occurs in some patients after a **concussion**. A concussion is a form of mild traumatic **head injury**. Often a concussion involves loss of consciousness for a brief period, but it is possible to have a concussion without ever losing consciousness. There are many different ways of defining PCS, but it is usually considered present if three or more symptoms (such as insomnia, headache, and dizziness) are present for at least three months. PCS is a controversial syndrome, because multiple studies have resulted in conflicting findings. Also, some experts believe that many of the symptoms are largely psychological, because usually no neurological causes for the symptoms can be found. Other experts, however, maintain that just because no one has been able to pinpoint neurological causes does not mean they do not exist.

Demographics

PCS occurs in approximately 23–93 percent of individuals with mild to severe head injuries. It is estimated that a neurologist (a physician who specializes in nerve and brain disorders) sees five patients with PCS per month. There is no accurate correlation between the severity of injury and the development of PCS symptoms, since signs of the disorder can occur in someone who was just dazed by an injury. Some studies suggest that PCS symptoms occur at a higher rate in individuals who were unconscious after trauma. Females may be more likely to develop PCS than males.

Causes and symptoms

PCS is most commonly caused by a minor head injury called a concussion. Many patients who have experienced minor head injury develop PCS with distinct symptoms. They may report problems with concentration, short and medium term memory, and abstract thinking. Additionally, patients may develop dizziness, irritability, fatigue, and personality changes. It is not known what causes these symptoms. No studies have been able to deduce definitively any kind of neurological basis for the syndrome.

When to call the doctor

If a child is displaying the signs and symptoms of post-concussion syndrome, especially if known to have recently experienced a head injury, a doctor should be consulted.

Diagnosis

There are no specific or reliable tests to diagnose PCS. A doctor will do a physical examination. A neuropsychologist can perform an in-depth neuropsychologic **assessment** that can determine presence or absence and extent of impairment. Sometimes tests used to measure memory or cognitive capacity will be performed. Doctors may recommend seeing a psychologist to determine if depression or **anxiety** is present. **Magnetic resonance imaging** (MRI) or **computed tomography** (CT) scans

KEY TERMS

Neuropsychologist—A clinical psychologist who specializes in assessing psychological status caused by a brain disorder.

may be done to ensure that a lesion or bleeding is not responsible for the symptoms.

Treatment

Treatment for PCS can be extensive. Medications for headache and **pain** may be indicated (**analgesics** and muscle relaxants). **Antidepressants** may be given to improve insomnia, irritability, anxiety, or depression. Pain control could be achieved with acupuncture, nerve blocks, or transcutaneous electrical nerve stimulation (TENS, electrical stimulation of muscle groups). It is important for clinicians to educate caretakers and to provide referrals for **family therapy** and cognitive rehabilitation for the affected child. The overall aim of treatment for PCS is to allow the child to return to school and to the activities that he or she participated in before the traumatic incident.

Prognosis

The overall outcome is difficult to assess. Most individuals who have PCS recover fully, although if recovery has not occurred in one year it is less likely that it will ever occur. Patient recovery is determined by cognitive function changes, subjective symptoms, and return to school or work. Cases of PCS can be a strain and threaten **family** stability. There may be compensation and litigation claims, which are often stressful and aggravate symptoms.

Prevention

The only way to prevent post-concussion syndrome is to prevent the original concussion. Wearing a helmet while riding a bike, rollerblading, or skateboarding can reduce the risk of head injury. Putting children in age-appropriate **safety** seats in the car can also help to prevent head trauma in the event of an automobile accident.

Parental concerns

Post-concussion syndrome can be very frustrating, because the symptoms are persistent and can affect the child's ability to perform in school or other activities. Counseling is thought to help the patient and family deal with the incapacitation and develop coping strategies. The best way to prevent PCS is to make sure that children avoid situations where they are likely to injure themselves.

See also Concussion.

Resources

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ORGANIZATIONS

*Brain Injury of America*8201 Greensboro Drive, Suite 611, McLean, VA 22102. Telephone: (703) 761-0750. Family Helpline: 1-800-444-6443. familyhelpline@biausa.org. <www.biausa.org>

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Postural drainage *see* **Chest physical therapy**

Potty training *see* **Toilet training**

Prader-Willi syndrome

Definition

Prader-Willi syndrome (PWS) is a genetic condition caused by the absence of chromosomal material from chromosome 15. Characteristics of the syndrome include developmental delays, poor muscle tone, short stature, small hands and feet, incomplete sexual development, and unique facial features. Insatiable appetite is a classic feature of PWS. This uncontrollable appetite can lead to morbid **obesity** and behavior disturbances.

Description

The first patients with features of PWS were described by Dr. Prader, Dr. Willi, and Dr. Lambert in 1956. Since that time, the complex genetic basis of PWS has begun to be understood. Initially, scientists found that individuals with PWS have a portion of genetic material deleted (erased) from chromosome 15. In order to have PWS, the genetic material must be deleted from the chromosome 15 received from one's father. If the deletion is on the chromosome 15 inherited from one's mother, a different syndrome develops. This was an important discovery, for it demonstrated for the first time that the genes inherited from one's mother can be expressed differently than the genes inherited from one's father.

Over time, scientists realized that some individuals with PWS do not have genetic material deleted from chromosome 15. Further studies found that these patients inherit both copies of chromosome 15 from their mother, which is not typical. Normally, an individual receives one chromosome 15 from his or her father and one chromosome 15 from his or her mother. When a person receives both chromosomes from the same parent it is called "uniparental disomy." When a person receives both chromosomes from his or her mother, it is called "maternal uniparental disomy."

Scientists are still discovering other causes of PWS. A small number of patients with PWS have a change (mutation) in the genetic material on the chromosome 15 inherited from their father. This mutation prevents certain genes on chromosome 15 from working properly. PWS develops when these genes do not work normally.

Newborns with PWS generally have poor muscle tone, (**hypotonia**) and do not feed well. This can lead to poor weight gain and **failure to thrive**. Genitalia can be smaller than normal, and a male with PWS may have undescended testicles. Hands and feet are also typically smaller than normal. Some patients with PWS have unique and subtle facial characteristics that are detectable only by physicians.

As children with PWS age, development is typically slower than normal. Developmental milestones, such as **crawling**, walking and talking occur later than usual. **Developmental delay** continues into adulthood for approximately 50 percent of individuals with PWS. At about one to two years of age, children with PWS develop an uncontrollable, insatiable appetite. These children, if not controlled, will eat until they suffer from life-threatening obesity, including respiratory failure with hypoxia (low blood oxygen levels), cor pulmonale (right-sided heart failure), and death. The desire to eat can also lead to significant behavior problems.

The symptoms and features of PWS require lifelong support and care. If food intake is strictly monitored and various therapies provided, individuals with PWS have a normal life expectancy.

Prader-Willi syndrome is also referred to as cryptochidism-dwarfism-subnormal mentality syndrome, Willi-Prader syndrome, Labhart-Willi syndrome, Prader-Labhart-Will Fancone syndrome, and hypotonia-hypomentia-hypogonadism-obesity syndrome.

Demographics

PWS affects approximately one in 12,000 to 15,000 live births. It is the most common genetic cause of life-threatening obesity. It affects both males and females and can be seen in all races and ethnic groups.

Causes and symptoms

Human beings have 46 chromosomes in the cells of their body. Chromosomes contain genes that regulate the function and development of the body. An individual's chromosomes are inherited from his or her parents. Each parent normally gives a child 23 chromosomes. A child receives 23 chromosomes from the egg and 23 chromosomes from the sperm.

The 46 chromosomes in the human body are divided into pairs based on their physical characteristics. Each pair is assigned a number or a letter. When viewed under a microscope, chromosomes within the same pair appear identical because they contain the same genes.

Most chromosomes have a constriction near the center called the centromere. The centromere separates the chromosome into long and short arms. The short arm of a chromosome is called the p arm; the long arm and is called the q arm.

Chromosomes in the same pair contain the same genes. However, some genes work differently depending on if they were inherited from the egg or the sperm. Sometimes, genes are silenced when inherited from the mother. Other times, genes are silenced when inherited from the father. When genes in a certain region on a chromosome are silenced, they are said to be imprinted. Imprinting is a normal process that does not typically cause disease. However, if normal imprinting is disrupted, a genetic disease can develop.

Individuals have two complete copies of chromosome 15. One chromosome 15 is inherited from the

mother, or is maternal in origin. The other chromosome 15 is inherited from the father, or is paternal in origin.

Chromosome 15 contains many different genes. There are several genes found on the q arm of chromosome 15 that are imprinted. A gene called SNPRN is an example of one of these genes. It is normally imprinted, or silenced, if inherited from the mother. The imprinting of this group of maternal genes does not typically cause disease. The genes in this region should not be imprinted if paternal in origin. Normal development depends on these paternal genes being present and active. If these genes are deleted, not inherited, or incorrectly imprinted, PWS develops.

Deletion in the paternally contributed chromosome 15

Seventy percent of the cases of PWS are caused when a piece of material is deleted, or erased, from the paternal chromosome 15. This deletion occurs in a specific region on the q arm of chromosome 15. The piece of chromosomal material that is deleted contains genes that must be present for normal development. These paternal genes must be working normally, because the same genes on the chromosome 15 inherited from the mother are imprinted. When these paternal genes are missing, the brain and other parts of the body do not develop as expected. This is what causes the symptoms associated with PWS.

In 99 percent of the cases of PWS, the deletion is sporadic. This means that it happens randomly, there is not an apparent cause, and the condition is not inherited. If a child has PWS due to a sporadic deletion in the paternal chromosome 15, the chance the parents could have another child with PWS is less than 1 percent. In fewer than 1 percent of the cases of PWS there is a chromosomal rearrangement in the **family** that causes the deletion. This chromosomal rearrangement is called translocation. If a parent has a translocation the risk of having a child with PWS is higher than 1 percent.

Maternal uniparental disomy

PWS can also develop if a child receives both chromosome 15s from his or her mother. This is seen in approximately 25 percent of the cases of PWS. Maternal uniparental disomy for chromosome 15 leads to PWS because the genes on chromosome 15 that should have been inherited from the father are missing, and the genes on both the chromosome 15s inherited from the mother are imprinted.

PWS caused by maternal uniparental is sporadic. This means that it occurs randomly and there is not an apparent cause. If a child has PWS due to maternal uniparental disomy, the chance the parents could have another child with PWS is less than 1 percent.

Error in imprinting process than renders paternal contribution non-functional

Approximately 2–5 percent of patients with PWS have a change (mutation) in a gene located on the q arm of chromosome 15. This mutation leads to incorrect imprinting. This mutation causes genes inherited from the father to be imprinted or silenced, which should not normally be imprinted. If a child has PWS due to a mutation that changes imprinting, the chance the parents could have another child with PWS is approximately 5 percent.

Signs of PWS can be seen at birth. Infants with PWS have weak muscle tone (hypotonia). This hypotonia causes problems with sucking and eating so that infants with PWS may initially have problems gaining weight. Consequently, some infants with PWS may be diagnosed with failure to thrive due to slow growth and development. Hypotonia may also During infancy, babies with PWS may also **sleep** more than normal and have problems controlling their temperature.

Some of the unique physical features associated with PWS can be seen during infancy. Genitalia (**hypogonadism**) that is smaller than normal is common. This may be more evident in males with PWS. Hands and feet may also be smaller than average. The unique facial features seen in some patients with PWS may be difficult to detect in infancy. These facial features are very mild and do not cause physical problems.

As early as six months, but more commonly at one to two years, a compulsive desire to eat develops. This uncontrollable appetite is a classic feature of PWS. Individuals with PWS lack the ability to feel full or satiated because of a flaw in the hypothalamus part of their brain, which normally registers feelings of hunger and satiety. Over-eating (hyperpahgia), a lack of a desire to **exercise**, and decreased calorie utilization (typically 1,000–1,200 calories per day for adults, due to low muscle mass and inactivity) places individuals with PWS at high risk for severe obesity. Obesity-related problems include hypoventilation, hyertension, right-sided heart failure, cellulitis, and skin problems with fat folds. Some individuals with PWS may also have a reduced ability to vomit.

Behavior problems are a common feature of PWS. Although infants and young children are typically happy and loving and exhibit few behavior problems, most older children and adults do have difficulties with behavior regulation, such as difficulties with transistions and unanticipated changes. Onset of behavioral problems

usually coincides with the onset of the compulsive eating. Difficulties peak in **adolescence** or early adulthood. Reported problems include obsessive/compulsive behaviors, depression, temper **tantrums** and violent outbursts, and tendencies to be argumentative, oppositional, rigid, manipulative, possessive, and stubborn. Individuals with PWS may also pick their own skin (skin picking). This unusual behavior may be due to a reduced **pain** threshold.

IQs range from 40 to 105, with an average of 70. Those with normal IQs typically have learning disabilities. Problem areas may include attention, short-term auditory memory, and abstract thinking. Common strengths include long-term memory, reading ability, and receptive language.

Puberty may occur early or late, but it is usually incomplete. In addition to the effects on sexual development and fertility, individuals do not undergo the normal adolescent growth spurt and may be short as adults. Muscles often remain underdeveloped and body fat is increased.

When to call the doctor

Parents should call a doctor if they notice symptoms that are characteristic of PWS.

Diagnosis

During infancy the diagnosis of PWS may be suspected if poor muscle tone, feeding problems, small genitalia, or the unique facial features are present. If an infant has these features, testing for PWS should be performed. This testing should also be offered to children and adults who display features commonly seen in PWS (developmental delays, uncontrollable appetite, small genitalia, etc.). There are several different genetic tests that can detect PWS. All of these tests can be performed usnig a blood sample.

Methylation testing detects 99 percent of the cases of PWS. Methylation testing can detect the absence of the paternal genes that should be normally active on chromosome 15. Although methylation testing can accurately diagnose PWS, it can not determine if the PWS is caused by a deletion, maternal uniparental disomy, or a mutation that disrupts imprinting. This information is important for genetic counseling. Therefore, additional testing should be performed.

Chromosome analysis can determine if the PWS is the result of a deletion in the q arm of chromosome 15. Chromosome analysis, also called karyotyping, involves staining the chromosomes and examining them under a microscope. In some cases the deletion of material from chromosome 15 can be easily seen. In other cases, further testing must be performed. FISH (fluorescence in-situ hybridization) is a special technique that detects small deletions that cause PWS.

More specialized DNA testing is required to detect maternal uniparental disomy or a mutation that disrupts imprinting. This DNA testing identifies unique DNA patterns in the mother and father. The unique DNA patterns are then compared with the DNA from the child with PWS.

PWS can be detected before birth if the mother undergoes amniocentesis testing or chorionic villus sampling (CVS). This testing is only recommended if the mother or father is known to have a chromosome rearrangement, or if they already have a child with PWS syndrome.

Treatment

There is currently not a cure for PWS. Treatment during infancy includes therapies to improve muscle tone. Some infants with PWS also require special nipples or tube feeding to improve weight gain.

Growth hormone therapy has been shown to improve the poor muscle tone and reduced height typically associated with PWS. Hypogonadism may be corrected at puberty with hormone replacement. Skin picking is best managed by ignoring the behavior, treating and bandaging sores, and providing substitute activities for the hands. Other behavioral problems can be managed through daily routines and structure, firm rules and limits, "time outs," positive rewards, and the use of psychotropic drugs.

Special education may be helpful in treating developmental delays and behavior problems. Individuals with PWS typically excel in highly structured environments. Physical and occupational therapies promote skill development and proper function. Exercise and sport activities should be encouraged, with adaptations made as necessary. Proficiency with jigsaw puzzles have been frequently reported, reflecting strong visual-perceptual skills. The need for speech therapy, due to speech difficulties caused by hypotonia, should be assessed. Sign language and picture communication boards can be used to reduce frustration and to aid communication. Products to increase saliva may help articulation problems. Social skills training can improve pragmatic language use. Verbal ability often becomes an area of strength for children with PWS.

KEY TERMS

Centromere—The constricted region of a chromosome. It performs certain functions during cell division.

Deletion—The absence of genetic material that is normally found in a chromosome. Often, the genetic material is missing due to an error in replication of an egg or sperm cell.

DNA—Deoxyribonucleic acid; the genetic material in cells that holds the inherited instructions for growth, development, and cellular functioning.

Fluorescence in situ hybridization (FISH)—A technique for diagnosing genetic disorders before birth by analyzing cells obtained by amniocentesis with DNA probes.

Gene—A building block of inheritance, which contains the instructions for the production of a particular protein, and is made up of a molecular sequence found on a section of DNA. Each gene is found on a precise location on a chromosome.

Hyperphagia—Over-eating.

Hypotonia—Having reduced or diminished muscle tone or strength.

Imprinting—A process that silences a gene or group of genes. The genes are silenced depending on whether they are inherited through the egg or the sperm.

Maternal uniparental disomy—A chromosome abnormality in which both chromosomes in a pair are inherited from one's mother.

Methylation testing—DNA testing that detects if a gene is active or if it is imprinted.

Mutation—A permanent change in the genetic material that may alter a trait or characteristic of an individual, or manifest as disease. This change can be transmitted to offspring.

Translocation—The transfer of one part of a chromosome to another chromosome during cell division. A balanced translocation occurs when pieces from two different chromosomes exchange places without loss or gain of any chromosome material. An unbalanced translocation involves the unequal loss or gain of genetic information between two chromosomes.

Uniparental disomy—Chromosome abnormality in which both chromosomes in a pair are inherited from the same parent.

Nutritional concerns

Treatment and management of PWS during childhood, adolescence, and adulthood is typically focused on weight control. Strict control of food intake is vital to prevent severe obesity. In many cases food must be made inaccessible. This may involve unconventional measures such as locking the refrigerator or kitchen cabinets. A lifelong balanced restricted-calorie diet with vitamin and calcium supplementation and a regular exercise program (at least 30 minutes per day) are also suggested. The best meal and snack plan is one that the family or caregiver is able to aply routinely and consistently. Unfortunately, diet medications nor surgery have not been shown to significantly prevent obesity in PWS or eliminate the need for strict dieting and supervision around food.

Prognosis

With help, people with PWS can expect to accomplish many of the things their "normal" peers do: complete school, achieve in their outside areas of interest, be successfully employed, and even move away from their family home. They do, however, need a significant amount of support from their families and from school, work, and residential service providers to both achieve these goals and to avoid obesity and the serious health consequences that accompany it. Even those with IQs in the normal range need lifelong diet supervision and protection from food availability.

Although in the past many people with PWS died in adolescence or young adulthood, prevention of obesity can enable those with the syndrome to live a normal lifespan. New medications, including psychotropic drugs and synthetic growth hormone, are already improving the quality of life for some people with PWS. Ongoing research offers the hope of new discoveries that will enable people affected by this unusual condition to live more independent lives.

Prevention

PWS currently cannot be prevented. Genetic counseling is recommended for parents who may be at risk for having a child with PWS.

Parental concerns

While there is no medical prevention or cure, early diagnosis of Prader-Willi syndrome gives parents time to learn about and prepare for the challenges that lie ahead and to establish family routines that will support their child's diet and behavior needs. The constant need for food restriction and behavior management may be stressful for family members. It is also important for the parents to provide basic sex education to promote good health and to protect against abuse. Knowing the cause of their child's developmental delays can facilitate a family's access to important early intervention services and may help program staff identify areas of specific need or risk. Additionally, a diagnosis of PWS opens the doors to a network of information and support from professionals and other families who are dealing with the syndrome. Adolescents and adults with PWS can function well in group and supported living programs, if the necessary diet control and structured environment are provided. Employment in sheltered workshops and other highly structured and supervised settings is successful for many. However, residential and vocational providers must be fully informed regarding management of PWS.

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Alliance of Genetic Support Groups. 4301 Connecticut Ave. NW, Suite 404, Washington DC 20008. (202) 966-5557. Fax: (202) 966-8553. <www.geneticalliance.org>.

International Prader-Willi Syndrome Organization. <www.ipwsp.org>.

National Organization for Rare Disorders, Inc. P.O. Box 8923, New Fairfield, CT 06812. (800) 999-6673. <www.rarediseases.org>.

Prader-Willi Foundation. 223 Main Street, Port Washington, NY 11050. (800)253- 7993. <www.prader-willi.org>.

Prader-Willi Syndrome Association (USA). 5700 Midnight Pass Rd., Sarasota, FL 34242. (800) 926-4797. <www.pwsausa.org>.

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Precocious puberty

Definition

Precocious puberty is sexual development before the age of eight in girls, and age 10 in boys.

Description

Precocious **puberty** often begins before age eight in girls, triggering the development of breasts and hair under the arms and in the genital region. The onset of ovulation and **menstruation** also may occur. In boys, the condition triggers the development of a large penis and testicles, with spontaneous erections and the production of sperm. Hair grows on the face, under arms and in the pubic area, and **acne** may become a problem.

While the early onset of puberty may seem fairly benign, in fact it can cause problems when hormones trigger changes in the growth pattern, essentially halting growth before the child has reached normal adult height. Girls may never grow above 5 ft (152 cm) and boys often stop growing by about 5 ft 2 in (157 cm).

The abnormal growth patterns are not the only problem, however. Children with this condition look noticeably different than their peers, and may feel rejected by their friends and socially isolated. Adults may expect these children to act more maturely simply because they look so much older. As a result, many of these children,

especially boys, are much more aggressive than others their own age, leading to behavior problems both at home and at school.

Demographics

Not every child reaches puberty at the same time, but in most cases it is safe to predict that sexual development will begin at about age 11 in girls and 12 or 13 in boys. However, occasionally a child begins to develop sexually much earlier. Between four to eight times more common in girls than boys, precocious puberty occurs in one out of every 5,000–10,000 U.S. children.

Causes and symptoms

Puberty begins when the brain secretes a hormone that triggers the pituitary gland to release gonadotropins, which in turn stimulate the ovaries or testes to produce sex hormones. These sex hormones (especially estrogen in girls and testosterone in boys) are what causes the onset of sexual maturity.

The hormonal changes of precious puberty are normal—it is just that the whole process begins a few years too soon. Especially in girls, there is not usually any underlying problem that causes the process to begin too soon. However, some boys do inherit the condition; the responsible gene may be passed directly from father to son, or inherited indirectly from the maternal grandfather through the mother, who does not begin early puberty herself. This genetic condition in girls can be traced in only about one percent of cases.

In about 15 percent of cases, there is an underlying cause for the precocious puberty, and it is important to search for these causes. The condition may result from a benign tumor in the part of the brain that releases hormones. Less commonly, it may be caused by other types of brain tumors, central nervous system disorders, or adrenal gland problems.

When to call the doctor

A pediatrician should be consulted when girls under age eight start to show signs breast development and menstruation, or if boys under age 10 show enlarged genitals and body hair.

Diagnosis

Physical exams can reveal the development of sexual characteristics in a young child. Bone **x rays** can reveal bone age, and pelvic ultrasound may show an enlarged uterus and rule out ovarian or adrenal tumors.

KEY TERMS

Puberty—The point in development when the ability to reproduce begins. The gonads begin to function and secondary sexual characteristics begin to appear.

Blood tests can highlight higher-than-normal levels of hormones. MRI or CAT scans should be considered to rule out intracranial tumors.

Treatment

Treatment aims to halt or reverse sexual development so as to stop the accompanying rapid growth that will limit a child's height. There are two possible approaches: either treat the underlying condition (such as an ovarian or intracranial tumor) or change the hormonal balance to stop sexual development. It may not be possible to treat the underlying condition; for this reason, treatment is usually aimed at adjusting hormone levels.

There are several drugs that have been developed to do this:

- histrelin (Supprelin)
- nafarelin (Synarel)
- synthetic gonadotropin-releasing hormone agonist
- deslorelin
- ethylamide
- triptorelin
- leuprolide

Prognosis

Drug treatments can slow growth to 2–3 in (5–7.5 cm) a year, allowing these children to reach normal adult height, although the long-term effects are not known.

Prevention

There is no way to prevent precocious puberty.

Parental concerns

Support and appropriate treatment of any underlying medical disorders are appropriate for parents. The vast majority of children experiencing precocious puberty become lost in the crowd of their peers when their age

peers enter puberty. Counseling may be useful for both parents and affected child.

Resources

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Prematurity

Definition

The average length of a normal pregnancy is 40 weeks (280 days) from the date of conception. Infants born before 37 weeks gestation are considered premature and may be at risk for complications.

Description

More than one out of every ten infants born in the United States is born prematurely. Advances in medical technology have made it possible for infants born as young as 23 weeks gestational age (17 weeks premature) to survive. These premature infants, however, are at higher risk for death or serious complications, which include heart defects, respiratory problems, blindness, and brain damage.

Demographics

According to the March of Dimes Foundation, there were 480,812 births in the United States in 2002 that occurred before 37 weeks gestation. This number represents 12.1 percent of live births that year. In an average week, approximately 9,200 infants are born prematurely, and approximately 1,500 are born before 32 weeks gestation. Black infants have the highest prematurity rate with 17.6 percent of live births; Native American (12.9%); Hispanic infants (11.4%); white infants (10.7%); and Asian infants (10.2%). Mothers younger than 20 years of age or older than 35 years of age have higher rates of preterm delivery.

Causes and symptoms

The birth of a premature baby can be brought on by several different factors, including the following:

- premature labor
- placental abruption, in which the placenta detaches from the uterus
- placenta previa, in which the placenta grows too low in the uterus
- premature rupture of membranes, in which the amniotic sac is torn, causing the amniotic fluid to leak out
- incompetent cervix, in which the cervix opens too soon
- maternal toxemia or preeclampsia

Prematurity is much more common in pregnancy of multiples and for mothers who have a history of miscarriages or prior premature birth. Another identifiable cause of prematurity is drug abuse (e.g. cocaine) by the mother.

Infants born prematurely may experience major complications due to their low birth weight and the immaturity of their organ systems. Some of the common problems among premature infants are **jaundice** (yellow discoloration of the skin and whites of the eyes), apnea (a long pause in breathing), and inability to breast or bottle feed. Body temperature, blood pressure, and heart rate may be difficult to regulate in premature infants. The lungs, digestive system, and nervous system (including the brain) are underdeveloped in premature babies and are particularly vulnerable to complications.

Complications

Respiratory distress syndrome (RDS) is the most common problem in premature infants. Babies born too soon have immature lungs that have not developed surfactant, a protective film that helps air sacs in the lungs

to stay open. With RDS, breathing is rapid and the center of the chest and rib cage pull inward with each breath. Extra oxygen can be supplied to the infant through tubes that fit into the nostrils of the nose or by placing the baby under an oxygen hood. In more serious cases, the baby may have to have a breathing tube inserted and receive air from a respirator or ventilator. A surfactant drug can be given in some cases. Extra oxygen may be needed for a few days or weeks. Bronchopulmonary dysplasia is the development of scar tissue in the lungs and can occur in severe cases of RDS.

Necrotizing enterocolitis (NEC) is another complication of prematurity. In this condition, part of the baby's intestine is destroyed as a result of bacterial infection. In cases where only the innermost lining of the bowel dies, the infant's body can regenerate it over time; however, if the full thickness of a portion dies, it must be removed surgically and an opening (ostomy) must be made for the passage of wastes until the infant is healthy enough for the remaining ends to be sewn together. Because NEC is potentially fatal, doctors are quick to respond to its symptoms, which include lethargy, **vomiting**, a swollen and/or red abdomen, **fever**, and blood in the stool. Measures include taking the infant off mouth feedings and feeding him or her intravenously, administering **antibiotics**, and removing air and fluids from the digestive tract via a nasal tube. Approximately 70 percent of NEC cases can be successfully treated without surgery.

Intraventricular hemorrhage (IVH) is another serious complication of prematurity. It is a condition in which immature and fragile blood vessels within the brain burst and bleed into the hollow chambers (ventricles) normally reserved for cerebrospinal fluid and into the tissue surrounding them. Physicians grade the severity of IVH according to a scale of I through IV, with I being bleeding confined to a small area around the burst vessels and IV being an extensive collection of blood in the ventricles and in the brain tissue itself. Grades I and II are not uncommon, and the baby's body usually reabsorbs the blood with no ill effects. However, more severe IVH can result in **hydrocephalus**, a potentially fatal condition in which too much fluid collects in the ventricles, exerting increased pressure on the brain and causing the baby's head to expand abnormally. To drain fluid and relieve pressure on the brain, doctors either perform lumbar punctures, a procedure in which a needle is inserted into the spinal canal to drain fluid; install a reservoir, a tube that drains fluid from a ventricle and into an artificial chamber under or on top of the scalp; or install a ventricular shunt, a tube that drains fluid from the ventricles and into the abdomen, where it is reabsorbed by the body. Infants who are at high risk for IVH usually have an ultrasound taken of their brain in the first week after birth, followed by others if bleeding is detected. IVH cannot be prevented; however, close monitoring can ensure that procedures to reduce fluid in the brain are implemented quickly to minimize possible damage.

Apnea of prematurity is a condition in which the infant stops breathing for periods lasting up to 20 seconds. It is often associated with a slowing of the heart rate. The baby may become pale, or the skin color may change to a blue or purplish hue. Apnea occurs most commonly when the infant is asleep. Infants with serious apnea may need medications to stimulate breathing or oxygen through a tube inserted in the nose. Some infants may be placed on a ventilator or respirator with a breathing tube inserted into the airway. As the baby gets older, and the lungs and brain tissues mature, the breathing usually becomes more regular. A group of researchers in Cleveland reported in 2003, however, that children who were born prematurely are three to five times more likely to develop sleep-disordered breathing by age 10 than children who were full-term babies.

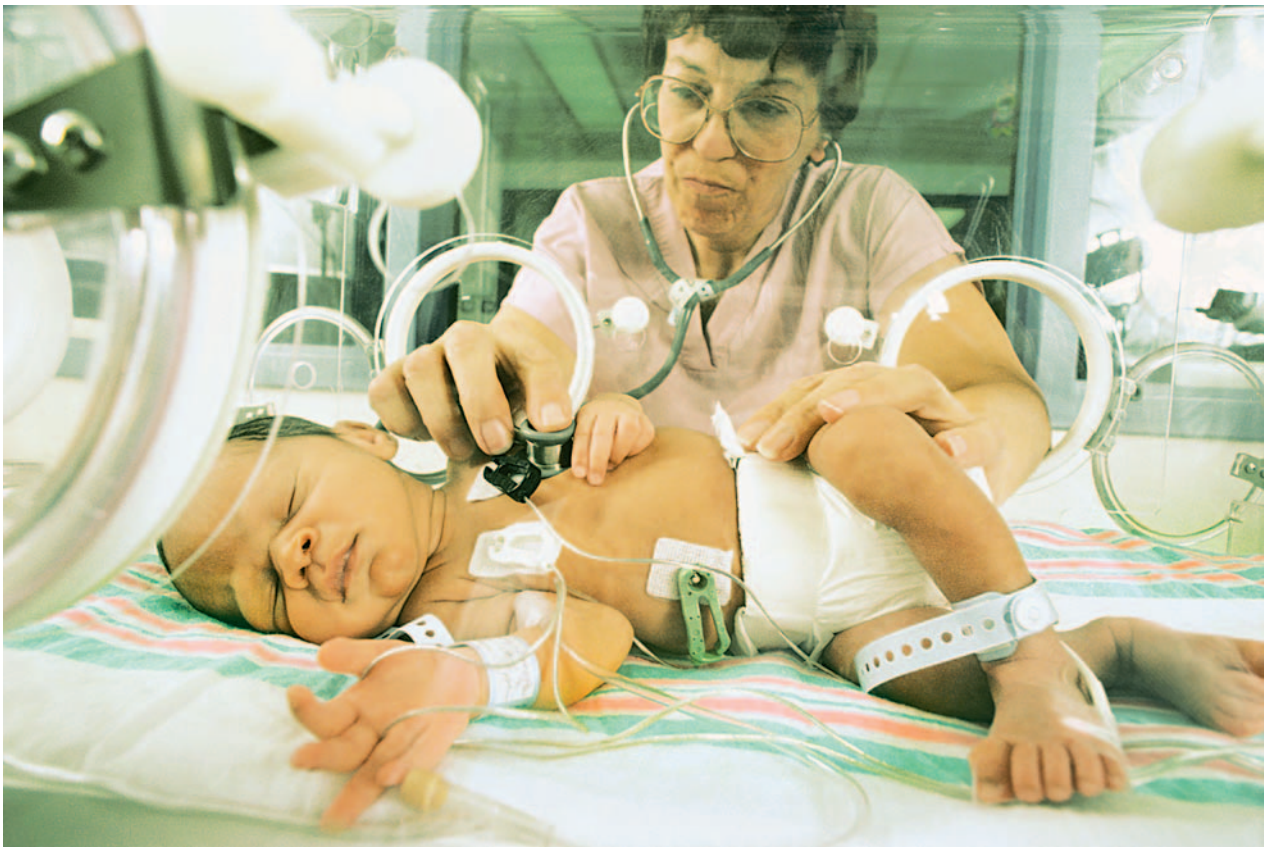
As the fetus develops, it receives the oxygen it needs from the mother's blood system. Most of the blood in the infant's system bypasses the lungs. Once the baby is born, its own blood must start pumping through the lungs to get oxygen. Normally, this bypass duct closes within the first few hours or days after birth. If it does not close, the baby may have trouble getting enough oxygen on its own. **Patent ductus arteriosus** is a condition in which the duct that channels blood between two main arteries does not close after the baby is born. In some cases, a drug called indomethacin can be given to close the duct. Surgery may be required if the duct does not close on its own as the baby develops.

Retinopathy of prematurity is a condition in which the blood vessels in the baby's eyes do not develop normally, and can, in some cases, result in blindness. Premature infants are also more susceptible to infections. They are born with fewer antibodies, which are necessary to fight off infections.

When to call the doctor

In some cases, healthcare professionals are able to stop or delay premature labor if treated early enough. A pregnant woman should contact her healthcare provider if she observes any of the signs of premature labor, including the following:

- contractions closer than 10 minutes apart
- leaking fluid or bleeding from the vagina
- menstrual-like cramps



Premature infant in an incubator. *(© Royalty-Free/Corbis.)*

- abdominal cramps, with or without diarrhea
- low, dull backache
- pelvic pressure

Diagnosis

Many of the problems associated with prematurity depend on how early the baby is born and how much it weighs at birth. The most accurate way of determining the gestational age of an infant in utero is calculating from a known date of conception or using ultrasound imaging to observe development. When a baby is born, doctors can use the Dubowitz exam to estimate gestational age. This standardized test scores responses to 33 specific neurological stimuli to estimate the infant's neural development. Once the baby's gestational age and weight are determined, further tests and **electronic fetal monitoring** may need to be used to diagnose problems or to track the baby's condition. A blood pressure monitor may be wrapped around the arm or leg. Several types of monitors can be taped to the skin. A heart monitor or cardiorespiratory monitor may be attached to the baby's chest, abdomen, arms, or legs with adhesive patches to monitor breathing and heart rate. A thermometer probe may be taped on the skin to monitor body temperature. Blood samples may be taken from a vein or artery. X-ray or ultrasound imaging may be used to examine the heart, lungs, and other internal organs.

Treatment

Treatment depends on the types of complications that are present. It is not unusual for premature infants to be placed in heat-controlled units (incubators) to maintain their temperature. Infants who are having trouble breathing on their own may need oxygen either pumped into the incubator, administered through small tubes placed in their nostrils, or through a respirator or ventilator, which pumps air into a breathing tube inserted into the airway. They may require fluids and nutrients to be administered through an intravenous line, in which a small needle is inserted into a vein in the hand, foot, arm, leg, or scalp. If the baby needs drugs or medications, these may also be administered through the intravenous line. Another type of line may be inserted into the baby's

umbilical cord. This can be used to draw blood samples or to administer medications or nutrients. If heart rate is irregular, the baby may have heart monitor leads taped to the chest. Many premature infants require time and support with breathing and feeding until they mature enough to breathe and eat unassisted. Depending on the complications, the baby may require drugs or surgery.

Alternative treatment

Research has shown that the risks of massaging preterm infants are minimal and that infants benefit from improved developmental scores, more rapid weight gain, and earlier discharge from the hospital. An additional benefit of **massage therapy** is closer **bonding** between the parents and their newborn child. Another method, called kangaroo care, entails placing a medically stable, diaper-clad premature infant on a parent's chest for periods of time so that the parent and child are touching skin-to-skin. A 2002 study published in *Pediatrics* found that both the parent and infant benefited from the practice: mothers reported lower rates of depression and more sensitivity to the infant's needs, and the infants showed improved cognitive and motor development.

Nutritional concerns

If a premature infant is unable to nurse at the breast or drink from a bottle, fluids and nutrients may be administered intravenously or with a tube in the nose or mouth that empties into the stomach (called gavage feeding). Even if a baby is unable to feed at the breast, a mother may pump her breast milk to be given to the infant via gavage feeding. Once the infant learns to suck and swallow effectively, breast or bottle feedings can commence.

Prognosis

Advances in medical care have made it possible for many premature infants to survive and develop normally. Whether a premature infant survives, however, is still intimately tied to his or her gestational age:

- 21 weeks or less: 0 percent survival rate
- 22 weeks: 0 to 10 percent survival rate
- 23 weeks: 10 to 35 percent survival rate
- 24 weeks: 40 to 70 percent survival rate
- 25 weeks: 50 to 80 percent survival rate
- 26 weeks: 80 to 90 percent survival rate
- 27 weeks: greater than 90 percent survival rate

Physicians cannot predict long-term complications of prematurity; some consequences may not become evident until the child is school age. Minor disabilities like learning problems, poor coordination, or short attention span may be the result of premature birth but can be overcome with early intervention. The risks of serious long-term complications depend on many factors, including how premature the infant was at birth, the weight at birth, and the presence or absence of breathing problems. Gender is an associated factor: a Swedish study published in 2003 found that boys are at greater risk of death or serious long-term consequences of prematurity than girls. For example, 60 percent of boys born at 24 weeks' gestation die compared to 38 percent mortality for girls. The development of infection or the presence of a birth defect can also affect long-term prognosis. Infants who have infections in prematurity and very low birth weight are at risk for later disorders of the nervous system; a study done at Johns Hopkins reported that 77 out of a group of 213 premature infants developed neurologic disorders. Severe disabilities such as brain damage, blindness, and chronic lung problems are possible and may require ongoing care.

Prevention

Some of the risks and complications of premature delivery can be reduced if the mother receives good prenatal care, follows a healthy diet, avoids alcohol or drug consumption, and refrains from cigarette **smoking**. In some cases of premature labor, the mother may be placed on bed rest or given drugs that can stop labor contractions for days or weeks, giving the developing infant more time to develop before delivery. The physician may prescribe a steroid medication to be given to the mother before the delivery to help speed up the baby's lung development. The availability of a neonatal intensive care unit (NICU), a special hospital unit equipped and trained to deal with premature infants, can also increase an infant's chances of survival.

A new medication may help to prevent spontaneous premature births. Researchers at Wake Forest University reported in June 2003 that a drug known as 17 alpha-hydroxyprogesterone caproate reduced the number of premature births in a group of women who received weekly injections of the drug compared to a placebo group and lowered the rates of necrotizing enterocolitis, intraventricular hemorrhage, and need for supplemental oxygen in their infants.

Nutritional concerns

Poor **nutrition** during pregnancy may lead to an increased risk of premature delivery. Research supported by the U.S. Public Health Service during the 1990s found that an inadequate diet during pregnancy was asso-

KEY TERMS

Apnea—The temporary absence of breathing. Sleep apnea consists of repeated episodes of temporary suspension of breathing during sleep.

Dubowitz exam—Standardized test that scores responses to 33 specific neurological stimuli to estimate an infant's neural development and, hence, gestational age.

Intraventricular hemorrhage (IVH)—A condition in which fragile blood vessels within the brain burst and bleed into the hollow chambers (ventricles) of the brain and into the tissue surrounding them.

Jaundice—A condition in which the skin and whites of the eyes take on a yellowish color due to an increase of bilirubin (a compound produced by the liver) in the blood. Also called icterus.

Necrotizing enterocolitis—A serious bacterial infection of the intestine that occurs primarily in sick or premature newborn infants. It can cause death of intestinal tissue (necrosis) and may progress to blood poisoning (septicemia).

Respiratory distress syndrome (RDS)—Also known as hyaline membrane disease, this is a condition of premature infants in which the lungs are imperfectly expanded due to a lack of a substance (surfactant) on the lungs that reduces tension.

Retinopathy of prematurity—A condition in which the blood vessels in a premature infant's eyes do not develop normally. It can, in some cases, result in blindness.

Surfactant—A protective film secreted by the alveoli in the lungs that reduces the surface tension of lung fluids, allowing gas exchange and helping maintain the elasticity of lung tissue. Surfactant is normally produced in the fetal lungs in the last months of pregnancy, which helps the air sacs to open up at the time of birth so that the newborn infant can breathe freely. Premature infants may lack surfactant and are more susceptible to respiratory problems without it.

ciated with premature rupture of amniotic sac membranes and premature birth. A well-balanced diet rich in nutrients such as **folic acid**, calcium, iron, zinc, vitamin D, and the B **vitamins** is recommended for pregnant women. Mothers are recommended to eat approximately 300 additional calories a day (above and beyond a normal non-pregnancy diet) to support the fetus's growth and development.

Parental concerns

Parents are often overwhelmed at the prospect of caring for a premature baby. Parents of infants being cared for in the NICU are often recommended to feed, change, and hold their child as long as he or she is medically stable. After the infant leaves the hospital, the parents can seek support from many professional and parent-to-parent resources, including books, web sites, support groups, and national organizations.

Resources

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ORGANIZATIONS

American Academy of Pediatrics (AAP). 141 Northwest Point Boulevard, Elk Grove Village, IL 60007. Web site: <www.aap.org>.

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Premenstrual syndrome

Definition

Premenstrual syndrome (PMS) refers to symptoms that occur between ovulation and the onset of **menstruation**. The symptoms include both physical symptoms, such as breast tenderness, back **pain**, abdominal cramps, **headache**, and changes in appetite, and psychological symptoms of **anxiety**, depression, and unrest. Severe forms of this syndrome are referred to as premenstrual dysphoric disorder (PMDD). These symptoms may be related to hormones and emotional disorders.

Description

Approximately 75 percent of all menstruating women experience some symptoms that occur before or during menstruation. PMS encompasses symptoms severe enough to interfere with daily life. About 3 to 7 percent of women experience the more severe PMDD. These symptoms can last four to ten days and can have a substantial impact on a woman's life. The reason some women get severe PMS while others have none was as of 2004, not understood.

Demographics

Not really a characteristic of adolescent girls, PMS symptoms usually begin between ages 20 and 30 years. The disease may run in families and is also more prone to occur in women with a history of psychological problems. Overall however, it is difficult to predict who is most at risk for PMS.

Causes and symptoms

Because PMS is restricted to the second half of a woman's menstrual cycle, after ovulation, it is thought that hormones play a role. During a woman's monthly menstrual cycle, which lasts 24 to 35 days, hormone levels change. The hormone estrogen gradually rises during the first half of a woman's cycle, the preovulatory phase, and falls dramatically at ovulation. After ovulation, the postovulatory phase, progesterone levels gradually increase until menstruation occurs. Both estrogen and progesterone are secreted by the ovaries, which are responsible for producing the eggs. The main role of these hormones is to cause thickening of the lining of the uterus (endometrium). However, estrogen and progesterone also affect other parts of the body, including the brain. In the brain and nervous system, estrogen can affect the levels of neurotransmitters, such as serotonin. Serotonin has long been known to have an effect on emotions, as well as eating behavior. It is thought that when estrogen levels go down during the postovulatory phase of the menstrual cycle, decreases in serotonin levels follow. Whether these changes in estrogen, progesterone, and serotonin are responsible for the emotional aspects of PMS was not, as of 2004, known with certainty. However, most researchers agree that the chemical transmission of signals in the brain and nervous system are in some way related to PMS. This belief is supported by the fact that the times following **childbirth** and menopause are also associated with both depression and low estrogen levels.

Symptoms for PMS are varied and many, including both physical and emotional aspects that range from mild to severe. The physical symptoms include: bloating, headaches, food cravings, abdominal cramps, headaches, tension, and breast tenderness. Emotional aspects include mood swings, irritability, and depression.

When to call the doctor

A physician or other healthcare provider should be called whenever a woman experiences symptoms of PMS that exceed her ability to cope.

Diagnosis

The best way to diagnose PMS is to review a detailed diary of a woman's symptoms for several months. PMS is diagnosed by the presence of physical, psychological, and behavioral symptoms that are cyclic and occur in association with the premenstrual period of time. PMDD, which is far less common, was officially recognized as a disease in 1987. Its diagnosis depends on the presence of at least five symptoms

related to mood that disappear within a few days of menstruation. These symptoms must interfere with normal functions and activities of the individual. The diagnosis of PMDD has caused controversy connected to the concern that it may be used against women, labeling them as being impaired by their menstrual cycles.

Treatment

There are many treatments for PMS and PMDD depending on the symptoms and their severity. For mild cases, treatment includes **vitamins**, diuretics, and pain relievers. Vitamins E and B6 may decrease breast tenderness and help with fatigue and mood swings in some women. Diuretics work for some women. For more severe cases and for PMDD, treatments available include antidepressant drugs, hormone treatment, or (only in extreme cases) surgery to remove the ovaries. Hormone treatment usually involves **oral contraceptives**. This treatment, as well as removal of the ovaries, is used to prevent ovulation and the changes in hormones that accompany ovulation. Some studies in the early 2000s, however, indicate that hormone treatment has little effect over placebo.

Antidepressants

The most progress in the treatment of PMS and PMDD has been through the use of antidepressant drugs. The most effective of these are sertraline (Zoloft), fluoxetine (Prozac), and paroxetine (Paxil). They are termed selective serotonin reuptake inhibitors (SSRIs) and act by indirectly increasing the brain serotonin levels, thus stabilizing emotions. Some doctors prescribe antidepressant treatment for PMS throughout the cycle, while others direct women to take the drug only during the latter half of the cycle. **Antidepressants** should be avoided by women who want to become pregnant. Sertraline appears to significantly improve productivity, social activities, and relationships compared. Side effects of sertraline were found to include **nausea**, **diarrhea**, and decreased libido.

There are alternative treatments that can both affect serotonin and hormone responses, as well as affect some of the physical symptoms of PMS.

Vitamins and minerals

Some women find relief with the use of vitamin and mineral supplements. Magnesium can reduce the fluid retention that causes bloating, while calcium may decrease both irritability and bloating. Magnesium and calcium also help relax smooth muscles, and this may reduce cramping. Vitamin E may reduce breast tenderness, nervous tension, fatigue, and insomnia. Vitamin B6 may decrease fluid retention, fatigue, irritability, and mood swings. Vitamin B5 supports the adrenal glands and may help reduce fatigue.

Phytoestrogens and natural progesterone

The Mexican wild yam (*Dioscorea villosa*) contains a substance that may be converted to progesterone in the body. Because this substance is readily absorbed through the skin, it can be found as an ingredient in many skin creams. (Some products also have natural progesterone added to them.) Some herbalists believe that these products can have a progesterone-like effect on the body and decrease some of the symptoms of PMS.

The most important way to alter hormone levels may be by eating more phytoestrogens. These plant-derived compounds have an effect similar to estrogen in the body. One of the richest sources of phytoestrogens is soy products, such as tofu. Additionally, many supplements can be found that contain black cohosh (*Cimicifugaracemosa*) or dong quai (*Angelica sinensis*), which are herbs high in phytoestrogens. Red clover (*Trifolium pratense*), alfalfa (*Medicago sativa*), licorice (*Glycyrrhiza glabra*), hops (*Humulus lupulus*), and legumes are also high in phytoestrogens. Increasing the consumption of phytoestrogens is also associated with decreased risks of osteoporosis, **cancer**, and heart disease.

Antidepressant alternatives

Many antidepressants act by increasing serotonin levels. An alternative means of achieving this result is to eat more carbohydrates. For instance, two cups of cereal or a cup of pasta have enough carbohydrates to effectively increase serotonin levels. An herb known as St. John's wort (*Hypericum perforatum*) has stood up to scientific trials as an effective antidepressant. As with the standard antidepressants, however, it must be taken continuously and does not show an effect until used for four to six weeks. There are also herbs, such as skullcap (*Scutellaria lateriflora*) and kava (*Piper methysticum*), that can relieve the anxiety and irritability that often accompany depression. An advantage of these herbs is that they can be taken when symptoms occur rather than continually. Chaste tree (*Vitex agnus-castus*) in addition to helping rebalance estrogen and progesterone in the body, also may relieve the anxiety and depression associated with PMS.

KEY TERMS

Antidepressant drug—A medication prescribed to relieve major depression. Classes of antidepressants include selective serotonin reuptake inhibitors (fluoxetine/Prozac, sertraline/Zoloft), tricyclics (amitriptyline/Elavil), MAOIs (phenelzine/Nardil), and heterocyclics (bupropion/Wellbutrin, trazodone/Desyrel).

Estrogen—Female hormone produced mainly by the ovaries and released by the follicles as they mature. Responsible for female sexual characteristics, estrogen stimulates and triggers a response from at least 300 tissues. After menopause, the production of the hormone gradually stops.

Neurotransmitter—A chemical messenger that transmits an impulse from one nerve cell to the next.

Phytoestrogens—Compounds found in plants that can mimic the effects of estrogen in the body.

Progesterone—The hormone produced by the ovary after ovulation that prepares the uterine lining for a fertilized egg.

Serotonin—A widely distributed neurotransmitter that is found in blood platelets, the lining of the digestive tract, and the brain, and that works in combination with norepinephrine. It causes very powerful contractions of smooth muscle and is associated with mood, attention, emotions, and sleep. Low levels of serotonin are associated with depression.

Prognosis

The prognosis for women with both PMS and PMDD is good. Most women who are treated for these disorders do well.

Prevention

Maintaining a good diet, one low in sugars and fats and high in phytoestrogens and complex carbohydrates, may prevent some of the symptoms of PMS. Women should try to **exercise** three times a week and keep in generally good health. Because PMS is often associated with stress, avoidance of stress or developing better means to deal with stress can be important.

Nutritional concerns

Consuming foods, such as soy products, that are good sources of phytoestrogens may provide relief of PMS symptoms. In general, eating a balanced diet is beneficial.

Parental concerns

Parents should be aware of the symptoms of PMS in their adolescent daughters. The condition is uncommon but can occur in women under the age of 20.

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Prenatal development

Definition

Prenatal development refers to the process in which a baby develops from a single cell after conception into an embryo and later a fetus.

Description

The average length of time for prenatal development to complete is 38 weeks from the date of conception. During this time, a single-celled zygote develops in a series of stages into a full-term baby. The three primary stages of prenatal development are the germinal stage, the embryonic stage, and the fetal stage.

Germinal stage

Conception occurs when the female egg (ovum) is fertilized by a the male sperm. Under normal circumstances, one egg is released approximately once a month from a woman's ovary during a process called ovulation. The egg makes its way into a fallopian tube, a structure that guides the egg away from the ovary toward the uterus. For fertilization to occur, sperm ejaculated during sexual intercourse (or introduced during artificial insemination) in a substance called semen must have made their way from the vagina into the uterus and subsequently into the fallopian tube where the ovum has been released. This process can take up to ten hours after ejaculation. For fertilization to occur, a sperm must penetrate the tough outer membrane of the egg called the zona pellucida. When one sperm successfully binds with the zona pellucida, a series of chemical reactions occurs to allow only that sperm to penetrate. Fertilization occurs when the sperm successfully enters the ovum's membrane. The genetic material of the sperm and egg then combine to form a single cell called a zygote and the germinal stage of prenatal development commences.

The zygote soon begins to divide rapidly in a process called cleavage, first into two identical cells called blastomeres, which further divide to four cells, then into eight, and so on. The group of diving cells begins to move along the fallopian tube toward the uterus. About sixty hours after fertilization, approximately sixteen cells have formed to what is called a morula, still enclosed by the zona pellucida; three days after fertilization, the morula enters the uterus. As cell division continues, a fluid-filled cavity called a blastocoele forms in the center of the group of cells, with the outer shell of cells called trophoblasts and an inner mass of cells called embryoblasts. The zona pellucida disappears and the morula becomes a blastocyst. At this stage the blastocyst consists of 200 to 300 cells and is ready for implantation.

Implantation, the process in which the blastocyst implants into the uterine wall, occurs approximately six days after conception. Hormones secreted from the mother's ovaries and a chemical secreted by the trophoblasts begin to prepare the uterine wall. The blastocyst first adheres to the wall then moves into the uterine tissue. Implantation marks the end of the germinal stage and the beginning of the embryonic stage.

Embryonic stage

The embryonic stage begins after implantation and lasts until eight weeks after conception. Soon after implantation, the cells continue to rapidly divide and clusters of cells begin to take on different functions (called differentiation). A process (gastrulation) leads to the formation of three distinct layers called germ layers: the ectoderm (outer layer), the mesoderm (middle layer), and the endoderm (inner layer). As the embryo develops, each germ layer differentiates into different tissues and structures. For example, the ectoderm eventually forms skin, nails, hair, brain, nervous tissue and cells, nose, sinuses, mouth, anus, tooth enamel, and other tissues. The mesoderm develops into muscles, bones, heart tissue, lungs, reproductive organs, lymphatic tissue, and other tissues. The endoderm forms the lining of lungs, bladder, digestive tract, tongue, tonsils, and other organs.

The process of differentiation takes place over a period of weeks with different structures forming simultaneously. Some of the major events that occur during the embryonic stage are as follows:

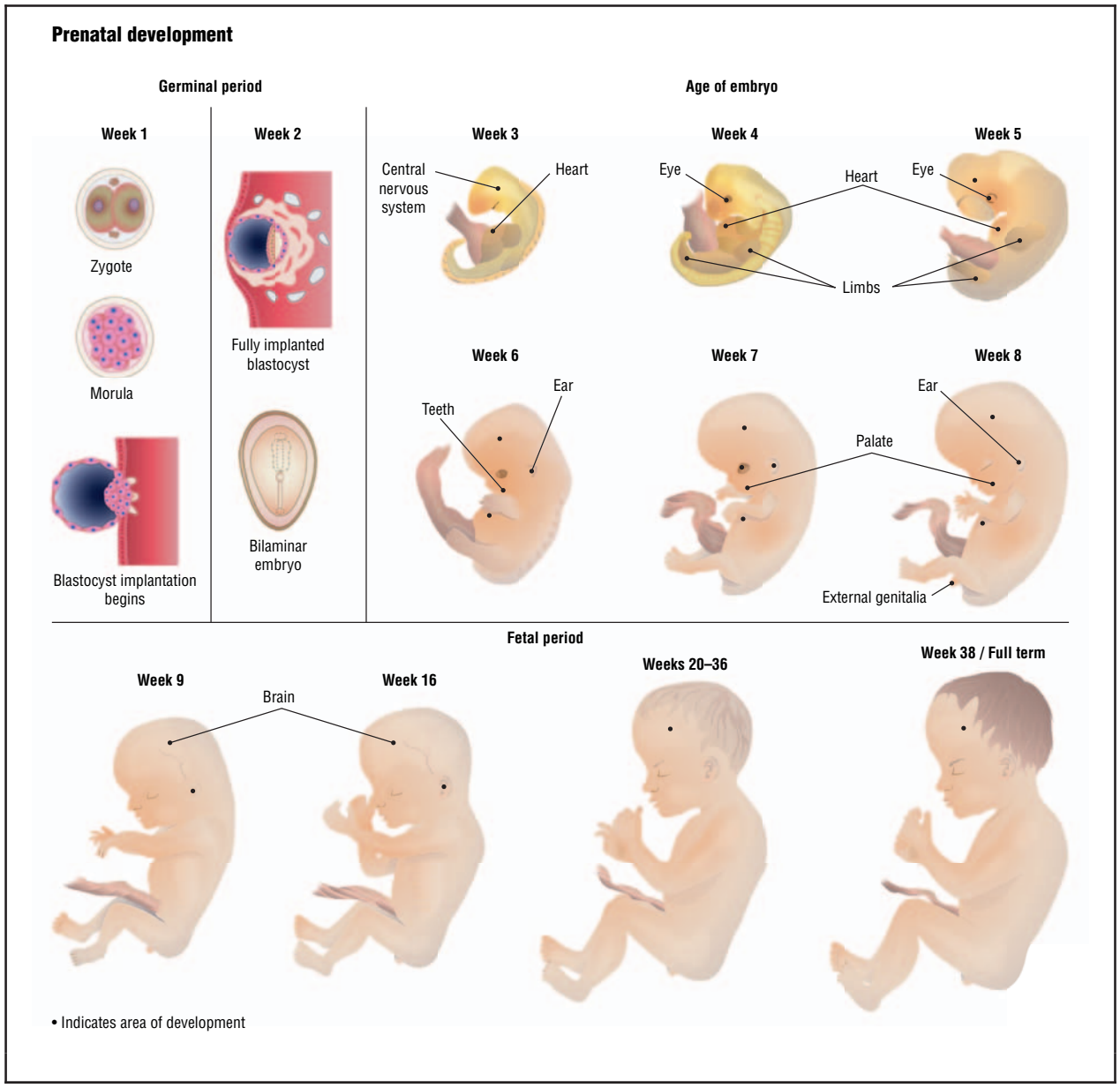

Illustration of prenatal development, from the two-cell, or zygote, stage through the embryonic stage, in which the major body systems develop, to the fetal stage, during which the baby's brain develops and the body adds size and weight. *(Illustration by GGS Information Services.)*

- Week 3: Beginning development of the brain, heart, blood cells, circulatory system, spinal cord, and digestive system.
- Week 4: Beginning development of bones, facial structures, and limbs (presence of arm and leg buds); continuing development of the heart (which begins to beat), brain, and nervous tissue.
- Week 5: Beginning development of eyes, nose, kidneys, lungs; continuing development of the heart (formation of valves), brain, nervous tissue, and digestive tract.
- Week 6: Beginning development of hands, feet, and digits; continuing development of brain, heart, and circulation system.

- Week 7: Beginning development of hair follicles, nipples, eyelids, and sex organs (testes or ovaries); first formation of urine in the kidneys and first evidence of brain waves.
- Week 8: Facial features more distinct, internal organs well developed, the brain can signal for muscles to move, heart development ends, external sex organs begin to form.

By the end of the embryonic stage, all essential external and internal structures have been formed. The embryo is now referred to as a fetus.

Fetal stage

Prenatal development is most dramatic during the fetal stage. When an embryo becomes a fetus at eight weeks, it is approximately 3 centimeters (1.2 inches) in length from crown to rump and weighs about 3 grams (0.1 ounce). By the time the fetus is considered full-term at 38 weeks gestation, he or she may be 50 centimeters (20 inches) or 3.3 kilograms (7.3 pounds). Although all of the organ systems were formed during embryonic development, they continue to develop and grow during the fetal stage. Examples of some of the major features of fetal development by week are as follows:

- Weeks 9–12: The fetus reaches approximately 8 cm. (3.2 in.) in length; the head is approximately half the size of the fetus. External features such as the face, neck, eyelids, limbs, digits, and genitals are well formed. The beginnings of teeth appear, and red blood cells begin to be produced in the liver. The fetus is able to make a fist.
- Weeks 13–15: The fetus reaches approximately 15 cm. (6 in.) in length. Fine hair called lanugo first develops on the head; structures such as the lungs, sweat glands, muscles, and bones continue to develop. The fetus is able to swallow and make sucking motions.
- Weeks 16–20: The fetus reaches approximately 20 cm. (8 in.) in length. Lanugo begins to cover all skin surfaces, and fat begins to develop under the skin. Features such as finger and toenails, eyebrows, and eyelashes appear. The fetus becomes more active, and the mother can sometimes begin to feel fetal movements at this stage.
- Weeks 21–24: The fetus reaches approximately 28.5 cm. (11.2 in.) in length and weighs approximately 0.7 kg (1 lb. 10 oz.). Hair grows longer on the head, and the eyebrows and eye lashes finish forming. The lungs continue to develop with the formation of air sac (alveoli); the eyes finish developing. A startle reflex develops at this time.
- Weeks 25–28: The fetus reaches approximately 38 cm. (15 in.) in length and weighs approximately 1.2 kg (2 lb. 11 oz.). The next few weeks mark a period of rapid brain and nervous system development. The fetus gains greater control over movements such as opening and closing eyelids and certain body functions. The lungs have developed sufficiently that air breathing is possible.
- Weeks 29–32: The fetus reaches approximately 38–43 cm. (15–17 in.) in length and weighs approximately 2 kg (4 lb. 6 oz.). Fat deposits become more pronounced under the skin. The lungs remain immature but breathing movements begin. The fetus's bones are developed but not yet hardened.
- Weeks 33–36: The fetus reaches approximately 41–48 cm. (16–19 in.) in length and weighs 2.6–3.0 kg (5 lb. 12 oz. to 6 lb. 12 oz.). Body fat continues to increase, lanugo begins to disappear, and fingernails are fully grown. The fetus has gained a high degree of control over body functions.
- Weeks 36–38: The fetus reaches 48–53 cm. (19–21 in.) in length is considered to be full-term by the end of this period. Lanugo has mostly disappeared and is replaced with thicker hair on the head. Fingernails have grown past the tips of the fingers. In a healthy fetus, all organ systems are functioning.

Common problems

Although 90 percent of babies born in the United States are considered healthy, abnormalities may arise during prenatal development that are considered congenital (inherited or due to a genetic abnormality) or environmental (such as material derived abnormalities). In other cases, problems may arise when a fetus is born prematurely.

Congenital abnormalities

In some cases abnormalities may arise during prenatal development that cause physical malformations or developmental delays or affect various parts of the body after the child is born. The cause may be a small mutation in or damage to the genetic material of cells, or a major chromosomal abnormality (each normal cell has two copies each of 23 strands [called chromosomes] of genetic material, and abnormalities can arise if there are three copies of a strand or only one). Sometimes the abnormality is inherited from one or both parents; in other cases, the defect occurs because of an error in prenatal development.

Some abnormalities are minor and do not affect the long-term prognosis once the child is born. At the other

end of the spectrum, abnormalities may be so severe that fetal demise is inevitable. Approximately 10 to 15 percent of pregnancies end before the twentieth week, a process called miscarriage or spontaneous abortion; congenital abnormalities account for a significant proportion of miscarriages. Genetic abnormalities account for approximately 5 percent of miscarriages.

Maternal derived abnormalities

The age, health status, nutritional status, and environment of the mother are all closely tied to the health of a growing embryo or fetus. Some examples of environmental factors that may lead to developmental abnormalities include:

- Age: As of 2004, research showed that babies born to mothers between the ages of seventeen and thirty-five tend to be healthier. One reason is that the risk of certain congenital abnormalities such as **Down syndrome** increases with mother's age (particularly mothers over forty). Another reason is that the risk of having pregnancy or birth complications is greater with women over the age of thirty-five.
- Health status: In some cases a mother may pass a viral or bacterial infection to the fetus, such as in human **immunodeficiency** virus (HIV). In other cases, a mother's illness may cause congenital malformations; an example is **rubella**, which can cause heart defects, deafness, developmental delays, and other problems in a fetus if the mother contracts it during pregnancy.
- Nutritional status: A well-balanced diet rich in nutrients such as **folic acid**, calcium, iron, zinc, vitamin D, and the B **vitamins** is recommended for pregnant women. Certain vitamin and mineral deficiencies can interfere with normal prenatal development. For example, a deficiency in folic acid during the early stages of pregnancy may lead to neural tube defects such as **spina bifida**. Mothers are recommended to eat approximately 300 additional calories a day (above and beyond a normal non-pregnancy diet) to support the fetus's growth and development.
- Other environmental factors: Exposure to certain substances called teratogens (agents that may interfere with prenatal development) during pregnancy may cause embryonic or fetal malformations. Examples of teratogens include alcohol, thalidomide, cocaine, certain seizure medications, diethylstilbestrol (DES), and the anti-acne drug Accutane.

Prematurity

Advances in medical care have made it possible for many infants born prematurely to survive and develop normally. The earlier the gestational age, the greater the chance of death or significant medical problems. Whether or not a premature infant will survive is intimately tied to his or her gestational age:

- 21 weeks or less: 0 percent survival rate
- 22 weeks: 0–10 percent survival rate
- 23 weeks: 10–35 percent survival rate
- 24 weeks: 40–70 percent survival rate
- 25 weeks: 50–80 percent survival rate
- 26 weeks: 80–90 percent survival rate
- 27 weeks: greater than 90 percent survival rate

KEY TERMS

Miscarriage—Loss of the embryo or fetus and other products of pregnancy before the twentieth week. Often, early in a pregnancy, if the condition of the baby and/or the mother's uterus are not compatible with sustaining life, the pregnancy stops, and the contents of the uterus are expelled. For this reason, miscarriage is also referred to as spontaneous abortion.

Ovary—One of the two almond-shaped glands in the female reproductive system responsible for producing eggs and the sex hormones estrogen and progesterone.

Teratogen—Any drug, chemical, maternal disease, or exposure that can cause physical or functional defects in an exposed embryo or fetus.

Uterus—The female reproductive organ that contains and nourishes a fetus from implantation until birth. Also called the womb.

Parental concerns

Many parents have questions or concerns about the prenatal development of an existing or anticipated child and what steps they should take to ensure their child's health. During prenatal visits to an obstetrician, a pregnant mother should be educated in proper **nutrition** and prenatal care; often, prenatal vitamins are prescribed to avoid nutritional deficiencies. Prenatal testing is often recommended to parents-to-be as a means of assessing the fetus's health and the risk of developing certain conditions. Some common prenatal tests that relate to prenatal development are as follows:

- blood tests to check for diseases that could affect the fetus, such as HIV, **hepatitis B**, or other sexually transmitted diseases
- blood tests to check if the mother carries a protein called Rh factor on her red blood cells; if she does not and her baby does (determined by whether the father is Rh-positive or not), she will require treatment to prevent a potentially damaging reaction to the baby
- chorionic villus sampling, a prenatal test that takes a tiny sample of the placenta with a needle to test for chromosomal abnormalities
- nuchal fold or nuchal translucency screening test, which measures a small space at the back of the fetus's neck using ultrasound; fetuses with larger nuchal folds have a greater risk of having a chromosomal abnormality
- amniocentesis, a test that takes a sample of the fluid that surrounds the fetus in the uterus to identify certain genetic disorders, congenital malformations, or the maturity of the fetus's lungs

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Prenatal surgery

Definition

Prenatal surgery is a surgical procedure performed on a fetus prior to birth.

Purpose

Prenatal surgery, also called fetal surgery, antenatal surgery, or maternal-fetal surgery, usually is performed under circumstances in which the fetus is not expected to survive delivery or to live long after birth without prenatal intervention. The most common prenatal surgeries are for conditions in which the child will not be able to breathe on its own after birth. There are only about 600 candidates for prenatal surgery in the United States each year and far fewer prenatal surgeries are performed. Most of these procedures are high-risk and may be considered experimental.

More common prenatal surgeries

Urinary tract obstructions in male fetuses are usually caused by a narrowing of the urinary tract. This can cause the urine, which normally flows out into the amniotic fluid surrounding the fetus, to back up and injure the kidneys. If only one kidney is affected and there is a normal amount of amniotic fluid, prenatal intervention is not required. However, in addition to kidney damage, urinary tract obstructions can lead to multiple abnormalities and depleted amniotic fluid, which endangers the fetus and prevents the lungs from growing. About 10 percent of fetuses with urinary tract obstructions may require prenatal surgery in which a device is placed in the fetus's bladder to drain the urine into the amniotic sac.

Congenital diaphragmatic **hernia** (CDH) occurs when the diaphragm does not form completely at about eight weeks of gestation, leaving a hole in this muscle that separates the chest and the abdomen. The stomach, intestines, liver, spleen, and kidneys can move into the chest cavity through this hole, which is most often on the left side. Most babies with CDH are treated after birth. However, about 50 percent of fetuses with CDH do not survive after birth because their lungs are too small (pulmonary hypoplasia). A fetus whose liver has moved into its chest, seriously restricting lung development, whose lung-to-head ratio is less than one in four, and whose chance of survival through delivery is less than 50 percent may be a candidate for surgery. CDH fetuses whose livers have not moved into the chest have a survival rate of about 90 percent without prenatal intervention.

Congenital cystic adenomatoid malformation (CCAM) occurs when one or more lobes of the lungs develop into fluid-filled sacs called cysts rather than into normal lung tissue. Most CCAMs disappear on their own or are small enough to not cause problems. Large CCAMs can limit lung development, causing pulmonary hypoplasia. About 10 percent of fetuses with CCAMs are at risk of heart failure because the cysts push into the heart. CCAMs also can push on the trachea and the esophagus where they prevent the fetus from ingesting amniotic fluid. Prenatal surgery to drain or remove the cyst is performed only on severely affected fetuses.

Sometimes prenatal surgery is performed to remove a tumor. Sacrococcygeal teratoma (SCT) is the most common tumor in newborns, occurring in one out of every 35,000 to 40,000 births. It is more common in girls than in boys. These tumors at the base of the tailbone can grow very large. With early diagnosis, most SCT babies are delivered normally and the tumor is removed after birth. However, a small percentage of these tumors are large, hard, and full of blood vessels and may stress the fetal heart. These may be treated with a surgical procedure that destroys the blood vessels leading to the tumor, thereby preventing its growth.

Up to 15 percent of **twins** who share a placenta (monochorionic twins) have twin-twin transfusion syndrome (TTTS). Because of abnormal blood vessel connections in the placenta, one twin pumps the circulating blood for both twins. As a result the pumping twin has reduced volumes of blood and amniotic fluid and the recipient twin as increased volumes, leading to a variety of problems, including the risk of heart failure in both fetuses. TTTS may be treated by removing fluid from the overfilled recipient amniotic sac and placing it into the depleted sac of the pumping twin. If this fails, prenatal surgery may be used to destroy the abnormal blood vessel connections in the placenta.

Twin-twin reverse arterial perfusion (TRAP) sequence occurs in about 1 percent of monochorionic twins. In a TRAP sequence one twin develops normally and the other lacks a heart. The normal twin pumps all the blood for both twins and is at risk for heart failure. If left untreated 50 to 75 percent of these normal twins die. In prenatal surgery for TRAP sequence, the connections between the twins are severed.

Other prenatal surgeries

Other conditions that may be treated by prenatal surgery include:

- various congenital defects that block air passages and will prevent the newborn from breathing on its own
- various lung malformations
- omphalocele, a birth defect in which an opening in the fetus's abdominal wall enables portions of the stomach, liver, and intestines to protrude
- fetal gastroschisis, a birth defect in which the abdominal wall muscles do not form correctly and the stomach and intestines protrude and float in the amniotic fluid, a condition that occurs primarily in fetuses of mothers in their late teens or early 20s
- bowel obstructions, usually caused by a narrowing in the small intestine
- experimental hematopoietic stem cell transplants for X-linked **severe combined immunodeficiency** syndrome

Hypoplastic left heart syndrome, in which the blood flow through the left side of the heart is obstructed, is the most common congenital heart defect that is a candidate for prenatal surgery. In this condition, which is often fatal, the left side of the heart is very small and stops working.

Spina bifida

Until the late 1990s, prenatal surgery was almost exclusively limited to life-threatening conditions. However in 1994 the first prenatal surgery to treat **spina bifida** (myelomeningocele) was performed. This is the second most common birth defect in the United States, affecting one out of every 2,000 newborns. It is not considered to be life-threatening.

Spina bifida occurs during the first month of fetal development when a small bit of bone and skin fails to fully enclose the nerves of the spinal cord, leaving a hole or lesion. Depending on the location of the lesion, spina bifida may not require treatment; the higher up the opening in the spinal cord, the more severe the condition, and myelomeningocele can cause severe deformities, paralysis, and **mental retardation**. The damage appears to be caused by leakage of fluid from the spinal cord and exposure of the cord to amniotic fluid. The risk of infant death is about 10 percent and after the age of one, about 1 percent of affected children die each year.

Surgery for spina bifida requires closing the opening in the cord. Since the damage from spina bifida occurs during fetal development, prenatal surgery may reduce the damage. However, prenatal surgery for spina bifida has become enmeshed in the politics of reproductive rights and fetal rights. As of 2004, prenatal surgery for spina bifida was available only as part of a prospective randomized clinical trial.

Description

The decision to have prenatal surgery is made on the basis of detailed ultrasound imaging of the fetus, including an echocardiogram that uses ultrasound to obtain images of the fetal heart, as well as other diagnostic tools. Consultations include a perinatologist, a neonatologist, a pediatric surgeon, a clinical nurse specialist, and a social worker. Usually only fetuses with a very poor prognosis are candidates for maternal-fetal surgery. Only about 10 percent of those referred for evaluation actually undergo the surgery. Since additional congenital defects preclude prenatal surgery, **amniocentesis** or chorionic villi sampling (CVS) are used to check for chromosomal abnormalities in the fetus. Prenatal surgeries usually are performed between 18 and 26 weeks of gestation.

Prenatal surgery usually requires a general anesthetic. The fetus receives the anesthetic via the mother's blood. During the operation the anesthesiologist controls the mother's breathing through a tube into her throat and airway. The anesthesiologist and a perinatologist monitor the heart rates of the mother and fetus. For some procedures an epidural anesthetic that numbs the abdominal region may be used instead of general anesthesia.

Open surgeries

Open prenatal surgery requires a procedure similar to a **cesarean section** (C-section) for delivering a baby through the mother's abdomen. Incisions are made through the mother's abdominal wall. In some procedures the fetus is partially removed from the uterus. In other procedures the entire uterus is removed from the mother's body cavity through her abdomen.

Using ultrasound as a guide, the surgeon feels for the affected fetal part. The fetus must be moved away from the placenta, the disk-shaped organ within the uterus that provides the blood supply to the fetus. The surgeon may knead and push on the uterus to move or flip the fetus. A narrow tube in placed through a tiny hole in the uterine wall, through which the amniotic fluid is drained and collected in syringes. Opening the uterus is the riskiest component of prenatal surgery. The first incision is made at a point away from the placenta to prevent damaging it. A special device simultaneously makes the incision and clamps the edges to prevent bleeding. Following the procedure the fetus is replaced in the uterus and the incision is stitched. Prior to the final stitch the amniotic fluid is re-injected into the uterus. The uterus is repositioned in the mother's body cavity and her abdominal wall is closed.

The first successful open fetal surgery was performed in 1981 for a urinary tract obstruction. A tiny hole was made in the bladder of the fetus and a catheter (a long, thin tube) was inserted to drain the urine directly into the amniotic fluid. The first successful open fetal surgery for CDH was performed in 1989.

Prenatal open surgery for CCAM requires opening the fetal chest. If a large cyst does not have a hard component, procedures called thoracoamniotic shunting or catheter decompression may be used to drain the cyst. Otherwise the surgeon must remove all or a portion of the mass. The first successful resection (removal) of a CCAM from a fetal lung was performed in 1990. The first resectioning of a fetal SCT was performed in 1992.

Between 1997 and 2004, more than 200 open surgeries were performed for myelomeningocele. An incision the size of a small fist is made in the uterus. The surgeon loosens and lifts the tissues of the spinal canal lesion and stitches them closed. The entire procedure takes about one hour.

Less invasive procedures

For urinary tract obstructions a needle may be used to insert a catheter through the mother's abdomen and uterus and into the fetal bladder where it drains the urine into the amniotic fluid. The catheter may have an expandable wire mesh that expands in the bladder to prevent the catheter from plugging up or dislodging.

The first successful fetoscopic temporary tracheal occlusion for CDH was performed in 1996. Small surgical openings are made in the uterus, and a tiny fiber optic fetoscope is inserted to guide the operation. A needle-like instrument enters the uterus through a small incision in the mother's abdomen. A balloon placed in the fetus's trachea prevents lung fluid from escaping through the mouth, enabling the lungs to expand, grow, and push the abdominal organs out of the chest and back into the abdomen. The balloon is removed at birth. In a successful procedure the lungs are developed enough that the baby will breathe on its own at birth.

Hypoplastic left heart syndrome is treated by passing a needle, guided by ultrasound, through the mother's abdominal wall, into the uterus and the fetal heart. A catheter is passed through the needle across the fetus's aortic valve. A balloon is inflated, opening the valve and allowing blood to flow through the left side of the heart.

Radiofrequency ablation

Radiofrequency ablation (RFA) sometimes can be used for SCT. Guided by ultrasound a needle is inserted through the mother's abdomen and uterus and into the tumor. Radiofrequency waves sent through the needle

destroy the blood supply to the tumor with heat. This slows the tumor's growth and may enable the fetus to survive until delivery. The first RFA of a SCT was performed in 1998.

A TRAP sequence also may be treated by RFA. A 3-mm needle targets the exact point where the blood enters the twin without a heart. Using an echocardiogram device, RFA is applied until the blood vessels and surrounding tissue are destroyed and the blood flow is halted. This procedure has eliminated the need for open surgery or larger fetoscopes to treat a TRAP sequence.

Laser treatment

TTTS is a progressive disorder and early intervention may prevent later complications. The most common treatment for TTTS is amnioreduction in which a syringe through the mother's abdomen is used to remove amniotic fluid from the overfilled sac and place it in the sac of the other twin. This procedure may need to be repeated during the course of the pregnancy. If TTTS does not respond to amnioreduction, laser treatment may be attempted to stop the abnormal blood circulation. Following detailed ultrasound, a thin fetoscope is inserted through the mother's abdominal and uterine walls and into the amniotic cavity of the recipient twin to examine the surface placental vessels. The abnormal blood vessel connections are located and eliminated with a laser beam. The first successful fetoscopic laser treatment for TTTS was performed in 1999.

EXIT

Ex utero intrapartum treatment (EXIT) is a surgery performed for a congenital defect that blocks the fetus's airway. The fetus is removed from the womb by cesarean section but the umbilical cord is left intact so that the mother's placenta continues to sustain the fetus. After the air passage is cleared, the umbilical cord is cut and the newborn can breathe on its own. The EXIT procedure is used for various types of airway obstruction including CCAM.

Precautions

The decision to undergo prenatal surgery is a difficult one. Considerations must include the following:

- serious risks for the mother and fetus
- time commitment for the surgery
- extended postoperative bed rest, perhaps until delivery
- travel to a hospital that performs the procedure
- possible need to stay near the hospital until delivery
- significant commitment of financial resources; some surgeries may not be covered by insurance
- arrangements for care of other children

Preparation

Prior to surgery the mother will need to do the following:

- arrange for help and support following the surgery since she will be on bed rest to prevent preterm labor
- prepare for the possibility of having to remain near the hospital until delivery
- possibly make arrangements for a blood donor in the unlikely event that she needs a blood transfusion
- take betamethasone, a steroid, in two intramuscular injections 12–24 hours apart to accelerate lung maturity in the fetus; if delivery occurs earlier than 34 weeks, the mother may have to take it again closer to the delivery date
- wear a fetal/uterine monitor after hospital admission
- have urine and blood samples taken
- have her blood typed and cross-matched in case of the need for a transfusion
- have an intravenous (IV) infusion of fluid and electrolytes
- take a sleeping medication
- sign a surgical consent
- wear thick elastic stockings
- take a medication for decreasing stomach acids

The mother usually is given medications called tocolytics to prevent contractions and labor during and after surgery. Tocolytics include:

- indocin suppositories before surgery and up to 48 hours after surgery
- magnesium sulfate for one to two days after surgery with careful monitoring
- nifedipine, a pill given every four to six hours as the indocin is being decreased; usually nifedipine is continued until 37 weeks of gestation or delivery
- terbutalin in pill form

Aftercare

Following the surgery the mother lies in bed on her side to provide the best circulation to the fetus and to help prevent contractions. The mother will have:

- an oxygen mask to provide post-anesthetic supplemental oxygen

- an IV catheter to supply fluids and **antibiotics** for at least 48 hours after surgery
- a urinary catheter to collect urine from the bladder for 48–72 hours
- an epidural catheter to supply constant **pain** medication, usually morphine, for several days after surgery, followed by oral pain medications
- a sequential compression device to improve blood circulation in the legs during bed rest
- a continuous electronic fetal/uterine monitor to check the fetal heart and the uterine response to the tocolytics and to check for signs of preterm labor
- a transparent dressing over the abdominal incision so that the fetus can be monitored and the incision site observed without removing the dressing

Fluids and food are not taken by mouth until the mother's digestive function returns. She usually remains in the hospital for four to seven days following surgery.

To prevent or treat postoperative lung or circulatory problems, the mother should:

- practice deep breathing to keep all airways clear
- perform incentive spirometer exercises, using a small simple device to assist deep breathing and opening of the lungs, five times during each waking hour
- be turned from side to side at least every two hours to increase circulation and relieve areas of pressure
- practice foot flexion exercises to improve circulation and help prevent blood clots

After discharge from the hospital the mother will be on modified bed rest, lying on her side until 37 weeks of gestation. This increases blood flow to the fetus and reduces pressure on the cervix to help prevent uterine contractions. She will be given bed rest exercises and prescribed a special diet. She will see a perinatologist once a week and have at least one ultrasound per week.

Risks

A major risk of prenatal surgery is nicking the placenta, causing blood hemorrhaging, uterine contractions, and birth of a premature infant who may not survive. Preterm labor is the most common complication of prenatal surgery. Fetoscopic surgeries are less dangerous and traumatic than open fetal surgery and reduce the risk of premature labor. Subsequent children of a mother who has undergone fetal surgery usually are delivered by cesarean section because of scarring of the uterus.

Other risks to the mother include:

- extensive blood loss
- complications from general anesthesia
- side effects, potentially fatal, from medications to control premature labor
- rupturing of the uterine incision
- infection of the wound or uterus
- psychological stress
- inability to have additional children
- death

All fetuses that undergo surgery are born prematurely. Infants born even six weeks early are at risk for delays in walking and talking and for learning problems. Infants born at 30 weeks of gestation or less are at risk for blindness, **cerebral palsy**, and brain hemorrhages.

About 25 percent of women undergoing prenatal surgery lose some amniotic fluid, often because of leakage at the uterine incision. Amniotic fluid is essential for lung development and protects the fetus from injury and infection. If all of the amniotic fluid is lost, the fetal lungs may not develop properly. Without the cushion that enables the fetus to float, the fetus may compress the umbilical cord causing death.

Other risks to the fetus include:

- birth during surgery
- membrane separation between the tissues surrounding the amniotic fluid sac and the uterus, causing early delivery or interference with blood flow to some fetal body part such as an arm or leg
- further damage to the spinal cord and nerves during prenatal surgery for spina bifida
- intrauterine infection requiring immediate birth of the fetus
- brain damage
- physical deformities
- death

Normal results

Although fetal surgeries heal without scarring, they are rare and risky, and it is difficult to predict the outcome. In general, the following usually occurs:

- Fetal surgery for CDH lessens the severity of the condition so that the fetus usually survives delivery and lives long enough to undergo corrective surgery.
- Thoracoamniotic shunting for CCAM usually results in survival.

KEY TERMS

Amniocentesis—A procedure performed at 16-18 weeks of pregnancy in which a needle is inserted through a woman's abdomen into her uterus to draw out a small sample of the amniotic fluid from around the baby for analysis. Either the fluid itself or cells from the fluid can be used for a variety of tests to obtain information about genetic disorders and other medical conditions in the fetus.

Amniotic fluid—The liquid in the amniotic sac that cushions the fetus and regulates temperature in the placental environment. Amniotic fluid also contains fetal cells.

Cesarean section—Delivery of a baby through an incision in the mother's abdomen instead of through the vagina; also called a C-section, Cesarean birth, or Cesarean delivery.

Chorion—The outer membrane of the amniotic sac. Chorionic villi develop from its outer surface early in pregnancy. The villi establish a physical connection with the wall of the uterus and eventually develop into the placenta.

Chorionic villus sampling—A procedure used for prenatal diagnosis at 10-12 weeks gestation. Under ultrasound guidance a needle is inserted either through the mother's vagina or abdominal wall and a sample of the chorionic membrane. These cells are then tested for chromosome abnormalities or other genetic diseases.

Congenital cystic adenomatoid malformation (CCAM)—A condition in which one or more lobes of the fetal lungs develop into fluid-filled sacs called cysts.

Congenital diaphragmatic hernia (CDH)—A condition in which the fetal diaphragm (the muscle dividing the chest and abdominal cavity) does not close completely.

Echocardiography—A non-invasive technique, using ultrasound waves, used to look at the various structures and functions of the heart.

Ex utero intrapartum treatment (EXIT)—A cesarean section in which the infant is removed from the uterus but the umbilical cord is not cut until after surgery for a congenital defect that blocks the air passage.

Fetoscope—A fiber optic instrument for viewing the fetus inside the uterus.

Monochorionic twins—Twins that share a single placenta.

Omphalocele—A birth defect where the bowel and sometimes the liver, protrudes through an opening in the baby's abdomen near the umbilical cord.

Placenta—The organ that provides oxygen and nutrition from the mother to the unborn baby during pregnancy. The placenta is attached to the wall of the uterus and leads to the unborn baby via the umbilical cord.

Pulmonary hypoplasia—Incomplete or defective development of the lungs.

Radiofrequency ablation (RFA)—A procedure in which radiofrequency waves are used to destroy blood vessels and tissues.

Sacrococcygeal teratoma (SCT)—A tumor occurring at the base of the tailbone in a fetus.

Spina bifida—A birth defect (a congenital malformation) in which part of the vertebrae fail to develop completely so that a portion of the spinal cord, which is normally protected within the vertebral column, is exposed. People with spina bifida can suffer from bladder and bowel incontinence, cognitive (learning) problems, and limited mobility.

Tocolytic drug—A compound given to women to stop the progression of labor.

Twin-twin transfusion syndrome (TTTS)—A condition in identical monochorionic twins in which there is a connection between the two circulatory systems so that the donor twin pumps the blood to the recipient twin without a return of blood to the donor.

Twin:twin reverse arterial perfusion (TRAP) sequence—A condition in which one fetus lacks a heart and the other fetus pumps the blood for both.

Ultrasonography—A medical test in which sound waves are directed against internal structures in the body. As sound waves bounce off the internal structure, they create an image on a video screen. Ultrasonography is often used to diagnose fetal abnormalities, gallstones, heart defects, and tumors. Also called ultrasound imaging.

- Removal of solid CCAM cysts has a survival rate of about 50 percent.
- RFA to slow the growth of a tumor usually enables the fetus to survive delivery, after which the tumor can be removed.
- Survival rates for prenatal treatment of TTTS are about 70 percent.

Prenatal surgery for spina bifida does not cure the condition. However, babies who survive the surgery appear to be 33 to 50 percent less likely to have **hydrocephalus**, a condition that requires surgically implanted tubes or shunts to remove fluid from the ventricles (cavities of the brain). The surgery also appears to reverse hindbrain herniation, in which the back of the brain slips down into the spinal canal. This condition can cause difficulties in breathing and swallowing and leads to death in 15 percent of children with spina bifida. In addition, children who have prenatal surgery for spina bifida appear to have better brain function than children who do not have surgery. However prenatal surgery does not prevent two of the most serious conditions associated with spina bifida: leg movement and bladder and bowel control. As of 2004, the long-term prognosis for these children was not known.

When to call the doctor

The doctor should be called if any of the following occurs:

- The abdominal incision become red, warm, tender to the touch, or is draining fluid.
- The mother's body temperature rises above 101°F (38.5°C).
- Fluid is leaking from the vagina.
- Vaginal bleeding occurs.
- The baby does not move daily.
- Persistent back pain, cramping, abdominal tightening, or pelvic pressure occurs.
- Chest pain or difficulty breathing develops.

Signs of preterm labor include:

- gas pain
- abdominal tightening
- cramping
- backache
- pelvic pressure
- change in vaginal discharge
- leakage of vaginal fluid
- bleeding

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Preschool

Definition

Preschool is an early childhood program in which children combine learning with **play** in a program run by professionally trained adults. Children are most commonly enrolled in preschool between the ages of three and five, though those as young as two can attend some schools. Preschools are different from traditional **day care** in that their emphasis is learning and development rather than enabling parents to work or pursue other activities.

Description

Before 1960, the education of young children was primarily regarded as the responsibility of families within the home. As of 2004, most young children in the United States spend some portion of their days apart from their parents. Most attend some sort of center-based program prior to kindergarten. In 2001, 52 percent of three- and four-year-olds were in a nursery school or preschool program. The enrollment rate for four-year-olds in 2001 was nearly the same as the enrollment rate for five-year-olds in 1970. There are several factors influencing this dramatic change, including a rise in the numbers of mothers working outside the home, a decline in the size of families (leading more parents to turn to preschools as a social outlet for their children), and a growing desire to give children a head start academically. The higher the income and educational level of the parents, the more likely it is that a child will attend preschool. This correlation remains true in spite of increasing governmental support for programs targeting children in low-income households.

In addition to being called preschool, these programs are known by other names, including child care, day care, and nursery school. They vary widely in their setting, format, and educational philosophy. Preschools may meet all-day or half-day, either every day or just a few days per week. They may be sponsored by a church, operate as an independent non-profit, or run for profit. They may be part of the public school system or part of the Federal Head Start program.

Types of preschool programs

PRIVATE PRESCHOOLS Private preschools operate as for-profits, independent nonprofits, and programs sponsored by religious organizations. Most are part-day programs. Some so-called lower schools are affiliated with private schools and maintain an educational philosophy in accord with the parent institution. Though the margin is small, private preschools still claimed the majority of total preschool enrollment in 2001. The educational quality of private preschools varies from program to program. Regulation is primarily by state child care agencies, but the arrangement varies from state to state.

HEAD START Since 1965, the federal Head Start program has provided free education for young children in many low-income families across the United States. In 2000, Head Start served 11 percent of all three- and four-year olds in the United States. In 2001, Head Start reported enrollment of over 900,000 children, at a cost of roughly $7,000 per child. **Head Start programs** are available in all 50 states and are offered in a variety of formats, including both all-day and half-day programs. Some of them are held at the public school the child will eventually attend.

Since its inception, there has been debate about Head Start's effectiveness. Research has shown that children enrolled in Head Start enjoy immediate, measurable gains in cognitive test scores; however, researchers disagree as to the long-term impact. Some research has shown that Head Start has long-term effects on academic ability and success that do not fade over time. These effects include: persistent gains in achievement test scores, fewer occurrences of grade retention, and less placement in **special education** programs. Other long-term benefits include higher high school graduation rates and decreased crime and delinquency rates. As adults, Head Start graduates are more likely to get better jobs and earn more money. On the other hand, some experts believe the research shows that disadvantaged children in Head Start start off a step behind and never catch up. One of the primary concerns about the program is with its teachers, who only subsequently were required to have a two-year degree and who made less than half the average salary of a public school teacher. To help determine Head Start's effectiveness, a research project called The National Head Start Impact Study was underway as of 2004. It intends to follow between 5,000 to 6,000 preschool aged children through 2006 to determine if Head Start is effective and how Head Start works best for children.

PUBLIC PRESCHOOLS A growing number of states have started to fund preschool programs offered at public schools, called pre-kindergarten (or pre-K) programs. They may be administered by the local school board or by an independent contractor paid by the state. Like private preschools, they may operate for a full day or just half a day.

Preschool class. *(© Ariel Skelley/Corbis.)*

Most state-run preschool programs began like Head Start and focused their services on children with the greatest needs, either children with disabilities or children from low-income families. Most states in the early 2000s choose to have their prekindergarten programs serve children in low-income families or children who have other risk factors that place them at greater risk of school failure or educational difficulties. These risk factors may include having a disability, being a child of teen parents, or having limited proficiency in the English language. Georgia was the first state to have a universally available pre-K program, which was started in 1995. It is still the only state to make preschool available to all students. Other states, including West Virginia and Florida, are making long-term plans to move toward universal prekindergarten.

Research tends to find that public preschool programs (public schools and Head Start) exhibit a greater effect on children than do private preschools. One of the reasons is public school programs provide the same quality of services whether children are rich or poor, while private provider quality is lower for children from lower-income families. It may be an issue of getting what a parent can pay for. Most of the long-term research on the effects of preschool focuses on low-income children. There is very little data on any long-term benefits for middle-class children.

Qualities of a good preschool

According to the National Institute for Early Education Research, the types of teaching activities and classroom emphases that contribute to a high-quality early education for children include the following:

- opportunities to learn persistence when working at tasks, direction following, and good listening skills
- focus on language and literacy skills, as well as interactive book reading
- emphasis on teaching children problem-solving skills
- helping children expand their knowledge and increase their vocabulary
- opportunities to learn beginning skills involving the alphabet, numbers, and spatial awareness

- focus on scientific thinking skills as well as information about the everyday environment, the world, and how things work
- emphasis on teaching early literacy and mathematics through a variety of activities and projects
- opportunity for preschoolers to engage in music, art, and dramatic play
- educational program in which parents are involved and have opportunities to watch and take part in classroom activities

Advantages of preschool

Many children who attend high-quality preschool programs have their lives changed for the better. In the first five years of life, children acquire the basic capabilities that prepare them for later success in school and life. Many studies show that high-quality preschools improve achievement, behavior, and school readiness for economically disadvantaged children. Follow-up research with these same children shows that they earn more money, experience more stable home lives, and become more responsible citizens than they would have if they had not attended preschool. Children who attend preschool are better prepared to enter kindergarten, both academically and socially. Whatever their format, preschools offer parents and children typical benefits. A good program can help children develop their gross and **fine motor skills**, improve their language and communication abilities, and exercise their **creativity**.

Disadvantages of preschool

The greatest academic and social progress seen in preschools is in children from deprived backgrounds. However, few programs have the quality necessary to bring about the benefits promised. The costs of a high-quality program can be far greater than the costs of education at some public universities. Most children in preschool, however, are not disadvantaged, and some researchers believe the same gains can be had at home by providing educational **toys**, games and books for the child. In some preschools, the emphasis on groups might mean that children will not receive the individual attention they require. This is a particular risk if the preschool does not follow the National Association for the Education of Young Children's recommended teacher-to-child ratio of no more than ten preschoolers per staff member. One-on-one instruction is an advantage parents will not likely find in any preschool. Opportunities for playing with other children exist in churches, clubs, and other outlets, where the child can learn social skills. Some believe that what children need most is lots of play and free time and close interaction with their parents, something that may be compromised if the child is away from home for long periods of time. Another disadvantage is that some children experience acute **separation anxiety**, indicating that they are not yet ready to make the transition to the preschool environment. Many programs also expect the child to be toilet-trained, a milestone that not all children have achieved at the preschool age.

KEY TERMS

Head Start—A federal program started in 1965 that provides free education for young children in many low-income families across the United States.

Preschool—An early childhood program in which children combine learning with play in a program run by professionally trained adults.

Common problems

When selecting a preschool for their child, parents should be aware of certain problems or warning signs that might make them decide to look at a different preschool provider. These problems or warning signs may include:

- negative reactions from other parents
- inattention to established rules and regulations (Schools should have clearly established written guidelines for everything from operating hours to managing emergencies.)
- lack of a sick-child policy (The preschool should require both staff and children to have current immunizations and regular checkups.)
- indicating they are hiding something, schools that balk at parents dropping by unannounced
- schools that either have no structure whatsoever or a structure that is inflexible
- lack of age-appropriate activities and toys
- an underqualified staff
- large class sizes
- dirty, unsafe facilities
- an expired license
- schools that promise to put a child on an academic fast track (These highly structured, intensive preschool academic programs create inappropriate expectations from children and may cause emotional stress.)

Parental concerns

Parents considering sending their child to preschool should investigate several different ones and consider many factors before choosing one. However, parents should realize that in spite of the potential advantages, preschool may not be for every child. Parents can be assured that there are alternative ways of introducing their child to early academic skills and social activities.

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Prickly heat

Definition

Prickly heat is also known as sweat retention syndrome or miliaria rubra. It is a common disorder of the sweat glands.

Description

The skin contains two types of glands. One type produces oil and the other produces sweat. Sweat glands are coil-shaped and extend deep into the skin. They are capable of plugging up at several different depths, producing four distinct skin **rashes**.

- Miliaria crystallina is the most superficial of the occlusions. At this level, only the thin upper layer of skin is affected. Little blisters of sweat that cannot escape to the surface form. A bad **sunburn** as it just starts to blister can look exactly like this condition.
- Deeper plugging causes miliaria rubra as the sweat seeps into the living layers of skin, where it irritates and itches.
- Miliaria pustulosis (a complication of miliaria rubra) occurs when the sweat is infected with pyogenic bacteria and turns to pus.
- Deeper still is miliaria profunda. The skin is dry and goose bumps may or may not appear.

There are two requirements for each of these phases of sweat retention: hot enough weather to induce sweating, and failure of the sweat to reach the surface.

Demographics

Infants are more likely to get miliaria rubra than adults. All the sweat retention rashes are also more likely to occur in hot, humid weather.

Causes and symptoms

As of 2004, the best evidence suggested that bacteria form the plugs in the sweat glands. These bacteria are probably normal inhabitants of the skin, and why they suddenly interfere with sweat flow is still not known.

Besides **itching**, these conditions prevent sweat from cooling the body, which it is supposed to do by evaporating from the skin surface. Sweating is the most important cooling mechanism available in hot environments. If it does not work effectively, the body can rapidly become too hot.

When to call the doctor

A doctor should be called when an infant's temperature rises above 100°F (37.8°C) and cannot be brought down within a few minutes. Infants whose temperatures exceed 102°F (38.9°:C) should be immersed in tepid or lukewarm water to reduce body temperature slowly.

A physician should be notified if a baby becomes dehydrated. Signs of **dehydration** include lethargy, poor skin tone, generalized weakness, and reduced urination.

Diagnosis

Rash and dry skin in hot weather are usually sufficient to diagnose these conditions.

Treatment

The rash itself may be treated with topical antipruritics (itch relievers). Preparations containing aloe, menthol, camphor, eucalyptus oil, and similar ingredients are available commercially. Even more effective, particularly for widespread itching in hot weather, are tepid baths with corn starch and/or oatmeal (about 0.5 lb [224 g] of each per bathtub-full).

Dermatologists can peel off the upper layers of skin using a special ultraviolet light. This procedure removes the plugs and restores sweating but is not necessary in most cases.

Much more important, however, is to realize that the body cannot cool itself adequately without sweating. Careful monitoring for symptoms of heat disease is important. If they appear, some decrease in the ambient temperature must be achieved by moving to the shade, taking a tepid bath or shower, or turning up the air conditioner.

Prognosis

The rash disappears in a day with cooler temperatures, but the skin may not recover its ability to sweat for two weeks—the time needed to replace the top layers of skin with new growth from below.

Prevention

Experimental application of topical antiseptics such as hexachlorophene almost completely prevent the rashes of prickly heat. Parents should consult their doctors before applying such antiseptics to their child's skin.

Nutritional concerns

Babies should receive adequate water and other liquids during periods of high heat. They should consume adequate amounts of electrolytes such as sodium, chloride, potassium, phosphate and bicarbonate during hot weather.

KEY TERMS

Ambient—Surrounding.

Pyogenic—Capable of generating pus. *Streptococcus, Staphocococcus,* and bowel bacteria are the primary pyogenic organisms.

Syndrome—A group of signs and symptoms that collectively characterize a disease or disorder.

A physician should be notified if a baby becomes dehydrated. Signs of dehydration include lethargy, poor skin tone, and generalized weakness. If oral replacement of fluid and electrolytes is suggested, then commercial or homemade preparations can be used. Commercial preparations such as Pedialyte are available. The World Health Organization has provided the following recipe for home preparation, which can be administered in small, frequent sips:

- table salt, 3/4 tsp
- baking powder, 1 tsp
- orange juice, 1 cup
- water, 1 quart

Parental concerns

Parents should carefully monitor their young children for symptoms of heat disease. Babies should be carefully bathed to maintain normal sweating, especially during periods of hot weather.

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Protein-energy malnutrition

Definition

Protein-energy **malnutrition** (PEM) is a potentially fatal body-depletion disorder. It is the leading cause of death in children in developing countries.

Description

PEM is also referred to as protein-calorie malnutrition. It develops in children whose consumption of protein and energy (measured by calories) is insufficient to satisfy their nutritional needs. While pure protein deficiency can occur when a person's diet provides enough energy but lacks an adequate amount of protein, in most cases deficiency will exist in both total calorie and protein intake. PEM may also occur in children with illnesses that leave them unable to absorb vital nutrients or convert them to the energy essential for healthy tissue formation and organ function.

Types of PEM

Primary PEM results from a diet that lacks sufficient sources of protein. Secondary PEM is more common in the United States, where it usually occurs as a complication of **AIDS**, **cancer**, chronic kidney failure, inflammatory bowel disease, and other illnesses that impair the body's ability to absorb or use nutrients or to compensate for nutrient losses. PEM can develop gradually in a child who has a chronic illness or experiences chronic semi-starvation. It may appear suddenly in a patient who has an acute illness.

Kwashiorkor, also called wet protein-energy malnutrition, is a form of PEM characterized primarily by protein deficiency. This condition usually appears at about the age of 12 months when breast-feeding is discontinued, but it can develop at any time during a child's formative years. It causes fluid retention (edema); dry, peeling skin; and hair discoloration.

Marasmus, a PEM disorder, is caused by total calorie/energy depletion rather than primarily protein calorie/energy depletion. Marasmus is characterized by stunted growth and wasting of muscle and tissue. Marasmus usually develops between the ages of six months and one year in children who have been weaned from breast milk or who suffer from weakening conditions such as chronic **diarrhea**.

Demographics

It is not entirely clear how common PEM is in the United States. Primary PEM is common in impoverished areas of developing countries. In the United States, secondary PEM is more common. Children at particular risk for secondary PEM are those who have diseases that involve diarrhea or that otherwise interfere with nutrient absorption. Children with chronic illnesses that require frequent **hospitalization** are more likely to develop PEM.

Causes and symptoms

Secondary PEM symptoms range from mild to severe, and can alter the form or function of almost every organ in the body. The type and intensity of symptoms depend on the patient's prior nutritional status, the nature of the underlying disease, and the speed at which the PEM is progressing.

Mild, moderate, and severe classifications for PEM have not been precisely defined, but patients who lose 10–20 percent of their body weight without trying may have moderate PEM. Some of the cause is replacement dependent (i.e. patients do not take in adequate protein during recovery from illness). This level of PEM is

characterized by a weakened grip and inability to perform high-energy tasks.

Losing 20 percent of body weight or more is generally classified as severe PEM. Children with this condition cannot eat normal-sized meals. They have slow heart rates and low blood pressure and body temperatures. Other symptoms of severe secondary PEM include baggy, wrinkled skin; **constipation**; dry, thin, or brittle hair; lethargy; pressure sores, and other skin lesions.

Children suffering from kwashiorkor often have extremely thin arms and legs, but liver enlargement and ascites (abnormal accumulation of fluid) can distend the abdomen and disguise weight loss. Hair may turn red or yellow. Anemia, diarrhea, and fluid and electrolyte disorders are common. The body's immune system is often weakened, behavioral development is slow, and **mental retardation** may occur. Children may grow to normal height but are abnormally thin.

Kwashiorkor-like secondary PEM usually develops in children who have been severely burned, suffered trauma, or had sepsis (massive tissue-destroying infection) or another life-threatening illness. The condition's onset is so sudden that body fat and muscle mass of normal-weight people may not change. Some patients even gain weight because of fluid retention.

Profound weakness accompanies severe marasmus. Since the body breaks down its own tissue to use for energy, children with this condition lose all their body fat and muscle strength, and acquire a skeletal appearance most noticeable in the hands and in the temporal muscle in front of and above each ear. Children with marasmus are small for their age. Since their immune systems are weakened, they suffer from frequent infections. Other symptoms include loss of appetite, diarrhea, skin that is dry and baggy, sparse hair that is dull brown or reddish yellow, mental retardation, behavioral retardation, low body temperature (hypothermia), and slow pulse and breathing rates.

The absence of edema (fluid retention) distinguishes marasmus-like secondary PEM, a gradual wasting process that begins with weight loss and progresses to mild, moderate, or severe malnutrition (cachexia). It is usually associated with cancer, chronic obstructive pulmonary disease (COPD), or another chronic disease that progresses very slowly.

Difficulty chewing, swallowing, and digesting food, **pain**, **nausea**, and lack of appetite are among the most common reasons that many hospital patients do not consume enough nutrients. Nutrient loss can be accelerated by bleeding, diarrhea, abnormally high blood sugar levels (glycosuria), kidney disease, malabsorption disorders, and other factors. **Fever**, infection, surgery, and benign or malignant tumors increase the amount of nutrients that hospitalized patients need. Trauma, **burns**, and some medications also increase caloric requirements.

When to call the doctor

The doctor should be consulted if a child has lost a significant amount of weight without trying, has persistent diarrhea, or has any other signs of PEM.

Diagnosis

When the physician suspects PEM, A thorough physical examination is performed, and these areas assessed:

- eating habits and weight changes
- body-fat composition and muscle strength
- gastrointestinal symptoms
- presence of underlying illness
- developmental delays and loss of acquired milestones in children
- nutritional status

Doctors further quantify a patient's nutritional status by:

- comparing height and weight to standardized norms
- calculating body mass index (BMI)
- measuring skinfold thickness or the circumference of the upper arm

Treatment

Treatment is designed to provide adequate **nutrition**, restore normal body composition, and cure the condition that caused the deficiency. Tube feeding or intravenous feeding is used to supply nutrients to patients who cannot or will not eat protein-rich foods.

In patients with severe PEM, the first stage of treatment consists of correcting fluid and electrolyte imbalances, treating infection with **antibiotics** that do not affect protein synthesis, and addressing related medical problems. The second phase involves replenishing essential nutrients slowly to prevent taxing the patient's weakened system with more food than it can handle. Physical therapy may benefit patients whose muscles have deteriorated significantly.

Prognosis

Most children can lose some of their body weight without side effects, but losing more than 40 percent is usually fatal. Death usually results from heart failure, an

KEY TERMS

Electrolytes—Salts and minerals that produce electrically charged particles (ions) in body fluids. Common human electrolytes are sodium chloride, potassium, calcium, and sodium bicarbonate. Electrolytes control the fluid balance of the body and are important in muscle contraction, energy generation, and almost all major biochemical reactions in the body.

electrolyte imbalance, or low body temperature. Patients with certain symptoms, including semi- consciousness, persistent diarrhea, **jaundice**, and low blood sodium levels, have a poorer prognosis than other patients. Recovery from marasmus usually takes longer than recovery from kwashiorkor. The long-term effects of childhood malnutrition are uncertain. Some children recover completely, while others may have a variety of lifelong impairments, including an inability to properly absorb nutrients in the intestines, as well as mental retardation. The outcome appears to be related to the length and severity of the malnutrition, as well as to the age of the child when the malnutrition occurred.

Prevention

Breastfeeding a baby for at least six months is considered the best way to prevent early-childhood malnutrition. Talking to a doctor before putting a child on any kind of diet, such as vegan, vegetarian, or low-carbohydrate, can help assure that the child gets the full supply of nutrients that he or she needs.

Every child being admitted to a hospital should be screened for the presence of illnesses and conditions that could lead to PEM. The nutritional status of patients at higher-than-average risk should be more thoroughly assessed and periodically reevaluated during extended hospital stays.

Parental concerns

Protein-energy malnutrition is fairly easily treated. If, however, it occurs for a prolonged period, it can have very serious and permanent health consequences. It is important to ensure that children are getting a healthy and balanced diet. Children should not be put on weight loss or other special diets without first consulting a pediatrician.

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ORGANIZATIONS

American Academy of Pediatrics.141 Northwest Point Boulevard, Elk Grove Village, IL 60007-1098. (847) 434-4000 Fax: (847) 434-8000. <www.aap.org>

American Dietetic Association. 120 South Riverside Plaza, Suite 2000 Chicago, IL 60606-6995. (800) 877-1600. <www.eatright.org>

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Prozac *see* **Antidepressants**

Pseudoephedrine *see* **Decongestants**

Psoriasis

Definition

Named for the Greek word *psōra* meaning itch, psoriasis is a chronic, non-contagious disease characterized by inflamed lesions covered with silvery-white scabs of dead skin.

Description

Normal skin cells mature and replace dead skin every 28 to 30 days. In psoriasis, the immune system triggers the immune system to make T cells, a type of white blood cell, that cause skin cells to mature in two to three days. Because the body cannot shed old skin as rapidly as the new cells appear, raised patches of dead skin form on the body.

Psoriasis is considered mild if it affects less than 5 percent of the surface of the body; moderate, if 5 to 30 percent of the skin is involved, and severe, if the disease affects more than 30 percent of the body surface.

There is no cure for psoriasis. The disease is managed through treatment. Psoriasis can seriously impact children's lives when the hands and feet are affected so the children cannot take notes or walk or **play**, or when the disease becomes so widespread that the immune system becomes compromised. Children also experience low **self-esteem** and depression because of the disfiguring aspects of the disease.

Types of psoriasis

Dermatologists distinguish different forms of psoriasis according to what part of the body is affected, how severe symptoms are, how long they last, and the pattern formed by the scales. Though children usually have only one form of the disease, some do experience two more types of psoriasis throughout their lifetimes.

PLAQUE PSORIASIS Plaque psoriasis (psoriasis vulgaris), the most common form of the disease, is characterized by small, red bumps that enlarge, become inflamed, and form scales. The top scales flake off easily and often, but those beneath the surface of the skin clump together. Removing these scales exposes tender skin, which bleeds and causes the plaques (inflamed patches of skin) to grow.

Plaque psoriasis can develop on any part of the body, but most often occurs on the elbows, knees, scalp, and trunk. Patches of psoriasis are found in the scalp for nearly half of all psoriasis sufferers.

GUTTATE PSORIASIS Named for the Latin word *gutta*, which means "a drop," guttate psoriasis is characterized by small, red, drop-like dots that enlarge rapidly and may be somewhat scaly. Often found on the arms, legs, trunk, scalp, and sometimes in the diaper area, guttate psoriasis can clear up without treatment or disappear and resurface in the form of plaque psoriasis.

Guttate psoriasis is the most common form of psoriasis in children. It usually first appears in children around four or five years old after a streptococcal infection.

PUSTULAR PSORIASIS Pustular psoriasis usually occurs in adults but can occur in children and adolescents. It is characterized by blister-like lesions filled with non-infectious pus and surrounded by reddened skin. Pustular psoriasis, which can be limited to one part of the body or can be widespread, may be the first symptom of psoriasis or develop in a patient with chronic plaque psoriasis.

Generalized pustular psoriasis is also known as Von Zumbusch pustular psoriasis. Widespread, acutely painful patches of inflamed skin develop suddenly. Pustules appear within a few hours, then dry, and peel within two days. It can make life-threatening demands on the heart and kidneys.

Palomar-plantar pustulosis (PPP) generally appears between the ages of 20 and 60.

INVERSE PSORIASIS Inverse psoriasis occurs in the armpits and groin, under the breasts, and in other areas where skin flexes or folds. This disease is characterized by smooth, inflamed lesions and can be debilitating.

ERYTHRODERMIC PSORIASIS Characterized by severe scaling, **itching**, and **pain** that affects most of the body, erythrodermic psoriasis disrupts the body's chemical balance and can cause severe illness or even death when the body's immune system becomes compromised. Erythrodermic psoriasis interferes with the body's ability to control temperature and prevent infections. This particularly inflammatory form of psoriasis can be the first sign of the disease but often develops in patients with a history of plaque psoriasis.

PSORIATIC ARTHRITIS About 10 percent of patients with psoriasis develop a complication called psoriatic arthritis. This type of arthritis can be slow to develop and mild, or it can develop rapidly. Symptoms of psoriatic arthritis include:

- joint discomfort, swelling, stiffness, or throbbing
- swelling in the toes and ankles
- pain in the digits, lower back, wrists, knees, and ankles
- eye inflammation or pink eye (conjunctivitis)

Children who have psoriatic arthritis also have nail deformations, usually pitting of the fingernails or toenails. Size, shape, and depth of the marks vary, and affected nails may thicken, yellow, or crumble. The skin around an affected nail is sometimes inflamed, and the nail may peel away from the nail bed.

Demographics

Psoriasis affects 4.5 million Americans and is slightly more common in women than in men. Although the disease can develop at any time, a third of all cases occur in childhood with 10 to 15 percent of them being diagnosed in children under ten. It appears between the ages of 15 and 35. It is rare in infants but does occur. Nearly 20,000 U.S. children are diagnosed with psoriasis every year. Psoriasis affects people of all ethnicities, but fair-skinned individuals have a slightly higher incidence.

About 1.5 million Americans have moderate to severe psoriasis. Of them, 75 percent report that their disease has a serious impact on their daily lives. One-third

report sleeping problems, disruptions with their normal routine, and negative self-image because of the disease.

In adults, psoriasis can be serious enough that four hundred people are granted disability by the Social Security Administration each year, and having psoriasis disqualifies individuals from serving in the military. Annually, three hundred and fifty people die annually from psoriasis or complications of treatment.

Nearly one million people in the United States have psoriatic arthritis. Though psoriatic arthritis usually develops between the age of 30 and 50, it does occur in children. About 10 to 30 percent of psoriasis patients have psoriatic arthritis, but the condition can occur before the characteristic scaly lesions occur.

Having one parent with psoriasis increases a child's risk of developing the disease to 20 to 25 percent. If both parents have psoriasis, the risk is doubled.

Patients with psoriasis make 2.4 million visits to dermatologists each year, with costs exceeding $3 million annually.

Causes and symptoms

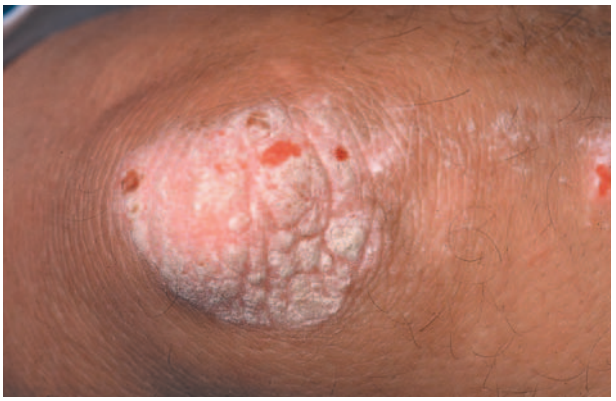
Causes

The cause of psoriasis is, as of 2004, unknown, but research suggests that it is genetic and is related to the immune-system. Having both parents with the disease increases a child's risk by 50 percent.

Psoriasis is usually cyclical, with episodes flaring up for weeks or months throughout the child's life and then receding. Certain factors, however, do seem to trigger bouts of the disease. Injury to the skin seems to precipitate many episodes of plaque psoriasis, usually within seven to ten days. This is called the Koebner reaction. **Streptococcal infections** are associated with guttate psoriasis and some plaque psoriasis cases. Both trauma and certain bacteria may also trigger psoriatic arthritis.

Environmental factors are also implicated in reoccurrence of psoriasis. Exposure to cold temperatures can trigger episodes of the disease. Though sunlight is usually beneficial to most patients, for a few children, too much sun can cause a flare up or worsen the condition.

Some drugs have been found to aggravate psoriasis. Antimalarial drugs, beta-blockers used to treat high blood pressure, and lithium, a drug used to treat depression and bi-polar disorder, can make episodes worse in some individuals. Non-steroid anti-inflammatory (NSAID) drugs, such as ibuprofen or naproxen used to manage pain and inflammation can also aggravate psoriasis.



Psoriasis, a chronic skin disorder, may appear on any area of the body, including the elbow, as shown above. *(Photograph by Scott Camazine. Photo Researchers, Inc.)*

During **puberty**, adolescents report more frequent flare ups and more severe ones. The hormonal changes within their bodies seem to trigger the immune system.

Stress is also a factor in increased frequency of psoriatic episodes. Because stress pumps large amounts of adrenalin, a hormone, into the body, the immune system is overstimulated and reacts by triggering flare ups of the disease.

Symptoms

The most common symptoms of psoriasis are skin **rashes** or red patches covered with white scales that may itch or burn. In plaque psoriasis, the skin may crack and bleed and is susceptible to infection. When the scales are removed, the skin underneath is deep red and shiny and may bleed. Psoriasis on the scalp is distinguished from **seborrheic dermatitis**, or dandruff, because the scales of psoriasis are dry, not greasy. There may be a red drop-like rash (guttate psoriasis) or patches of scaly skin that crack and ooze pus (pustular psoriasis).

In young children, the scaly patches in plaque psoriasis do not appear as thick or as scaly as those of adults. Psoriasis appears often in the diaper area and affects the face more in children than adolescents or adults.

When to call the doctor

Many children routinely see their doctors to supervise their regime of treatment for psoriasis flare ups. Others only see their doctors at the first sign of a recurrence of the disease. There are circumstances, however, when the doctor should be notified. If a treatment does

not seem to be working, episodes worsen with treatment, or the child experiences a serious side effect to medications give, the doctor should be consulted to discuss alternative treatment. If there are signs of infections, such as red streaks on the skin or pus, or if there is **fever** or increased pain, the doctor should be called immediately.

Diagnosis

A complete medical history and examination of the skin, nails, and scalp are the basis for a diagnosis of psoriasis. In some cases, a microscopic examination of skin cells is also performed.

Blood tests can distinguish psoriatic arthritis from other types of arthritis. Rheumatoid arthritis, in particular, is diagnosed by the presence of a particular antibody present in the blood. That antibody is not present in the blood of patients with psoriatic arthritis.

Treatment

Age, general health, lifestyle, and the severity and location of symptoms influence the type of treatment used to reduce inflammation and decrease the rate at which new skin cells are produced. Because the course of this disease varies with each individual, doctors must experiment with or combine different treatments to find the most effective therapy for a particular patient.

Treating children with this disease with drugs is problematic. Though treatment regimes have been developed that are effective on adults, research has not been conducted sufficiently on children, except in the area of psoriatic arthritis. Treatment in children is usually not aggressive because of their small, developing bodies. Long-term use can produce toxicity so potent drugs, such as methotrexate (MTX) and cyclosporine, are not used with children. Although MTX is sometimes used in extreme cases for brief amounts of time. Topical steriods are also not used on children because their bodies can absorb the steriods in the medication.

Mild psoriasis

Typically, steroid creams and ointments are commonly used to treat mild or moderate psoriasis in adults. These topical ointments are not generally used with children for mild psoriasis. However, new creams that are used in treating eczema appear to be effective in treating psoriasis as well and do not appear to have long-term problems. In addition, tazarotene (Tazorac), a drug approved by the United States Food and Drug Administration (FDA) in 1997, is proving to be effective for mild-to-moderate plaque psoriasis. This water-based gel has chemical properties similar to vitamin A.

A more subdued approach is undertaken with children who have less severe psoriasis. Brief daily doses of natural sunlight can significantly relieve most symptoms. **Sunburn**, however, has the opposite effect.

Moisturizers and bath oils are used to loosen scales, soften skin, and eliminate the itch. Adding a cup of oatmeal to a tub of bath water is also helpful. Salicylic acid (an ingredient in aspirin) can be used to remove dead skin or increase the effectiveness of other therapies.

Moderate psoriasis

Administered under medical supervision, ultraviolet light B (UVB) is used to control psoriasis that covers many areas of the body or that has not responded to topical preparations. Doctors combine UVB treatments with topical medications to treat some patients and sometimes prescribe home phototherapy, in which the parent administers the UVB treatments.

Tanning beds use ultraviolet A and produce a more intense experience. Adolescents should avoid tanning salons and should sunbathe but without tanning. Any sun exposure or UVB treatment should be coordinated with a dermatologist.

Severe psoriasis

Methotrexate (MTX), given as a pill or as an injection, is sometimes used in extreme cases to alleviate symptoms of severe psoriasis or psoriatic arthritis. Patients who take MTX must be carefully monitored to prevent liver damage.

Enbrel is another drug dermatologists prescribe for children. It appears to be very safe when used for long periods of time.

A new self-injected medication called efalizumab (Raptiva) has the potential to be effective for severe cases of psoriasis. Since it is suppresses the immune system, its use with children or over the long-term is cautioned because it can increase the risk of infection.

Psoriatic arthritis can also be treated with NSAIDs, such as **acetaminophen** (Tylenol) or aspirin. Hot compresses and warm water soaks may also provide some relief for painful joints.

Photochemotherapy (PUVA) is a medically supervised procedure that combines medication with exposure to ultraviolet light (UVA) to treat localized or widespread psoriasis. An individual with widespread psoriasis that has not responded to treatment may enroll in one

of the day treatment programs conducted at special facilities throughout the United States. Psoriasis patients who participate in these intensive sessions are exposed to UVA and given other treatments for six to eight hours a day for two to four weeks.

Alternative treatment

Non-traditional psoriasis treatments include:

- soaking in warm water and German chamomile (*Matricaria recutita*) or bathing in warm salt water
- drinking as many as three cups a day of hot tea made with one or a combination of the following herbs: burdock (*Arctium lappa*) root, dandelion (*Taraxacum mongolicum*) root, Oregon grape (*Mahonia aquifolium*), sarsaparilla (*Smilax officinalis*), and balsam pear (*Momardica charantia*)
- taking two 500-mg capsules of evening primrose (*Oenothera biennis*) oil a day (Pregnant women should not use evening primrose oil, and patients with liver disease or **high cholesterol** should use it only under a doctors supervision.)
- eating a diet that includes plenty of fish, turkey, celery (for cleansing the kidneys), parsley, lettuce, lemons (for cleansing the liver), limes, fiber, and fruit and vegetable juices
- eating a diet that eliminates animal products high in saturated fats, since they promote inflammation
- drinking plenty of water (at least eight glasses) each day
- taking nutritional supplements including **folic acid**, lecithin, vitamin A, vitamin E, selenium, and zinc
- regularly imagining clear, healthy skin

Other helpful alternative approaches include identifying and eliminating food allergens from the diet, enhancing the function of the liver, augmenting the hydrochloric acid in the stomach, and completing a detoxification program. Constitutional homeopathic treatment, if properly prescribed, can also help resolve psoriasis.

Prognosis

Most cases of psoriasis can be controlled, and most people who have psoriasis can live normal lives. However, some people who have psoriasis are so self-conscious and embarrassed about their appearance that they become depressed and withdrawn. Others may become disabled because of psoriatic arthritis or because their psoriasis affects their hands and feet so that they cannot walk or handle objects.

Prevention

Psoriasis cannot be prevented. However, recurrences can be avoided or minimized by maintaining a healthy lifestyle by getting plenty of **sleep**, eating a balanced diet, participating in regular **exercise**, and minimizing stress. Avoiding overexposure to cold temperatures, sunburn, and skin irritants, such as drying soaps and lotions, can also minimize flare-ups. Not **smoking** or drinking alcohol can also prevent or minimize some episodes.

Parental concerns

Children living with psoriasis often find the disease overwhelming. It is an emotionally charged disease that can have a child feeling anger one minute and deep depression the next. Because the disfigurement of their skin, though often temporary, is sometimes quite pronounced, children will turn inward, avoiding contact with friends or relatives. School can be particularly traumatizing due to teasing by other children. Teenagers, who already feel awkward and ugly, may feel worse during flare-ups of the disease. Complicating this already emotional situation is the discouragement of treatments that do not work as expected and the uncertainty of finding something that will work.

Parents can help their children by providing education about the psoriasis. This is the first step in managing the disease and feeling some control in their lives. Sometimes this education includes discussing the disease with the childs teachers or the parents of their friends so that these adults will understand more about the emotional state of the child.

Parents can listen to their children when they are able to talk about their feelings about the disease. Emphasizing their childrens strengths, especially when these children appear sad or depressed, and encouraging them to stay active and see their friends can help a child cope with the disease.

Sometimes, participating in a childrens psoriasis support group may be helpful. In addition, sending the child to a special camp for school age children with childhood skin diseases can help them learn tools for coping with the disease as well as establish a support system.

Children can often feel shame as well as guilt, thinking that they have somehow brought on the disease. Coupled with anger and resentment, these powerful emotions can contribute to stress, which can trigger the recurrence of the disease. Stress reduction techniques, such as exercise, **yoga**, and meditation, are also helpful.

KEY TERMS

Adrenaline—Another name for epinephrine, the hormone released by the adrenal glands in response to stress. It is the principal blood-pressure raising hormone and a bronchial and intestinal smooth muscles relaxant.

Arthritis—A painful condition that involves inflammation of one or more joints.

Plaque—Inflamed patches of skin present in some forms of psoriasis.

T cell—A type of white blood cell that is produced in the bone marrow and matured in the thymus gland. It helps to regulate the immune system's response to infections or malignancy.

Parents should be available to their child and offer as much tangible and emotional support they can. However, they should not encourage the child to become too dependent on the parent or others. Parents can also help children find creative solutions to deal with teasing, camouflage their lesions, and educate their peers about the disease. One of the most important lessons parents can teach their child, who is living with psoriasis, is not to be embarrassed because of the disease. Psoriasis can be treated matter-of-factly as people do diabetes, another chronic disorder.

See also Depressive disorders; Itching; Self-esteem.

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American Academy of Dermatology. 930 N. Meacham Road, PO Box 4014, Schaumburg, IL 601684014. Web site: <www.aad.org>.

American Skin Association Inc. 150 E. 58th St., 3rd floor, New York, NY 101550002. Web site: <www.americanskin.org>.

National Psoriasis Foundation. 6600 SW 92nd Ave., Suite 300, Portland, OR 97223. Web site: <www.psoriasis.org>.

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Psychological tests

Definition

Psychological tests are written, visual, or verbal evaluations administered to assess the cognitive and emotional functioning of children and adults.

Purpose

Psychological tests are used to assess a variety of mental abilities and attributes, including achievement and ability, personality, and neurological functioning.

For children, academic achievement, ability, and **intelligence** tests may be used as tools in school placement, in determining the presence of a learning disability or a **developmental delay**, in identifying giftedness, or in tracking intellectual development. Intelligence testing may also be used with teens and young adults to determine vocational ability (e.g., in career counseling).

Personality tests are administered for a wide variety of reasons, from diagnosing psychopathology (e.g., personality disorder, depressive disorder) to screening job candidates. They may be used in an educational setting to determine personality strengths and weaknesses.

Description

Psychological tests are formalized measures of mental functioning. Most are objective and quantifiable; however, certain projective tests may involve some level of subjective interpretation. Also known as inventories, measurements, questionnaires, and scales, psychological tests are administered in a variety of settings, including preschools, primary and secondary schools, colleges and

universities, hospitals, outpatient healthcare settings, and social agencies. They come in a variety of formats, including written, verbal, and computer administered.

Achievement and ability tests

Achievement and ability tests are designed to measure the level of a child's intellectual functioning and cognitive ability. Most achievement and ability tests are standardized, meaning that norms were established during the design phase of the test by administering the test to a large representative sample of the test population. Achievement and ability tests follow a uniform testing protocol, or procedure (i.e., test instructions, test conditions, and scoring procedures) and their scores can be interpreted in relation to established norms. Common achievement and ability tests include the Wechsler intelligence scale for children (WISC-III) and the **Stanford-Binet intelligence scales**.

Personality tests

Personality tests and inventories evaluate the thoughts, emotions, attitudes, and behavioral traits that comprise personality. The results of these tests can help determine a child's personality strengths and weaknesses, and may identify certain disturbances in personality, or psychopathology. Tests such as the **Minnesota Multiphasic Personality Inventory** for Adolescents (MMPI-A) and the Millon Pre-Adolescent Clinical Inventory III (M-PACI), are used to screen children for specific psychopathologies or emotional problems.

Another type of personality test is the projective personality **assessment**. A projective test asks a child to interpret some ambiguous stimuli, such as a series of inkblots. The child's responses provide insight into his or her thought processes and personality traits. For example, the Holtzman Ink blot Test (HIT) uses a series of inkblots that the test subject is asked to identify. Another projective assessment, the **Thematic Apperception Test** (TAT), asks the child to tell a story about a series of pictures. Some consider projective tests to be less reliable than objective personality tests. If the examiner is not well-trained in psychometric evaluation, subjective interpretations may affect the evaluation of these tests.

Neuropsychological tests

Children and adolescents who have experienced a traumatic brain injury, brain damage, or other organic neurological problems, are administered neuropsychological tests to assess their level of functioning and identify areas of mental impairment. Neuropsychological tests may also be used to evaluate the progress of a patient who has undergone treatment or rehabilitation for a neurological injury or illness. In addition, certain neuropsychological measures may be used to screen children for developmental delays and/or learning disabilities.

Precautions

Psychological testing requires a clinically trained examiner. All psychological tests should be administered, scored, and interpreted by a trained professional, preferably a psychologist or psychiatrist with expertise in the appropriate area.

Psychological tests are only one element of a psychological assessment. They should never be used as the sole basis for a diagnosis. A detailed clinical and personal history of the child and a review of psychological, medical, educational, or other relevant records are required to lay the groundwork for interpreting the results of any psychological measurement.

Cultural and language differences among children may affect test performance and may result in inaccurate test results. The test administrator should be informed before psychological testing begins if the test taker is not fluent in English and/or belongs to a minority culture. In addition, the child's level of motivation may also affect test results.

Preparation

Prior to the administration of any psychological test, the administrator should provide the child and the child's parent with information on the nature of the test and its intended use, complete standardized instructions for taking the test (including any time limits and penalties for incorrect responses), and information on the confidentiality of the results. After these disclosures are made, informed consent should be obtained from the child (as appropriate) and the child's parent before testing begins.

Normal results

All psychological and neuropsychological assessments should be administered, scored, and interpreted by a trained professional. When interpreting test results, the test administrator will review with parents what the test evaluates, its precision in evaluation, any margins of error involved in scoring, and what the individual scores mean in the context of overall test norms and the specific background of the individual child.

Risks

There are no significant risks involved in psychological testing.

KEY TERMS

Norms—A fixed or ideal standard; a normative or mean score for a particular age group.

Psychopathology—The study of mental disorders or illnesses, such as schizophrenia, personality disorder, or major depressive disorder.

Quantifiable—A result or measurement that can be expressed as a number. The results of quantifiable psychological tests can be translated into numerical values, or scores.

Representative sample—A random sample of people that adequately represents the test-taking population in age, gender, race, and socioeconomic standing.

Standardization—The process of determining established norms and procedures for a test to act as a standard reference point for future test results.

Parental concerns

Test anxiety can have an impact on a child's performance, so parents should not place undue emphasis on the importance of any psychological testing. They should speak with their child before any scheduled tests and reassure them that their best effort is all that is required. Parents can also ensure that their children are well-rested on the testing day and have a nutritious meal beforehand.

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American Psychological Association. Testing and Assessment Office of the Science Directorate. 750 First St., N.E., Washington, D.C. 20002–4242. (202)336–6000. Web site: <www.apa.org/science/testing.html>.

National Association of School Psychologists. 4340 East West Highway, Suite 402, Bethesda, MD 20814. (301) 657–0270.

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Paula Ford-Martin

Psychosocial personality disorders

Definition

A psychosocial disorder is a mental illness caused or influenced by life experiences, as well as maladjusted cognitive and behavioral processes.

Description

The term psychosocial refers to the psychological and social factors that influence mental health. Social influences such as **peer pressure**, parental support, cultural and religious background, socioeconomic status, and interpersonal relationships all help to shape personality and influence psychological makeup. Children and adolescents with psychosocial disorders frequently have difficulty functioning in social situations and may have problems effectively communicating with others.

In the *Diagnostic and Statistical Manual of Mental Disorders 4th edition, text revision (DSM-IV-TR)* , the American Psychiatric Association distinguishes 16 different subtypes (or categories) of mental illness. Although psychosocial variables arguably have some degree of influence on all subtypes of mental illness, the major categories of mental disorders thought to involve significant psychosocial factors include:

- Substance-related disorders. Disorders related to alcohol and drug use, abuse, dependence, and withdrawal.
- **Schizophrenia** and other psychotic disorders. These include the schizoid disorders (schizophrenia, schizophreniform, and schizoaffective disorder), delusional disorder, and psychotic disorders.
- Mood disorders. Affective disorders such as depression (major, dysthymic) and bipolar disorders.
- **Anxiety** disorders. Disorders in which a certain situation or place triggers excessive **fear** and/or anxiety symptoms (i.e., **dizziness**, racing heart), such as panic disorder, agoraphobia, social phobia, **obsessive-compulsive disorder**, post-traumatic stress disorder, and generalized anxiety disorders. **Separation anxiety**

disorder is one permutation of anxiety disorders that is common in children.

- Somatoform disorders. Somatoform disorders involve clinically significant physical symptoms that cannot be explained by a medical condition (e.g., somatization disorder, conversion disorder, **pain** disorder, hypochondriasis, and body dysmorphic disorder).
- Factitious disorders. Disorders in which an individual creates and complains of symptoms of a non-existent illness in order to assume the role of a patient (or sick role).
- Sexual and **gender identity** disorders. Disorders of sexual desire, arousal, and performance. It should be noted that the categorization of gender identity disorder as a mental illness has been a point of some contention among mental health professionals.
- Eating disorders. Anorexia and bulimia nervosa.
- **Adjustment disorders**. Adjustment disorders involve an excessive emotional or behavioral reaction to a stressful event.
- Personality disorders. Maladjustments of personality, including conduct, paranoid, narcissistic, avoidant, dependent, and obsessive-compulsive personality disorder (not to be confused with the anxiety disorder OCD).
- Disorders usually first diagnosed in infancy, childhood, or **adolescence**. Some learning and developmental disorders (i.e., ADHD) may be partially psychosocial in nature.

Demographics

According to the National Institute of Mental Health, an estimated one in 10 children and adolescents in the United States suffers from mental illness severe enough to cause significant impairment in their day-to-day living. The MECA Study (Methodology for Epidemiology of Mental Disorders in Children and Adolescents) put the number even higher, estimating that nearly 21 percent of U.S. children between the ages of nine and 17 had a diagnosable mental disorder associated with at least minimum impairment.

Causes and symptoms

It is important to note that the causes of mental illness are diverse and not completely understood. The majority of psychological disorders are thought to be caused by a complex combination of biological, genetic (hereditary), familial, and social factors or biopsychosocial influences. In addition, the role that each of these plays can differ from person to person, so that a disorder such as depression that is caused by genetic factors in one person may be caused by a traumatic life event in another.

The symptoms of psychosocial disorders vary depending on the diagnosis in question. In addition to disorder-specific symptoms, children with psychosocial dysfunction usually have difficulty functioning normally in social situations and may have trouble forming and maintaining close interpersonal relationships.

When to call the doctor

Any child or adolescent that exhibits symptoms of psychosocial personality disorder should be taken to his or her health care provider as soon as possible for evaluation and possible referral to a mental health care professional. If a child or teen reveals at any time that he or she has had recent thoughts of self-injury or **suicide**, or if he or she demonstrates behavior that compromises personal **safety** or the safety of others, professional assistance from a mental health care provider or care facility should be sought immediately.

Diagnosis

Children with symptoms of psychosocial disorders or other mental illness should undergo a thorough physical examination and patient history to rule out an organic cause for the illness (such as a neurological disorder). If no organic cause is suspected, a psychologist or other mental healthcare professional will meet with the child and her parents or guardians to conduct an interview and take a detailed social and medical history. Interviews with caretakers and teachers may also be part of the diagnostic process.

The child and/or the child's parents may be asked to complete one or more psychological questionnaires or tests (also called clinical inventories, scales, or assessments). These may include the Children's Depression Inventory (CDI), the Diagnostic Interview Schedule for Children (DISC), Youth Self-Report, the School Social Behavior Scales (SSBS), the Overt Aggression Scale (OAS), Behavioral Assessment System for Children (BASC), Child Behavior Checklist (CBCL), the Nisonger Child Behavior Rating Form (N-CBRF), Clinical Global Impressions scale (CGI), the Minnesota Multiphasic Personality Inventory-2 (MMPI-2), and the Millon Adolescent Personality Inventory (MAPI).

Treatment

Counseling is typically a front-line treatment for psychosocial disorders. A number of counseling or talk therapy approaches exist, including psychotherapy, cognitive therapy, behavioral therapy, and group therapy. **Family therapy** may be recommended to help parents and siblings understand and cope with a child's mental illness. Therapy or counseling may be administered by social workers, nurses, licensed counselors and therapists, psychologists, or psychiatrists.

Psychotropic medication may also be prescribed for symptom relief in patients with mental disorders considered psychosocial in nature. For disorders such as major depression or **bipolar disorder**, which may have psychosocial aspects but also have known organic causes, drug therapy is a primary treatment approach. In cases such as personality disorder that are thought to not have biological roots, psychoactive medications are usually considered a secondary, or companion treatment to psychotherapy. It is important to note that there is limited data on the long-term repercussions of the use of most psychotropic medications in children and teens; the prescribing physician should present parents with an analysis of the risks and benefits of drug therapy before a course of treatment begins.

In some cases, treating mental illness requires **hospitalization**. This hospitalization, also known as inpatient treatment, is usually employed in situations where a controlled therapeutic environment is critical for the patient's recovery (e.g., rehabilitation treatment for **alcoholism** or other drug addictions), or when there is a risk that the patient may harm himself (suicide) or others. It may also be necessary when the patient's physical health has deteriorated to a point where life-sustaining treatment is necessary, such as with severe **malnutrition** associated with **anorexia nervosa**.

Adolescents may be successful in treating psychosocial disorders through regular attendance in self-help groups or 12-step programs such as Alcoholics Anonymous. This approach, which allows them to seek advice and counsel from others in similar circumstances, can be extremely effective.

Alternative treatment

Therapeutic approaches, such as art therapy, which encourages self-discovery and empowerment, may be useful in treating psychosocial disorders. Art therapy, the use of the creative process to express and understand emotion, encompasses a broad range of humanistic disciplines, including visual arts, dance, drama, music, film, writing, literature, and other artistic genres. This use of the creative process is believed to provide the patient/artist with a means to gain insight to emotions and thoughts they might otherwise have difficulty expressing. After the artwork is created, the patient/artist continues the therapeutic journey by interpreting its meaning under the guidance of a trained therapist.

KEY TERMS

Affective disorder—An emotional disorder involving abnormal highs and/or lows in mood. Now termed mood disorder.

Bipolar disorder—A severe mental illness, also known as manic depression, in which a person has extreme mood swings, ranging from a highly excited state, sometimes with a false sense of well being, to depression.

Bulimia nervosa—An eating disorder characterized by binge eating and inappropriate compensatory behavior, such as vomiting, misusing laxatives, or excessive exercise.

Cognitive processes—Thought processes (i.e., reasoning, perception, judgment, memory).

Learning disorders—Academic difficulties experienced by children and adults of average to above-average intelligence that involve reading, writing, and/or mathematics, and which significantly interfere with academic achievement or daily living.

Schizophrenia—A severe mental illness in which a person has difficulty distinguishing what is real from what is not real. It is often characterized by hallucinations, delusions, and withdrawal from people and social activities.

Prognosis

According to the National Institute of Mental Health, fewer than one in five of those children suffering from mental illness receive treatment for the problem. Because of the diversity of types of mental disorders influenced by psychosocial factors, and the complexity of diagnosis and treatment, the prognosis for psychosocial disorders is highly variable. In some cases, they can be effectively managed with therapy and/or medication. In others, mental illness can cause long-term disability.

The U.S. Centers for Disease Control reports that suicide is the third leading cause of death among children and youth between the ages of 10 and 24. Because more than 90 percent of those who commit suicide have a

diagnosable mental disorder, seeking swift and appropriate treatment as soon as symptoms appear is critical

Prevention

Patient education (i.e., therapy or self-help groups) can encourage patients to take an active part in their treatment program and to recognize symptoms of a relapse of their condition. In addition, educating friends and **family** members on the nature of the psychosocial disorder can assist them in knowing how and when to provide support to the patient.

Parental concerns

While seeking help for their child, parents must remain sensitive to the emotional needs and physical well being of their other children. This may mean adjusting regular routines to avoid leaving siblings alone together, getting assistance with childcare, or even seeking residential or hospital treatment for the child if the safety of other family members is in question. Parents should also maintain an open dialog with their child's teachers to ensure that their child receives appropriate educational assistance.

See also Anorexia nervosa; Bipolar disorder; Depressive disorders.

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National Institute of Mental Health. 6001 Executive Boulevard, Rm. 8184, MSC 9663, Bethesda, MD 20892–9663. (301) 443–4513.

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Puberty

Definition

Puberty is the period of human development during which physical growth and sexual maturity occurs.

Description

The word puberty is derived from the Latin *pubertas*, which means adulthood. Puberty is initiated by hormonal changes triggered by a part of the brain called the hypothalamus, which stimulates the pituitary gland, which in turn activates other glands as well. These changes begin about a year before any of their results are visible. Both the male reproductive hormone testosterone and female hormone estrogen are present in children of both sexes. However, their balance changes at puberty, with girls producing relatively more estrogen and boys producing more testosterone.

Beginning as early as age eight in girls—and two years later, on average, in boys—the hypothalamus signals hormonal change that stimulates the pituitary. In turn, the pituitary releases its own hormones called gonadotrophins that stimulate the gonads and adrenals. From these glands come a flood of sex hormones—androgen and testosterone in the male, estrogen and progestin in the female—that regulate the growth and function of the sex organs. It is interesting to note that the gonadotrophins are the same for males and females, but the sex hormones they induce are different.

The experience of puberty is new and unusual for both boys and girls. It is not something that happens overnight, but rather it is a process that occurs in stages and at different ages for different people. It is perfectly normal, for example, for one person to have already started developing while one's best friend of the same age has not. The age at which puberty begins can vary widely between individuals. Timing of onset is affected by genetic factors, body mass, nutritional state, and general health.

School age

The average age for first signs of **breast development** in girls is about 10.5 years, with **menstruation** and fertility following about two years later. Average age for first signs of testicle enlargement in boys is 11.5 years. Puberty may not begin until age 16 in boys and continue in a random fashion beyond age 20. In contrast to puberty, **adolescence** is more a social/cultural term that refers to the interval between childhood and adulthood. The duration of puberty, from time of onset to completion, varies less between children than does the age of onset. Duration of puberty in girls from onset of breast development to cessation of growth is roughly five years. Duration of puberty in boys from first testicle enlargement to cessation of growth is about six years.

Puberty has been divided into five Sexual Maturity Rating (SMR) stages by two doctors, W. Marshall and J. M. Tanner. These ratings are often referred to as Tanner Stages one through five. Staging is based on pubic hair growth, on genital development, and female breast development. Staging helps determine whether development is normal for a given age. Both sexes also grow armpit hair and develop pimples. Males develop muscle mass, a deeper voice, and facial hair. Females redistribute body fat. Along with the maturing of the sex organs, there is a pronounced growth spurt averaging three to four inches (8–10 centimeters) and culminating in full adult stature. Puberty can be early or delayed.

PUBERTY STAGES IN GIRLS

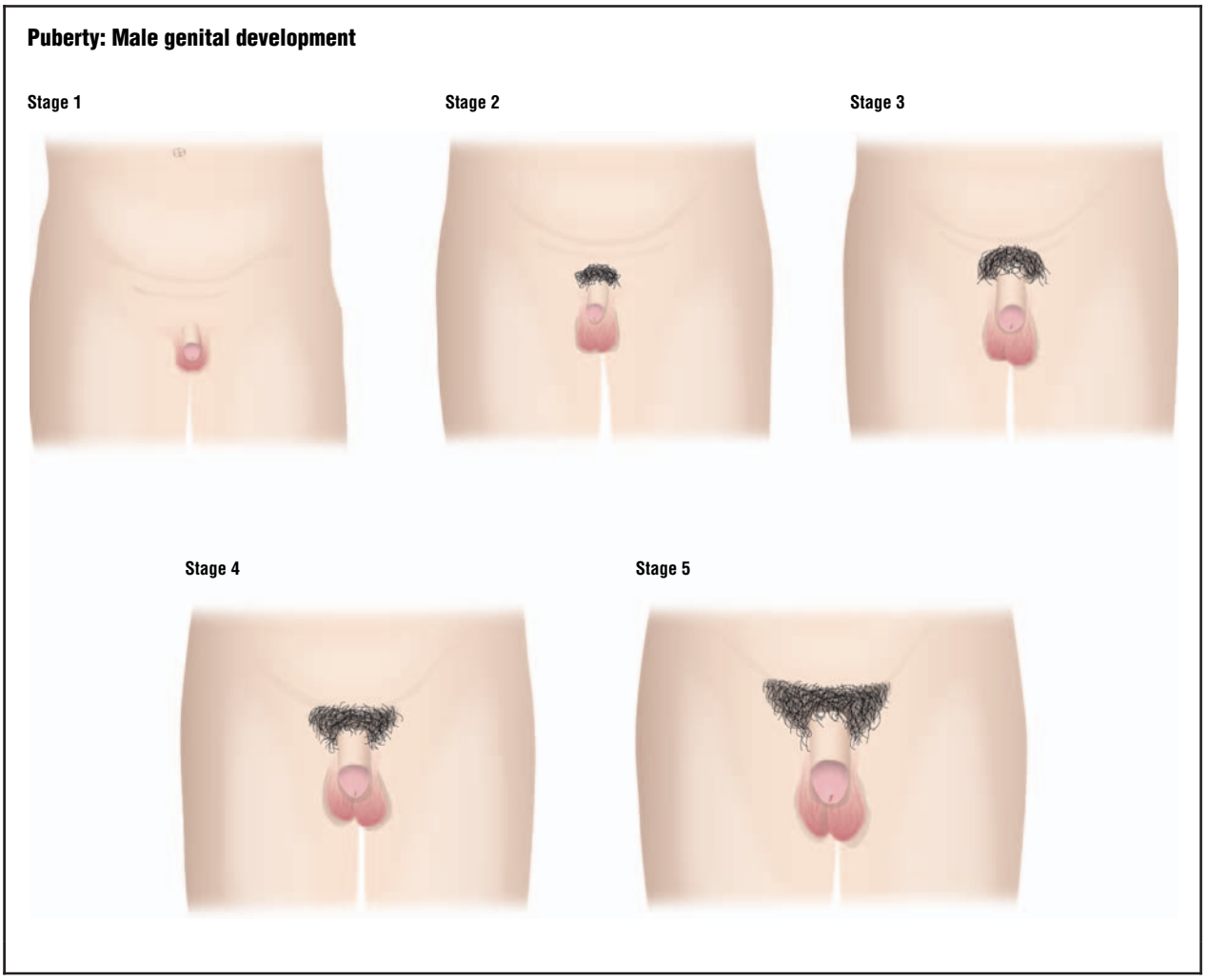
- Stage One (approximately between the ages of eight and eleven): The ovaries enlarge and hormone production starts, but external development is not yet visible.
- Stage Two (approximately between the ages of eight and fourteen): The first external sign of puberty is usually breast development. At first breast buds develop. The nipples will be tender and elevated. The area around the nipple (the aureole) will increase in size. The first stage of pubic hair may also be present at this time. It may be coarse and curly or fine and straight. Height and weight increase at this time. The body gets rounder and curvier.
- Stage Three (approximately between the ages of nine and 15): Breast growth continues and pubic hair gets coarser and darker. During this stage, whitish discharge from the vagina may be present. For some girls, this is the time that the first menstrual period begins.
- Stage Four (approximately from ages 10 to 16): Some girls notice that their aureoles get even darker and separate into a little mound rising above the rest of the breast. Pubic hair may begin to have a more adult triangular pattern of growth. If it did not happen in Stage Three, menarche (first menstruation) should start now. Ovulation may start now, too. But it will not necessarily occur on a regular basis. (It is possible to have regular periods even if ovulation does not occur every month.)
- Stage Five (approximately between ages 12 and 19): This is the final stage of development. Full height is reached, and young women are ovulating regularly. Pubic hair is filled in, and the breasts are developed fully for the body.

PUBERTY STAGES IN BOYS

- Stage One (approximately between ages nine and 12): No visible signs of development occur, but, internally, male hormones become a lot more active. Sometimes a growth spurt begins at this time.
- Stage Two (approximately between ages nine to 15): Height increases and the shape of the body changes. Muscle tissue and fat develop at this time. The aureole, the dark skin around the nipple, darkens and increases in size. The testicles and scrotum grow, but the penis probably does not. A little bit of pubic hair begins to grow at the base of the penis.
- Stage Three (approximately between ages 11 and 16): The penis starts to grow during this stage. It tends to grow in length rather than width. Pubic hair is getting darker and coarser and spreading to where the legs meet the torso. Also, boys continue to grow in height, and even their faces begin to appear more mature. The shoulders broaden, making the hips look smaller. Muscle tissue increases and the voice starts to change and deepen. Finally, facial hair begins to develop on the upper lip.
- Stage Four (approximately 11 to 17): At this time, the penis starts to grow in width, too. The testicles and scrotum also continue to grow. Hair may begin to grow on the anus. The texture of the penis becomes more adult-looking. Underarm and facial hair increases as well. Skin gets oilier, and the voice continues to deepen.
- Stage Five (approximately 14 to 18): Boys reach their full adult height. Pubic hair and the genitals look like an adult man's do. At this point, too, shaving is a necessity. Some young men continue to grow past this point, even into their twenties.

Common problems

When puberty occurs outside the age limits considered normal parents may be prompted to search for the cause. As health and **nutrition** have improved over the past few generations, there has been a gradual decrease



The five stages of male genital development. Stage 1 shows the undeveloped genitals of childhood. In Stage 2, pubic hair growth begins and the testicles begin to enlarge. By Stage 3, the penis grows longer and wider. The testicles continue to enlarge. In Stage 4, the penis and testicles continue to enlarge while the head of the penis becomes more developed. In Stage 5, the genitals have become their adult size, and pubic hair covers the region. *(Illustration by GGS Information Services.)*

in the average age for the onset of puberty. These causes of early or late puberty may include the following:

- Excess hormone stimulation is the cause for early puberty. It can come from the brain in the form of gonadotrophins or from the gonads and adrenals. Functioning tumors may cause overproduction of sex hormones. Brain overproduction of factors promoting sex hormone production can also be the result of brain infections or injury.
- Likewise, delayed puberty is due to insufficient hormone. If the pituitary output is inadequate, so will be the output from the gonads and adrenals. By contrast, a normal pituitary overproduces if it senses there are not enough hormones in the circulation.
- There are several congenital disorders called polyglandular deficiency syndromes that include failure of hormone output. Children with these syndromes do not experience normal puberty, but it may be induced by giving them hormones at the proper time.
- Finally, there are in females abnormalities in hormone production that produce male characteristics, so called virilizing syndromes. Should one of these appear during adolescence, it will disturb the normal progress of puberty. Notice that virilizing requires abnormal hormones in the female, while feminizing results from absent hormones in the male. Each embryo starts out life as female. Male hormones transform it if they are present.

Delayed or early puberty requires measurement of the several hormones involved to determine which are lacking or which are in excess. There are blood tests for each one. If a tumor is suspected, imaging of the suspect

organ needs to be done with x rays, **computed tomography** scans (CT scans), or **magnetic resonance imaging** (MRI).

Puberty is a period of great stress, both physically and emotionally. The psychological changes and challenges of puberty are made infinitely greater if its timing is off.

In early puberty, the offending gland or tumor may require surgical attention, although there are several drugs as of 2004 that counteract hormone effects. If delayed, puberty can be stimulated with the correct hormones. Treatment should not be delayed because necessary bone growth is also affected.

Early puberty often begins before age eight in girls, triggering the development of breasts and hair under the arms and in the genital region. The onset of ovulation and menstruation also may occur. In boys, the condition triggers the development of a large penis and testicles, with spontaneous erections and the production of sperm. Hair grows on the face, under arms, and in the pubic area, and **acne** may become a problem.

Several studies indicate an increase in incidences of early puberty and other forms of early sexual development in the United States. Sexual development in children seven years of age and younger should be evaluated by a physician. In some cases, early sexual development can be caused by a tumor or other pathological conditions. Properly administered hormones can restore the normal growth pattern.

Parental concerns

Most experts suggest that parents begin short and casual discussions about the body changes that occur in puberty with their children by the age of seven or eight. Offering the child reading materials about puberty can impart information to the young person without the awkwardness that may characterize the parent-child conversations. Parents can then offer their children opportunities to ask questions or to discuss any aspects of puberty and sexuality that may arise from their reading.

It is also a good idea for parents to talk to their children about proper hygiene at the onset and during puberty. While good hygiene is important for everyone at any age, it can require greater care at the onset of puberty. Hormones produced by the maturing body bring about physical changes that require greater attention when it comes to hygiene. For a young girl or boy, this means taking more time to clean the body, especially the sexual organs, to treat acne, use mouthwash for bad breath, and deodorant for stronger body odor.

Puberty

	Boys	Girls
Stage one	Prepubertal: no sexual development	Prepubertal: no sexual development
Stage two	Testes enlarge Body odor	Breast budding First pubic hair Body odor Height spurt
Stage three	Penis enlarges Pubic hair starts growing Ejaculation (wet dreams)	Breasts enlarge Pubic hair darkens, becomes curlier Vaginal discharge
Stage four	Continued enlargement of testes and penis Penis and scrotal sac deepen in color Pubic hair curlier and coarser Height spurt Male breast development	Onset of menstruation Nipple is distinct from areola
Stage five	Fully mature male Pubic hair extends to inner thighs Increases in height slow, then stop	Fully mature female Pubic hair extends to inner thighs Increases in height slow, then stop

SOURCE: Child Development Institute. http://www.childdevelopmentinfo.com. 2005.

(Table by GGS Information Services.)

When a boy or girl begins to go through puberty, the body produces more perspiration because sweat glands, some of which are located near the underarms, become more active. More perspiration means a different type of body odor, one that is stronger and similar to an adult's. Daily bathing and showering are enough to control body odor, along with deodorants and antiperspirants.

Boys should be instructed to wash their genitals every day. This includes washing the penis, the scrotum that holds the testicles, the anus, and pubic hair with water and mild soap. Uncircumcised boys need to be instructed that the foreskin should be pulled down daily to expose the tip of the penis, which should then be washed with mild soap and water.

In girls, it is perfectly natural to have a slight sweet smell from the vagina that is inoffensive. A strong, foul odor indicates a possible infection. With treatment, the infection goes away and so does the strong odor. Vaginal discharge is a necessary part of the body's regular functioning. Normal discharge, usually clear to white, is part of the body's self-cleaning process. As discharge leaves the body, it takes bacteria with it, which helps prevent vaginal infections. Parents should stress that girls clean the vaginal area with a mild soap and water

KEY TERMS

Adrenal gland—A small gland located above the kidney (one on each side) that secretes various hormones.

Circumcision—A surgical procedure, usually with religious or cultural significance, where the prepuce or skin covering the tip of the penis on a boy, or the clitoris on a girl, is cut away.

Estrogen—Female hormone produced mainly by the ovaries and released by the follicles as they mature. Responsible for female sexual characteristics, estrogen stimulates and triggers a response from at least 300 tissues. After menopause, the production of the hormone gradually stops.

Estrus—A regular period of sexual excitement in females.

Gonadotrophin—Hormones that stimulate the ovary and testicles.

Gonads—Organs that produce gametes (eggs or sperm), i.e., the ovaries and testes.

Hypothalamus—A part of the forebrain that controls heartbeat, body temperature, thirst, hunger, body temperature and pressure, blood sugar levels, and other functions.

Menstruation—The periodic discharge from the vagina of blood and tissues from a nonpregnant uterus.

Pituitary gland—The most important of the endocrine glands (glands that release hormones directly into the bloodstream), the pituitary is located at the base of the brain. Sometimes referred to as the "master gland," it regulates and controls the activities of other endocrine glands and many body processes including growth and reproductive function. Also called the hypophysis.

Testosterone—Male hormone produced by the testes and (in small amounts) in the ovaries. Testosterone is responsible for some masculine secondary sex characteristics such as growth of body hair and deepening voice. It also is sometimes given as part of hormone replacement therapy to women whose ovaries have been removed.

Virilizing syndromes—Abnormalities in female hormone production that produce male characteristics.

on a regular basis to help control bacteria growth and limit infections.

When to call the doctor

Parents should consult a pediatrician or physician when their child shows signs of either early or delayed puberty.

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Pulled elbow *see* **Nursemaid's elbow**

Pulmonary function tests

Definition

Pulmonary function tests are a group of procedures that measure how well the lungs are functioning.

Purpose

Pulmonary function tests help a doctor to diagnose respiratory diseases and disorders such as **asthma**, chronic obstructive pulmonary disease (COPD), and emphysema, and mechanical injury by measuring the degree of lung impairment. These tests are also done before major lung surgery to make sure the patient will not be disabled by having a reduced lung capacity. When performed over time, these tests are helpful in evaluating how a lung disease is progressing, and how serious the lung disease has become. They are also be used to assess how a patient is responding to different treatments.

A cystic fibrosis patient receives a pulmonary function test. *(Custom Medical Stock Photo Inc.)*

Description

There are many types of pulmonary function tests. The most common are:

- peak expiratory flow rate (PEFR) measures airflow during forced expirations
- forced vital capacity (FVC) measures the maximum amount of air exhaled after taking a deep breath
- forced expiratory volume in one second (FEV1) measures the amount of air that can be exhaled in one second
- maximum voluntary volume (MVV) measures the amount of air a person can breathe in and out in one minute
- total lung capacity (TLC) is the measure of the amount of air the lungs can hold
- residual volume (RV) is the amount of air left in the lungs after forced expiration
- arterial blood gas (ABG) measures the amount of oxygen and carbon dioxide in the blood and gives a picture of how efficiently the lungs are functioning
- pulse oximetry measures the percentage of oxygen in the blood

With the exception of arterial blood gas, pulse oximetry, and total lung capacity, pulmonary function tests are performed using spirometry (from the Greco-Latin term meaning "to measure breathing"). Spirometry tests can be done a hospital or doctor's office. The patient places a clip over the nose and breathes through the mouth into a tube connected to a machine called a spirometer. The patient breathes in deeply, and then exhales as quickly and forcefully as possible into the tube. The machine records the volume of air that moves through the tube. The exhalation must last at least six seconds for the machine to work properly. Usually the patient repeats this test three times, and the best of the three results is considered the measure of the lung function. A similar

KEY TERMS

Artery—A blood vessel that carries blood away from the heart to the cells, tissues, and organs of the body.

Bronchodilator—A drug that when inhaled helps to expand the airways.

Carbon dioxide—A heavy, colorless gas that dissolves in water.

Forced exhalation—Blowing as much air out of the lungs as possible.

machine called a peak flow meter may be used to measure PERF. Sometimes when airways are obstructed, the patient is given a bronchodilator, and the test is performed again.

Total lung capacity is measured by body plethysmography. The patient sits in a sealed box that resembles a telephone booth and breathes against a mouthpiece. A device measures the changes in air pressure in the box during inhalation and exhalation. From these air pressure measurements, the total capacity of the lungs can be calculated.

Arterial blood gases are measured on a blood sample that is taken from an artery. Pulse oximetry uses a sensor placed on the earlobe or fingertip to measure the amount of oxygen in the blood.

Taken together, pulmonary function tests give a good picture of how much air is moving in and out of the lungs and how efficiently oxygen is moved into the blood and carbon dioxide is moved out. Some of these tests are performed as part of a routine health screening, while others are used most often to evaluate the condition of diseased or damaged lungs.

Precautions

Except for the arterial blood gas tests and pulse oximetry, pulmonary function tests should not be given to patients who have had a recent heart attack, or who have certain other types of heart disease. Conditions that cause **pain** on breathing, such as broken ribs, may interfere with the performance of the tests and produce inaccurate results. Children must be old enough to follow directions and inhale and exhale as instructed.

Preparation

The patient should not eat a heavy meal before the test, nor smoke for four to six hours beforehand. The doctor will give specific instructions about whether or not to use medications before the test.

Aftercare

No special aftercare is needed following these tests.

Risks

Risks with these tests are minimal. However, some people become lightheaded or faint. The tests may also trigger an asthma attack in individuals with asthma.

Parental concerns

Normal results are based on a person's age, height, and gender. Normal results are expressed as a percentage of the predicted lung capacity. Results of 80 percent or less suggest some sort of lung impairment.

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Punishment *see* **Discipline**

Purging *see* **Bulimia nervosa**

R

Rabies

Definition

Rabies is an acute viral disease of the central nervous system that is transmitted through saliva from the bite of an infected animal.

Description

Rabies affects humans and other mammals but is most common in carnivores (flesh eaters). It is sometimes referred to as a zoonosis, or disease of animals that can be communicated to humans. Rabies is almost exclusively transmitted through saliva from the bite of an infected animal. Another name for the disease is hydrophobia, which literally means **fear** of water, a symptom shared by half of all people infected with rabies. Other symptoms include **fever**, depression, confusion, painful **muscle spasms**, sensitivity to touch, loud noise, and light, extreme thirst, painful swallowing, excessive salivation, and loss of muscle tone. If rabies is not prevented by immunization, it is almost always fatal.

In late 2002, rabies re-emerged as an important public health issue. Charles E. Rupprecht, director of the World Health Organization (WHO) Collaborating Center for Rabies Reference and Research, listed several factors responsible for the increase in the number of rabies cases worldwide:

- Rapid evolution of the rabies virus. Bats in the United States have developed a particularly infectious form of the virus.
- Increased diversity of animal hosts for the disease.
- Changes in the environment that are bringing people and domestic pets into closer contact with infected wildlife.
- Increased movement of people and animals across international borders. In one case, a man who had contracted rabies in the Philippines was not diagnosed until he began to feel ill in the United Kingdom.
- Lack of advocacy about rabies.

Demographics

Cases of rabies in humans are very infrequent in the United States, averaging one or two a year (down from over 100 cases annually in 1900), but the worldwide incidence is estimated to be between 30,000 and 50,000 cases each year. These figures are based on data collected by the World Health Organization (WHO) in 1997 and updated in 2002. Rabies is most common in developing countries in Africa, Latin America, and Asia, particularly India. Dog **bites** are the major origin of infection for humans in developing countries, but other important host animals are the wolf, mongoose, and bat. Worldwide, the highest risk groups for contracting rabies are boys under the age of fifteen. Most deaths from rabies in the United States result from bat bites.

People whose work frequently brings them in contact with animals are also considered to be at higher risk than the general population. This group includes those in the fields of veterinary medicine, animal control, wildlife work, and laboratory work involving live rabies virus. People in these occupations and residents of or travelers to areas where rabies is a widespread problem should consider being immunized.

Causes and symptoms

Rabies is caused by a rod- or bullet-shaped virus that belongs to the family Rhabdoviridae. The virus is usually transmitted via an animal bite; however, cases have also been reported in which the virus penetrated the body through infected saliva, moist tissues such as the eyes or lips, a scratch on the skin, or the transplantation of infected tissues. Inhalation of the virus from the air, as might occur in a highly populated bat cave, is also thought to occur.

From the bite or other area of penetration, the virus multiplies as it spreads along nerves that travel away from the spinal cord and brain (efferent nerves) and into the salivary glands. The rabies virus may lie dormant in the body for several weeks or months, but rarely much longer, before symptoms appear. Initially, the area

around the bite may burn and be painful. Early symptoms may also include a **sore throat**, low-grade fever, headaches, loss of appetite, **nausea and vomiting**, and **diarrhea**. Painful spasms develop in the muscles that control breathing and swallowing. The individual may begin to drool thick saliva and may have dilated or irregular pupils, increased tears and perspiration, and low blood pressure.

As the disease progresses, the patient becomes agitated and combative and may exhibit increased mental confusion. The affected person usually becomes sensitive to touch, loud noises, and bright lights. The victim also becomes extremely thirsty but is unable to drink because swallowing is painful. Some patients begin to dread water because of the painful spasms that occur. Other severe symptoms during the later stage of the disease are excessive salivation, **dehydration**, and loss of muscle tone. Death usually occurs three to 20 days after symptoms have developed. Recovery is very rare.

Diagnosis

After the onset of symptoms, blood tests and **cerebrospinal fluid (CSF) analysis** tests will be conducted. **CSF** will be collected during a procedure called a lumbar puncture in which a needle is used to withdraw a sample of CSF from the area around the spinal cord. The CSF tests do not confirm diagnosis but are useful in ruling out other potential causes for the patient's altered mental state.

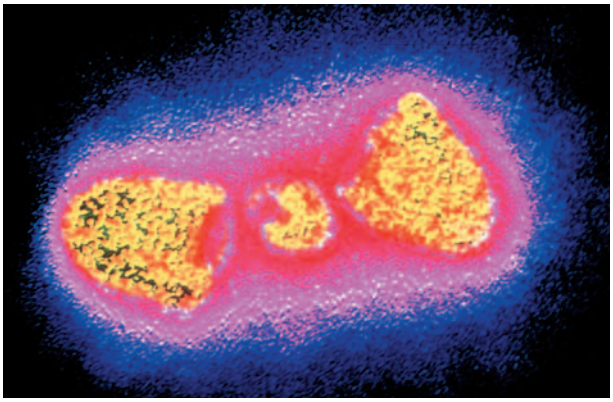
The two most common diagnostic tests are the fluorescent antibody test and isolation of the rabies virus from an individual's saliva or **throat culture**. The fluorescent antibody test involves taking a small sample of skin (biopsy) from the back of the neck of the patient. If specific proteins, called antibodies, that are produced only in response to the rabies virus are present, they will bind with the fluorescent dye and become visible. Another diagnostic procedure involves taking a corneal impression in which a swab or slide is pressed lightly against the cornea of the eye to determine whether viral material is present.

Treatment

Because of the extremely serious nature of a rabies infection, the need for rabies immunizations should be carefully considered for anyone who has been bitten by an animal, based on a personal history and results of diagnostic tests.

If necessary, treatment includes the following:

- The wound is washed thoroughly with medicinal soap and water. Deep puncture **wounds** should be flushed with a catheter and soapy water. Unless absolutely necessary, a wound should not be sutured.
- Tetanus toxoid and **antibiotics** will usually be administered.
- Rabies **vaccination** may or not be given, based on the available information. If the individual was bitten by a domestic animal and the animal was captured, the animal will be placed under observation in quarantine for ten days. If the animal does not develop rabies within four to seven days, then no immunizations are required. If the animal is suspected of being rabid, it is killed, and the brain is examined for evidence of rabies infection. In cases involving bites from domestic animals in which the animal is not available for examination, the decision for vaccination is made based on the prevalence of rabies within the region where the bite occurred. If the bite was from a wild animal and the animal was captured, it is generally killed because the incubation period of rabies is unknown in most wild animals.
- If necessary, the patient is vaccinated immediately, generally through the administration of human rabies immune globulin (HRIG) for passive immunization, followed by human diploid cell vaccine (HDCV) or **rabies vaccine** adsorbed (RVA) for active immunization. Passive immunization is designed to provide the individual with antibodies from an already immunized individual, while active immunization involves stimulating the individual's own immune system to produce antibodies against the rabies virus. These rabies vaccines are equally effective and carry a lower risk of side effects than some earlier treatments. Unfortunately, however, in underdeveloped countries, these vaccines are usually not available. Antibodies are administered to the patient in a process called passive immunization. To do so, the HRIG vaccine is administered once, at the beginning of treatment. Half of the



Rabies virus shown in yellow. *(Custom Medical Stock Photo Inc.)*

dose is given around the bite area, and the rest is given in the muscle. Inactivated viral material (antigenic) is then given to stimulate the patient's own immune system to produce antibodies against rabies. For active immunization, either the HDCV or RVA vaccine is given in a series of five injections. Immunizations are typically given on days 1, 3, 7, 14, and 28.

In those rare instances in which rabies has progressed beyond the point where immunization would be effective, the patient is given medication to prevent seizures, relieve some of the **anxiety**, and relieve painful muscle spasms. **Pain** relievers are also given. In the later stages, aggressive supportive care will be provided to maintain breathing and heart function. Survival is rare but can occur.

Prognosis

If preventative treatment is sought promptly, rabies need not be fatal. Immunization is almost always effective if started within two days of the bite. Chance of effectiveness declines, however, the longer vaccination is put off. It is, however, important to start immunizations, even if it has been weeks or months following a suspected rabid animal bite, because the vaccine can be effective even in these cases. If immunizations do not prove effective or are not received, rabies is nearly always fatal within a few days of the onset of symptoms.

Prevention

The following precautions should be observed in environments where humans and animals are likely to come into contact:

- Domesticated animals, including household pets, should be vaccinated against rabies. If a pet is bitten by an animal suspected to have rabies, its owner should contact a veterinarian immediately and notify the local animal control authorities. Domestic pets with current vaccinations should be revaccinated immediately; unvaccinated dogs, cats, or ferrets are usually euthanized (killed).
- Wild animals should not be touched or petted, no matter how friendly they may appear. It is also important not to touch an animal that appears ill or passive or whose behavior seems odd, such as failing to show the normal fear of humans. These are all possible signs of rabies. Many animals, such as raccoons and skunks, are nocturnal and their activity during the day should be regarded as suspicious.
- People should not interfere in fights between animals.
- Because rabies is transmitted through saliva, a person should wear rubber gloves when handling a pet that has had an encounter with a wild animal.
- Garbage or pet food should not be left outside the house or camp site because it may attract wild or stray animals.

KEY TERMS

Active immunization—Treatment that provides immunity by challenging an individual's own immune system to produce antibody against a particular organism.

Antibody—A special protein made by the body's immune system as a defense against foreign material (bacteria, viruses, etc.) that enters the body. It is uniquely designed to attack and neutralize the specific antigen that triggered the immune response.

Biopsy—The surgical removal and microscopic examination of living tissue for diagnostic purposes or to follow the course of a disease. Most commonly the term refers to the collection and analysis of tissue from a suspected tumor to establish malignancy.

Efferent nerves—Peripheral nerves that carry signals away from the brain and spinal cord.

Fluorescent antibody test—A test in which a fluorescent dye is linked to an antibody for diagnostic purposes.

Lumbar puncture—A procedure in which the doctor inserts a small needle into the spinal cavity in the lower back to withdraw spinal fluid for testing. Also known as a spinal tap.

Passive immunization—Treatment that provides immunity through the transfer of antibodies obtained from an immune individual.

Rhabdovirus—A family of viruses named for their rod- or bullet-like shapes. The rabies virus is a rhabdovirus.

Vector—A carrier organism (such as a fly or mosquito) which serves to deliver a virus (or other agent of infection) to a host. Also refers to a retrovirus that had been modified and is used to introduce specific genes into the genome of an organism.

Zoonosis—Any disease of animals that can be transmitted to humans. Rabies is an example of a zoonosis.

- Windows and doors should be screened. Some victims of rabies have been attacked by infected animals, particularly bats, that entered through unprotected openings.
- State or county health departments should be consulted for information about the prevalence of rabies in an area. Some areas, such as New York City, have been rabies-free, only to have the disease reintroduced at a later time.
- Preventative vaccination against rabies should be considered if one's occupation involves frequent contact with wild animals or non-immunized domestic animals.
- Bites from mice, rats, or squirrels rarely require rabies prevention because these rodents are typically killed by any encounter with a larger, rabid animal, and would, therefore, not be carriers.
- Travelers should ask about the prevalence of the disease in countries they plan to visit.

Parental concerns

Parents should speak with their children about the importance of avoiding contact with wild animals and reporting strange behavior in any animal, even a pet.

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Rabies vaccine

Definition

Rabies vaccine is an injection that provides protection against the rabies virus that can be transmitted to humans via the saliva of an infected animal. Rabies is fatal in humans unless it is prevented with a vaccine.

Description

Rabies are caused by viruses of the genus *Lyssavirus* in the family Rhabdoviridae. Although all mammals are thought to be susceptible to rabies infection, the primary hosts are carnivores and bats. Most human exposure to rabies occurs via an animal bite in which the skin is broken and the virus is transmitted from the infected animal's saliva to the blood and tissues of the victim. The rabies virus infects the human nervous system causing acute encephalomyelitis, an inflammation of the brain and spinal cord. Death, usually by respiratory failure, occurs within seven to 10 days after appearance of the first symptoms. The average incubation period before symptoms of the disease appear is three to seven weeks, with a range of 10 days to seven years.

Prevalence of rabies

UNITED STATES Cases of human rabies are very rare in the United States due to the routine **vaccination** of domestic animals. In the past most human rabies resulted

from **bites** by infected dogs. However as the incidence of rabies in dogs has decreased dramatically, rabies among wildlife has increased across the continental United States. Bat bites are now the most common source of human rabies infection. Hawaii remains rabies-free.

Between 1990 and 2003, there were 39 diagnosed cases of rabies among Americans. Every year an estimated 18,000 Americans receive rabies pre-exposure prophylaxis and an additional 16,000-39,000 receive post-exposure prophylaxis as a result of animal bites.

WORLDWIDE Rabies is common in some parts of the world, particularly in the developing countries of Africa, Asia, and Latin America. Rabies has been eradicated in the United Kingdom. Rabies is considered to be a reemerging viral disease because it is poorly controlled in many developing countries despite widely available human and animal vaccines. WHO estimates that every year about 10–12 million people worldwide receive post-exposure prophylaxis and that about 35,000 people—primarily children—die of rabies every year. However the incidence of rabies in the developing world is believed to be severely underreported. Most rabies exposures are from bites by unvaccinated dogs.

Vaccine development

The French scientist Louis Pasteur developed the first vaccine against rabies. In 1885, he injected his attenuated (weakened) virus into a nine-year-old boy who had been bitten by a rabid dog. The child's life was saved. Over the following century several generations of rabies vaccines were developed.

Although there is no cure for rabies once symptoms of the disease have appeared, in the 1980s scientists developed a highly effective vaccine that provides protection from the virus both before exposure—pre-exposure prophylaxis—or after exposure—post-exposure prophylaxis. The vaccine consists of killed rabies virus that, when injected, induces the child's immune system to produce antibodies that bind to and destroy the virus. The antibody response develops within seven to 10 days of vaccination and provides protection for up to two years. A second type of rabies vaccine, rabies immune globulin (RIG), provides immediate, short-term protection after exposure to the virus.

Pre-exposure prophylaxis

Routine rabies vaccination and booster immunizations are necessary only for those in high-risk professions such as veterinarian medicine and laboratory workers. However pre-exposure prophylaxis of children who are at risk of being exposed to rabid animals eliminates the need for RIG and decreases the number of required vaccinations after exposure. Pre-exposure prophylaxis is particularly important for children who may be exposed to rabies in places where vaccines, if available, may cause adverse reactions. Pre-exposure prophylaxis also may be helpful for children who are exposed unknowingly or do not report the exposure.

Children traveling internationally are at particular risk for rabies exposure because they may not exhibit caution in approaching animals. Such children may be considered for pre-exposure prophylaxis if they will be:

- in an area where rabies is prevalent or endemic
- camping in rural areas
- in an area where appropriate rabies vaccines and RIG may not be available

Post-exposure prophylaxis

After exposure to a potentially rabid animal, a child's risk of contracting rabies is assessed based on:

- the rabies vaccination status of the animal
- the type of animal
- whether the animal can be captured and tested for rabies
- the geographical location of the exposure
- whether the contact was provoked or unprovoked Unprovoked attacks are more likely to come from a rabid animal. Provoked attacks can include bites received while feeding or handling an animal.

Post-exposure prophylaxis usually is recommended when a child has been:

- bitten by any animal, including a pet dog or cat, that has not been vaccinated against rabies
- scratched or bitten by a wild animal, particularly a bat, raccoon, skunk, fox, or coyote (Some animals, particularly bats, may not leave obvious bite marks.)

When a child is bitten by a healthy domestic dog, cat, or ferret, the animal is usually confined for 10 days and observed for signs of rabies prior to initiating post-exposure prophylaxis. The rabies status of an animal also can be determined by testing for antibodies against rabies in its blood or by killing the animal and testing its brain tissue.

Post-exposure prophylaxis should be considered following any contact between a child and a bat, even if there is no evidence of a bite or scratch, since the child may be unaware of the contact and marks may not be apparent. For example, post-exposure prophylaxis

should be considered if an unattended child is found in a room with a bat and the bat cannot be tested for rabies.

Vaccine types

Four formulations of three inactivated rabies virus vaccines are licensed for use by the U.S. Food and Drug Administration (FDA). Two RIG formulations are also FDA-licensed.

HUMAN DIPLOID CELL VACCINE (HDCV) Human diploid cell vaccines (HDCVs) use inactivated rabies viruses. HDCV comes in two formulations: one for intramuscular (IM) injection and one for intradermal (ID) injection into a deep layer of skin.

PURIFIED CHICK EMBRYO CELL VACCINE (PCEC) Purified chick embryo cell (PCEC) vaccine became available in the United States in 1997. PCEC is made from rabies virus grown in cultures of chicken embryos and then inactivated. The drug is formulated for IM administration only.

RABIES VACCINE ADSORBED (RVA) Rabies vaccine adsorbed (RVA) is manufactured from virus grown in cell cultures of fetal rhesus monkey lung cells and then inactivated.

RABIES IMMUNE GLOBULIN (RIG) Human rabies immune globulin (RIG, HRIG) is a vaccine made from human serum that contains high levels of antibodies against rabies. It is used in conjunction with an inactivated-rabies vaccine for post-exposure prophylaxis. RIG provides immediate but short-lived protection against rabies. Approximately one-half of the antibodies are lost within 21 days after administration.

RIG is separated from the blood plasma of hyperimmunized human donors. Numerous procedures are used to clear the serum of rabies virus.

OTHER VACCINES Although the four types of inactivated-rabies vaccines and the two RIGs are the only rabies vaccines available in the United States, various other rabies vaccines are produced throughout the world. Although inactivated-rabies vaccines from diploid cell cultures are safe and effective, they are expensive. In developing countries, rabies vaccines often contain nerve tissue which can cause adverse effects. Various less expensive but safe and effective vaccines are under development.

General use

Inactivated-rabies vaccines are injected, either before or after exposure to the virus, in 1.0-ml. doses containing at least 2.5 IU/ml. of rabies virus antigen. This is the recommended standard of the World Health Organization (WHO). The size and number of vaccine doses are the same for children and adults. Although the same rabies vaccine usually is used throughout an immunization series, there is no evidence of adverse reactions or loss of effectiveness when two different vaccines are used in the same series. Modern rabies vaccines are relatively painless.

Pre-exposure prophylaxis

For preventative rabies immunization in an unexposed child, an inactivated-rabies vaccine is administered in three 1.0-ml. doses, with the second dose seven days after the first, and the third dose 21 or 28 days after the first. The vaccine is injected into the upper arm. Studies have found that this regimen produces adequate antibodies against rabies in the blood serum of all subjects.

Post-exposure prophylaxis

Following an animal bite or contact between a child's mucous membranes and an animal's saliva, an attempt is usually made to determine whether the animal has rabies. If there is a threat of rabies, an unvaccinated child receives RIG and a series of five rabies vaccinations over a 28-day period. Ideally, treatment should begin within two days of exposure, however it may be started at any time thereafter.

The wound is cleaned thoroughly and, if possible, RIG is injected into the wound and the surrounding tissues to block the virus's entry into the central nervous system. The recommended dose is 20 IU/kg (1 kg = 2.2 lb) of body weight. This is equivalent to 22 mg of the antibody immunoglobulin G (IgG) per kilogram of body weight. Any remaining RIG is injected intramuscularly at a site removed from the vaccination site. RIG also may be injected into the buttocks. RIG is never injected with the same syringe or at the same site as the vaccine. RIG is used only once to provide antibodies until the child's immune system begins producing its own antibodies in response to the vaccine. RIG is administered concurrently with the first dose of inactivated-rabies vaccine or up to seven days thereafter. Additional treatment with RIG may interfere with antibody production in response to the inactivated-rabies vaccine.

Inactivated-rabies vaccine is administered in 1.0-ml. doses, at three, seven, 14, and 28 days after the first vaccination. It is injected intramuscularly in the upper arm or the upper thigh. If an animal is found to be rabies-free after the vaccination series has been initiated, the series can be discontinued.

Exposure following vaccination

Children exposed to rabies following vaccination receive a 1.0-ml. dose of vaccine immediately and a second dose three days later. These children do not receive RIG because it will diminish the rapid antibody response resulting from the previous vaccination.

Precautions

Precautions should be taken before vaccinating a child who has:

- a weakened immune system due to HIV/AIDS or other disease or condition
- **cancer**
- had a life-threatening reaction to a previous rabies vaccine or to any component of the vaccine

Children with suppressed immune systems should not receive pre-exposure prophylaxis against rabies. Medical conditions and medications that suppress the immune system can interfere with antibody production in response to a rabies vaccine. If a child has exhibited a serious hypersensitivity to a previous rabies vaccine, **antihistamines** may be used concurrently. Children who are allergic to eggs should not be give vaccines cultured in chicken embryos.

A minor illness, such as a cold, does not preclude rabies vaccination. However pre-exposure vaccination should be postponed if the child has a moderate or severe illness. Post-exposure prophylaxis should be administered regardless of any other illness or condition. Children should not be vaccinated against **measles** or **chickenpox** (varicella) for four months after being treated with RIG. Children receiving post-exposure prophylaxis outside of the United States should have their antibody levels against rabies measured after their return.

Side effects

Side effects from the rabies vaccines currently used in the United States are much less common and less severe than the side effects of earlier rabies vaccines. However side effects may vary with the brand of vaccine and adverse reactions to rabies vaccines used in some other countries are quite common. The risk of side effects also increases with the number of vaccine doses. However a vaccination series should not be interrupted because of localized or mild side effects.

Mild side effects from rabies vaccines include:

- soreness, redness, swelling, **itching**, or **pain** at the site of the injection in 30–74 percent of recipients
- headache, **nausea**, abdominal pain, muscle aches, or **dizziness** in 5–40 percent of recipients

More serious side effects of rabies vaccines include:

- hives, joint pain, or **fever** in about 6 percent of those receiving a booster vaccination
- very rarely, an illness resembling Guillain-Barré syndrome, a disorder of the motor nerves that can result in temporary paralysis, lasting no longer than 12 weeks and resulting in complete recovery Other nervous system disorders occur so rarely following rabies vaccination that they may not be related to the vaccine. However, a physician should be consulted if a high fever or behavioral changes occur following rabies vaccination.

Reported side effects of RIG include:

- local pain
- low-grade fever

Although any vaccine is capable of inducing an allergic reaction, serious reactions to rabies vaccine are very rare. Signs of an allergic reaction include:

- paleness
- weakness
- dizziness
- hoarseness or wheezing
- difficulty breathing
- a fast heartbeat

In case of a serious reaction to a rabies vaccine:

- A doctor should be consulted immediately.
- The date, time, and type of reaction should be recorded.
- Medical personnel or the local health department should file a Vaccine Adverse Event Report.

Interactions

Immune system-suppressing treatments, including cancer drugs and radiation and steroids, can interfere with the antibody response to rabies vaccination. If possible, immunosuppressive medications should be suspended during the vaccination series, and the vaccine injections should be intramuscularly. Alternatively, the child's serum can be checked for antibody production to determine if the vaccination was successful.

Chloroquine phosphate or similar anti-malarial drugs such as mefloquine may interfere with the response to HDCV. Children who will be taking anti-

malarial drugs while traveling in areas with endemic rabies should begin the three-dose regimen of ID vaccine one month prior to travel, before they begin taking drugs to prevent malaria. However, a three-dose, pre-exposure regimen of IM vaccine provides an adequate response even in the presence of anti-malarial drugs.

Parental concerns

Preparing a child for an injection

Most children are afraid of injections; however there are simple methods for easing a child's **fear**. Prior to the vaccination parents should:

- Tell children that they will be getting a shot.
- Explain to children that the shot will prevent them from becoming sick.
- Have older siblings comfort and reassure a younger child.
- Bring along the child's favorite toy or blanket.
- Never threaten children by telling them they will get a shot.
- Read the vaccination information statement (VIS) and ask questions of the medical practitioner.

During the vaccination parents should:

- Hold the child.
- Make eye contact with the child and smile.
- Talk softly and comfort the child.
- Distract the child by using a hand puppet or pointing out pictures or objects.
- Sing or tell the child a story.
- Have the child tell a story.
- Teach the child to focus on something other than the shot.
- Help the child take deep breaths.
- Allow the child to cry.
- Stay calm.

After the injection

Following an injection parents should:

- Hold and caress a child or breastfeed an infant.
- Talk soothingly and reassuringly.
- Hug and praise the child for doing well.
- Use a cool, wet cloth to reduce soreness or swelling at the injection site.

Parents should also be aware that:

- The child may eat less during the first 24 hours following a vaccination.
- The child should drink plenty of fluids.
- The medical practitioner may suggest a non-aspirin-containing pain reliever for the child.

KEY TERMS

Antibody—A special protein made by the body's immune system as a defense against foreign material (bacteria, viruses, etc.) that enters the body. It is uniquely designed to attack and neutralize the specific antigen that triggered the immune response.

Antigen—A substance (usually a protein) identified as foreign by the body's immune system, triggering the release of antibodies as part of the body's immune response.

Booster immunization—An additional dose of a vaccine to maintain immunity to the disease.

Encephalomyelitis—Encephalitis or another acute inflammation of the brain and spinal cord that can be caused by the rabies virus.

Human diploid cell vaccine (HDCV)—A rabies vaccine in which the virus is grown in cultures of human cells, concentrated, and inactivated for IM or ID injection.

Intracutaneous—Into the skin, in this case directly under the top layer of skin.

Intramuscular(IM)—An injection into a muscle.

Prophylaxis—Protection against or prevention of a disease. Antibiotic prophylaxis is the use of antibiotics to prevent a possible infection.

Purified chicken embryo cell vaccine (PCEC)—A rabies vaccine in which the virus is grown in cultures of chicken embryo cells, inactivated, and purified for IM injection.

Rabies immune globulin (RIG or HRIG)—A human serum preparation containing high levels of antibodies against the rabies virus; used for post-exposure prophylaxis.

Rabies virus adsorbed (RVA)—A rabies vaccine in which the virus is grown in cultures of lung cells from rhesus monkeys, inactivated, and adsorbed to aluminum phosphate.

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Rape and sexual assault

Definition

Rape and sexual assault are crimes that involve the use of threats, **fear** tactics, and/or physical violence to force a child or adolescent to submit to sexual intercourse or to engage in other sexual activity (e.g., oral sex, anal sex).

Description

Rape and sexual assault are violent crimes, and children and adolescents constitute a large number of the victims of these crimes. Rape and sexual assault are defined according to the degree of sexual interaction. Rape and sexual assault can involve contact between the penis and vagina or penis and anus that involves penetration; contact between the mouth and genitals or anus; penetration of the vagina or anus with an object; or direct touching (not through clothing). Sexual assault is defined as intentional touching of the genitals, breasts, buttocks, anus, inner thigh, or groin with no sexual penetration that is forced upon the victim. Rape and sexual assault that recurs is considered sexual abuse. When the perpetrator is a **family** member, these crimes are also referred to as incest.

Rape and sexual assault are especially traumatic for children and adolescents, who often do not fully understand normal sexual activity. Studies and statistics have shown that **adolescence** is the riskiest life stage for sexual assault, and the time when the most psychological trauma can result.

Demographics

The 2000 Victim, Incident, and Offender Characteristics, published by the National Center for Juvenile Justice (NCJJ), analyzed sexual assault data collected by law enforcement agencies over a five-year span. The following characteristics were found to be significant among victims of sexual assault:

- Age: Over two-thirds of reported victims of sexual assault were juveniles under the age of 18. Adolescents aged 12 to 18 years represented the largest group of victims at 33 percent; 20 percent were between the ages of six and 11; children younger than five years old and adults between 18 and 24 years of age each constituted 14 percent of victims. One out of every seven victims surveyed in the study was under the age of six.
- Gender: Females were more than six times more likely to be a victim of sexual assault than males; more than 86 percent of victims were females. The great majority (99%) of the victims of forcible rapes were women, while men constituted the majority (54%) of the victims of forcible sodomy (oral or anal intercourse). Females are most likely to be the victim of sexual assault at age 14, while males are at most risk at age four.
- Location: The residence of the victim was the most commonly noted location of sexual assault (70%). Other common locations included schools, hotels/motels, fields, woods, parking lots, roadways, and commercial/office buildings.

Similar statistics were gathered by the NCJJ regarding the perpetrators of rape and sexual assault. These characteristics included the following:

- Age: Over 23 percent of offenders were under the age of 18; juveniles were more likely to be perpetrators of forcible sodomy and fondling. The remaining 77 percent of offenders were adults and were responsible for 67 percent of juvenile victims. For younger juvenile victims (under the age of 12), juvenile offenders were responsible for approximately 40 percent of assaults.
- Gender: The great majority of all reported offenders were male (96%). The number of female offenders rose for victims under the age of six (12%), in contrast to 6 percent for victims aged six through 12, 3 percent for victims aged 12 through 17, and 1 percent for adult victims.
- Relationship with offender: Approximately 59 percent of offenders were acquaintances of their victims, compared to family members (27%) or strangers (14%). Family members were more likely to be perpetrators against juveniles (34%) than against adults (12%). In contrast, strangers accounted for 27 percent of adult victims and 7 percent of juveniles.
- Past offenses: In 19 percent of juvenile cases, the victim was not the only individual to be assaulted by the offender, compared to only 4 percent of adult cases.

Of particular importance are the number of rapes and sexual assaults that go unreported, especially in adolescents. Although one in five sexual assault reports occurs for adolescents between 12 and 17 years of age, and adolescents between ages 16 and 19 years have the highest rate of reported sexual assault, anonymous school surveys have revealed that only 5 percent of sexually assaulted adolescents actually report the crime to law enforcement.

When to call the doctor

Many children and adolescents are reluctant to report rape and sexual assault for a number of reasons. Often the victim fears retaliation from the offender. He or she may be afraid that family, friends, the community, or the media may learn about the offense. There may be a concern about being judged or blamed by others. The victim may think that no one will believe the assault occurred or that they were somehow at fault. Unreported rape and sexual assault are especially common when the offender is known to the victim, such as a family member or respected member of the community (e.g., clergy, teacher).

Parents who suspect that their child or adolescent has been raped or sexually assaulted should take the child to see a doctor and psychologist or psychiatrist. Signs that a child or adolescent may have been raped or sexually assaulted include shying away from physical affection, unexplained bleeding from the rectum and/or vagina, bruising around the breasts and genitals, and hiding or throwing away undergarments. Any child or adolescent who is raped or sexually assaulted should be taken to an emergency room immediately so that evidence against the perpetrator can be gathered, and medical treatment can be given.

Diagnosis

Rape and sexual assault are diagnosed by interviewing the patient and parents, physical and gynecological examination, and laboratory tests for the presence of seminal fluid. In many cases, children or adolescents do not report the rape or sexual assault, but they do show obvious signs of physical violence. When rape is suspected, diagnosis may be made by a psychiatrist or psychologist based on sessions with the victim. In cases where obvious signs of the crime are not visible, and immediate treatment is not received, the victim may develop post-traumatic stress disorder (PTSD), also known as rape trauma syndrome, which is a mental health disorder that describes a range of symptoms often experienced by someone who has undergone a severely traumatic event. In such cases, diagnosis of rape or sexual assault is revealed through therapy sessions for PTSD.

Approximately 31 percent of rape victims develop PTSD as a result of their assault. The symptoms of PTSD include:

- recurrent memories or flashbacks of the incident
- nightmares
- insomnia
- mood swings
- difficulty concentrating
- panic attacks
- emotional numbness
- depression
- **anxiety**

Treatment

Once a victim of rape or sexual assault reports the crime to local authorities, calls a rape crisis hotline, or arrives at the emergency room to be treated for injuries, a multidisciplinary team is often formed to address his or her physical, psychological, and judicial needs. This team usually includes law enforcement officers, physicians, nurses, mental health professionals, victim advocates, and/or prosecutors.

The victim may continue to feel fear and anxiety for some time after the incident, and in some instances this may significantly impact his or her personal and academic life. Follow-up counseling should, therefore, be provided for the victim, particularly if symptoms of PTSD become evident.

Forensic medical examination

Because rape is a crime, there are certain requirements for medical evaluation of the patient and for record keeping. The forensic medical examination is an invaluable tool for collecting evidence against a perpetrator that may be admissible in court. Since the great majority of victims know their assailant, the purpose of the medical examination is often not to establish identity but to establish nonconsensual sexual contact. The Sexual Assault Nurse Examiner program is an effective model that is used in many U.S. hospitals and clinics to collect and document evidence, evaluate and treat for **sexually transmitted diseases** (STDs) and pregnancy, and refer victims to follow-up medical care and counseling. Many nurse examiners are specially trained to handle cases that involve children and adolescents. The "Sexual Assault Nurse Examiner Development and Operation Guide," prepared by the Sexual Assault Resource Service, describes the ideal protocol for collecting evidence from a sexual assault victim. This protocol includes the following:

- performing the medical examination within 72 hours of the assault
- taking a history of the assault
- documenting the general health of the victim, including menstrual cycle, potential **allergies**, and pregnancy status
- assessing for trauma and taking photographic evidence of injuries
- taking fingernail clippings or scrapings
- taking samples for sperm or seminal fluid
- combing head/pubic hair for foreign hairs, fibers, and other substances
- collecting bloody, torn, or stained clothing
- taking samples for blood typing and DNA screening

After evidence is collected, rape victims are treated with appropriate medical care for their injuries. In female children and adolescents, vaginal tears and injuries may require suturing; in male children and adolescents, anal tears and injuries are common and may require suturing and other treatment.

Prognosis

Children and adolescents who have been raped or sexually assaulted are three times more likely to experience another rape in adulthood. Victims of rape and sexual assault who report their attack greater than one month afterwards are more likely to suffer from PTSD, mood swings, and major depression than victims who report their attack immediately, most likely because victims who report their attacks immediately receive appropriate interventional care, particularly mental health support and counseling. For adolescents, untreated rape and sexual assault can result in serious long-term psychological effects. One in 10 sexually assaulted adolescents attempt **suicide**, and about 50 percent are diagnosed with **phobias**, depression, substance abuse, and other psychological disorders. Compared to those who have never been victimized, rape victims are three times more likely to have a major depressive episode, four times more likely to have contemplated suicide, 13 times more likely to develop alcohol dependency problems, and 26 times more likely to develop substance abuse problems. In addition, school performance in many sexually assaulted adolescents declines, and many eventually fail academically. Even when treated, rape and sexual assault can cause poor **self-esteem**, sexual dysfunction and impaired sexual and personal relations, insomnia,

anxiety, eating disorders, and other psychological symptoms that last into adulthood.

Lasting psychological trauma is especially serious in male children and adolescents who are raped or sexually assaulted. Young boys may be more reluctant to discuss their attack and may harbor feelings of resentment and anxiety over potential **homosexuality**. Assaulted young boys may, in turn, commit sexual assault themselves in the future. Appropriate psychological therapy is necessary for improved long-term outcomes.

Prevention

Usually, rape and sexual assault cannot be prevented, and it is important that children and adolescents, who often think they are at fault after an attack, be told that there was nothing they could have done to prevent the attack. However, measures to reduce the likelihood of a rape or sexual assault and to increase the chances of an assailant being caught can be taken:

- Children and adolescents can be instructed on **safety** and strangers and inappropriate touching, and the importance of telling parents about any uncomfortable situation.
- Parents can monitor social activities, particularly for older adolescents, who may attend events (e.g., parties with no adult supervision) without their parents' knowledge.
- Adolescents can be educated about "date-rape" drugs and methods to prevent their consumption (e.g., never leaving drinks unattended at a party). And they can be informed about the dangers of alcohol consumption.
- Parents can encourage open communication regarding normal sexual development and activity and emphasize the importance of saying no in compromising or uncomfortable situations.

Sexually transmitted disease (STD) prevention

STDs are a source of concern for many victims of sexual assault. The most commonly transmitted diseases are gonorrhea, chlamydia, genital **warts**, and acquired **immunodeficiency** syndrome (AIDS)/human immunodeficiency virus (HIV). STDs are transmitted in up to 30 percent of rapes. Treatment involves **antibiotics** and antiviral medications, depending on the STD. In some instances, cultures may be taken during the medical examination and at time point afterward to test for gonorrhea or chlamydia. It is important that the victim receive information regarding the symptoms of STDs and be counseled to return for further examination if any of these symptoms occur.

KEY TERMS

Aggravated sexual abuse—When an individual is forced to submit to sexual acts by use of physical force; threats of death, injury, or kidnapping; or substances that render that individual unconscious or impaired.

Forcible sodomy—Forced oral or anal intercourse.

Forensic—Pertaining to courtroom procedure or evidence used in courts of law.

Incest—Unlawful sexual contact between persons who are biologically related. Many therapists, however, use the term to refer to inappropriate sexual contact between any members of a family, including stepparents and stepsiblings.

Post-traumatic stress disorder (PTSD)—A disorder that occurs among survivors of extremely stressful or traumatic events, such as a natural disaster, an airplane crash, rape, or military combat. Symptoms include anxiety, insomnia, flashbacks, and nightmares. Patients with PTSD are unnecessarily vigilant; they may experience survivor guilt, and they sometimes cannot concentrate or experience joy.

Sexual abuse—Forced sexual contact through the use of threats or other fear tactics, or instances in which an individual is physically unable to decline sexual activity.

Sexual assault nurse examiner—A registered nurse who is trained to collect and document evidence from a sexual assault victim, evaluate and treat for STDs and pregnancy, and refer victims to follow-up medical care and counseling.

Pregnancy prevention

Female adolescents at risk of becoming pregnant after an assault should be counseled on the availability of emergency **contraception**. According to the Food and Drug Administration (FDA), emergency contraception (in the form of a course of pills) is not effective if there is a pregnancy but works to prevent pregnancy from occurring by delaying or preventing ovulation, by affecting the transport of sperm, and/or by thinning the inner layer of the uterus (endometrium) so that implantation is prevented.

Parental concerns

Parents of children and adolescents who are raped or sexually assaulted are understandably upset, angry, and

even violent toward perpetrators. For the best mental health of the victim, parents should strive to listen to their children, use healthy coping strategies, and reassure the victim that he/she was not at fault. Parents should resist letting anger toward the assailant take precedence over attention to their child or adolescent.

Because rape and sexual assault cause long-term psychological trauma, parents should be aware of symptoms of PTSD, depression, substance abuse, high-risk behaviors, and anxiety in their children. Long-term therapy with a counselor experienced in rape and sexual assault trauma can benefit both the victim and parents.

Parents should also be aware that PTSD and other psychological effects of rape can manifest as poor school performance. Impaired concentration, **acting out** in school, diminished energy, embarrassment, and frustration may all occur in traumatized children and adolescents. School officials and counselors should be contacted to help students with academic problems; temporary homebound instruction or day therapy programs may be necessary.

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Rape, Abuse, and Incest National Network (RAINN). 635-B Pennsylvania Ave. SE, Washington, DC 20003. National Sexual Assault Hotline: 1–800–656-HOPE. Web site: <www.rainn.org/>.

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Jennifer E. Sisk, M.A.

Rashes

Definition

The popular term for a group of spots or red, inflamed skin that is usually a symptom of an underlying condition or disorder. Often temporary, a rash is only rarely a sign of a serious problem.

Description

A rash may occur on only one area of the skin, or it may cover almost all of the body. Also, a rash may or may not be itchy. Depending on how it looks, a rash may be described as having the following characteristics:

- blistering (raised oval or round collections of fluid within or beneath the outer layer of skin)
- macular (flat spots)
- nodular (small, firm, knotty rounded mass)
- papular (small solid slightly raised areas)
- pustular (pus-containing skin blister)

Demographics

Most persons experience rashes at many times in their lives. Rashes are not reportable events. As such, their prevalence is not precisely known. Rashes are common among infants, and most are harmless.

Causes and symptoms

There are many theories about the development of skin rashes, but experts are not completely sure what causes some of them. Generally a skin rash is an intermittent symptom, fading and reappearing. Rashes may accompany a range of disorders and conditions, such as the following:

- Infectious illness: A rash is a symptom of many different kinds of childhood infectious illnesses, including **chickenpox** and **scarlet fever**. It may be triggered by other infections, such as **Rocky Mountain spotted fever** or ringworm.

- Allergic reactions: One of the most common symptoms of an allergic reaction is an itchy rash. **Contact dermatitis** is a rash that appears after the skin is exposed to an allergen, such as metal, rubber, some cosmetics or lotions, or some types of plants (e.g. **poison ivy**). Drug reactions are another common allergic cause of rash; in this case, a rash is only one of a variety of possible symptoms, including **fever**, seizures, **nausea and vomiting**, **diarrhea**, heartbeat irregularities, and breathing problems. This rash usually appears soon after the first dose of the course of medicine is taken.
- Autoimmune disorders: Conditions in which the immune system turns on the body itself, such as systemic lupus erythematosus or purpura, often have a characteristic rash.
- Nutritional disorders: For example, scurvy, a disease caused by a lack of vitamin C, has a rash as one of its symptoms.
- **Cancer**: A few types of cancer, such as chronic lymphocytic leukemia, can be the underlying cause of a rash.

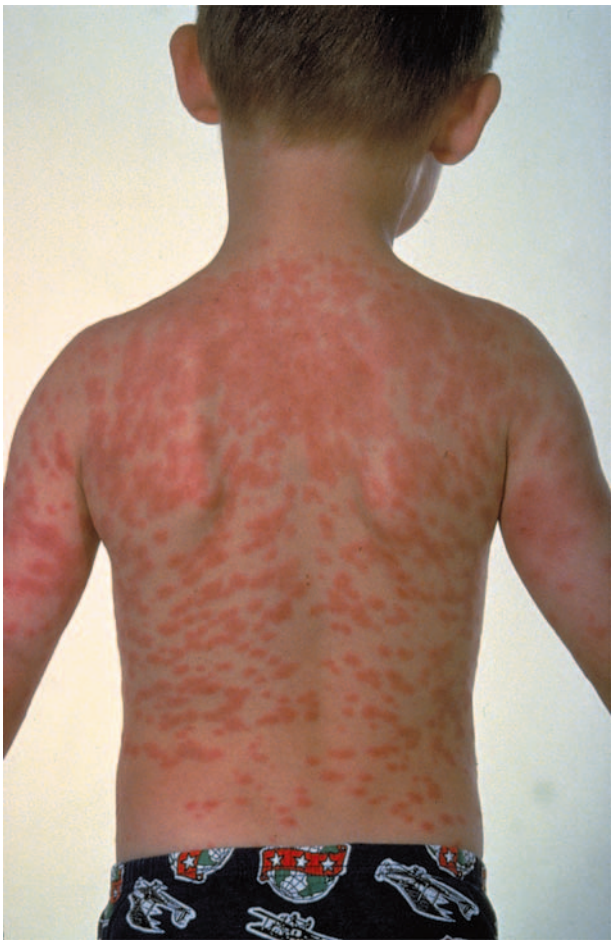
Rashes in infancy

Rashes are extremely common in infancy. They are usually not serious at all and can be treated at home.

Diaper rash is caused by prolonged skin contact with bacteria and the baby's waste products in a damp diaper. This rash has red, spotty sores, and there may be an ammonia smell. In most cases the rash will respond within three days to drying efforts. A diaper rash that does not improve in this time may be a yeast infection requiring prescription medication. A doctor should be consulted if the rash is solid, bright red, causes fever, or the skin develops blisters, boils, or pus.

Infants also can get a rash on cheeks and chin caused by contact with food and stomach contents. This rash will come and go, but usually responds to a good cleaning after meals. About one-third of all infants develop **acne** usually after the third week of life in response to their mothers' hormones before birth. This rash can last a few weeks to a few months. Heat rash is a mass of tiny pink bumps on the back of the neck and upper back caused by blocked sweat glands. The rash usually appears during hot, humid weather, although a baby with a fever can also develop the rash.

A baby should be seen by a doctor immediately if the rash appears suddenly and looks purple or blood-colored, looks like a burn, or appears while the infant seems to be sick.



An unidentified rash on young boy's back. *(Custom Medical Stock Photo Inc.)*

When to call the doctor

A doctor or other healthcare provider should be called when a rash that cannot accurately be identified appears or when an identified rash does not disappear in two to three days.

Diagnosis

A physician can make a diagnosis based on the medical history and the appearance of the rash, where it appears, and any other accompanying symptoms.

Treatment

Treatment of rashes focuses on resolving the underlying disorder and providing relief of the **itching** that often accompanies them. Soothing lotions or oral **antihistamines** can provide some relief, and **topical antibiotics** may be administered if the person, particularly a

KEY TERMS

Purpura—A group of disorders characterized by purplish or reddish brown areas of discoloration visible through the skin. These areas of discoloration are caused by bleeding from broken capillaries.

Scurvy—A nutritional disorder caused by vitamin C deficiency that is characterized by tiredness, muscle weakness, joint and muscle aches, a rash on the legs, bleeding gums, and skin bruising.

child, has caused a secondary infection by scratching. The rash triggered by **allergies** should disappear as soon as the allergen is removed; drug rashes will fade when the person stops taking the drug causing the allergy. For the treatment of diaper rash, the infant's skin should be exposed to the air as much as possible; ointments are not needed unless the skin is dry and cracked. Experts also recommend switching to cloth diapers and cleaning affected skin with plain water.

Prognosis

Most rashes that have an acute cause, such as an infection or an allergic reaction, will disappear as soon as the infection or irritant is removed from the body's system. Rashes that are caused by chronic conditions, such as autoimmune disorders, may remain indefinitely or fade and return periodically.

Prevention

Some rashes can be prevented, depending on the triggering factor. A person known to be allergic to certain drugs or substances should avoid those things in order to prevent a rash. Diaper rash can be prevented by using cloth diapers, keeping the diaper area very clean, changing diapers often, and by breastfeeding.

Nutritional concerns

Foods that are known to trigger rashes in persons should be avoided.

Parental concerns

Parents should monitor the foods that their children eat so that they will be able to identify foods that cause rashes and avoid their consumption. In addition, children who are prone to skin allergies should avoid contact with strong, perfumed soaps; nickel-based jewelry; and irritating fabrics (such as wool).

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American Academy of Dermatology. 930 N. Meacham Road, PO Box 4014, Schaumburg, IL 60168–4014. Web site: <www.aad.org/>.

American Academy of Family Physicians. 11400 Tomahawk Creek Parkway, Leawood, KS 66211–2672. Web site: <www.aafp.org/>.

American Academy of Pediatrics. 141 Northwest Point Blvd., Elk Grove Village, IL 60007–1098. Web site: <www.aap.org/>.

American College of Occupational and Environmental Medicine. 55 West Seegers Rd., Arlington Heights, IL 60005. Web site: <www.acoem.org/>.

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L. Fleming Fallon, Jr., MD, DrPH

RAST test *see* **Allergy tests**

Rat-bite fever

Definition

Rat-bite fever refers to an infection which develops in a person after being bitten or scratched by an infected animal.

Description

Rat-bite fever occurs most often among laboratory workers who handle lab rats in their jobs, and among people who live in poor conditions, with rodent infestation. Children are particularly likely to be bitten by rodents infesting their home and are, therefore, most likely to contract rat-bite fever. Other animals that can carry the types of bacteria responsible for this illness are mice, squirrels, weasels, dogs, and cats. One kind of bacteria can cause the same illness if it is ingested in unpasteurized milk or in water that has been contaminated with rat waste.

Demographics

About half of all cases of rat-bite fever occur in children.

Causes and symptoms

There are two variations of rat-bite fever, caused by two different organisms. In the United States, the bacteria *Streptobacillus moniliformis* is the most common cause (causing streptobacillary rat-bite fever). In other countries, especially Africa, *Spirillum minus* causes a different form of the infection (called spirillary rat-bite fever).

Streptobacillary rat-bite fever occurs up to 22 days after the initial bite or scratch. The patient becomes ill with fever, chills, **nausea and vomiting**, **headache**, and **pain** in the back and joints. A rash made up of tiny pink bumps develops, covering the palms of the hands and the soles of the feet. Without treatment, the patient is at risk of developing serious infections of the lining of the heart (endocarditis), the sac containing the heart (pericarditis), the coverings of the brain and spinal cord (**meningitis**), or lungs (**pneumonia**). Any tissue or organ throughout the body may develop a pocket of infection and pus, called an abscess.

Spirillary rat-bite fever occurs some time after the initial injury has already healed, up to about 28 days after the bite or scratch. Although the wound had appeared completely healed, it suddenly grows red and swollen again. Lymph nodes in the area become swollen and tender, and the patient develops fever, chills, and headache. The skin in the area of the original wound sloughs off. Although rash is less common than with streptobacillary rat-bite fever, there may be a lightly rosy, itchy rash all over the body. Joint and muscle pain rarely occur. If left untreated, the fever usually subsides, only to return again in repeated two- to four-day cycles. Though these cycles can last for a year, the illness usually resolves without treatment in four to eight weeks. This can go on for up to a year although, even without treatment, the illness usually resolves within four to eight weeks.

Diagnosis

In streptobacillary rat-bite fever diagnosis can be made by taking a sample of blood or fluid from a painful

KEY TERMS

Abscess—A localized collection of pus in the skin or other body tissue caused by infection.

Endocarditis—Inflammation of the inner membrane lining heart and/or of the heart valves caused by infection.

Meningitis—An infection or inflammation of the membranes that cover the brain and spinal cord. It is usually caused by bacteria or a virus.

Pasteurization—A process during which milk is heated and maintained at a particular temperature for the purpose of killing, or retarding the development of, pathogenic bacteria.

Pericarditis—Inflammation of the pericardium, the sac that surrounds the heart and the roots of the great blood vessels.

joint, which can be cultured to allow the growth of organisms. Examination under a microscope will then allow identification of the bacteria *Streptobacillus moniliformis*.

In spirillary rat-bite fever, diagnosis can be made by examining blood or a sample of tissue from the wound for evidence of *Spirillum minus*.

Treatment

Either injections of procaine penicillin G or penicillin V by mouth are effective against both streptobacillary and spirillary rat-bite fever. When a patient is allergic to the **penicillins**, either erythromycin may be given by mouth for streptobacillary infection or tetracycline by mouth for spirillary infection.

Prognosis

With treatment, prognosis is excellent for both types of rat-bite fever. Without treatment, the spirillary form usually resolves on its own, although it may take up to a year to do so.

The streptobacillary form, found in the United States, however, can progress to cause extremely serious, potentially fatal complications. In fact, before **antibiotics** were available to treat the infection, streptobacillary rat-bite fever frequently resulted in death.

Prevention

Prevention involves avoiding contact with those animals capable of passing on the causative organisms. This can be a difficult task for people whose economic situations do not allow them to move out of rat-infested buildings. Because streptobacillary rat-bite fever can occur after drinking contaminated milk or water, only pasteurized milk, and water from safe sources, should be ingested.

Parental concerns

The parents of children living in rodent-infested conditions, or who have pet rodents (mice, rats, gerbils) should be vigilant to illness in their children.

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Red blood cell indices

Definition

Red blood cell (RBC) indices are calculations derived from the complete blood count that aid in the diagnosis and classification of anemia. Measurements needed to calculate indices are the red blood cell count, hemoglobin, and hematocrit. The hematocrit is the percentage of blood by volume that is occupied by the red cells. The three RBC indices are:

- Mean corpuscular volume (MCV). The average size of the red blood cells expressed in femtoliters. MCV is calculated by dividing the hematocrit (as percent) by the RBC count in millions per microliter of blood, then multiplying by 10.
- Mean corpuscular hemoglobin (MCH). The average amount of hemoglobin inside an RBC expressed in picograms. The MCH is calculated by dividing the hemoglobin concentration in grams per deciliter by the RBC count in millions per microliter, then multiplying by 10.
- Mean corpuscular hemoglobin concentration (MCHC). The average concentration of hemoglobin in the RBCs expressed as a percent. It is calculated by dividing the hemoglobin in grams per deciliter by the hematocrit, then multiplying by 100.

Purpose

Red blood cell indices help classify types of anemia, a decrease in the oxygen carrying capacity of the blood. Healthy people have an adequate number of correctly sized red blood cells containing enough hemoglobin to carry sufficient oxygen to all the body's tissues. Anemia is diagnosed when either the hemoglobin or hematocrit of a blood sample is too low.

The mechanisms by which anemia occurs will alter the RBC indices in a predictable manner. Therefore, the RBC indices permit the physician to narrow down the possible causes of an anemia. The MCV is an index of the size of the RBCs. When the MCV is below normal, the RBCs will be smaller than normal and are described as microcytic. When the MCV is elevated, the RBCs will be larger than normal and are termed macrocytic. RBCs of normal size are termed normocytic. Failure to produce hemoglobin results in smaller than normal cells. This occurs in many diseases, including **iron deficiency anemia**, **thalassemia** (an inherited disease in which globin chain production is deficient), and **anemias** associated with chronic infection or disease. Macrocytic cells occur when division of RBC precursor cells in the bone marrow is impaired. The most common causes of macrocytic anemia are vitamin B_{12} deficiency, folate deficiency, and liver disease. Normocytic anemia may be caused by decreased production (e.g. malignancy and other causes of bone marrow failure), increased destruction (hemolytic anemia), or blood loss. The RBC count is low, but the size and amount of hemoglobin in the cells are normal.

A low MCH indicates that cells have too little hemoglobin. This is caused by deficient hemoglobin production. Such cells will be pale when examined under the microscope and are termed hypochromic. Iron deficiency is the most common cause of a hypochromic anemia. The MCH is usually elevated in macrocytic anemias associated with vitamin B_{12} and folate deficiency.

The MCHC is the ratio of hemoglobin mass in the RBC to cell volume. Cells with too little hemoglobin are lighter in color and have a low MCHC. The MCHC is low in microcytic, hypochromic anemias such as iron deficiency, but is usually normal in macrocytic anemias. The MCHC is elevated in hereditary spherocytosis, a condition with decreased RBC survival caused by a structural protein defect in the RBC membrane.

Description

Cell indices are usually calculated from tests performed on an automated electronic cell counter. However, these counters measure the MCV, which is directly proportional to the voltage pulse produced as each cell passes through the counting aperture. Electronic cell counters calculate the MCH, MCHC, hematocrit, and an additional parameter called the red cell distribution width (RDW). The RDW is a measure of the variance in red blood cell size. It is calculated by dividing the standard deviation of RBC volume by the MCV and multiplying by 100. A large RDW indicates abnormal variation in cell size, termed anisocytosis. The RDW aids in differentiating anemias that have similar indices. For example, thalassemia minor and iron deficiency anemia are both microcytic and hypochromic anemias, and overlap in MCV and MCH. However, iron deficiency anemia has an abnormally wide RDW, but thalassemia minor does not.

Precautions

Certain prescription medications may affect the test results. These drugs include zidovudine (Retrovir), phenytoin (Dilantin), and azathioprine (Imuran). When the hematocrit is determined by centrifugation, the MCV and MCHC may differ from those derived by an electronic cell counter, especially in anemia. Plasma trapped between the RBCs tends to cause an increase in the hematocrit, giving rise to a somewhat higher MCV and lower MCHC.

RBC indices require 3–5 mL of blood collected by venipuncture. A nurse or phlebotomist usually collects the sample following standard precautions for the prevention of transmission of bloodborne pathogens.

Aftercare

Discomfort or bruising may occur at the puncture site. Pressure to the puncture site until the bleeding stops reduces bruising; warm packs relieve discomfort. Some people feel dizzy or faint after blood has been drawn and should be treated accordingly.

KEY TERMS

Anemia—A condition in which there is an abnormally low number of red blood cells in the bloodstream. It may be due to loss of blood, an increase in red blood cell destruction, or a decrease in red blood cell production. Major symptoms are paleness, shortness of breath, unusually fast or strong heart beats, and tiredness.

Hypochromic—A descriptive term applied to a red blood cell with a decreased concentration of hemoglobin.

Macrocytic—A descriptive term applied to a larger than normal red blood cell.

Mean corpuscular hemoglobin concentration (MCHC)—A measurement of the average concentration of hemoglobin in a red blood cell.

Mean corpuscular hemoglobin (MCH)—A measurement of the average weight of hemoglobin in a red blood cell.

Mean corpuscular volume (MCV)—A measurement of the average volume of a red blood cell.

Microcytic—A descriptive term applied to a smaller than normal red blood cell.

Normochromic—A descriptive term applied to a red blood cell with a normal concentration of hemoglobin.

Normocytic—A descriptive term applied to a red blood cell of normal size.

Red blood cell indices—Measurements that describe the size and hemoglobin content of red blood cells. The indices are used to help in the differential diagnosis of anemia. Also called red cell absolute values or erythrocyte indices.

Red cell distribution width (RDW)—A measure of the variation in size of red blood cells.

Risks

The risks are potential bruising at the puncture site, and mild **dizziness**. Rarely excess bleeding, or infection of the puncture site occurs.

Normal results

Normal results for red blood cell indices are as follows:

- MCV: 78–102 fl (femtoliters) for ages 12–18 years, 77–95 fl for ages six to 12 years, 75–87 fl for ages two to six years, 70–86 fl for ages six months to two years, 85–123 fl for age one month
- MCH: 25–35 pg (picograms) for ages 12–18 years, 25–33 pg for ages six to 12 years, 24–30 pg for ages two to six years, 23–31 pg for ages six months to two years, 28–40 pg for age one month
- MCHC: 31–37 g/dL for ages two to 18 years, 30–36 g/dL for ages six months to two years, 29–37 pg for age one month
- RDW: 12–15 percent

Parental concerns

The **pain** from the needle puncture only lasts a moment. The parent should comfort a child as needed. Older children can be prepared for the test ahead of time, and the reason why the test is being given should also be explained if the child is old enough to understand.

When to call a doctor

If the bleeding does not stop at the needle puncture site, or if hours to days later, there appears to be infection (redness and swelling), then parents should contact a doctor.

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Reflex tests

Definition

Reflex tests are simple physical tests of nervous system function.

Purpose

A reflex is a simple nerve circuit. A stimulus, such as a light tap with a rubber hammer, causes sensory

Doctor performing a reflex test on a young girl's elbow. *(Photo Researchers, Inc.)*

neurons (nerve cells) to send signals to the spinal cord. Here, the signals are conveyed both to the brain and to nerves that control muscles affected by the stimulus. Without any brain intervention, these muscles may respond to an appropriate stimulus by contracting. Newborn babies have a particular set of reflexes not present in older babies, children, and adults.

Reflex tests measure the presence and strength of a number of reflexes. In so doing, they help to assess the integrity of the nerve circuits involved. Reflex tests are performed as part of a neurological exam, either a mini-exam done to quickly confirm integrity of the spinal cord or a more complete exam performed to diagnose the presence and location of **spinal cord injury** or neuromuscular disease.

Deep tendon reflexes are responses to muscle stretch. The familiar knee-jerk reflex is an example; this reflex tests the integrity of the spinal cord in the lower back region. The usual set of deep tendon reflexes tested, involving increasingly higher regions of the spinal cord, are:

- ankle
- knee
- abdomen
- forearm
- biceps
- triceps

Another type of reflex test is called the Babinski test, which involves gently stroking the sole of the foot to assess proper development of the spine and cerebral cortex.

Description

The examiner places the person in a comfortable position, usually seated on the examination table with legs hanging free. The examiner uses a rubber mallet to strike different points on the individual's body and observes the response. The examiner may position, or hold, one of the limbs during testing, and may require exposure of the ankles, knees, abdomen, and arms. Reflexes can be difficult to elicit if the person is paying too much attention to the stimulus. To compensate for this, the person may be asked to perform some muscle contraction, such as clenching teeth or grasping and pulling the two hands apart. When performing the Babinski reflex test, the doctor will gently stroke the outer soles of the person's feet with the mallet while checking to see whether the big toe extends out as a result.

Risks

Reflex tests are entirely safe, and no special precautions are needed.

Normal results

The strength of the response depends partly on the strength of the stimulus. For this reason, the examiner will attempt to elicit the response with the smallest stimulus possible. Learning the range of normal responses requires some clinical training. Responses should be the same for both sides of the body. A normal response to the Babinski reflex test depends upon the age of the person being examined. In children under the age of one-and-a-half years, the big toe will extend out with or without the other toes. This is due to the fact that the fibers in the spinal cord and cerebral cortex have not been completely covered in myelin, the protein and lipid sheath that aids in processing neural signals. In adults and children over

KEY TERMS

Babinski sign—Dorsiflexion (curling) of the big toe on stimulating the sole of the foot.

Neuron—The fundamental nerve cell of the nervous system.

the age of one-and-a-half years, the myelin sheath should be completely formed, and, as a result, all the toes will curl under (planter flexion reflex).

Parental concerns

Parents should expect reflex tests to be included in every examination given to their children by a doctor or other healthcare provider. Reflex tests present no risks. Parents should only be concerned when they are told of abnormal reflex test results.

See also Neonatal reflexes.

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American Academy of Neurology. 1080 Montreal Avenue, St. Paul, MN 55116. Web site: <www.aan.com/>.

American Academy of Pediatrics. 141 Northwest Point Blvd., Elk Grove Village, IL 60007–1098. Web site: <www.aap.org/default.htm>.

American Academy of Physical Medicine and Rehabilitation. One IBM Plaza, Suite 2500, Chicago, IL 60611–3604. Web site: <www.aapmr.org/>.

American College of Emergency Physicians. PO Box 619911, Dallas, TX 75261–9911. Web site: <www.acep.org/>.

American College of Physicians. 190 N Independence Mall West, Philadelphia, PA 19106–1572. Web site: <www.acponline.org/>.

American College of Sports Medicine. 401 W. Michigan St., Indianapolis, IN 46202–3233. Web site: <www.acsm.org/>.

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Reflexes, neonatal *see* **Neonatal reflexes**

Refsum disease *see* **Peroxisomal disorders**

Rehydration therapy *see* **Intravenous rehydration**

Remarriage *see* **Stepfamilies**

Renal vein thrombosis

Definition

Renal vein thrombosis develops when a blood clot forms in the renal vein, which is the blood vessel that carries blood from the kidneys back to the heart. The disorder is not common.

Description

Normally, kidneys rid the body of wastes by filtering the wastes into the bladder where they exit the body through the urine. When one or more blood vessels in the kidneys become narrowed (renal artery stenosis) because of debris and plaque build-up, or blocked because of a blood clot (renal vein thrombosis), the kidneys are unable to function properly. There is usually a rise in blood pressure, and kidney failure can occur without prompt treatment.

The onset of renal vein thrombosis can be rapid (acute) or gradual.

Blood clots in the renal arteries are uncommon, but when they do occur, there is a risk of pulmonary embolism, a dangerous condition that occurs when the clot or a portion of the clot dislodges and travels to the lungs. There is also an increased risk of congestive heart failure, a condition in which the heart's pumping power is weaker than normal.

Demographics

Renal vein thrombosis occurs in both infants and adults. The number of people who suffer from renal vein thrombosis is difficult to determine, as many do not show symptoms, and the disorder is diagnosed only by specific tests. Ninety percent of pediatric cases of renal vein thrombi occur in infants less than one year old; 75 percent occur in infants under one month of age.

Causes and symptoms

Causes

In children, most cases of renal vein thrombosis are thought to be caused by an episode of severe **dehydration**. Severe dehydration decreases blood volume and causes the blood to clot more readily. Symptoms occur rapidly.

In adults, renal vein thrombosis can be caused by injury to the abdomen or back, malignant kidney tumors growing into the renal vein, scar formation (stricture) and other blockages in the vein, or kidney diseases that cause degenerative changes in the cells of the renal tubules (nephrotic syndrome).

Renal vein thrombosis is more common in patients with nephrotic syndrome, although studies have shown high variability among these patients, with rates of 5 to 62 percent reported. Nephrotic syndrome is marked by abnormally low levels of albumin (hypoalbuminemia), abnormally high levels of cholesterol in the blood (hypercholesterolemia), and fluid retention (edema). Minimal change disease is a form of nephrotic syndrome seen in children, characterized by swelling and weight (from fluid retention), foamy urine, and loss of appetite.

Symptoms

Acute onset of renal vein thrombosis at any age causes **pain** in the lower back and sides of the abdomen, **fever**, bloody urine, decreased urine output, and sometimes kidney failure.

Other symptoms include high blood pressure or a "whooshing" sound heard by the physician when he or she places a stethoscope on the abdomen. This sound is the result of blood attempting to pass through the blocked vessel. The doctor may also feel an enlarged kidney during a physical exam. Some patients have no symptoms.

When to call the doctor

If the child has any of these symptoms, the parent should seek emergency medical care:

- a high fever of 102° F, or 38.9°C, or above
- sudden onset of lower back pain
- sudden, severe leg swelling
- difficulty breathing
- blood in the urine
- decreased urination

If the child has any of these symptoms of dehydration, the parent should give the child clear fluids and an oral rehydrating solution, such as Pedialyte, and contact the child's pediatrician:

- dry mouth
- increased or excessive thirst
- few or no tears when crying
- dark yellow urine
- irritability
- low energy or severe weakness
- lightheadedness or fainting
- sunken abdomen, eyes, and cheeks

Diagnosis

A physician makes the diagnosis of renal vein thrombosis based on the presence of symptoms and the results of a medical examination and diagnostic tests. When examining the child, the doctor will palpate (feel) the child's abdomen to detect kidney enlargement. The doctor will listen to the child's heartbeat with a stethoscope. He or she will also place the stethoscope over the child's abdomen; when renal vein thrombosis is present, the doctor may hear an abnormal ''whooshing'' as blood tries to flow through the blocked vessel.

Urine tests and blood tests are usually performed. If nephrotic syndrome is present, the urine test may indicate an abnormally large quantity of protein, and the blood test may show abnormally high levels of cholesterol.

Vascular ultrasound is a non-invasive ultrasound method used to examine blood circulation and detect the presence of blood clots. During a vascular ultrasound, an ultrasound transducer (small hand-held device) is placed over the area being examined. The transducer generates high-frequency sound waves through the tissues. These sound waves reflect off blood cells moving within the blood vessels, allowing the radiologist to calculate their speed. The sound waves are measured, recorded, and displayed on a computer screen.

Other tests that may be used to detect a blood clot include **computed tomography** scans (CT scans) and **magnetic resonance imaging** (MRI).

A less common test used to diagnose renal vein thrombosis is renal venography, also called renal angiography, an x-ray examination of the renal veins after a contrast material (dye) has been injected. This test may be performed to locate the narrowing or blockage in the renal vein. During this test, a catheter (long, thin tube) is inserted into the vein in the groin area (femoral vein) and threaded first into the right kidney and then into the vein of the other kidney. Blood samples may be taken from each kidney for further testing. The contrast material is injected through the catheter into each vein and **x rays** are taken. This test is not common, since other less invasive imaging tests, including MRI and CT scans, are available to aid the physician in making an accurate diagnosis.

Treatment

One of the major goals of treatment is to prevent the blood clot in the renal vein from detaching and moving into the lungs (pulmonary embolism), where it can cause serious complications.

Clot-busting medications, such as tissue plasminogen activator (t-PA, also called streptokinase enzymes or thrombolytic drugs), may be given to help dissolve the renal clot. Clot busters must be administered quickly and properly through several specifically timed intravenous infusions according to a rigid protocol established for each drug and the body weight of each patient. Research has shown that these medications are most effective when given within two hours of the onset of symptoms.

Anticoagulant medications, including heparin or warfarin and low-dose aspirin, may be prescribed to prevent existing blood clots from enlarging and to prevent the formation of new clots. The use of these medications in children remains controversial because of the risk of **Reye's syndrome**. Sometimes the potential benefits of these medications outweigh the risk of side effects. Researchers agree that more studies are needed to determine the proper dosage and effectiveness of aspirin and other anticoagulant medications in children.

Bedrest or limited activity may be recommended for a brief period.

Severe dehydration requires medical treatment with intravenous (IV) fluids and may require **hospitalization**. IV therapy can be followed with oral rehydration as the child's condition improves.

If the renal artery is partially or completely blocked, an interventional catheter-based procedure may be performed. During the catheterization, a long, slender tube called a catheter is inserted into a vein or artery and slowly directed to the blocked blood vessel, using x-ray guidance (angiography). A specialized balloon tip or other device at the end of the catheter can be used to open the blocked or narrowed vessel. The balloon is rapidly inflated and deflated to open or widen the area. In some patients, a stent (metal mesh tube) can be placed to act as a scaffold and hold the area open.

Intra-arterial thrombolysis involves threading a catheter with clot-busting medication through a large blood vessel in the groin to the site of the clot. Using renal angiography, doctors pinpoint the precise location of the blockage and deliver the medication to that spot.

Endarterectomy and surgical bypass are two surgical treatment options, but they are not common in children. In a renal endarterectomy, a vascular surgeon removes the blockage from the inner lining of the renal artery. Bypass surgery reroutes the blood flow around the narrowed or blocked sections of the renal artery. Rarely, when there is a complete blockage of the renal vein in infants, the kidney must be surgically removed.

KEY TERMS

Albumin—A blood protein that is made in the liver and helps to regulate water movement in the body.

Anticoagulant drug—A drug used to prevent clot formation or to prevent a clot that has formed from enlarging. Anticoagulant drugs inhibit clot formation by blocking the action of clotting factors or platelets. They fall into three groups: inhibitors of clotting factor synthesis, inhibitors of thrombin, and antiplatelet drugs.

Antiplatelet drug—Drugs that inhibit platelets from aggregating to form a plug. They are used to prevent clotting and alter the natural course of atherosclerosis.

Arteriosclerosis—A chronic condition characterized by thickening, loss of leasticity, and hardening of the arteries and the build-up of plaque on the arterial walls. Arteriosclerosis can slow or impair blood circulation. It includes atherosclerosis, but the two terms are often used synonymously.

Artery—A blood vessel that carries blood away from the heart to the cells, tissues, and organs of the body.

Blood clotting—Also called coagulation. A natural process in which blood cells and fibrin strands clump together to stop bleeding after a blood vessel has been injured.

Clot—A soft, semi-solid mass that forms when blood coagulates.

Clot busters—Also called thrombolytics. Medications used to break up a blood clot.

Clotting factors—Substances in the blood, also known as coagulation factors, that act in sequence to stop bleeding by triggering the formation of a clot. Each clotting factor is designated with a Roman numeral I through XIII.

Coagulate—To clot or cause hemostasis; in electrosurgery, to cause tissue dehydration without cutting.

Computed tomography (CT)—An imaging technique in which cross-sectional x rays of the body are compiled to create a three-dimensional image of the body's internal structures; also called computed axial tomography.

Dehydration—An excessive loss of water from the body. It may follow vomiting, prolonged diarrhea, or excessive sweating.

Embolism—A blood clot, air bubble, or mass of foreign material that travels and blocks the flow of blood in an artery. When blood supply to a tissue or organ is blocked by an embolism, infarction, or death of the tissue the artery feeds, occurs. Without immediate and appropriate treatment, an embolism can be fatal.

Embolus—Plural, emboli. An embolus is something that blocks the blood flow in a blood vessel. It may be a gas bubble, a blood clot, a fat globule, a mass of bacteria, or other foreign body that forms somewhere else and travels through the circulatory system until it gets stuck.

Hypercoagulable state—(Also called thromboembolic state or thrombophilia.) A condition characterized by excess blood clotting.

Hypertension—Abnormally high arterial blood pressure, which if left untreated can lead to heart disease and stroke.

Intravenous (IV) therapy—Administration of fluids or medications through a vein, usually in the hand or arm.

Magnetic resonance imaging (MRI)—An imaging technique that uses a large circular magnet and radio waves to generate signals from atoms in the body. These signals are used to construct detailed images of internal body structures and organs, including the brain.

Nephrologist—A physician who specializes in treating diseases of the kidney.

Platelet—A cell-like particle in the blood that plays an important role in blood clotting. Platelets are activated when an injury causes a blood vessel to break. They change shape from round to spiny, "sticking" to the broken vessel wall and to each other to begin the clotting process. In addition to physically plugging breaks in blood vessel walls, platelets also release chemicals that promote clotting.

Stenosis—A condition in which an opening or passageway in the body is narrowed or constricted.

Thrombolysis—The process of dissolving a blood clot.

Thrombolytics—Drugs that dissolve blood clots. Thrombolytics are used to treat embolisms.

Thrombus—A blood clot that forms within a blood vessel or the heart.

KEY TERMS (contd.)

Ultrasonography—A medical test in which sound waves are directed against internal structures in the body. As sound waves bounce off the internal structure, they create an image on a video screen. Ultrasonography is often used to diagnose fetal abnormalities, gallstones, heart defects, and tumors. Also called ultrasound imaging.

Urologist—A physician who specializes in the anatomy, physiology, diseases, and care of the urinary tract (in men and women) and male reproductive tract.

Blood pressure medications may be prescribed to treat high blood pressure, and other medications may include diuretics, beta blockers, ACE inhibitors, and calcium channel blockers. Some of these drugs have not been extensively studied in children, and a specific pediatric dose has not been established.

Nutritional concerns

Impaired kidneys cause an increased level of phosphorus in the blood, which interferes with calcium absorption. In addition, damaged kidneys cannot activate vitamin D, which is needed to absorb calcium. Dietary changes may include limiting foods high in phosphorus, such as dairy products, meat, and poultry. A phosphate binder may be recommended to keep phosphorus in the bowel (so it does not interfere with calcium absorption) where it is excreted during a bowel movement. Calcium or vitamin D supplements also may be recommended. To maintain adequate **nutrition**, a registered dietitian can help parents and children implement specific dietary changes.

Prognosis

Most cases of renal vein thrombosis resolve over time, without permanent injury to the kidneys. Acute renal failure can occur with severe dehydration. Death from renal vein thrombosis is rare, and is often caused by the blood clot detaching and lodging in the heart or lungs.

Prevention

Renal vein thrombosis cannot be prevented. Preventing dehydration by maintaining fluids in the body may help reduce the risk of renal vein thrombosis.

Parental concerns

Most cases of renal vein thrombosis resolve without complication. When a child has been diagnosed with renal vein thrombosis, it is important to follow the doctor's recommendations for follow-up checkups to closely monitor his or her condition. If an anticoagulant medication has been prescribed, it is important to keep all scheduled laboratory appointments so the effectiveness of the medication can be evaluated.

If the child has developed any new symptoms, the parents should call the child's doctor.

Resources

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American Kidney Fund. 6110 Executive Blvd., Suite 1010, Rockville, MD 20852. (800) 638-8299. <www.akfinc.org>

National Institute of Diabetes and Digestive and Kidney Diseases (NIDDK). 2 Information Way, Bethesda, MD 20892-3570. (800) 891- 5389. nddic@info.niddk.nih.gov. <www.niddk.nih.gov>

National Kidney and Urologic Diseases Information Clearinghouse. 3 Information Way, Bethesda, MD 20892-3580. (800) 891-5390. nkudic@info.niddk.nih.gov. <http://kidney.niddk.nih.gov>

National Kidney Foundation. 30 East 33rd St., New York, NY 10016. (800) 622-9010 or (212) 889-2210. info@kidney.org. <www.kidney.org>

WEB SITES

VascularWeb. Provided by the Society for Vascular Surgery. <www.vascularweb.org>

Angela M. Costello

Rendu-Osler-Weber disease *see* **Hereditary hemorrhagic telangiectasia**

Respiratory distress syndrome

Definition

Respiratory distress syndrome (RDS) of the newborn, also known as infant RDS, is an acute lung disease

present at birth, which usually affects premature babies. Layers of tissue called hyaline membranes keep the oxygen that is breathed in from passing into the blood. The lungs are said to be airless. Without treatment, the infant will die within a few days after birth, but if oxygen can be provided, and the infant receives modern treatment in a neonatal intensive care unit, complete recovery with no after-effects can be expected.

Description

If a newborn infant is to breathe properly, the small air sacs (alveoli) at the ends of the breathing tubes must remain open so that oxygen in the air can get into the tiny blood vessels that surround the alveoli. Normally, in the last months of pregnancy, cells in the alveoli produce a substance called surfactant, which keep the surface tension inside the alveoli low so that the sacs can expand at the moment of birth, and the infant can breathe normally. Surfactant is produced starting at about 34 weeks of pregnancy and, by the time the fetal lungs mature at 37 weeks, a normal amount is present.

If an infant is born prematurely, enough surfactant might not have formed in the alveoli causing the lungs to collapse and making it very difficult for the baby to get enough air (and the oxygen it contains). Sometimes a layer of fibrous tissue called a hyaline membrane forms in the air sacs, making it even harder for oxygen to get through to the blood vessels. RDS in newborn infants used to be called hyaline membrane disease.

Demographics

According to the National Heart, Lung, and Blood Institute, in 2003, approximately 40,000 infants and 150,000 adults were reported to have RDS. Translated, these figures means RDS affected about one person in 6,800.

Causes and symptoms

RDS nearly always occurs in premature infants, and the more premature the birth, the greater is the chance that RDS will develop. RDS also is seen in some infants whose mothers are diabetic. Paradoxically, RDS is less likely in the presence of certain states or conditions which themselves are harmful: abnormally slow growth of the fetus; high blood pressure, a condition called preeclampsia in the mother; and early rupture of the birth membranes.

Labored breathing (the respiratory distress of RDS) may begin as soon as the infant is born, or within a few hours. Breathing becomes very rapid, the nostrils flare, and the infant grunts with each breath. The ribs, which are very flexible in young infants, move inwards each time a breath is taken. Before long the muscles that move the ribs and diaphragm, so that air is drawn into the lungs, become fatigued. When the oxygen level in the blood drops severely the infant's skin turns bluish in color. Tiny, very premature infants may not even have signs of trouble breathing. Their lungs may be so stiff that they cannot even start breathing when born.

There are two major complications of RDS. One is called pneumothorax, which means "air in the chest." When the infant itself or a breathing machine applies pressure on the lungs in an attempt to expand them, a lung may rupture, causing air to leak into the chest cavity. This air causes the lung to collapse further, making breathing even harder and interfering with blood flow in the lung arteries. The blood pressure can drop suddenly, cutting the blood supply to the brain. The other complication is called intraventricular hemorrhage; this is bleeding into the cavities (ventricles) of the brain, which may be fatal.

When to call the doctor

A doctor attending a birth should recognize respiratory distress and immediately begin appropriate treatment. A doctor should be called if a baby delivered outside of a hospital setting is observed to have any difficulty in breathing or whose skin becomes blue in color (cyanotic).

Diagnosis

When a premature infant has obvious trouble breathing at birth or within a few hours of birth, RDS is an obvious possibility. If premature birth is expected, or there is some condition that calls for delivery as soon as possible, the amount of surfactant in the amniotic fluid will indicate how well the lungs have matured. If little surfactant is found in an amniotic fluid sample taken by placing a needle in the uterus (**amniocentesis**), there is a definite risk of RDS. Often this test is done at regular intervals so that the infant can be delivered as soon as the lungs are mature. If the membranes have ruptured, surfactant can easily be measured in a sample of vaginal fluid.

The other major diagnostic test is a chest x ray. Collapsed lung tissue has a typical appearance, and the more lung tissue is collapsed, the more severe the RDS. An x ray also can demonstrate pneumothorax, if this complication has occurred. The level of oxygen in the blood can be measured by taking a blood sample from an artery, or, more easily, using a device called an oximeter, which is

clipped to an earlobe. Pneumothorax may have occurred if the infant suddenly becomes worse while on ventilation; x rays can help make the diagnosis.

Treatment

If only a mild degree of RDS is present at birth, placing the infant in an oxygen hood may be enough. It is important to guard against too much oxygen, as this may damage the retina and cause loss of vision. Using an oximeter to keep track of the blood oxygen level, repeated artery punctures or heel sticks can be avoided. In more severe cases a drug very like natural surfactant (Exosurf Neonatal or Survanta) can be dripped into the lungs through a fine tube (endotracheal tube) placed in the infant's windpipe (trachea). Typically, the infant will be able to breathe more easily within a few days at the most, and complications such as lung rupture are less likely to occur. The drug is continued until the infant starts producing its own surfactant. There is a risk of bleeding into the lungs from surfactant treatment; about 10 percent of the smallest infants are affected.

Infants with severe RDS may require treatment with a ventilator, a machine that takes over the work of the lungs and delivers air under pressure. In tiny infants who do not breathe when born, ventilation through a tracheal tube is an emergency procedure. Assisted ventilation must be closely supervised, as too much pressure can cause further lung damage. A gentler way of assisting breathing, continuous positive airway pressure (CPAP), delivers an oxygen mixture through nasal prongs or a tube placed through the nose rather than an endotracheal tube. CPAP may be tried before resorting to a ventilator or after an infant placed on a ventilator begins to improve. Drugs that stimulate breathing may speed the recovery process.

Pneumothorax is an emergency condition that must be treated right away. Air may be removed from the chest using a needle and syringe. A tube then is inserted into the lung cavity, and suction applied.

Prognosis

If an infant born with RDS is not promptly treated, lack of an adequate oxygen supply will damage the body's organs and eventually cause them to stop functioning altogether. Death is the result. The central nervous system in particular—made up of the brain and spinal cord—is very dependent on a steady oxygen supply and is one of the first organ systems to feel the effects of RDS. By contrast, if the infant's breathing is supported until the lungs mature and make their own surfactant, complete recovery within three to five days is the pattern.

KEY TERMS

Alveoli—The tiny air sacs clustered at the ends of the bronchioles in the lungs in which oxygen-carbon dioxide exchange takes place.

Amniotic fluid—The liquid in the amniotic sac that cushions the fetus and regulates temperature in the placental environment. Amniotic fluid also contains fetal cells.

Endotracheal tube—A hollow tube that is inserted into the trachea (windpipe) through the nose or mouth. It is used to administer anesthesia, to deliver oxygen under pressure, or to deliver medications (e.g. surfactants).

Hyaline membrane—A fibrous layer that settles in the alveoli in respiratory distress syndrome and prevents oxygen from escaping from inhaled air to the bloodstream.

Pneumothorax—A collection of air or gas in the chest or pleural cavity that causes part or all of a lung to collapse.

Preeclampsia—A condition that develops after the twentieth week of pregnancy and results in high blood pressure, fluid retention that doesn't go away, and large amounts of protein in the urine. Without treatment, it can progress to a dangerous condition called eclampsia, in which a woman goes into convulsions.

Steroid—A class of drugs resembling normal body substances that often help control inflammation in the body tissues.

Surfactant—A protective film secreted by the alveoli in the lungs that reduces the surface tension of lung fluids, allowing gas exchange and helping maintain the elasticity of lung tissue. Surfactant is normally produced in the fetal lungs in the last months of pregnancy, which helps the air sacs to open up at the time of birth so that the newborn infant can breathe freely. Premature infants may lack surfactant and are more susceptible to respiratory problems without it.

Ventilator—A mechanical device that can take over the work of breathing for a patient whose lungs are injured or are starting to heal. Sometimes called a respirator.

If an air leak causes pneumothorax, immediate removal of air from the chest allows the lungs to re-expand. Bleeding into the brain is a very serious condition that worsens the outlook for an infant with RDS.

Prevention

The best way of preventing RDS is to delay delivery until the fetal lungs have matured and are producing enough surfactant, generally at about 37 weeks of pregnancy. If delivery cannot be delayed, the mother may be given a steroid hormone, similar to a natural substance produced in the body, which crosses the barrier of the placenta and helps the fetal lungs to produce surfactant. The steroid should be given at least 24 hours before the expected time of delivery. If the infant does develop RDS, the risk of bleeding into the brain will be much less if the mother has been given a dose of steroid.

If a very premature infant is born without symptoms of RDS, it may be wise to deliver surfactant to its lungs. This may prevent RDS or make it less severe if it does develop. An alternative is to wait until the first symptoms of RDS appear and then immediately give surfactant. Pneumothorax may be prevented by frequently checking the blood oxygen content and limiting oxygen treatment under pressure to the minimum needed.

Parental concerns

Parents should monitor their newborn infant's breathing status closely for the first week of life. Premature infants are of particular concern, although many may be hospitalized through the neonatal period. While a newborn is hospitalized, parents should try to maintain as much physical contact with the infant as is allowed by the hospital, and let the infant frequently hear the familiar voices of the parents, especially the mother, when physical contact is not recommended.

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American Academy of Family Physicians. 11400 Tomahawk Creek Parkway, Leawood, KS 66211–2672. Web site: <www.aafp.org/>.

American Academy of Pediatrics. 141 Northwest Point Boulevard, Elk Grove Village, IL 60007–1098. Web site: <www.aap.org/default.htm>.

American College of Emergency Physicians. PO Box 619911, Dallas, TX 75261–9911. Web site: <www.acep.org/>.

American Lung Association. 1740 Broadway, New York, NY 10019. Web site: <www.lungusa.org/diseases/lungtb.html>.

American Thoracic Society. 1740 Broadway, New York, NY 10019. Web site: <www.thoracic.org/>.

Canadian Cystic Fibrosis Foundation. 2221 Yonge St., Suite 601, Toronto, Ontario, M4S 2B4, Canada. Web site: <www.ccff.ca/home.cfm>.

Cystic Fibrosis Foundation. 6931 Arlington Road, Bethesda, MD 0814. Web site: <www.cff.org/>.

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Respiratory syncytial virus infection

Definition

Respiratory syncytial virus (RSV) is a virus that can cause severe lower respiratory infections in children younger than two years of age and milder upper respiratory infections in older children and adults. RSV infection in young children is also called **bronchiolitis**, because it is marked by inflammation of the bronchioles, the narrow airways that lead from the large airways (bronchi) to the tiny air sacs (alveoli) in the lungs. The symptoms include wheezing, difficulty breathing, and sometimes respiratory failure.

Description

RSV infection is caused by a group of viruses found worldwide. There are two different subtypes of the virus with numerous different strains. Taken together, these viruses account for a significant number of deaths in infants.

RSV infection shows distinctly different symptoms, depending on the age of the infected person. In young children, the virus causes a serious lower respiratory infection in the lungs. In older children and healthy adults, it causes a mild upper respiratory infection often mistaken for the **common cold**.

Although anyone can get this disease, infants suffer the most serious symptoms and complications. Breastfeeding seems to provide partial protection from the virus. Conditions in infants that increase their risk of infection include:

- premature birth
- lower socio-economic environment
- congenital heart disease
- chronic lung diseases, such as cystic fibrosis
- immune system deficiencies, including HIV infection
- immunosuppressive therapy, such as that given to organ transplant or **cancer** patients

Many older children and adults get RSV infection, but the symptoms are so similar to the common cold that the true cause is undiagnosed. People of any age with compromised immune systems, either from such diseases as **AIDS** or leukemia, or as the result of **chemotherapy** or corticosteroid medications, and patients with chronic lung disease are more at risk for serious RSV infections.

Demographics

RSV infection is primarily a disease of winter or early spring, with waves of illness sweeping through a community. The rate of RSV infection is estimated to be 11.4 cases for every 100 children during their first year of life. In the United States, RSV infection occurs most frequently in infants between the ages of two months and six months.

Respiratory syncytial virus is spread through close contact with an infected person. It has been shown that if a person with RSV infection sneezes, the virus can be carried to others within a radius of 6 feet (1.8 m). This group of viruses can live on the hands for up to half an hour and on **toys** or other inanimate objects for several hours.

Scientists had, as of 2004, not understood why RSV viruses attack the lower respiratory system in infants and the upper respiratory system in adults. In infants, RSV begins with such cold symptoms as a low **fever**, runny nose, and **sore throat**. Soon, other symptoms appear that suggest an infection that involves the lower airways. Some of these symptoms resemble those of **asthma**. RSV infection is suggested by the following characteristics:

- wheezing and high-pitched, whistling breathing
- rapid breathing (more than 40 breaths per minute)
- shortness of breath
- labored breathing out (exhalations)
- bluish tinge to the skin (cyanosis)
- croupy, seal-like, barking **cough**
- high fever

Breathing problems occur in RSV infections because the bronchioles swell, making it difficult for air to get in and out of the lungs. If the child is having trouble breathing, immediate medical care is needed. Breathing problems are most common in infants under one year of age; they can develop rapidly.

Diagnosis

Physical examination and imaging studies

RSV infection is usually diagnosed during a physical examination by the pediatrician or primary care doctor. The doctor listens with a stethoscope for wheezing and other abnormal lung sounds in the patient's chest. The doctor will also take into consideration whether there is a known outbreak of RSV infection in the area. Chest x rays give some indication of whether the lungs are hyperinflated from an effort to move air in and out.

X rays may also show the presence of a secondary bacterial infection, such as **pneumonia**.

Laboratory tests

A nasal swab can be obtained to isolate the virus or antibodies to the virus in secretions. If infants are hospitalized, other tests such as an arterial blood gas analysis are done to determine if the child is receiving enough oxygen.

Treatment

Home care

Home treatment for RSV infection is primarily supportive. It involves taking steps to ease the child's breathing. **Dehydration** can be a problem, so children should be encouraged to drink plenty of fluids. **Antibiotics** have no effect on viral illnesses. In time, the body will make antibodies to fight the infection and return itself to health.

Home care for keeping a child with RSV comfortable and breathing more easily includes:

- use of a cool mist room humidifier to ease congestion and sore throat
- elevation of that baby's head by putting books under the head end of the crib
- acetaminophen (Tylenol, Pandol, Tempra) for fever (Aspirin should not be given to children because of its association with **Reye's syndrome**, a serious disease.)
- For babies too young to blow their noses, suctioning mucus with an infant nasal aspirator

Hospital treatment

In the United States, RSV infections are responsible for 90,000 hospitalizations and 4,500 deaths each year. Children who are hospitalized receive oxygen and humidity through a mist tent or vaporizer. They also are given intravenous fluids to prevent dehydration. Mechanical ventilation may be necessary. Blood gases are monitored to assure that the child is receiving enough oxygen.

Medications

Bronchiodilators, such as albuterol (Proventil, Ventilin), may be used to keep the airways open. Ribavirin (Virazole) is used for desperately ill children to stop the growth of the virus. Ribavirin is both expensive and has toxic side effects, so its use is restricted to the most severe cases.

KEY TERMS

Alveoli—The tiny air sacs clustered at the ends of the bronchioles in the lungs in which oxygen-carbon dioxide exchange takes place.

Antibody—A special protein made by the body's immune system as a defense against foreign material (bacteria, viruses, etc.) that enters the body. It is uniquely designed to attack and neutralize the specific antigen that triggered the immune response.

Reye's syndrome—A serious, life-threatening illness in children, usually developing after a bout of flu or chickenpox, and often associated with the use of aspirin. Symptoms include uncontrollable vomiting, often with lethargy, memory loss, disorientation, or delirium. Swelling of the brain may cause seizures, coma, and in severe cases, death.

Prognosis

RSV infection usually runs its course in seven to 14 days. The cough may linger for weeks. There are no medications that can speed the body's production of antibodies against the virus. Opportunistic bacterial infections that take advantage of a weakened respiratory system may cause ear, sinus, and throat infections or pneumonia.

Hospitalization and death are much more likely to occur in children whose immune systems are weakened or who have underlying diseases of the lungs and heart. People do not gain permanent immunity to respiratory syncytial virus and can be infected many times. Children who suffer repeated infections seem to be more likely to develop asthma in later life.

Prevention

As of 2004, there were no vaccines against RSV. Respiratory syncytial virus infection is so common that prevention is impossible. However, steps can be taken to reduce a child's contact with the disease. People with RSV symptoms should stay at least six feet away from young children. Frequent hand washing, especially after contact with respiratory secretions, and the correct disposal of used tissues help keep the disease from spreading. Parents should try to keep their children under 18 month of age away from crowded environments where they are likely to come in contact with older people who have only mild symptoms of the disease. Childcare centers should regularly disinfect surfaces that children touch.

Parental concerns

Because symptoms of severe respiratory distress may be subtle in very young babies, parents need to keep a high level of suspicion when young babies contract a respiratory illness, particularly young babies with a history of **prematurity** or other risk factor for severe RSV infection.

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Retention in school

Definition

The term "retention" in regards to school means repeating an academic year of school. Retention in school is also called grade retention, being held back, or repeating a grade. Grade retention is the opposite of social promotion, in which children continue with their age peers regardless of academic performance.

Description

According to the National Association of School Psychologists, in 2003 as many as 15 percent or more than 2.4 million American students are held back and repeat a grade each year. Other studies have found that between 30 percent and 50 percent of all students are retained at least once by the time they are freshmen in high school (about age 14). In most cases, teachers recommend retention for one of three reasons: developmental immaturity that has resulted in learning difficulties; emotional immaturity that has resulted in severely disruptive behavior; or failure to pass standardized proficiency or achievement tests at the end of specific years. Another less common reason for retention is poor attendance due either to **truancy** or medical absences. Grade retention has become increasingly controversial as early 2000s education initiatives such as No Child Left Behind have pressed schools to meet certain standards defined by scores on standardized tests.

Students at highest risk of being retained share certain characteristics:

- They tend to be boys.
- They tend to be African American or Hispanic.
- They are young or immature for their grade.
- They show developmental delays.
- They show attention, behavioral, or emotional problems.
- They are not proficient in English (English language learners).
- They have problems reading.
- They have changed schools often.
- They live in families with incomes below the poverty level.
- They live in single-parent families.
- They live with adults who are uninvolved in their education.

Preschool

Sometimes **preschool** teachers will recommend that a child attend an extra year of preschool before enrolling in kindergarten. This practice is more common in suburban school districts than in urban ones. The theory behind this practice is to allow children, especially those who would be young compared to their peers in kindergarten (birthdays falling near the cutoff date for school entry), to gain maturity and a greater likelihood of success in kindergarten. One 1984 study found that more than 11 percent of six year olds were enrolled in kindergarten or pre-first classes rather than in first grade.

In some athletically competitive families, children are held back and start school one year later because parents believe this will give them an edge in high school **sports** that require strength and size. Studies have found that as a group students who begin kindergarten a year late do no better or worse academically than their younger classmates.

Elementary school

Retention is most likely to be recommended by teachers in grades one through three. The most common reason for retention is poor reading skills. As a group, students who are retained in these grades show initial improvement in academics. However, this improvement disappears after two to three years, after which retained students do no better or even slightly worse than

similarly achieving students who were promoted. Studies also show that most elementary school teachers overestimate the academic benefits of retention. It has been suggested that this occurs because lower grade teachers see only the initial gains made by the student in the first few years after retention but do not follow the student's progress through middle and high school.

Retention in early elementary school does not appear to have an immediate effect on **self-esteem** or adjustment to school. However, by junior and senior high school, retained students tend to have more behavior problems, more difficulties with peer relationships, lower self-esteem, and poorer attendance.

Middle school

Retention can be emotionally traumatic for middle school students. A 1990 study found that being held back a grade was the third most stressful life event for sixth grade students topped only by the death of a parent or going blind. When this study was repeated in 2001, sixth grade students ranked flunking a grade as first in stress among these three events.

Middle school students who have been retained have more negative behaviors than their peers in academic ability who were not retained. These behaviors include **smoking** cigarettes, alcohol use, early sexual activity, and aggressive or violent behaviors. The retained group also had worse academic performance than similar students who were not retained.

In some school districts red shirting of student athletes is tacitly endorsed. This practice occurs when students are retained to improve performance in a nonacademic area, namely sports. Regardless of academic performance, a student is retained, usually in junior high school, to increase his or her likelihood of winning a college athletic scholarship. In addition, retention of strong athletes allows the school to build teams of older, bigger athletes. In these cases, retention is usually carried out with the knowledge and support of the student and his **family** and is not likely to carry a social stigma, as would be the case if the retention were for academic reasons.

High school

Grade retention is an excellent predictor of who will drop out of high school. Studies spanning several decades suggest that being retained one grade increases the risk of dropping out by 40 to 50 percent. Being retained twice or more almost guarantees the student will drop out. High school students who have been retained, even in earlier years, have the same unhealthful behaviors as retained middle-school students as well as more incidents of driving while using alcohol, marijuana use, suicidal behaviors, and high-risk sexual behavior. Individuals who have repeated a grade are more likely as adults to be unemployed, live on welfare, or be in prison than adults who did not repeat a grade.

Alternatives to retention

Given research finding that retention does not help learning difficulties, the question remains regarding what to do with a child who is, for whatever reason, unprepared to move to the next grade. Schools feel pressure to adhere to academic standards, while at the same time being fully aware of studies that show retention is counter-productive. However, the social promotion policies common in the 1970s, where students were kept with their age peers regardless of readiness for the next grade, does not produce academic success for at-risk students either.

Strong evidence indicates that at-risk students need remedial intervention, not simply more time or the repetition of material that retention provides. Potential remediations that can serve as alternatives to retention include:

- mixed-age classes where students advance at their own rate without grade-level labeling
- individual instruction and/or tutoring
- smaller classes for students who are struggling academically
- intensive early reading programs in lower grades for students who fail to achieve reading fluency
- early evaluation for learning disorders/deficits and emotional disorders followed by appropriate modifications in instruction
- extended day and summer school programs
- transfer to an alternative school
- programs to educate and involve parents in their child's academic program

Common problems

It is difficult to separate the effects of retention and the influence of other socioeconomic and family factors that affect children. Research suggests that social promotion and grade retention are not educationally effective policies. As of 2004, some people in the educational field believed that better educational gains may be made by linking the community organizations that deliver social services (health, mental health, family support services) with the school system in order to serve the child and family as a unit.

KEY TERMS

Social promotion—Passing a child on to the next grade regardless of readiness in order for the child to remain with his or her age peers.

Parental concerns

Research evaluates outcomes for groups, not individual students. Parents may have valid reasons for believing that their child may benefit or suffer from retention. Most experts support the idea that parents should be involved in the decision to promote or retain their child and should make their concerns known to the teacher and school. Parents need to understand their school district's policy on retention and request evidence supporting a retention decision, including details of their child's academic performance, standardized test results, or other pertinent factors, such as the student's emotional maturity and behavior in class. Parents also need to advocate for early evaluation of learning disabilities if their child is falling behind.

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National Association of School Psychologists. 4340 East West Highway, Suite 402, Bethesda, MD 20814. Web site: <www.nasponline.org>.

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Tish Davidson, A.M.

Retin-A *see* **Antiacne drugs**

Retinoblastoma

Definition

Retinoblastoma is a malignant tumor of the retina that occurs predominantly in young children.

Description

The eye has three layers, the sclera, the choroid, and the retina. The sclera is the outer protective white coating of the eye. The choroid is the middle layer and contains blood vessels that nourish the eye. The front portion of the choroid is colored and is called the iris. The opening in the iris is called the pupil. The pupil is responsible for allowing light into the eye and usually appears black. When the pupil is exposed to bright light it contracts (closes), and when it is exposed to low light conditions it dilates (opens) so that the appropriate amount of light enters the eye. Light that enters through the pupil hits the lens of the eye. The lens then focuses the light onto the retina, the innermost of the three layers. The job of the retina is to transform the light into information that can be transmitted to the optic nerve, which will transmit this information to the brain. It is through this process that people are able to see the world around them.

Occasionally a tumor, called a retinoblastoma, develops in the retina of the eye. Usually this tumor forms in young children, but it can occasionally occur in adults. Most people with retinoblastoma develop only one tumor (unifocal) in only one eye (unilateral). Some, however, develop multiple tumors (multifocal) in one or both eyes. When retinoblastoma occurs independently in both eyes, it is then called bilateral retinoblastoma.

Occasionally, children with retinoblastoma develop trilateral retinoblastoma, which results from the development of an independent brain tumor that forms in a part of the brain called the pineal gland. In order for retinoblastoma to be classified as trilateral retinoblastoma, the tumor must have developed independently and not as the result of the spread of the retinal **cancer**. The prognosis for trilateral retinoblastoma is quite poor.

The retinal tumor which characterizes retinoblastoma is malignant, meaning that it can metastasize (spread) to other parts of the eye and eventually other parts of the body. In most cases, however, retinoblastoma is diagnosed before it spreads past the eye to other parts of the body (intraocular) and the prognosis is quite good. The prognosis is poorer if the cancer has spread beyond the eye (extraocular).

Retinoblastoma can be inherited or can arise spontaneously. Approximately 40 percent of people with

retinoblastoma have an inherited form of the condition and approximately 60 percent have a sporadic (not inherited) form. Individuals with multiple independent tumors, bilateral retinoblastoma, or trilateral retinoblastoma are more likely to be affected with the inherited form of retinoblastoma.

Demographics

Approximately one in 15,000 to one in 30,000 infants are born with retinoblastoma, making it the most common childhood eye cancer. It is, however, a relatively rare childhood cancer and accounts for approximately 3 percent of childhood cancers. Retinoblastoma is found mainly in children under the age of five but can occasionally be seen in older children and adults. Retinoblastoma is found in individuals of all ethnic backgrounds and is found equally frequently in males and females.

Causes and symptoms

Causes

Retinoblastoma is caused by changes in or absence of a gene called RB1. RB1 is located on chromosome 13. Cells of the body, with the exception of the egg and sperm cells, contain 23 pairs of chromosomes. All of the cells of the body excluding the egg and the sperm cells are called the somatic cells. The somatic cells contain two of each chromosome 13 and, therefore, two copies of the RB1 gene. Each egg and sperm cell contains only one copy of chromosome and, therefore, only one copy of the RB1 gene.

RB1 produces a tumor suppressor protein that normally helps to regulate the cell cycle of cells such as those of the retina. A normal cell of the retina goes through a growth cycle during which it produces new cells. Genes such as tumor suppressor genes tightly regulate this growth cycle.

Cells that lose control of their cell cycle and replicate out of control are called cancer cells. These undergo many cell divisions, often at a quicker rate than normal cells and do not have a limited lifespan. A group of adjacent cancer cells can form a mass called a tumor. Malignant (cancerous) tumors can spread to other parts of the body. A malignant tumor of the retina (retinoblastoma) can result when just one retinal cell loses control of it cell cycle and replicates out of control.

Normally, the tumor suppressor protein produced by RB1 prevents a retinal cell from becoming cancerous. Each RB1 gene produces tumor suppressor protein. Only one functioning RB1 gene in a retinal cell is necessary to prevent the cell from becoming cancerous. If both RB1 genes in a retinal cell become non-functional, then a retinal cell can become cancerous and retinoblastoma can result. An RB1 gene is non-functional when it is changed or missing (deleted) and no longer produces normal tumor suppressor protein.

Approximately 40 percent of people with retinoblastoma have inherited a non-functional or deleted RB1 gene from either their mother or father. Therefore, they have a changed/deleted RB1 gene in every somatic cell. A person with an inherited missing or non-functional RB1 gene will develop a retinal tumor if the remaining RB1 gene becomes changed or deleted in a retinal cell. The remaining RB1 gene can become non-functional when exposed to environmental triggers such as chemicals and radiation. In most cases, however, the triggers are unknown. Approximately 90 percent of people who inherit a changed or missing RB1 gene develop retinoblastoma.

People with an inherited form of retinoblastoma are more likely to have a tumor in both eyes (bilateral) and are more likely to have more than one independent tumor (multifocal) in one or both eyes. The average age of onset for the inherited form of retinoblastoma is one year, which is earlier than the sporadic form of retinoblastoma. Although most people with the inherited form of retinoblastoma develop bilateral tumors, approximately 15 percent of people with a tumor in only one eye (unilateral) are affected with an inherited form of retinoblastoma.

A person with an inherited missing or non-functional RB1 gene has a 50 percent chance of passing on this abnormal gene to his or her offspring. The chance that the children will inherit the changed/deleted gene and actually develop retinoblastoma is approximately 45 percent.

Some people with retinoblastoma have inherited a non-functioning or missing RB1 gene from either their mother or father even though their parents have never developed retinoblastoma. It is possible that one parent has a changed or missing RB1 gene in every somatic cell but has not developed retinoblastoma because his or her remaining RB1 gene has remained functional. It is also possible that the parent had developed a retinal tumor that was destroyed by the body. In other cases, one parent has two normal RB1 genes in every somatic cell, but some egg or sperm cells contain a changed or missing RB1 gene. This is called gonadal mosaicism.

Retinoblastoma can also result when both RB1 genes become spontaneously changed or deleted in a retinal cell but the RB1 genes are normal in all the other

cells of the body. Approximately 60 percent of people with retinoblastoma have this type of disease, called sporadic retinoblastoma. A person with sporadic retinoblastoma does not have a higher chance of having children with the disease. His or her relatives do not have a higher risk of developing retinoblastoma or having children who develop retinoblastoma. Sporadic retinoblastoma is usually unifocal and has an average age of onset of approximately two years.

Symptoms

The most common symptom of retinoblastoma is leukocoria. Leukocoria results when the pupil reflects a white color rather than the normal black or red color that is seen on a flash photograph. It is often most obvious in flash photographs; since the pupil is exposed to a lot of light and the duration of the exposure is so short, the pupil does not have time to constrict. Children with retinoblastoma can also have problems seeing and this can cause them to appear cross-eyed (**strabismus**). People with retinoblastoma may also experience red, painful, and irritated eyes, inflamed tissue around the eye, enlarged pupils, and possibly different-colored eyes.

Diagnosis

Children who have symptoms of retinoblastoma are usually first evaluated by their pediatrician. The pediatrician will often perform a red reflex test to diagnose or confirm leukocoria. Prior to this test the doctor inserts medicated eye drops into the child's eyes so that the pupils remain dilated and not contract when exposed to bright light. The doctor then examines the eyes with an ophthalmoscope, which shines a bright light into the eyes and allows the doctor to check for leukocoria. Leukocoria can also be diagnosed by taking a flash Polaroid photograph of a patient who has been in a dark room for three to five minutes.

If the pediatrician suspects retinoblastoma on the basis of these evaluations, he or she will most likely refer the patient to an ophthalmologist (eye doctor) who has experience with retinoblastoma. The ophthalmologist will examine the eye using an indirect ophthalmoscope. The opthalmoscope shines a bright light into the eye, which helps the doctor to visualize the retina. This evaluation is usually done under general anesthetic, although some very young or older patients may not require it. Prior to the examination, medicated drops are put into the eyes to dilate the pupils, and anesthetic drops may also be used. A metal clip is used to keep the eyes open during the evaluation. During the examination, a cotton swab or a metal instrument with a flattened tip is used to press on the outer lens of the eye so that a better view of the front areas of the retina can be obtained. Sketches or photographs of the tumor as seen through the ophthalmoscope are taken during the procedure.

An ultrasound evaluation is used to confirm the presence of the tumor and to evaluate its size. Computed axial tomography (CT scan) is used to determine whether the tumor has spread outside of the eye and to the brain. Sometimes **magnetic resonance imaging** (MRI) is also used to look at the eyes, eye sockets, and the brain to see if the cancer has spread.

In most cases, the cancer has not spread beyond the eye, and other evaluations are unnecessary. If the cancer appears to have spread beyond the eye, then other assessments such as a blood test, spinal tap (lumbar puncture), and/or bone marrow biopsy may be recommended. During a spinal tap, a needle is inserted between the vertebrae of the spinal column and a small sample of the fluid surrounding the spinal cord is obtained. In a bone marrow biopsy, a small amount of tissue (bone marrow) is taken from inside the hip or breast bone for examination.

Genetic testing

Establishing whether someone is affected with an inherited or non-inherited form of retinoblastoma can help to determine whether other **family** members such as siblings, cousins, and offspring are at increased risk for developing retinoblastoma. It can also sometimes help guide treatment choices, since patients with an inherited form of retinoblastoma may be at increased risk for developing recurrent tumors or other types of cancers, particularly when treated with radiation. It is helpful for the families of a child diagnosed with retinoblastoma to meet with a genetic specialist such as a genetic counselor and/or geneticist. These specialists can help to ascertain the chances that the retinoblastoma is inherited and facilitate genetic testing if desired.

If a patient with unilateral or bilateral retinoblastoma has a relative or relatives with retinoblastoma, it can be assumed that they have an inherited form of retinoblastoma. However, it cannot be assumed that a patient without a family history of the disease has a sporadic form.

Even when there is no family history, most cases of bilateral and trilateral retinoblastoma are inherited, as are most cases of unilateral, multifocal retinoblastoma. However, only 15 percent of unilateral, unifocal retinoblastoma cases are inherited.

The only way to establish whether someone has an inherited form of retinoblastoma is to see if the retinoblastoma gene is changed or deleted in the blood cells obtained from a blood sample. Approximately 5 to 8 per-

cent of individuals with retinoblastoma possess a chromosomal abnormality involving the RB1 gene that can be detected by looking at their chromosomes under the microscope. The chromosomes can be seen by obtaining a blood sample. If this type of chromosomal abnormality is detected in a child, then analysis of the parents' chromosomes should be performed. If one of the parents possesses a chromosomal abnormality, then they are at higher risk for having other offspring with retinoblastoma. Chromosome testing would be recommended for the blood relatives of the parent with the abnormality.

Usually, however, a chromosomal abnormality is not detected in a child with retinoblastoma. In this case, specialized DNA tests that look for small RB1 gene changes need to be performed on the blood cells. DNA testing can be difficult, time consuming, and expensive, since there are many possible RB1 gene changes that can cause the gene to become nonfunctional.

If a sample of tumor is available, then it is recommended that DNA testing be performed on the tumor cells prior to DNA testing of the blood cells. This testing can usually identify the gene changes/deletions in the RB1 genes that caused the tumor to develop. In some cases, RB1 gene changes/deletions are not found in the tumor cells (approximately 20% of RB1 gene changes or deletions are not detectable). In these cases, DNA testing of the blood cells will not be able to ascertain whether someone is affected with an inherited or non-inherited form of retinoblastoma.

If the changes in both RB1 genes are detected in the tumor cell, then these same changes can be looked for in the blood cells. If an RB1 gene is deleted or changed in all of the blood cells tested, the patient can be assumed to have been born with a changed/deleted RB1 gene in all of his or her cells. This person has a 50 percent chance of passing the RB1 gene change/deletion on to his or her children. Most of the time, this change/deletion has been inherited from a parent. Occasionally the gene change/deletion occurred spontaneously in the original cell that was formed when the egg and sperm came together at conception (de novo).

If an RB1 gene change/deletion is found in all of the blood cells tested, both parents should undergo blood testing to check for the same RB1 gene change/deletion. If the RB1 gene change/deletion is identified in one of the parents, it can be assumed that the retinoblastoma was inherited and that siblings have a 50 percent chance of inheriting the altered gene. More distant blood relatives of the parent with the identified RB1 gene change/deletion may also be at risk for developing retinoblastoma. Siblings and other relatives could undergo DNA testing to see if they have inherited the RB1 gene change/deletion.

If the RB1 gene change/deletion is not identified in either parent, then the results can be more difficult to interpret. In this case, there is a 90 to 94 percent chance that the retinoblastoma was not inherited.

In some cases, a person with retinoblastoma will have an RB1 gene change/deletion detected in some of their blood cells and not others. It can be assumed that this person did not inherit the retinoblastoma from either parent. Siblings and other relatives would, therefore, not be at increased risk for developing retinoblastoma. Offspring would be at increased risk since some of the egg or sperm cells could have the changed/deleted RB1 gene. The risks to offspring would probably be less than 50 percent.

In families where there are multiple family members affected with retinoblastoma, blood samples from multiple family members are often analyzed and compared through DNA testing. Ninety-five percent of the time, this type of analysis is able to detect patterns in the DNA that are associated with a changed RB1 gene in that particular family. When a pattern is detected, at-risk relatives can be tested to establish whether they have inherited an RB1 gene change/deletion.

PRENATAL TESTING If chromosome or DNA testing identifies an RB1 gene/deletion in someone's blood cells, then prenatal testing can be performed on this person's offspring. An **amniocentesis** or chorionic villus sampling can be used to obtain fetal cells which can be analyzed for the RB1 gene change/deletion or chromosomal abnormality.

Treatment

A number of different classification (staging) systems are used to establish the severity of retinoblastoma and aid in choosing an appropriate treatment plan. The most widely used staging system is the Reese-Ellsworth system. This system is used to classify intraocular tumors and predict which tumors are favorable enough that sight can be maintained. The Reese-Ellsworth classification system is divided into several groups:

- Group I (very favorable for maintenance of sight): small solitary or multiple tumors, less than 6.4 mm in size (1 inch equals 25.4 mm), located at or below the equator of the eye.
- Group II (favorable for maintenance of sight): solitary or multiple tumors, 6.4 to 16 mm in size, located at or behind the equator of the eye.

- Group III (possible for maintenance of sight): any tumor located in front of the equator of the eye, or a solitary tumor larger than 16 mm in size and located behind the equator of the eye.
- Group IV (unfavorable for maintenance of sight): multiple tumors, some larger than 16 mm in size, or any tumor extending in front of the outer rim of the retina (ora serrata).
- Group V (very unfavorable for maintenance of sight): large tumors involving more than half of the retina, or vitreous seeding, in which small pieces of tumor are broken off and floating around the inside of the eye.

When choosing a treatment plan, the first criterion is to determine whether the cancer is localized within the eye (intralocular) or has spread to other parts of the body (extralocular). An intraocular retinoblastoma may only involve the retina or could involve other parts of the eye. An extraocular retinoblastoma could involve only the tissues around the eye or could result from the spread of cancer to the brain or other parts of the body.

It is also important to establish whether the cancer is unilateral (one eye) or bilateral (both eyes), multifocal or unifocal. In order for the tumors to be considered multifocal, they must have arisen independently and not as the result of the spread of cancer cells. It is also important to check for trilateral retinoblastoma.

The treatment chosen depends on the size and number of tumors, whether the cancer is unilateral or bilateral, and whether the cancer has spread to other parts of the body. The goal of treatment is to cure the cancer and prevent as much loss of vision as possible.

TREATMENT OF INTRAOCULAR TUMORS Surgical removal of the affected eye (enucleation) is performed when the tumor(s) are so large and extensive that preservation of sight is not possible. This surgery is performed under general anesthetic and usually takes less than an hour. Most children who have undergone this surgery can leave the hospital on the same day. A temporary ball is placed in the eye socket after the surgery. Approximately three weeks after the operation, a plastic artificial eye (prosthesis) that looks like the normal eye is inserted into the eye socket.

Radiation therapy is often used for treatment of large tumors when preservation of sight is possible. External beam radiation therapy involves focusing a beam of radiation on the eye. If the tumor has not spread extensively, the radiation beam can be focused on the cancerous retinal cells. If the cancer is extensive, radiation treatment of the entire eye may be necessary. External beam radiation is performed on an outpatient basis and usually occurs over a period of three to four weeks. Some children may need sedatives prior to the treatment. This type of therapy can result in a temporary loss of a patch of hair on the back of the head and a small area of "sun-burned" skin. Long-term side effects of radiation treatment can include cataracts, vision problems, bleeding from the retina, and decreased growth of the bones on the side of the head. People with an inherited form of retinoblastoma have an increased risk of developing other cancers as a result of this therapy. Some consideration should, therefore, be given to alternative treatment therapies for those with an inherited form of retinoblastoma.

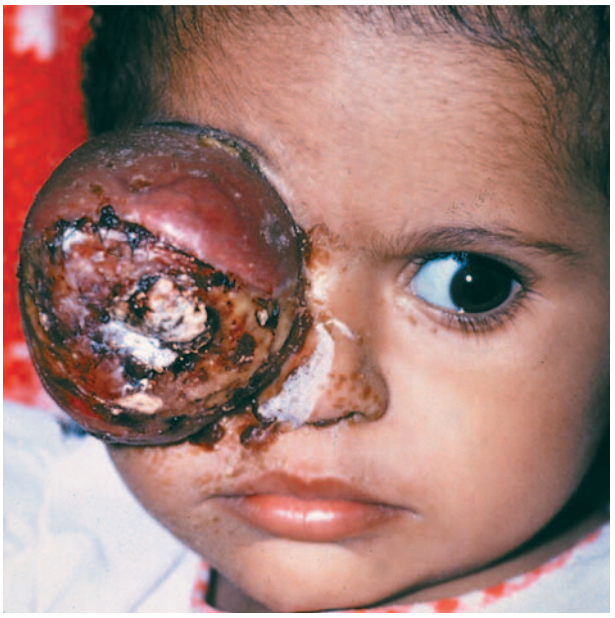
Photocoagulation therapy is often used in conjunction with radiation therapy but may be used alone to treat small tumors that are located on the back of the eye. Photocoagulation involves using a laser to destroy the cancer cells. This type of treatment is performed under local or general anesthesia and is usually not associated with post-procedural **pain**.

Thermotherapy is also often used in conjunction with radiation therapy or drug therapy (**chemotherapy**). Thermotherapy involves the use of heat to help shrink tumor cells. The heat is either used on the whole eye or localized to the tumor area. It is performed under local or general anesthesia and is usually not painful.

Cryotherapy is a treatment often used in conjunction with radiation therapy but can also be used alone on small tumors located on the front part of the retina. Cryotherapy involves the use of intense cold to destroy cancer cells and can result in harmless, temporary swelling of the external eye and eyelids that can last for up to five days. Eye drops or ointment are sometimes provided to reduce the swelling.

Brachytherapy involves the application of radioactive material to the outer surface of the eye at the base of the tumor. It is generally used for tumors of medium size. A patient undergoing this type of procedure is usually hospitalized for three to seven days. During that time, he or she undergoes one surgery to attach the radioactive material and one surgery to remove it. Eye drops are often administered for three to four weeks following the operation to prevent inflammation and infection. The long-term side effects of this treatment can include cataracts and damage to the retina, which can lead to impaired vision.

Intravenous treatment with one or more drugs (chemotherapy) is often used for treatment of both large and small tumors. Chemotherapy is sometimes used to shrink tumors prior to other treatments such as radiation therapy or brachytherapy. Occasionally, it is also used alone to treat very small tumors.



Child's right eye completely covered with a tumor associated with retinoblastoma. *(Custom Medical Stock Photo Inc.)*

TREATMENT OF INTRAOCULAR AND UNILATERAL RETINOBLASTOMA Often, by the time that unilateral retinoblastoma is diagnosed, the tumor is so large that useful vision cannot be preserved. In these cases removal of the eye (enucleation) is the treatment of choice. Other therapies are unnecessary if enucleation is used to treat intraocular unilateral retinoblastoma. If the tumor is small enough, other therapies such as external beam radiation therapy, photocoagulation, cryotherapy, thermotherapy, chemotherapy, and brachytherapy may be considered.

TREATMENT OF INTRAOCULAR AND BILATERAL RETINOBLASTOMA If vision can be preserved in both eyes, radiation therapy of both eyes may be recommended. Smaller, more localized tumors can sometimes be treated by local therapies such as cryotherapy, photocoagulation therapy, thermotherapy or brachytherapy. Some centers may use chemotherapy in place of radiation therapy when the tumors are too large to be treated by local therapies or are found over the optic nerve of the eye. As of the early 2000s, many centers are moving away from radiation treatment and toward chemotherapy because it is less likely to induce future tumors. Enucleation is performed on the more severely affected eye if sight cannot be preserved in both.

EXTRAOCULAR RETINOBLASTOMA There is no proven effective therapy for the treatment of extraocular retinoblastomas. Commonly, radiation treatment of the eyes and chemotherapy is provided.

Alternative treatment

As of 2004 there are no alternative or complementary therapies specific to the treatment of retinoblastoma. Since most people diagnosed with retinoblastoma are small children, most drug-based alternative therapies designed to treat general cancer would not be recommended. Many specialists would, however, stress the importance of establishing a well-balanced diet, including certain fruits, vegetables, and vitamin supplements, to ensure that the body is strengthened in its fight against cancer. Some advocate the use of visualization strategies, in which patients are encouraged to visualize the immune cells of their body attacking and destroying the cancer cells.

Prognosis

Individuals with intraocular retinoblastoma who do not have trilateral retinoblastoma usually have a good survival rate with a 90 percent chance of disease-free survival for five years. Those with extraocular retinoblastoma have less than a 10 percent chance of disease-free survival for the same amount of time. Trilateral retinoblastoma generally has a very poor prognosis. Patients with trilateral retinoblastoma who receive treatment have an average survival rate of approximately eight months, while those who remain untreated have an average survival rate of approximately one month. Patients with trilateral retinoblastoma who are asymptomatic at the time of diagnosis may have a better prognosis then those who experience symptoms.

Patients with an inherited form of unilateral retinoblastoma have a 70 percent chance of developing retinoblastoma in the other eye. Retinoblastoma reoccurs in the other eye in approximately 5 percent of people with a non-inherited form of retinoblastoma, so it is advisable for even these patients to be closely monitored. People with an inherited form of retinoblastoma who have not undergone radiation treatment have approximately a 26 percent chance of developing cancer in another part of the body within 50 years of the initial diagnosis. Those with an inherited form who have undergone radiation treatment have a 58 percent chance of developing a secondary cancer within 50 years after the initial diagnosis. Most of the secondary cancers are skin cancers, bone tumors (osteosarcomas), and soft-tissue **sarcomas**. Soft-tissue sarcomas are malignant tumors of the muscle, nerves, joints, blood vessels, deep skin tissues, or fat.

Prevention

Although retinoblastoma cannot be prevented, appropriate screening and surveillance should be applied to

KEY TERMS

Amniocentesis—A procedure performed at 16–18 weeks of pregnancy in which a needle is inserted through a woman's abdomen into her uterus to draw out a small sample of the amniotic fluid from around the baby for analysis. Either the fluid itself or cells from the fluid can be used for a variety of tests to obtain information about genetic disorders and other medical conditions in the fetus.

Benign tumor—An abnormal proliferation of cells that does not spread to other parts of the body.

Bilateral—Occurring on two sides. For example, a patient with bilateral retinoblastoma has this retinal tumor in both eyes.

Brachytherapy—A method of treating cancers, such as prostate cancer, involving the implantation near the tumor of radioactive seeds.

Chorionic villus sampling—A procedure used for prenatal diagnosis at 10–12 weeks gestation. Under ultrasound guidance a needle is inserted either through the mother's vagina or abdominal wall and a sample of the chorionic membrane. These cells are then tested for chromosome abnormalities or other genetic diseases.

Chromosome—A microscopic thread-like structure found within each cell of the human body and consisting of a complex of proteins and DNA. Humans have 46 chromosomes arranged into 23 pairs. Chromosomes contain the genetic information necessary to direct the development and functioning of all cells and systems in the body. They pass on hereditary traits from parents to child (like eye color) and determine whether the child will be male or female.

Cryotherapy—The use of a very low-temperature probe to freeze and thereby destroy tissue. Cryotherapy is used in the treatment skin lesions, Parkinson's disease, some cancers, retinal detachment, and cataracts. Also called cryosurgery.

DNA—Deoxyribonucleic acid; the genetic material in cells that holds the inherited instructions for growth, development, and cellular functioning.

DNA testing—Analysis of DNA (the genetic component of cells) in order to determine changes in genes that may indicate a specific disorder.

Enucleation—Surgical removal of the eyeball.

Equator—Imaginary line encircling the eyeball and dividing the eye into a front and back half.

Extraocular retinoblastoma—Cancer that has spread from the eye to other parts of the body.

Gene—A building block of inheritance, which contains the instructions for the production of a particular protein, and is made up of a molecular sequence found on a section of DNA. Each gene is found on a precise location on a chromosome.

Intraocular retinoblastoma—Cancer of the retina that is limited to the eye and has not spread to other parts of the body.

Malignant tumor—An abnormal proliferation of cells that can spread to other sites.

Multifocal—Having many focal points. When referring to a disease, it means that damage caused by the disease occurs at multiple sites. When referring to a cancer, it means that more than one tumor is present.

Oncologist—A physician specializing in the diagnosis and treatment of cancer

Ophthalmologist—A physician who specializes in the anatomy and physiology of the eyes and in the diagnosis and treatment of eye diseases and disorders.

Optic nerve—A bundle of nerve fibers that carries visual messages from the retina in the form of electrical signals to the brain.

Photocoagulation—A type of cancer treatment in which cancer cells are destroyed by an intense beam of laser light.

Prenatal testing—Testing for a disease, such as a genetic condition, in an unborn baby.

Protein—An important building blocks of the body, a protein is a large, complex organic molecule composed of amino acids. It is involved in the formation of body structures and in controlling the basic functions of the human body.

Retina—The inner, light-sensitive layer of the eye containing rods and cones. The retina transforms the image it receives into electrical signals that are sent to the brain via the optic nerve.

Somatic cells—All the cells of the body with the exception of the egg and sperm cells.

Tumor—A growth of tissue resulting from the uncontrolled proliferation of cells.

Tumor-suppressor gene—A gene involved in controlling normal cell growth and preventing cancer.

Unifocal—Only one tumor present in one eye.

Unilateral—Refers to one side of the body or only one organ in a pair.

Vitreous—The transparent gel that fills the back part of the eye.

Vitreous seeding—Small pieces of tumor have broken off and are floating around the vitreous.

all at-risk individuals to ensure that the tumor(s) are diagnosed at an early stage. The earlier the diagnosis, the more likely that an eye can be salvaged and vision maintained.

Screening of people diagnosed with retinoblastoma

Children who have been diagnosed with retinoblastoma should receive periodic dilated retinal examinations until the age of five. Young children will need to undergo these evaluations under anesthetic. After five years of age, periodic eye examinations are recommended. It may be advisable for patients with bilateral retinoblastoma or an inherited form of retinoblastoma to undergo periodic screening for the brain tumors found in trilateral retinoblastoma. There are no specific screening protocols designed to detect non-ocular tumors. All lumps and complaints of bone pain, however, should be thoroughly evaluated.

Screening of relatives

When a child is diagnosed with retinoblastoma, it is recommended that parents and siblings receive a dilated retinal examination by an ophthalmologist who is experienced in the diagnosis and treatment of the disease. It is also recommended that siblings continue to undergo periodic retinal examinations under anesthetic until they are three years of age. For children three to seven years of age, periodic eye examinations are recommended. The retinal examinations can be avoided if DNA testing indicates that the patient has a non-inherited form of retinoblastoma or if the sibling has not inherited the RB1 gene change/deletion. Any relatives who are found through DNA testing to have inherited an RB1 gene change/deletion should undergo the same surveillance procedures as siblings.

The children of someone diagnosed with retinoblastoma should also undergo periodic retinal examinations under anesthetic. Retinal surveillance should be performed unless DNA testing proves that their child does not possess the RB1 gene change/deletion. If desired, prenatal detection of tumors using ultrasound may also be performed. During the ultrasound procedure, a handheld instrument is placed on the maternal abdomen or inserted vaginally. The ultrasound produces sound waves that are reflected back from the body structures of the fetus, producing a picture that can be seen on a video screen. If a tumor is detected through this evaluation, the affected baby may be delivered a couple of weeks earlier. This can allow for earlier intervention and treatment.

Parental concerns

Careful attention to a child's diet can be very helpful for patients with cancer. This can be difficult when the cancer and/or the treatments are affecting the appetite, however. Whole foods, including grains, beans, fresh fruits and vegetables, and high quality fats, should be emphasized in the diet, while processed foods should be avoided. Increased consumption of fish, especially cold-water fish like salmon, mackerel, halibut, and tuna, provides a good source of omega-3 fatty acids. Nutritional supplements can build strength and help maintain it during and following chemotherapy, radiation, or surgery.

Guided imagery and relaxation techniques can be helpful for children undergoing difficult treatments. Support groups for the child and the family can be very helpful and can provide an important emotional outlet for the child, the parents, and the siblings.

Resources

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ORGANIZATIONS

Institute for Families with Blind Children. PO Box 54700, Mail Stop 111, Los Angeles, CA 90054–0700. Web site: <www.instituteforfamilies.org>.

Retinoblastoma International. 4650 Sunset Blvd., Mail Stop #88, Los Angeles, CA 90027. Web site: <www.retinoblastoma.net>.

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Reye's syndrome

Definition

Reye's syndrome is a disorder principally affecting the liver and brain, marked by rapid development of life-threatening neurological symptoms.

Description

Reye's syndrome is an emergency illness chiefly affecting children and teenagers. It almost always follows a viral illness such as a cold, the flu, or **chickenpox**. Reye's syndrome may affect all the organs of the body, but it most seriously affects the brain and liver. Brain swelling and massive accumulations of fat in the liver and other organs lead to the rapid development of severe neurological symptoms, including lethargy, confusion, seizures, and coma. Reye's syndrome is a life-threatening emergency, with a fatality rate of about 30 to 50 percent.

Demographics

Reye's syndrome is a rare illness, even rarer in the early 2000s than when it was first described in the early 1970s. The incidence of the disorder peaked in 1980, with 555 cases reported. The number of cases declined rapidly thereafter once researchers made the association between aspirin use and the development of Reye's syndrome. Cases dropped precipitously when parents and healthcare providers were clearly warned against using any aspirin-containing products in children. As of 2004, fewer than 20 cases of Reye's syndrome are reported annually. Because of the rarity of Reye's syndrome, it is often misdiagnosed as **encephalitis**, **meningitis**, diabetes, or **poisoning**, and the true incidence may be higher than the number of reported cases indicates.

Causes and symptoms

Reye's syndrome causes fatty accumulation in the organs of the body, especially the liver. In the brain, it causes fluid accumulation (edema), which leads to a rise in pressure in the brain (intracranial pressure). This pressure compresses blood vessels, preventing blood from entering the brain. Untreated, this pressure increase leads to brain damage and death.

Although as of 2004 the cause remains unknown, Reye's syndrome appears to be linked to an abnormality in the energy-converting structures (mitochondria) within the body's cells.

Reye's syndrome usually occurs after a viral illness with **fever**, most often an upper respiratory tract infection. It is most often associated with use of aspirin during the fever, and for this reason aspirin and aspirin-containing products are not recommended for people under the age of 19 during fever. Although rare, Reye's syndrome may occur without aspirin use and in adults.

After the beginning of recovery from the viral illness, the affected person suddenly becomes more ill again, with the development of persistent **vomiting**. This may be followed rapidly by quietness, lethargy, agitation or combativeness, seizures, and coma. In infants, **diarrhea** may be more common than vomiting. Fever is usually absent at this point.

Diagnosis

Reye's syndrome may be suspected in a child who begins vomiting three to six days after a viral illness, followed by an alteration in consciousness. Diagnosis involves blood tests to determine the levels of certain liver enzymes, which are highly elevated in Reye's syndrome. Other blood changes may occur as well, including an increase in the level of ammonia and amino acids, a drop in blood sugar, and an increase in clotting time. A liver biopsy may also be done after clotting abnormalities are corrected with vitamin K or blood products. A lumbar puncture (spinal tap) may be needed to rule out other possible causes, including meningitis or encephalitis.

Treatment

Reye's syndrome is a life-threatening emergency that requires intensive management. The likelihood of recovery is greatest if it is recognized early and treated promptly. Children with Reye's syndrome should be managed in an intensive-care unit.

Treatment in the early stages includes intravenous sugar to return blood sugar levels to normal and plasma transfusion to restore normal clotting time in the blood. Intracranial pressure is monitored and, if elevated, is treated with drugs such as mannitol and barbiturates placing the patient on a ventilator so that hyperventilation can be used.

Prognosis

The mortality rate for Reye's syndrome is between 30 and 50 percent. The likelihood of recovery is increased to 90 percent by early diagnosis and treatment. Almost all children who survive Reye's syndrome

KEY TERMS

Acetylsalicylic acid—Aspirin; an analgesic, antipyretic, and antirheumatic drug prescribed to reduce fever and to relieve pain and inflammation.

Edema—The presence of abnormally large amounts of fluid in the intercellular tissue spaces of the body.

Mitochondria—Spherical or rod-shaped structures of the cell. Mitochondria contain genetic material (DNA and RNA) and are responsible for converting food to energy.

recover fully, although recovery may be slow. In some patients, permanent neurologic damage may remain, requiring physical or educational special services and equipment.

Prevention

Because Reye's syndrome is so highly correlated with use of aspirin for fever in young people, avoidance of aspirin use by children is strongly recommended. Aspirin is in many over-the-counter and prescription drugs, including drugs for **headache**, fever, menstrual cramps, muscle **pain**, **nausea**, upset stomach, and arthritis. It may be used in drugs taken orally or by suppository.

Any of the following ingredients indicates that aspirin is present:

- aspirin
- acetylsalicylate
- acetylsalicylic acid
- salicylic acid
- salicylate

Teenagers who take their own medications without parental consultation should be warned not to take aspirin-containing drugs.

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ORGANIZATIONS

National Reye's Syndrome Foundation. PO Box 829, Bryan, OH 43506-0829. Web site: <www.reyessyndrome.org>.

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Rh disease *see* **Erythroblastosis fetalis**

Rheumatic fever

Definition

Rheumatic fever (RF) is an illness that arises as a complication of untreated or inadequately treated **strep throat** infection. Rheumatic fever can seriously damage the valves of the heart.

Description

Throat infection with a member of the Group A streptococcus (strep) bacteria is a common problem among school-aged children. It is easily treated with a 10-day course of **antibiotics** by mouth. However, when such a throat infection occurs without symptoms, or when a course of medication is not taken for the full ten days, there is a 3 percent chance the person will develop rheumatic fever. Other types of strep infections (such as of the skin) do not put the patient at risk for RF.

Demographics

Children between the ages of five and 15 are most susceptible to strep throat, and therefore most susceptible to rheumatic fever. Other risk factors include poverty, overcrowding (as in military camps), and lack of access to good medical care. Just as strep throat occurs most frequently in fall, winter, and early spring, so does rheumatic fever. Rheumatic fever used to be a leading cause of death and disability in children. Since 1960, it has become much less common in the United States, partially because of increasingly accurate and swift diagnosis of strep throat. It is still a large problem in many developing countries. Moreover, children who have **family** members who have had rheumatic fever are more likely to get rheumatic fever themselves.

Causes and symptoms

Two different theories exist about how a bacterial throat infection can develop into rheumatic fever. One theory suggests that the bacteria produce some kind of poisonous chemical (toxin). This toxin is sent into circulation throughout the bloodstream, thus affecting other systems of the body.

Research seems to point to a different theory, however. The second theory suggests that the disease is caused by the body's immune system acting inappropriately. The body produces immune cells (called antibodies), that are specifically designed to recognize and destroy invading agent—in this case, streptococcal bacteria. The antibodies are able to recognize the bacteria because the bacteria contain special markers called antigens. Due to a resemblance between Group A streptococcus bacteria's antigens and antigens present on the body's own cells, the antibodies may mistakenly attack the body itself.

It is interesting to note that members of certain families seem to have a greater tendency to develop rheumatic fever than do others. This statistical fact could be related to the above theory, in that these families may have cell antigens that more closely resemble streptococcal antigens than do members of other families.

In addition to fever, in about 75 percent of all cases of RF one of the first symptoms is arthritis. The joints (especially those of the ankles, knees, elbows, and wrists) become red, hot, swollen, shiny, and extraordinarily painful. Unlike many other forms of arthritis, this arthritis may not occur symmetrically (affecting a particular joint on both the right and left sides, simultaneously). The arthritis of RF rarely strikes the fingers, toes, or spine. The joints become so tender that even the touch of bed sheets or clothing is terribly painful.

A particular type of involuntary movement, coupled with emotional instability, occurs in about 10 percent of all RF patients. The patient begins experiencing a change in coordination, often first noted by changes in handwriting. The arms or legs may flail or jerk uncontrollably. The patient seems to develop a low threshold for anger and sadness. This feature of RF is called Sydenham's chorea or St. Vitus' dance.

A number of skin changes are common to RF. A rash called erythema marginatum often develops (especially in those patients who will develop heart problems from their illness), composed of pink splotches that may eventually spread into each other. The rash does not itch. Bumps the size of peas may occur under the skin. These are called subcutaneous nodules. They are hard to the touch, but not painful. These nodules most commonly occur over the knee and elbow joint, as well as over the spine.

The most serious problem occurring in RF is called pancarditis ("pan" means total; "carditis" refers to inflammation of the heart). Pancarditis is an inflammation that affects all aspects of the heart, including the lining of the heart (endocardium), the sac containing the heart (pericardium), and the heart muscle itself (myocardium). About 40 to 80 percent of all RF patients develop pancarditis. This RF complication has the most serious, long-term effects. The valves within the heart (structures that allow the blood to flow only in the correct direction and only at the correct time in the heart's pumping cycle) are frequently damaged during the course of pancarditis. This effect may result in blood that either leaks back in the wrong direction or has a difficult time passing a stiff, poorly moving valve. Either way, damage to a valve can result in the heart having to work very hard in order to move the blood properly. The heart may not be able to "work around" the damaged valve, which may result in a consistently inadequate amount of blood entering the circulation.

When to call the doctor

The doctor should be contacted if the child is displaying any of the signs or symptoms of rheumatic fever. If they are not indications of rheumatic fever, they could be indicative of another disease or disorder. The doctor should also be contacted if the child has had a **sore throat** and fever for more than 24 hours. The doctor will do a strep test, and if the child does have strep throat the doctor can administer antibiotics that will help prevent rheumatic fever.

Diagnosis

There are no laboratory tests that can determine with complete certainty if a child has rheumatic fever. Some laboratory tests may be used in conjunction with careful examination of the patient to determine if the child has RF. A list of diagnostic criteria has been created. These "Jones Criteria" are divided into major and minor criteria. A patient can be diagnosed with RF if he or she has either two major criteria (conditions) or one major and two minor criteria. In either case, it must also be proved that the individual has had a previous infection with streptococcus.

The major criteria include:

- carditis
- arthritis
- chorea
- subcutaneous nodules
- erythema marginatum

The minor criteria include:

- fever
- joint **pain** (without actual arthritis)
- evidence of electrical changes in the heart (determined by measuring electrical characteristics of the heart's functioning during a test called an electrocardiogram, or EKG)
- evidence (through a blood test) of the presence in the blood of certain proteins that are produced early in an inflammatory/infectious disease

Tests are also performed to provide evidence of recent infection with group A streptococcal bacteria. A swab of the throat can be taken and smeared on a gel-like substance in a petri dish to see if bacteria will multiply and grow over 24 to 72 hours. These bacteria can then be specially processed and examined under a microscope to identify streptococcal bacteria. Other tests can be performed to see if the patient is producing specific antibodies that are only made in response to a recent strep infection.

Treatment

A 10-day course of penicillin by mouth or a single injection of penicillin G is usually the first line of treatment for RF. If the child does not tolerate or is allergic to penicillin, other antibiotics can be used effectively. These antibiotics are given to help cure a strep infection, if the child still has one. Patients will need to remain on some regular dose of antibiotic to prevent recurrence of RF. This can mean a small daily dose of antibiotic by mouth or an injection every three to four weeks. Some practitioners keep patients on this regimen for five years or until they reach 18 years of age whichever comes first. Other practitioners prefer to continue treating those patients who will be regularly exposed to streptococcal bacteria (teachers, medical workers), as well as those patients with known RF heart disease.

Arthritis quickly improves when the patient is given a preparation containing aspirin or some other anti-inflammatory agent (e.g. ibuprofen). Mild carditis also improves with such anti-inflammatory agents, although more severe cases of carditis require steroid medications. A number of medications are available to treat the involuntary movements of chorea, including diazepam for mild cases and haloperidol for more severe cases.

Prognosis

The long-term prognosis of an RF patient depends primarily on whether he or she develops carditis. This manifestation of RF is the only one that can have permanent effects. Those patients with no or mild carditis have an excellent prognosis. Those with more severe carditis have a risk of heart failure, as well as a risk of future heart problems that may lead to the need for valve replacement surgery. Patients who have had rheumatic fever are at an increased risk of getting it again.

KEY TERMS

Antibody—A special protein made by the body's immune system as a defense against foreign material (bacteria, viruses, etc.) that enters the body. It is uniquely designed to attack and neutralize the specific antigen that triggered the immune response.

Antigen—A substance (usually a protein) identified as foreign by the body's immune system, triggering the release of antibodies as part of the body's immune response.

Arthritis—A painful condition that involves inflammation of one or more joints.

Autoimmune disorder—One of a group of disorders, like rheumatoid arthritis and systemic lupus erythematosus, in which the immune system is overactive and has lost the ability to distinguish between self and non-self. The body's immune cells turn on the body, attacking various tissues and organs.

Chorea—Involuntary movements in which the arms or legs may jerk or flail uncontrollably.

Immune system—The system of specialized organs, lymph nodes, and blood cells throughout the body that work together to defend the body against foreign invaders (bacteria, viruses, fungi, etc.).

Inflammation—Pain, redness, swelling, and heat that develop in response to tissue irritation or injury. It usually is caused by the immune system's response to the body's contact with a foreign substance, such as an allergen or pathogen.

Pancarditis—Inflammation of the lining of the heart, the sac around the heart, and the muscle of the heart.

Prevention

Prevention of the development of RF involves proper diagnosis of initial strep throat infections and adequate treatment within 10 days with an appropriate anti-

biotic. Prevention of RF recurrence requires continued antibiotic treatment, perhaps for life. Prevention of complications of already-existing RF heart disease requires that the patient always take a special course of antibiotics when he or she undergoes any kind of procedure (even dental cleanings) that might allow bacteria to gain access to the bloodstream.

Parental concerns

Rheumatic fever can be life-threatening if not treated. It can also lead to lifelong heart problems. The best way for parents to prevent rheumatic fever is to take seriously sore throats that are accompanied with fever and to take the child to a doctor to test for strep throat. Children who have had rheumatic fever need to take extra precautions to ensure they do not have repeat attacks triggered by strep infections.

See also Strep throat.

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ORGANIZATIONS

American Heart Association. 7272 Greenville Ave., Dallas, TX 75231. Web site: <www.americanheart.org>.

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Rhinitis

Definition

Rhinitis is inflammation of the mucous lining of the nose.

Description

Rhinitis is a nonspecific term that covers infections, **allergies**, and other disorders whose common feature is the location of their symptoms. In rhinitis, the mucous membranes become infected or irritated, producing a discharge, congestion, and swelling of the tissues. The most widespread form of infectious rhinitis is the **common cold**. Doctors sometimes designate two different forms of rhinitis. These are **allergic rhinitis** and nonallergic rhinitis. Allergic rhinitis is cause by allergies, and nonallergic rhinitis is caused by other conditions such as the common cold.

Transmission

Nonallergic rhinitis is generally transmitted in the same ways as the common cold. It is transmitted from person to person. The sick person touches his or her nose and then another person's hands. If that person then touches his nose, mouth, or eyes, the infection is transmitted. Infection can also be transmitted through sharing of cups, silverware, or eating utensils, or by coughing or sneezing. Allergic rhinitis cannot be transmitted from person to person.

Demographics

The most frequent cause of nonallergic rhinitis is the common cold. The common cold is the most frequent viral infection in the general population, causing more absenteeism from school or work than any other illness. Colds are self-limited, lasting about three to 10 days, although they are sometimes followed by a bacterial infection. Children are more susceptible than adults; teenage boys more susceptible than teenage girls; and adult women more susceptible than adult men. In the United States, colds are most frequent during the late fall and winter. Allergic rhinitis is less common that nonallergic rhinitis. Allergic rhinitis affects between 20 and 40 million people in the United States. Children are more at risk for allergic rhinitis if one or both parents has allergies.

Causes and symptoms

The onset of a cold is usually sudden. The virus causes the lining of the nose to become inflamed and produce large quantities of thin, watery mucus. Children sometimes develop a **fever** with a cold. The inflammation spreads from the nasal passages to the throat and upper airway, producing a dry **cough**, **headache**, and watery eyes. Some people develop muscle or joint aches and feel generally tired or weak. After several days, the nose becomes less inflamed and a thick, sticky mucus

replaces the watery discharge. This change in the appearance of the nasal discharge helps to distinguish rhinitis caused by a viral infection from rhinitis caused by an allergy.

Allergic rhinitis is caused by allergens such as pollen, animal dander, dust mites, or grass. The symptoms of allergic rhinitis are similar to those of nonallergic rhinitis, except that they are usually much longer lasting and are rarely accompanied by a fever. These symptoms often occur at specific times of year if they are not constant.

When to call the doctor

If the symptoms of rhinitis persist for more than a week, or it they frequently occur in specific situations or during specific times of year, a doctor should be consulted. The doctor can then do tests to determine if the rhinitis is viral, bacterial, or caused by allergies and treat it accordingly.

Diagnosis

There is no specific test for viral rhinitis. The diagnosis is based on the symptoms. In children, the doctor will examine the child's throat and glands to rule out other childhood illnesses that have similar early symptoms. If the symptoms last for more than a week, the child may be tested further to rule out bacterial infections or allergies. Allergies can be evaluated by blood tests, skin testing for specific substances, or nasal smears.

Treatment

There is no cure for viral nonallergic rhinitis; treatment is given for symptom relief. Medications include aspirin or **nonsteroidal anti-inflammatory drugs** (NSAIDs) for headache and muscle **pain**, and **decongestants** to relieve stuffiness or runny nose. Patients should be warned against overusing decongestants, because they can cause a rebound effect. Over-the-counter (OTC) **antihistamines** are also available; however, most antihistamines carry warnings of drowsiness and the inability to do some tasks while medicated. Claritin is a prescription-strength OTC non-drowsy antihistamine that helps relieve symptoms of rhinitis. **Antibiotics** are not given for viral nonallergic rhinitis because they do not kill viruses. Supportive care includes bed rest and drinking plenty of fluid. Treatments under investigation, as of 2004, included the use of ultraviolet light and injections of interferon. If the nonallergic rhinitis has a bacterial cause at its root, antibiotics can be given; however, bacterial causes of rhinitis are not very common.

Allergic rhinitis is treated in a number of ways, including seasonal allergy medication, nasal sprays, and decongestants. If the cause of the allergies is determined to be an indoor allergen such as dust mites or pet dander, steps can be taken to rid the home of some of the allergens. Injections, or **allergy shots**, are also sometimes used to treat allergic rhinitis. A small amount of the allergen is injected at first, with tolerance built up over weeks or months. The shots are given frequently at first, but when a maintenance level of the allergen is reached, they are given less frequently. After a few years, they are no longer given at all. These shots have been found to be very effective in some cases, but there are often problems with compliance. Children may also find the experience of regular injections over such a long period to be distressing.

Alternative treatment

Homeopaths might prescribe any of 10 different remedies, depending on the appearance of the nasal discharge, the patients emotional state, and the stage of infection. Naturopaths may recommend vitamin A and zinc supplements, together with botanical preparations made from echinacea (Echinacea spp.), goldenseal (*Hydrastis canadensis*), licorice (*Glycyrrhiza glabra*), or astragalus (*Astragalus membraneceus*) root.

Prognosis

Most rhinitis caused by a cold resolves completely in about a week. Complications are unusual but may include **sinusitis** (inflammation of the nasal sinuses), bacterial infections, or infections of the middle ear. Allergic rhinitis can usually be treated very effectively. Bacterial causes of rhinitis can usually be resolved fairly quickly with the use of antibiotics.

Prevention

There is no known way to successfully prevent allergic rhinitis. The only way to prevent viral and bacterial nonallergic rhinitis is to take the steps which prevent transmission of the common cold. These include:

- washing hands often, especially before touching the face
- minimizing contact with people already infected
- not sharing hand towels, eating utensils, or water glasses

KEY TERMS

Allergen—A foreign substance that provokes an immune reaction or allergic response in some sensitive people but not in most others.

Interferon—A potent immune-defense protein produced by virus-infected cells; used as an anti-cancer and anti-viral drug.

Parental concerns

Rhinitis causes symptoms such as runny nose, **itching**, and sneezing that may be uncomfortable for the child. Nonallergic rhinitis is not thought to have any significant long-term consequences. Children who have allergic rhinitis may be at increased risk for developing **asthma**.

See also Allergic rhinitis; Allergies; Common cold.

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ORGANIZATIONS

American Academy of Allergy, Asthma, Immunology. 555 East Wells Street, Suite 1100, Milwaukee, WI 53202-3823. Web site: <www.aaaai.org>.

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Rickets *see* **Vitamin D deficiency**

Rickettsia infection *see* **Rocky Mountain spotted fever**

Ringworm

Definition

Ringworm is a common fungal infection of the skin. The name is a misnomer because the disease is not caused by a worm.

Description

Ringworm is characterized by patches of rough, reddened skin. Raised eruptions usually form the circular pattern that gives the condition its name. As lesions grow, the centers start to heal. The inflamed borders expand and spread the infection. Ringworm may also be referred to as dermatophyte infection. It is more common in males than females, and is most common among children ages three to nine years.

Types of ringworm

Ringworm is a term that is commonly used to encompass several types of fungal infection. Sometimes, however, only body ringworm is classified as true ringworm.

Body ringworm (tinea corporis) can affect any part of the body except the scalp, feet, and facial area where a man's beard grows. The well-defined, flaky sores can be dry and scaly or moist and crusty.

Scalp ringworm (tinea capitis) is most common in children. It causes scaly, swollen blisters or a rash that looks like black dots. Sometimes inflamed and filled with pus, scalp ringworm lesions can cause crusting, flaking, and round bald patches. Most common in black children, scalp ringworm can cause scarring and permanent hair loss.

Ringworm of the groin (tinea cruris or jock itch) produces raised red sores with well-marked edges. It can spread to the buttocks, inner thighs, and external genitals.

Ringworm of the nails (tinea unguium) generally starts at the tip of one or more toenails, which gradually thicken and discolor. The nail may deteriorate or pull away from the nail bed. Fingernail infection is far less common.

Demographics

Ringworm can affect people at any age. It is more common among children, athletes, and people with poor hygiene habits.

Causes and symptoms

Ringworm can be transmitted by infected people or pets or by towels, hairbrushes, or other objects contaminated by them. Symptoms include inflammation, scaling, and sometimes, **itching**.

Diabetes mellitus increases susceptibility to ringworm. Dampness, humidity, and dirty, crowded living areas also increase susceptibility. Braiding hair tightly and using hair gel also raise the risk.

When to call the doctor

A health professional should be consulted when signs of ringworm appear or if exposure to someone with ringworm is suspected.

Diagnosis

Diagnosis is based on microscopic examination of scrapings taken from lesions. A dermatologist may also study the scalp of a person with suspected tinea capitis under ultraviolet light.

Treatment

Some infections disappear without treatment. Others respond to such topical antifungal medications as naftifine (Caldesene Medicated Powder) or tinactin (Desenex) or to griseofulvin (Fulvicin), which is taken by mouth. Medications should be continued for two weeks after lesions disappear.

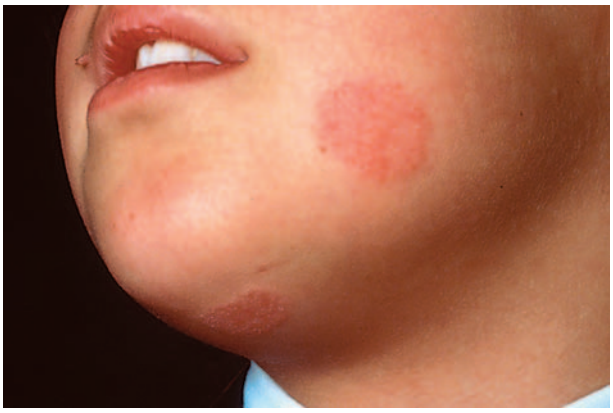
A person with body ringworm should wear loose clothing and check daily for raw, open sores. Wet dressings applied to moist sores two or three times a day can lessen inflammation and loosen scales. The doctor may suggest placing special pads between folds of infected skin, and anything the person has touched or worn should be sterilized in boiling water.

Infected nails should be cut short and straight and carefully cleared of dead cells with an emery board.

People with jock itch should:

- wear cotton underwear and change it more than once a day
- keep the infected area dry
- apply antifungal ointment over a thin film of antifungal powder

Shampoo containing selenium sulfide can help prevent spread of scalp ringworm, but prescription shampoo or oral medication is usually needed to cure the infection.



Child with a patch of rough, reddened skin in a circular pattern on his jaw caused by ringworm. *(© NMSB/Custom Medical Stock Photo, Inc.)*

Alternative treatment

The fungal infection ringworm can be treated with homeopathic remedies. Among the homeopathic remedies recommended are:

- sepia for brown, scaly patches
- tellurium for prominent, well-defined, reddish sores
- graphites for thick scales or heavy discharge
- sulfur for excessive itching

Topical applications of antifungal herbs and essential oils also can help resolve ringworm. Tea tree oil (*Melaleuca* spp.), thuja (*Thuja occidentalis*), and lavender (*Lavandula officinalis*) are the most common. Two drops of essential oil in 0.25 oz (7 ml) of carrier oil is the dose recommended for topical application. Essential oils should not be applied to the skin undiluted. Botanical medicine can be taken internally to enhance the body's immune response. A person must be susceptible to exhibit this overgrowth of fungus on the skin. Echinacea (*Echinacea* spp.) and astragalus (*Astragalus membranaceus*) are the two most common immune-enhancing herbs. A well-balanced diet, including protein, complex carbohydrates, fresh fruits and vegetables, and good quality fats, is also important in maintaining optimal immune function.

Prognosis

Ringworm can usually be cured, but recurrence is common. Chronic infection develops in one person in five.

It can take six to 12 months for new hair to cover bald patches, and three to 12 months to cure infected fin-

KEY TERMS

Dermatophyte—A type of fungus that causes diseases of the skin, including tinea or ringworm.

Diabetes mellitus—The clinical name for common diabetes. It is a chronic disease characterized by the inability of the body to produce or respond properly to insulin, a hormone required by the body to convert glucose to energy.

gernails. Toenail infections do not always respond to treatment.

Prevention

Likelihood of infection can be lessened by avoiding contact with infected people or pets or contaminated objects and staying away from hot, damp places.

Parental concerns

Parents should monitor the children with whom their own children interact or **play**. Children should not be allowed to play with other children who have open skin sores or scratch excessively.

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American Academy of Dermatology, 930 N. Meacham Road, PO Box 4014, Schaumburg, IL 60168-4014. (847) 330-0230. Fax: (847) 330-0050. Web site: <www.aad.org>.

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Ritalin *see* **Methlphenidate**

Rocky Mountain spotted fever

Definition

Rocky Mountain spotted fever (RMSF) is a tick-borne illness caused by a bacteria, resulting in a high fever and a characteristic rash.

Description

The bacteria causing RMSF is passed to humans through the bite of an infected tick. The illness begins within about two weeks of such a bite. RMSF is the most widespread tick-borne illness in the United States, occurring in every state except Alaska and Hawaii. The states in the south-Atlantic region, (Delaware, Maryland, Washington DC, Virginia, West Virginia, North Carolina, South Carolina, Georgia, and Florida) have a great deal of tick activity during the spring and summer months, and the largest number of RMSF cases come from those states. About 5 percent of all ticks carry the causative bacteria.

Demographics

About 90 percent of all cases of RMSF occur between the months of April and September. Children under the age of 15 years have the majority of RMSF infections (about 66% of all infections). The peak incidence of RMSF occurs in five to nine year old children, with boys more likely to be infected than girls. A higher risk of infection seems to occur in individuals who spend time with dogs or who live near wooded or grassy areas.

Causes and symptoms

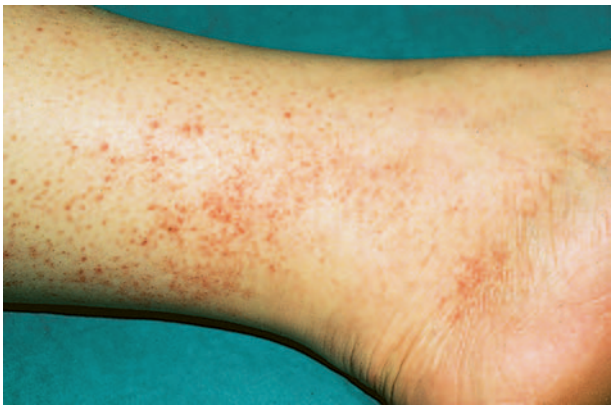
The bacterial culprit in RMSF is *Rickettsia rickettsii*. It causes no illness in the tick carrying it and can be passed on to the tick's offspring. When a tick attaches to a human, the bacteria are passed. The tick must be attached to the human for about six hours for this passage to occur. Although prompt tick removal will cut down on the chance of contracting RMSF, removal requires great care. If the tick's head and body are squashed during the course of removal, the bacteria can be inadvertently rubbed into the tiny bite wound.

Symptoms of RMSF begin within two weeks of the bite of the infected tick. Symptoms usually begin suddenly, with high fever, chills, **headache**, severe weakness, and muscle **pain**. Pain in the large muscle of the calf is very common, and may be particularly severe. The patient may be somewhat confused and delirious. Without treatment, these symptoms may last two weeks or more.

The rash of RMSF is quite characteristic. It usually begins on the fourth day of the illness and occurs in at least 90 percent of all patients with RMSF. It starts around the wrists and ankles, as flat pink marks (called macules). The rash spreads up the arms and legs, toward the chest, abdomen, and back. Unlike **rashes** that accompany various viral infections, the rash of RMSF does spread to the palms of the hands and the soles of the feet. Over a couple of days, the macules turn a reddish-purple color. In this new stage they are called petechiae, which are tiny areas of bleeding under the skin (pinpoint hemorrhages). Over the next several days, the individual petechiae may spread into each other, resulting in larger patches of hemorrhage.

The most severe effects of RMSF occur due to damage to the blood vessels, which become leaky. This action accounts for the production of petechiae. As blood and fluid leak out of the injured blood vessels, other tissues and organs may swell and become damaged. Other symptoms that may occur are as follows:

- breathing difficulties as the lungs are affected
- heart rhythms abnormal
- kidney failure in very ill patients
- liver function decrease
- nausea, **vomiting**, abdominal pain, and diarrhea
- brain inflammation (**encephalitis**) in about 25 percent of RMSF patients (Brain injury can result in seizures, changes in consciousness, actual coma, loss of coordination, imbalance on walking, **muscle spasms**, loss of bladder control, and various degrees of paralysis.)



Rash caused by Rocky Mountain spotted fever. *(Photograph by Ken E. Greer. Visuals Unlimited.)*

- the clotting system impaired and blood evident in the stools or vomit

Diagnosis

Diagnosis of RMSF is almost always made on the basis of the characteristic symptoms, coupled with either a known tick bite (noted by about 60 to 70 percent of patients) or exposure to an area known to harbor ticks. Complex tests exist to determine conclusively the diagnosis of RMSF, but these are performed in only a few laboratories. The results of these tests take so long to obtain that they are seldom used; delaying treatment is the main cause of death in patients with RMSF.

Treatment

It is essential to begin treatment absolutely as soon as RMSF is seriously suspected. Delaying treatment can result in death.

Antibiotics are used to treat RMSF. The first choice is doxycycline; the second choice is chloramphenicol. If the patient is well enough, treatment by oral intake of medicine is perfectly effective. Sicker patients may need to be given the medication through a needle in the vein (intravenously). Penicillin and sulfa drugs are not suitable for treatment of RMSF, and their use may increase the death rate by delaying the use of truly effective medications.

Very ill patients need to be hospitalized in an intensive care unit. Depending on the types of complications a particular patient experiences, a variety of treatments may be necessary, including intravenous fluids, blood transfusions, anti-seizure medications, kidney dialysis, and mechanical ventilation (a breathing machine).

KEY TERMS

Encephalitis—Inflammation of the brain, usually caused by a virus. The inflammation may interfere with normal brain function and may cause seizures, sleepiness, confusion, personality changes, weakness in one or more parts of the body, and even coma.

Macule—A flat, discolored area on the skin.

Petechia—Plural, petechiae. A tiny purple or red spot on the skin resulting from a hemorrhage under the skin's surface.

Prognosis

Prior to the regular use of antibiotics to treat RMSF, the death rate was about 25 percent. Although the death rate from RMSF has improved greatly with an understanding of the importance of early use of antibiotics, there is still a 5 percent death rate. This rate is believed to be due to delays in the administration of appropriate medications.

Certain risk factors suggest a worse outcome in RMSF. Death rates are higher in males and increase as people age. It is considered a bad prognostic sign to develop symptoms of RMSF within only two to five days of a tick bite.

Prevention

The mainstay of prevention involves avoiding areas known to harbor ticks. However, because many people enjoy recreational activities in just such areas, the following preventative steps can be taken:

- wearing light colored clothing (so that attached ticks are more easily noticed)
- wearing long sleeved shirts and long pants and tucking pant legs into socks
- spraying clothing with appropriate tick repellents
- examining oneself (Anybody who has been outside for any amount of time in an area known to have a population of ticks should examine his or her body carefully for ticks. Parents should examine their children at the end of the day.)
- removing any ticks using tweezers, so that infection does not occur due to handling the tick. (Parents should grasp the tick's head with the tweezers and pull gently but firmly so that the head and body are entirely removed.)
- keeping areas around homes clear of brush, which may serve to harbor ticks

Parental concerns

When children have been playing outside, it is important to carefully examine them for ticks when they come indoors. Rapidly yet carefully removing any ticks may help prevent or decrease the injection of infection-causing material. Dogs that are kept as **family** pets should also be examined for the presence of ticks and treated regularly with tick-killing products.

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ORGANIZATIONS

Centers for Disease Control and Prevention. 1600 Clifton Rd., NE, Atlanta, GA 30333. Web site: <www.cdc.gov>.

Rosalyn Carson-DeWitt, MD

Roseola

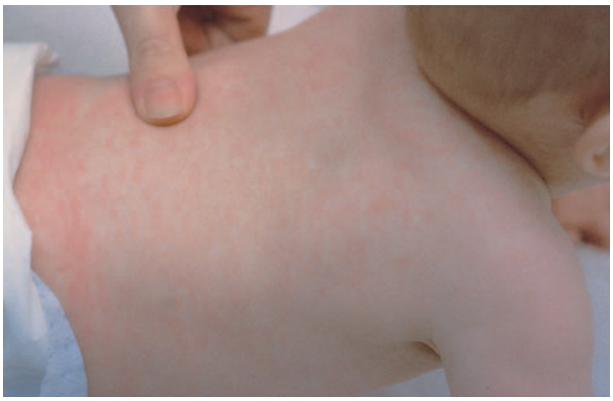
Definition

Roseola is a common disease of babies or young children, in which several days of very high **fever** are followed by a characteristic rash.

Demographics

Roseola is an extraordinarily common infection, caused by a virus. About 90 percent of all children have been exposed to the virus, with about 33 percent actually demonstrating the syndrome of fever followed by rash.

The most common age for a child to contract roseola is between six and twelve months. Roseola infection strikes boys and girls equally. The infection may occur at any time of year, although late spring and early summer seem to be peak times for it.



Roseola rash on infant's back and shoulders. *(Photograph by Keith. Custom Medical Stock Photo, Inc.)*

KEY TERMS

Jaundice—A condition in which the skin and whites of the eyes take on a yellowish color due to an increase of bilirubin (a compound produced by the liver) in the blood. Also called icterus.

Mononucleosis—An infection, caused by the Epstein-Barr virus, that causes swelling of lymph nodes, spleen, and liver, usually accompanied by extremely sore throat, fever, headache, and intense long-lasting fatigue. Also called infectious mononucleosis.

Causes and symptoms

About 85 percent of the time, roseola is caused by a virus called human herpesvirus 6 (HHV-6). Although the virus is related to those herpesviruses known to cause sores on the lips or genitalia, HHV-6 causes a very different type of infection. HHV-6 is believed to be passed between people via infected saliva. A few other viruses (called enteroviruses) can produce a similar fever-then-rash illness, which is usually also called roseola.

Researchers believe that it takes about five to 15 days to develop illness after having been infected by HHV-6. Roseola strikes suddenly, when a previously well child spikes an impressively high fever. The temperature may reach 106°F (41°C) . As is always the case with sudden fever spikes, the extreme change in temperature may cause certain children to have seizures. About 5 to 35 percent of all children with roseola have **febrile seizures**.

The most notable thing about this early phase of roseola is the absence of symptoms, other than the high fever. Although some children have a slightly reddened throat or a slightly runny nose, most children have no symptoms whatsoever, other than the sudden development of high fever. This fever lasts for between three and five days.

Somewhere around the fifth day, a rash begins on the body. The rash is usually composed of flat pink patches or spots, although there may be some raised patches as well. The rash usually starts on the chest, back, and abdomen then spreads out to the arms and neck. It may or may not reach the legs and face. The rash lasts for about three days then fades.

Very rarely, roseola causes more serious disease. Patients so afflicted experience significant swelling of the lymph nodes, the liver, and the spleen. The liver may become sufficiently inflamed to interfere with its functioning, resulting in a yellowish color to the whites of the eyes and the skin (**jaundice**). This syndrome (called a mononucleosis-like syndrome, after the disease mononucleosis that causes many of the same symptoms) has occurred in both infants and adults.

Diagnosis

The diagnosis of roseola is often made by carefully examining the feverish child to make sure that other illnesses are not causing the temperature spike. Once it is clear that no **pneumonia**, ear infection, **strep throat**, or other common childhood illness is present, the practitioner usually feels comfortable waiting to see if the characteristic rash of roseola begins.

Treatment

As of 2004, there were no treatments available to stop the course of roseola. **Acetaminophen** or ibuprofen is usually given to try to lower the fever. Children who are susceptible to seizures may be given a sedative medication when the fever first spikes in an attempt to prevent such a seizure.

Prognosis

Children recover quickly and completely from roseola. The only complications are those associated with seizures or the rare mononucleosis-like syndrome.

Prevention

Other than the usual good hygiene practices always recommended to decrease the spread of viral illness, no

methods as of 2004 are available to specifically prevent roseola.

Parental concerns

Roseola is usually a benign illness, from which the child recovers uneventfully. One of the more major potential complications is the development of febrile seizures secondary to the rapid, high rise in fever. Rare cases of **encephalitis** or meningoencephalitis have also been reported.

Resources

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Rosalyn Carson-DeWitt, MD

Rotavirus infections

Definition

Rotavirus is the major cause of **diarrhea** and **vomiting** in young children worldwide. The infection is highly contagious and may lead to severe **dehydration** (loss of body fluids) and even death.

Description

Gastroenteritis (inflammation of the stomach and the intestine) is the second most common illness in the United States, after the **common cold**. More than one-third of such cases are caused by viruses. Many different viruses can cause gastroenteritis, but the most common ones are the rotavirus and the Norwalk virus.

The name rotavirus comes from the Latin word "rota" for wheel and is given because the viruses have a distinct wheel-like shape. Rotavirus infection is also known as infantile diarrhea or winter diarrhea, because it mainly targets infants and young children. The outbreaks are usually in the cooler months of winter.

The virus is classified into different groups (Group A through group G), depending on the type of protein marker (antigen) that is present on its surface. The diarrheal infection of children is caused by the group A rotaviruses. Group B rotaviruses have caused major epidemics of adult diarrhea in China. Group C rotavirus has been associated with rare cases of diarrheal outbreaks in Japan and England. Groups D through G have not been detected in humans.

Demographics

In the United States, more than 50,000 children are hospitalized and up to 125 die each year as a result of rotavirus infection. Moreover, worldwide, rotavirus is thought to be responsible for more than 5 to 10 million deaths in children every year. Children in developing countries are particularly hard-hit by this infection, which is thought to be the leading cause of childhood death globally.

Causes and symptoms

The main symptoms of the rotavirus infection are **fever**, stomach cramps, vomiting, and diarrhea (which can lead to severe dehydration). The symptoms last from four to six days. Symptoms of dry lips and tongue, dry skin, sunken eyes, and fewer than six diapers wet per day indicate dehydration, and a physician needs to be notified. Because of excellent U.S. healthcare, rotavirus is rarely fatal to American children. In developing countries, however, with insufficient means to rehydrate children, rotavirus is oftentimes fatal.

The virus is usually spread by the fecal-oral route. In other words, a child can catch a rotavirus infection if she puts her finger in her mouth after touching **toys** or things that have been contaminated by the stool of another infected child. This usually happens when children do not wash their hands after using the toilet or before eating food.

The viruses can also spread by way of contaminated food and drinking water. Infected food handlers who prepare salads, sandwiches, and other foods that require no cooking can spread the disease. Generally, symptoms appear within four to 48 hours after exposure to the contaminated food or water.

Children between the ages of six months and two years, especially in a daycare setting, are the most susceptible to this infection. Breastfed babies may be less likely to become infected, because breast milk contains antibodies (proteins produced by the white blood cells of the immune system) that fight the illness. Nearly every child by the age of four has been infected by this virus and has rotavirus antibodies in their body. The disease

also targets the elderly and people who have weak immune systems.

Children who have been infected once can be infected again. However, second infections are less severe than the first infections. By the time a child has had two infections, the chance of subsequent severe infection is remote.

Diagnosis

The rotavirus infection is diagnosed by identifying the virus in the patient's stool. This is done using electron microscopy. Immunological tests such as Enzyme-linked immunosorbent assay (ELISA) are also widely used for diagnosis, and several commercial kits are available.

Treatment

Oral rehydration therapy (drinking enough fluids to replace those lost through bowel movements and vomiting) is the primary aim of the treatment. Electrolyte and fluid replacement solutions are available over the counter in food and drug stores. Dehydration is one of the greatest dangers for infants and young children. If the diarrhea becomes severe, it may be necessary to hospitalize the patient so that fluids can be administered intravenously.

Anti-diarrheal medication should not be given to children unless the parent or caregiver is directed to do so by the physician. Antibiotic therapy is not useful in viral illness. Specific drugs for the virus were, as of 2004, not available.

Prognosis

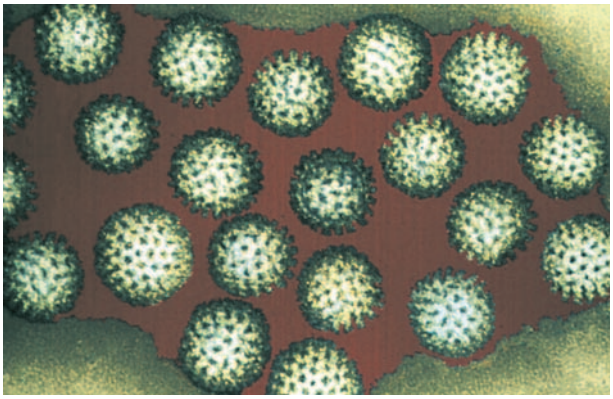
Most of the infections resolve spontaneously. Dehydration due to severe diarrhea is one of the major complications.

Prevention

The best way to prevent the disease is by proper food handling and thorough hand washing, after using the toilet and whenever hands are soiled. In childcare centers and hospital settings, the staff should be educated about personal and environmental hygiene. All dirty diapers should be regarded as infectious and disposed of in a sanitary manner.

Parental concerns

As with any illness that may cause dehydration, the primary parental concern is using an appropriate rehydration solution. Several balanced electrolyte rehydration solutions are available. Some healthcare providers also give their patients a homemade recipe for such a solution. It is crucial that parents and children use excellent hand-washing technique after toileting and diaper changes to prevent further spread of rotavirus diarrhea throughout the **family**. The healthcare provider should also give guidance concerning how long a child with rotavirus diarrhea should be kept home from daycare or school.



Rotaviruses are probably the most common viruses to infect humans and animals. These viruses are associated with gastroenteritis and diarrhea in humans and other animals. *(Photograph by Dr. Linda Stannard. Photo Researchers, Inc.)*

Resources

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Lata Cherath, PhD
Rosalyn Carson-DeWitt, MD

RSV infection *see* **Respiratory syncytial virus infection**

Rubella

Definition

Rubella, also called German **measles** or three-day measles, is a highly contagious viral disease that in most

children and adults causes mild symptoms of low **fever**, swollen glands, joint **pain**, and a fine red rash. Although rubella causes only mild symptoms in child and adult sufferers, the infection can have severe complications for the fetus of a woman who becomes infected with the virus during the first trimester of pregnancy. These complications include severe birth defects or death of the fetus.

Description

Rubella is spread through contact with fluid droplets expelled from the nose or throat of an infected person. A person infected with the rubella virus is contagious for about seven days before any symptoms appear and continues to be able to spread the disease for about four days after the appearance of symptoms. Rubella has an incubation period of 12 to 23 days.

Although rubella is generally considered a childhood illness, people of any age who have not been vaccinated or previously caught the disease can become infected. Having rubella once or being immunized against rubella normally gives lifetime immunity. For this reason **vaccination** is highly effective in reducing the number of rubella cases.

Women of childbearing age who do not have immunity against rubella should be particularly concerned about getting the disease. Rubella infection during the first three months of pregnancy can cause a woman to miscarry or cause her baby to be born with birth defects. Although it has been practically eradicated in the United States, rubella is still common in less developed countries because of poor immunization penetration, creating a risk to susceptible travelers. Some countries have chosen to target rubella vaccination to females only and outbreaks in foreign-born males have occurred on cruise ships and at U.S. summer camps.

Demographics

Although rubella was once a common childhood illness, its occurrence has been drastically reduced since vaccine against it became available in 1969. According to statistics for 1964–1965, prior to routine rubella immunization in the United States, there were 2,100 newborn deaths and 11,250 miscarriages attributed to rubella infection of pregnant women. In addition, about 20,000 infants were born with birth defects attributable to rubella infection in utero. Of these babies, 11,600 were born deaf; 3,580 were born blind; and 1,800 suffered severe **developmental delay**. In the 20 years following the introduction of the vaccine, reported rubella cases dropped 99.6 percent. In 2000, there were only 152 reported cases of rubella infection and seven reported cases of congenital rubella.

Causes and symptoms

Rubella is caused by the rubella virus (*Rubivirus*). Symptoms are generally mild, and complications are rare in anyone who is not pregnant.

The first visible sign of rubella is a fine red rash that begins on the face and rapidly moves downward to cover the whole body within 24 hours. The rash lasts about three days, which is why rubella is sometimes called the three-day measles. A low fever and swollen glands, especially in the head (around the ears) and neck, often accompany the rash. Joint pain and sometimes joint swelling can occur, more often in women. It is quite common to get rubella and not show any symptoms (subclinical infection).

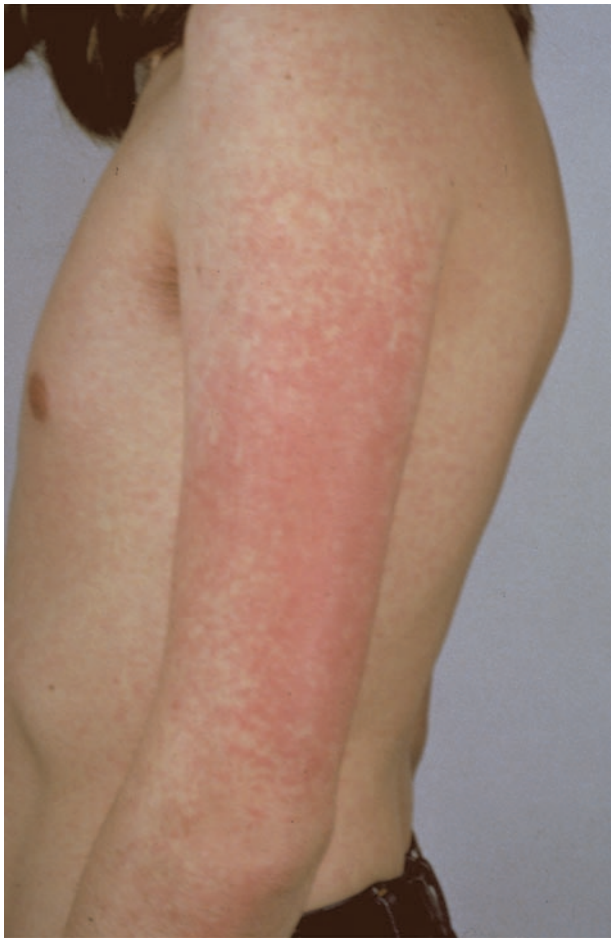
Symptoms disappear within three to four days, except for joint pain, which may linger for a week or two. Most people recover fully with no complications. However, severe complications may arise in the unborn children of women who get rubella during the first three months of their pregnancy. These babies may be miscarried or stillborn. A high percentage is born with birth defects. Birth defects are reported to occur in 50 percent of women who contract the disease during the first month of pregnancy, 20 percent of those who contract it in the second month, and 10 percent of those who contract it in the third month.

The most common birth defects resulting from congenital rubella infection are eye defects such as cataracts, glaucoma, and blindness; deafness; congenital heart defects; and **mental retardation**. Taken together, these conditions are called congenital rubella syndrome (CRS). The risk of birth defects drops after the first trimester, and by the twentieth week, there are rarely any complications.

Diagnosis

The rash caused by the rubella virus and the accompanying symptoms are so similar to other viral infections that it is impossible for a physician to make a confirmed diagnosis on visual examination alone. The only sure way to confirm a case of rubella is by isolating the virus with a blood test or in a laboratory culture.

A blood test is done to check for rubella antibodies. When the body is infected with the rubella virus, it produces both immunoglobulin G (IgG) and immunoglobulin M (IgM) antibodies to fight the infection. Once IgG exists, it persists for a lifetime, but the special IgM



A red rash is one characteristic of rubella, or German measles, as seen on this teenager's arm. *(Custom Medical Stock Photo, Inc.)*

antibody usually wanes over six months. A blood test can be used either to confirm a recent infection (IgG and IgM) or determine whether a person has immunity to rubella (IgG only). The lack of antibodies indicates that a person is susceptible to rubella.

All pregnant women should be tested for rubella early in pregnancy, whether they have a history of vaccination. If the woman lacks immunity, she is counseled to avoid anyone with the disease and to be vaccinated after giving birth.

Treatment

There is no drug treatment for rubella. Bed rest, fluids, and **acetaminophen** for pain and temperatures over 102°F (38.9°C) are usually all that is necessary.

Babies born with suspected CRS are isolated and cared for only by people who are sure they are immune to rubella. Congenital heart defects are treated with surgery.

KEY TERMS

Incubation period—The time period between exposure to an infectious agent, such as a virus or bacteria, and the appearance of symptoms of illness. Also called the latent period.

Trimester—The one of three periods of about 13 weeks each into which a pregnancy is divided.

Prognosis

Complications from rubella infection are rare in children, pregnant women past the twentieth week of pregnancy, and other adults. For women in the first trimester of pregnancy, there is a high likelihood of the child being born with one or more birth defect. Unborn children exposed to rubella early in pregnancy are also more likely to be miscarried, stillborn, or have a low birth weight. Although the symptoms of rubella pass quickly for the mother, the consequences to the unborn child can last a lifetime.

Prevention

Vaccination is the best way to prevent rubella and is normally required by law for children entering school. Rubella vaccine is usually given in conjunction with measles and **mumps** vaccines in a shot referred to as MMR (mumps, measles, and rubella). Children receive one dose of **MMR vaccine** at 12 to 15 months and another dose at four to six years.

Pregnant women should not be vaccinated, and women who are not pregnant should avoid conceiving for at least three months following vaccination. As of 2004, however, accidental rubella vaccinations during pregnancy had not clearly been associated with the same risk as the natural infection itself. Women may be vaccinated while they are breastfeeding. People whose immune systems are compromised, either by the use of drugs such as steroids or by disease, should discuss possible complications with their doctor before being vaccinated.

Parental concerns

While rubella infection in an older child or adult is rarely complicated, the risks of not immunizing a child

against rubella are highest in the unborn. Congenital rubella is a serious, life-changing condition, and adherence to immunization recommendations is crucial to the public health.

Resources

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ORGANIZATIONS

March of Dimes Birth Defects Foundation. 1275 Mamaroneck Ave., White Plains, NY 10605. Web site: <www.modimes.org>.

National Organization for Rare Disorders. PO Box 8923, New Fairfield, CT 06812-8923. Web site: <www.rarediseases.org>.

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Rosalyn Carson-DeWitt, MD

Rubeola *see* **Measles**

Running away

Definition

Running away involves being voluntarily absent from home at least overnight without permission from a parent or caretaker.

Description

Every year about 800,000 children in the United States are reported missing and another estimated 500,000 go missing without being reported. Not all of these children are runaways. This number also includes children abducted by **family** members, usually in custody disputes, and a very small number of stranger abductions. In addition, when children run away, each time it is reported as a separate event. Some children are repeat runaways, so it is difficult to know the exact number of runaway children. What is clear is that the number is large. Runaways include "throwaways," who leave with the overt or tacit approval of parents or caretakers, and "push-outs," who are turned out by parents who do not want them, as well as teens who leave because they are dissatisfied with their home life.

The 2002 White House Conference on Missing, Exploited, and Runaway Children estimated that there were about 1.3 million American children living on the streets each day and that one in seven children between the ages of 10 and 18 will run away. Most runaways return voluntarily within a few days. Many go to homes of friends or relatives who encourage them to return. Some are aided by police and social agencies and eventually return home or are placed in alternative stable environments. Children who remain on the street are exposed to sexual exploitation, drug **addiction**, violent crime, and the other harmful mental and physical effects of homelessness.

Why children run away

Rather than seeking adventures, most runaways in the early 2000s are running from intolerable domestic situations. It has been estimated that at least 60 to 70 percent of these young people are fleeing from families in which they have been mentally, physically, or sexually abused. Historically, attention to the role played by a child's family environment in the treatment of a runaway is relatively new. In past eras, runaways themselves were uniformly blamed for their situation and seen as hostile and destructive lawbreakers who needed to be reformed. In the nineteenth century, they were generally sent to reform schools that were similar to prisons. Even after the establishment of the juvenile justice system toward the end of the nineteenth century, most runaways were regarded as delinquents, and the home situations from which they had fled received little scrutiny. In the early and mid-twentieth century, the prevailing view of runaways underwent a partial shift in emphasis from crime to pathology. Early versions of the American Psychiatric Association's *Diagnostic and Statistical Manual* included "runaway reaction" as a mental disorder.

As of 2004, researchers had identified several common characteristics of the abusive family environments that prompt young people to run away. These include financial troubles, sexual abuse, alcohol and drug abuse, physical and verbal abuse, and intolerance of deviant behavior. Besides outright physical or sexual abuse, runaways may be reacting to persistent tension between family members, including parental fighting or competition among siblings (especially step-siblings), feelings of rejection by their families, or authoritarian parenting that allows too little room for normal self-expression or social life.

Other events may also prompt children to run away. They may have done something to get into trouble (for

example, become pregnant or been arrested) and feel unable to face their families. Still other children flee out of romantic notions of being with a girlfriend or boyfriend. Cyber predators who meet young people in Internet chat rooms and convince them to leave home to meet or live with them constitutes a relatively new, but growing, problem. Some children leave with friends for adventure. These children are usually ones who have had difficulties with parents, school, and authority figures in the past. Running away is one component of **conduct disorder**, a diagnosis recognized by the American Psychiatric Association.

Legislation affecting the treatment of runaways

In the 1960s, with the growth of the hippie counterculture and the associated "youth rebellion," the number of teen runaways increased dramatically, drawing attention to the risks these youths faced on the streets. Growing public concern over their fate was reflected in the 1974 passage of the Runaway Youth Act, which funded a program to establish a network of centers for runaways. Increased attention to the plight of these young people revealed the dangers of child prostitution and pornography, which they faced on the streets. It also began to change the public image of runaways from that of thrill-seekers to that of young people from families in crisis fleeing intolerable conditions with no place else to go. This perception of runaways has become influential in both public opinion and government policy.

The 1982 Missing Children's Act enabled the entry of missing child information into the FBI's national crime computer (NCIC). The 1984 Missing Children's Assistance Act mandated a national resource center to address child abduction and exploitation. The private, nonprofit National Center for Missing and Exploited Children (NCMEC) was established in cooperation with the United States Department of Justice, to find missing children and prevent child victimization. It created a 24-hour hotline (1–800–THE-LOST). In 1990, the National Child Search Assistance Act eliminated the waiting time for law enforcement action on missing children, mandating an immediate police report and NCIC entry for missing children cases.

Common problems

What happens to runaway children?

Young people who run away and do not return home may remain on the street, go to a shelter, or be placed in foster homes by welfare agencies. Some eventually join the armed services or take jobs that keep them on the road, such as carnival or sales work. Others end up in jails or mental institutions. Those who remain on the streets have few options that would provide them with decent living conditions. Their age, lack of work experience, and uncompleted education make it difficult for them to find a job, especially one that pays more than minimum wage. It is common for both male and female runaways living on the streets to steal, panhandle, deal and abuse drugs, engage in prostitution, and pose for pornographic pictures. For shelter they may stay with strangers, spend nights in bus stations, all-night coffee shops, and other public places, or stow away in empty or abandoned buildings or even in stairwells. Many never get off the streets, becoming part of the adult homeless population.

There are an estimated 750 runaway shelters and youth crisis centers in the United States. These offer safe shelter, food, counseling, and advocacy services to help young people deal with parents, police, and the courts. Many also provide educational and vocational assistance. However, shelters do set certain conditions for accepting runaways, the most common being parental notification. This is an obstacle for some young people who do not want their parents contacted, even though the shelter does not press them to return home. One problem that has occurred at some shelters is sexual molestation by other runaways and staff members. Nevertheless, many young people have had positive experiences at shelters, which they either find on their own or are sent to by the legal or welfare systems.

Since the 1970s, hotlines have been available to help runaways and their families. The Runaway Hotline and the National Runaway Switchboard (1–800–621–4000) have become widely used 24-hour help lines that offer crisis counseling and referrals to service agencies that can provide food, shelter, medical aid, and other types of help. The National Runaway Switchboard will put runaways and their parents in touch without revealing the location from which the teenager is calling.

Parental concerns

Parents are often emotionally devastated when their child runs away. Their fluctuating emotions may include anger, grief, guilt, and **fear**. Sometimes they are not sure if their child has run away or been abducted. A parent's first concern is to find his or her child and/or make sure he or she is in a safe environment. To help achieve this, the National Center for Missing and Exploited Children recommends these steps for parents.

- They should check with friends and relatives to try to locate the child and enlist their help in thinking about where the child might be.

- They should check diaries and e-mails for clues about the child's plans. They can ask the child's friends if they know the child's online passwords.
- They should report to local law enforcement immediately that the child has run away or is missing. There is no waiting period to report a missing or runaway minor or to enter their information into the FBI NCIC database.
- They should provide a description and photograph of the child to law officers.
- They should check local places where the child may be hanging out.
- They should check again with the child's friends. They may know something but initially be reluctant to tell the parents.
- They should call the National Runaway Switchboard (1–800–621–4000) and see if the child has left a message for them. They can leave a message for the child here in case the child calls the hotline.

If the child contacts the parents and refuses to return home, the parents should encourage him or her to contact the National Runaway Switchboard and ask for assistance or encourage the child to go to a friend or relative. Parents can ask their child to stay in touch and make a plan about when the child will call again. If the child returns home, parents need to try to respond with concern and love, rather than anger. Children who have been away for more than a few days should have a complete medical examination. They also can benefit from seeing a mental health practitioner for help dealing with the distress that drove them away from home. **Family therapy** to help resolve whatever family problems may have driven the child away from home initially can also be beneficial in preventing a repeat running away incident.

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Covenant House. Telephone: toll-free 800/999–9999 (Referrals and counseling for youth in need); Web site: <www.covenanthouse.org>.

National Center for Missing and Exploited Children (NCMEC). Charles B. Wang International Children's Building, 699 Prince St., Alexandria, VA 22314–2175. Telephone: 24-hour toll-free hotline 800/THE-LOST [800/843–5678]; Web site: <www.missingkids.com>.

National Runaway Switchboard. Telephone: toll-free hotline 800/621–4000; Web site: <www.nrscrisisline.org>.

The Runaway Hotline. PO Box 12428, Austin, TX 78711. Telephone: toll-free hotline 800/231–6946; Web site: <www.nrscrisisline.org>.

Tish Davidson, A.M.

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INFANCY THROUGH ADOLESCENCE

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Safety

Definition

The safety of children is potentially at risk from accidents and injuries, as well as crime. Providing a safe environment, putting prevention measures into practice, and teaching children methods of self-protection are all ways to reduce the potential for harm to children.

Description

Accidents are the leading cause of death for children aged 14 and under in the United States, claiming more than 5600 lives each year, or an average of 15 children per day. More than 16 percent of all hospitalizations for accidental injuries among children lead to permanent disability. Although the accidental injury death rate declined among children ages 14 and under by almost 40 percent from 1987 to 2000, accidental injury remained in the early 2000s the number one killer of this age group. In 2000, the leading cause of fatal accidental injury among children was motor vehicle occupant injury (28%), followed by drowning (16%) and airway obstruction injury (14%). Falls (36%) were the leading cause of nonfatal, hospital emergency room-treated childhood injury in 2001. Other frequent causes of accidental injuries and deaths are fire and burn injury, accidental firearm injury, and **poisoning**.

Another way children may have their safety jeopardized is by becoming victims of crime. Child abductions are often publicized widely and cause parents to experience a great deal of **anxiety** and **fear** regarding this possibility. Another relatively new place children face potential dangers is on the "information highway." Though the Internet opens a world of possibilities to children, there are individuals who may attempt to exploit and harm children through this technology.

Though the idea of the number of potential risks children face may seem overwhelming to parents, there are a variety of measures parents can take to reduce those risks.

Motor vehicle occupant injury

In 2001, motor vehicle accidents resulted in 36 percent of accidental deaths in children ages one to four. In the early 2000s an estimated 14 percent of children ages 14 and under continued to ride unrestrained, however, and 55 percent of those children killed in motor vehicle accidents were not restrained. Also, at that time, nearly one-third of children rode in the wrong restraint for their zage and size, and an estimated 82 percent of child safety seats were installed or used incorrectly. The following measures will help parents keep their children safe:

- Car seats need to meet federal safety standards. A car seat with a five-point harness will provide the best protection. In addition, the car seat needs to be the correct size for the child and needs to fits properly into the vehicle.
- The Lower Anchors and Tethers for Children (LATCH) system in cars manufactured after September 1, 2002, should be used. Some car seats require that parents attach additional hardware for maximum protection.
- The child must face in the right direction. Infants should ride in a car seat that faces the rear of the car until they are one year of age and weigh approximately 20 lbs (9 kg). Infants who weigh 20 lbs (9 kg) before they are one need a restraint approved for the higher weight and should also be rear-facing.
- Car seats should be installed correctly. The car seat should be held tightly against the car's back seat. After installing the car seat, parents need to make certain they cannot move it more than one inch from side to side or front to back. Police departments and community organizations frequently hold child restraint inspections, during which parents can discover if they have installed their car seats properly.
- Rear-facing car seats should not be placed in the front seat of a car that has air bags. Children 12 and under should ride in the back seat in order to avoid being hurt by inflating airbags. Generally, the back seat of the car is the safest place in a crash.

- Children need to stay in a safety seat with a full harness for as long as possible, at least until they weigh 40 lbs (18 kg). Afterwards, they can use a belt-positioning booster seat, which provides a taller sitting height so that the adult lap and shoulder belts fit correctly.
- When older children are 57 inches (1.45 m) tall and weigh 80 lbs (36 kg), they may use adult lap belts.
- Children need to be restrained every time they ride in a car.
- Children should never be left alone in or around a vehicle. Unattended children can quickly die from heat stroke or carbon monoxide poisoning.

Drowning

Drowning remains the second leading cause of accidental injury-related deaths among children ages 14 and under, claiming 943 children in 2000. An estimated 4700 children required treatment in hospital emergency departments for drowning-related incidents in 2001. As many as 20 percent of children who survive **near-drowning** suffer severe, permanent neurological disability. Children ages one to four are at the highest risk of drowning. The following measures may significantly reduce the drowning risk for your child:

- Parents and caregivers should never, even for a moment, leave children alone or in the care of another young child while in bathtubs, pools, spas, or wading pools or near any other open standing water. Infant bath seats are not a substitute for adult supervision. Parents should remove all water from containers, such as pails and buckets, immediately after use.
- If the home has a swimming pool, it should be surrounded by a fence that prevents children from having direct access to the pool from the house. Remove **toys** from in and around the pool, as toys can attract children to the pool.
- Parents should enroll their child in swimming lessons when they are old enough (usually not before age four), but should remember that these lessons do not provide protection against drowning for children of any age.
- Children should be taught to always swim with a buddy. In addition, they should be instructed never to dive into an unknown body of water, but instead jump in feet first to avoid hitting their heads.
- When boating, every person must wear a U.S. Coast Guard approved life jacket.
- Air-filled swimming aids (such as water wings) cannot take the place of life preservers.
- Parents need to teach their children about the risks of drowning in the cold weather months. Children should not walk, skate, or ride on thawing ice on any body of water.
- Parents should learn CPR and keep a telephone close to the area where their children are swimming.

Poisoning

Poisoning is a common cause of home accidents, with toddlers being the ones most vulnerable. Children are at risk of poisoning from household and personal care products, medicines, **vitamins**, indoor plants, lead, and carbon monoxide. In 2000, 91 children ages 14 and under died as a result of accidental poisoning. Approximately 114,000 children in this age group were treated in the emergency room for accidental poisonings in 2001. People can keep children safe by being aware of the potential hazards in the home and by following these guidelines:

- Medications and cleaning solutions need to be stored in locked cabinets.
- Medication lids need to be tightly closed with child-resistant caps.
- Parents should avoid taking medicine in front of children and never refer to pills as candy, as children often mimic the behavior of adults.
- Parents should check the garage for any toxic chemicals and gasoline containers. Items such as windshield washer fluid, antifreeze, and pesticides are poisonous and should be placed where children cannot reach them. In addition, these kinds of items should never be kept in juice or milk bottles.
- Poisonous plants in the home need to be identified and either removed or placed where children cannot reach them.
- Carbon monoxide detectors/alarms should be installed in homes and recreational vehicles. These should be placed in the hallway near every separate sleeping area of the home.
- Insect sprays should not be used around food.
- All painted furniture and toys should be checked for non-toxic finishes.
- The Poison Control Center phone number should be posted in a prominent place, where **family** members and other caregivers can find it quickly. Caregivers should call the Poison Control Center (1–800–222–1222) immediately when a poisoning incident is suspected. The experts at the Poison Center provide directions on the appropriate actions to take.

Fire and burn injuries

Fire and burn injury is the fifth leading cause of child accidental injury-related death. Children make up 20 percent of all fire deaths, and over 30 percent of all fires that kill children are set by children playing with fire. Children of all ages set over 100,000 fires each year, and approximately 20,000 of these are set in homes. Children aged four and under are at the greatest risk, with a fire- and burn-related death rate nearly twice that of all children. This circumstance occurs for several reasons. Young children have a less acute perception of danger and a limited ability to properly respond to a life-threatening burn or fire situation. They are also more susceptible to fire-related asphyxiation, as well as more prone to **burns** than adults. The United States Fire Administration (USFA) encourages parents to teach children at an early age about the dangers of playing with fire in order to help prevent child injuries, fire deaths, and the number of fires set in homes. The following suggestions will aid in keeping children safe from fires:

- Young children need to be supervised closely. They should not be left alone even for short periods of time.
- Lighters and matches should be kept in a secured area and children taught to tell an adult if they find lighters or matches.
- Parents should look for indications that children may be playing with fire, for burnt matches under beds or in closets.
- Families need a home fire escape plan and to practice it with the children. A meeting place outdoors should be designated.
- Children should be taught that if a fire occurs, they should crawl low on the floor, below the smoke, and get out of the house according to the escape plan. They should not attempt to get back in the house.
- Children need to know how to stop, drop to the ground, and roll if their clothes catch on fire.
- Parents should install smoke alarms on every level in the home, and familiarize children with the sound of the smoke alarm. They should test the alarm monthly and replace the battery at least yearly. Having a working smoke alarm dramatically increases residents' chances of surviving a fire.
- The thermostat on the hot water heater should be set to 120°F (49°C) or lower. The water temperature should be checked when bathing or showering children.
- Do not drink or carry very hot beverages or soup when holding a child.
- Access to the stove should be blocked if possible. Foods should be cooked on the back burners with pot handles turned away from the front of the stove. Parents and caregivers should avoid holding a baby or small child while they are cooking.

Falls

Each year, nearly 3 million children in the United States are injured in falls. For those under five, falls cause more than half of all injuries. Even close supervision is not adequate, as falls can happen very quickly. They can occur at home as well as away from home. Although most falls result in only mild bumps and **bruises**, many cause serious injuries that require immediate medical attention. Following these guidelines may help to prevent children from becoming injured in a fall:

- Playgrounds should have soft surfaces to cushion children if they should fall. Examples of soft surfaces are those made of items like bark mulch, wood chips, sand, pea gravel, or shredded tires. Avoid concrete, asphalt, and dirt surfaces. Even sod can be too hard under certain weather conditions.
- Chairs and other pieces of furniture in the home should be kept away from windows. Windows should be closed and locked when children are around. Residential windows in tall buildings should have bars or window guards. Window screens may not prevent children from falling out a window.
- Stairways must be clear to prevent children from tripping over clutter.
- Throw rugs should be secured to the floor with a rubber pad, double-sided tape, or a piece of foam carpet backing.
- Safety gates can keep toddlers away from stairs. Gates should be attached to the wall if they are used at the top of a staircase.
- Safety belts keep children from falling from shopping carts.
- As children get older and start riding a bike, a scooter, or using skates, they should always wear a correctly fitting helmet. If a child falls from one of these while wearing a helmet, the risk of a brain injury is reduced by 88 percent. A properly fitting helmet sits evenly on top of the head (low on the forehead, no more than two finger widths above the eyebrows), should be comfortable but snug, and have straps firm enough so that the helmet will not rock forward, backward, or side to side.

Airway obstruction injury

Children, especially those under the age of three, are quite vulnerable to airway obstruction injury because

they have small upper airways and have relative inexperience with chewing. They also have a tendency to place objects in their mouths. On average, infants account for approximately 64 percent of **choking** deaths among children ages 14 and under. Causes of choking or airway obstruction-injury deaths include suffocation by things such as pillows, choking on food or small objects, and strangulation from window blind and clothing strings. Anything children can place in their mouths can be dangerous. Taking the following steps will help protect children:

- Parents should avoid giving children under age four any hard, smooth foods that may block or partially block their airway. These include all nuts, sunflower seeds, watermelon with seeds, cherries with pits, popcorn, hard candy, raw carrots, raw peas, and raw celery.
- Certain soft foods, such as hot dogs, grapes, and link sausages, should be chopped into small pieces. These foods can cause choking because they are the right shape to block the windpipe.
- When babies start to eat solid food, parents need to beware of foods such as raw apples or pears. Raw fruit is difficult for babies to chew properly because their teeth are just developing.
- Children should sit still while eating and chew food thoroughly.
- Children should not run, ride in the car, or **play sports** with gum, lollipops, or candy in their mouths.
- Buttons, beads, and other small objects need to be stored safely out of children's reach.
- Drawstrings should be removed from children's coats and sweatshirts. Also window blind cords that pose a risk for strangulation should be removed.
- Parents should follow manufacturer's recommendations regarding toys and check toys frequently for loose or broken parts.
- Older children should not to leave toys with small pieces or loose game parts where younger children can reach them.
- A latex balloon should not be given to a child younger than age eight. Children can choke by inhaling the balloon or a portion of it into their windpipes.
- Parents should obtain and use a "small parts tester," an inexpensive child safety device that shows if an object is small enough to fit in a child's mouth.

Accidental firearm injury

In the year 2000, 193 children in the United States ages new infant to 19 died from accidental injuries involving firearms. A child as young as three has the finger strength to pull a trigger. Some studies show that by age eight, 90 percent of children are capable of firing a gun. Whether people are gun collectors, hunters, or fierce gun control advocates, they need to ensure their families' safety by talking with their children about the potential dangers of guns and what to do if one is found. Parents should assume that their children may come across a gun at some point in their youth and proactively teach them about gun safety. There are a number of programs available that instruct children, including the very popular "Eddie Eagle," a program of the National Rifle Association (NRA). This program offers a four-step approach to gun safety: stop, don't touch, leave the area, and tell an adult. People who own firearms should follow these guidelines to prevent accidental shootings:

- Guns need to be stored unloaded in a securely locked case and out of children's reach.
- Trigger locks and other safety features should be used.
- Ammunition should be stored in a separate place from the firearms, locked in a container that is out of children's reach.
- Gun owners should take a firearms safety course to learn the correct and safe way to use the firearm, and they should practice firearm safety. Children need to be taught that guns are not toys. They need to be taught to always tell an adult about any gun they happen to find.

Online risks

While online computer exploration opens a world of possibilities to children, it also may expose them to a variety of dangers. Teenagers are particularly at risk because they are more likely to go online unsupervised and are more likely than younger children to participate in online discussions. Risks posed by the Internet include the following:

- Exposure to inappropriate material that is sexual, violent, hateful, or that encourages activities that are dangerous or illegal.
- Exposure to information or arrangements for an encounter that could risk children's safety or the safety of other family members. In some cases child molesters have used chat rooms, email, and instant messaging to gain a child's trust and then to arrange a face-to-face meeting.

There are several signs that children may be at risk online. These include their spending large amounts of time online, especially in the evenings; the presence of pornography on their computers; their making or receiv-

ing calls from men parents do not know; their receiving mail, gifts, or packages from people parents do not know; their turning off the monitor or quickly changing the screen on the monitor when parents enter the room; their becoming withdrawn from the family; and their using an online account that belongs to someone else.

Parents should not feel powerless in the face of these online risks. There are a variety of measures they can take to minimize the chances of an online exploiter victimizing their child. These include the following:

- Children need to be warned about the potential dangers online and about sexual victimization.
- Parents should spend time online with children.
- Computer should be kept in a common room in the house, not in the child's bedroom. It is more difficult for a predator to communicate with a child when the computer screen is visible to other members of the household.
- Parental controls and/or blocking software should be used.
- Parents should maintain access to the child's online account and randomly check his or her email. They should be open with children about parental access and state the reasons for it.
- Children should be instructed never to arrange a face-to-face meeting with someone they meet online; never to upload pictures of themselves onto the Internet to people they do not know; never to give out identifying information such as their name, address, school name, or telephone number; never to download pictures from an unknown source; and never to respond to messages that are suggestive, obscene, or harassing.

Abduction

Publicized crimes involving childhood abductions, although rare occurrences, frighten many parents and make them unsure about how best to protect their children. According to one study, in 57 percent of the cases, the victims of child-abduction murder were victims of opportunity. The tips noted below will help parents lessen the opportunity for abduction and kidnapping and better safeguard their children:

- Parents should teach children to run away from danger, never towards it. Danger is defined as anything or anyone that invades their personal space. Children should be taught to yell loudly, as their safety is more important than being polite.
- Children should not be allowed to go places alone, and they should always be supervised directly by parents or by another trusted adult. Older children should always take a friend along when they go somewhere.
- Parents should know where and with whom children are at all times. They should know children's friends and be clear about the places and homes they may visit. Children should habitually contact their parents when they arrive and leave a destination and if their plans change.
- Parents should talk openly with children about safety and encourage them to report to trusted adults anything or anyone makes them feel uncomfortable or frightened. Children should know they have the right to say no to any unwelcome, confusing, or uncomfortable attention by others and that they should tell parents immediately whenever such an experience occurs.
- Babysitters and caregivers should be screened and their references checked.
- Instead of confusing children with messages about avoiding strangers, Parents should identify adults to whom children may talk. Parents should list the people by name whom they permit their children to visit.
- Parents should avoid using code words but instead use the "check first" method. Children should be taught not to talk to anyone, go with anyone, or accept gifts or candy from anyone without first checking with their parents or trusted adults in charge.

Common problems

In spite of taking precautions and putting safety measures into place, accidents, injuries, and crime may still take place. All children should be taught how to call for help in an emergency. Instruct them to dial 911 when emergency assistance is needed and to remain on the phone as long as they are directed to do so.

Parental concerns

Children can injure themselves in the blink of an eye. Parents can turn their heads away for a moment, and a child could pull down a boiling pot of food or swallow something and choke on it. It is impossible for new parents to anticipate all the potential dangers or safety problems around babies and children. The trick to keeping an environment safe for children is to stay one step ahead of them at all times. By spotting dangers before an injury happens, parents can protect their children from harm and protect themselves from stress and heartache. As children develop, some of the potential dangers may change. What does not change is the responsibility parents have to provide a safe, trusting environment in which their children can thrive.

Safety rules for parents

Know where your children are at all times.

Be sensitive to changes in your child's behavior.

Talk with your child about their schoolwork and activities regularly.

Get to know your child's teachers, friends, and friends' families.

Listen sincerely to your children.

Be alert to a teenager or adult who is paying an unusual amount of attention to your child.

Make sure your children know what to do when approached by a stranger.

Don't put your child's name on clothing in a way that is visible to others.

Be aware of your child's time and activities online.

SOURCE: MetLife, "Protecting Your Child," http://www.metlife.com, 2003-5.

(Table by GGS Information Services.)

See also Childproofing.

Resources

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National Center for Injury Prevention and Control. Mailstop K65, 4770 Buford Highway NE, Atlanta, GA 30341–3724. Web site: <www.cdc.gov/ncipc/ncipchm.htm>.

National Center for Missing & Exploited Children. Charles B. Wang International Children's Building, 699 Prince Street, Alexandria, VA 22314–3175. Web site: <www.missingkids.org>

National Highway Traffic Safety Administration (NHTSA). 400 7th Street, SW, Washington, DC 20590. Web site: <www.nhtsa.gov>.

National SAFE KIDS Campaign. 1301 Pennsylvania Ave., NW, Suite 1000, Washington, DC 20004. Web site: <www.safekids.org>.

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Deanna M. Swartout-Corbeil, RN

KEY TERMS

Airway obstruction injury—An injury that obstructs the airway and prevents proper breathing, either through strangulation, suffocation, or choking.

Safety, infant and toddler *see* **Childproofing**

Salmonella food poisoning

Definition

Salmonella **food poisoning** is a bacterial infection that causes inflammation (swelling) of the lining of the stomach and intestines (**gastroenteritis**). The causative bacteria is called *Salmonella*. While domestic and wild animals, including poultry, pigs, cattle, and pets such as turtles, iguanas, chicks, dogs, and cats can transmit this illness, most people become infected by ingesting foods contaminated with significant amounts of the causative bacteria.

Description

Improperly handled or undercooked poultry and eggs are the foods which most frequently cause salmonella food poisoning. Chickens are a major carrier of salmonella bacteria, which accounts for its prominence in poultry products. However, identifying foods which may be contaminated with salmonella is particularly difficult

because infected chickens typically show no signs or symptoms. Since infected chickens have no identifying characteristics, these chickens go on to lay eggs or to be used as meat.

At one time, it was thought that salmonella bacteria were only found in eggs which had cracked, thus allowing the bacteria to enter. Ultimately, it was learned that, because the egg shell has tiny pores, even uncracked eggs which sat for a time on a surface (nest) contaminated with salmonella could themselves become contaminated. It is known also that the bacteria can be passed from the infected female chicken directly into the substance of the egg before the shell has formed around it.

Anyone may contract salmonella food poisoning, but the disease is most serious in infants, the elderly, and individuals with weakened immune systems. In these individuals, the infection may spread from the intestines to the blood stream and then to other body sites, causing death unless the person is treated promptly with **antibiotics**. In addition, people who have had part or all of their stomach or their spleen removed or who have **sickle cell anemia**, cirrhosis of the liver, leukemia, lymphoma, malaria, louse-borne relapsing fever, or acquired **Immunodeficiency** syndrome (**AIDS**) are particularly susceptible to salmonella food poisoning.

Demographics

Although salmonella food poisoning occurs worldwide, it is most frequently reported in North America and Europe. Only a small proportion of infected people are tested and diagnosed, and as few as 1 percent of cases are actually reported. While the infection rate may seem relatively low, even an attack rate of less than 0.5 percent in such a large number of exposures results in many infected individuals. The poisoning typically occurs in small, localized outbreaks in the general population or in large outbreaks in hospitals, restaurants, or institutions for children or the elderly. In the United States, salmonella is responsible for about 15 percent of all cases of food poisoning.

Causes and symptoms

Salmonella food poisoning can occur when someone drinks unpasteurized milk or eats undercooked chicken or eggs, or salad dressings or desserts which contain raw eggs. Even if salmonella-containing foods such as chicken are thoroughly cooked, any food can become contaminated during preparation if conditions and equipment for food preparation are unsanitary.

Other foods can then be accidentally contaminated if they come into contact with infected surfaces. In addition, children have become ill after playing with turtles or iguanas and then eating without washing their hands. Because the bacteria are shed in the feces for weeks after infection with salmonella, poor hygiene can allow such a carrier to spread the infection to others.

Symptoms appear about one to two days after infection and include fever (in 50% of patients), **nausea and vomiting**, **diarrhea**, and abdominal cramps and **pain**. The diarrhea is usually very liquid and rarely contains mucus or blood. Diarrhea usually lasts for about four days. The illness usually ends in about five to seven days.

Serious complications are rare, occurring most often in individuals with other medical illnesses. Complications occur when the salmonella bacteria make their way into the bloodstream (bacteremia). Once in the bloodstream, the bacteria can enter any organ system throughout the body, causing disease. Other infections which can be caused by salmonella include:

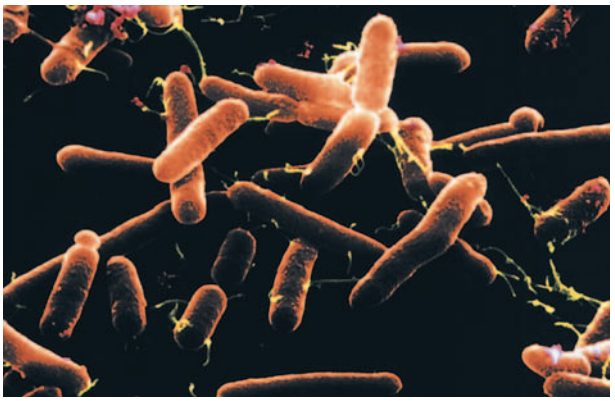
- bone infections (osteomyelitis)
- joint infections (arthritis)
- infection of the sac containing the heart (pericarditis)
- infection of the tissues which cover the brain and spinal cord (meningitis)
- infection of the liver (hepatitis)
- lung infections (pneumonia)
- infection of aneurysms (aneurysms are abnormal outpouchings which occur in weak areas of the walls of blood vessels)
- infections in the center of already-existing tumors or cysts

Diagnosis

Under appropriate laboratory conditions, salmonella can be grown and then viewed under a microscope for identification. Early in the infection, the blood is far more likely to positively show a presence of the salmonella bacterium when a sample is grown on a nutrient substance (culture) for identification purposes. Eventually, however, positive cultures can be obtained from the stool and in some cases from a urine culture.

Treatment

Even though salmonella food poisoning is a bacterial infection, most practitioners do not treat simple cases with antibiotics. Studies have shown that using antibio-



Exposure to the *Salmonella enteritidis* bacterium usually occurs by contact with contaminated food. *(Photograph by Oliver Meckes. Photo Researchers, Inc.)*

tics does not usually reduce the length of time that the patient is ill. Paradoxically, it appears that antibiotics do, however, cause the patient to shed bacteria in their feces for a longer period of time. In order to decrease the length of time that a particular individual is a carrier who can spread the disease, antibiotics are generally not given.

In situations where an individual has a more severe type of infection with salmonella bacteria, a number of antibiotics may be used. Chloramphenicol was the first antibiotic successfully used to treat salmonella food poisoning. It is still a drug of choice in developing countries because it is so inexpensive, although some resistance has developed to it. Ampicillin and trimethoprim-sulfonamide have been used successfully in the treatment of infections caused by chloramphenicol-resistant strains. Newer types of antibiotics, such as cephalosporin or quinolone, are also effective. These drugs can be given by mouth or through a needle in the vein (intravenously) for very ill patients. With effective antibiotic therapy, patients feel better in 24 to 48 hours, the temperature returns to normal in three to five days, and the patient is generally recovered by ten to 14 days.

Prognosis

The prognosis for uncomplicated cases of salmonella food poisoning is excellent. Most people recover completely within a week's time. In cases in which other medical problems complicate the illness, prognosis depends on the severity of the other medical conditions, as well as the specific organ system infected with salmonella.

KEY TERMS

Carrier—A person who possesses a gene for an abnormal trait without showing signs of the disorder. The person may pass the abnormal gene on to offspring. Also refers to a person who has a particular disease agent present within his/her body, and can pass this agent on to others, but who displays no symptoms of infection.

Gastroenteritis—Inflammation of the stomach and intestines that usually causes nausea, vomiting, diarrhea, abdominal pain, and cramps.

Prevention

Prevention of salmonella food poisoning involves the proper handling and cooking of foods likely to carry the bacteria. This means that recipes utilizing uncooked eggs (Caesar salad dressing, meringue toppings, mousses) need to be modified to eliminate the raw eggs. Not only should chicken be cooked thoroughly, until no pink juices flow, but all surfaces and utensils used on raw chicken must be carefully cleaned to prevent salmonella from contaminating other foods. Careful hand washing is a must before, during, and after all food preparation involving eggs and poultry. Hand washing is also important after handling and playing with pets such as turtles, iguanas, chicks, dogs and cats.

Parental concerns

Because children are notoriously bad at hand washing, parents want to be particularly vigilant to make sure that careful hand washing is followed, especially if someone in the home is actually ill with salmonella food poisoning. In this case, extra precautions should be taken. Children should not share foods, utensils, beverages, etc. Hand washing after toileting or diaper changes should be undertaken with extra care to avoid spreading the infection to others. The healthcare provider should give the **family** guidance regarding when a recovering child should return to school or daycare.

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Rosalyn Carson-DeWitt, MD

Sanfilippo's syndrome *see* **Mucopolysaccharidoses**

Sarcomas

Definition

A sarcoma is a cancerous (malignant) bone tumor.

Description

A primary bone tumor originates in or near a bone. Most primary bone tumors are benign, and the cells that compose them do not spread (metastasize) to nearby tissue or to other parts of the body.

A sarcoma is a type of malignant primary bone tumor. Malignant primary bone tumors account for less than 1 percent of all cancers diagnosed in the United States. They can infiltrate nearby tissues, enter the bloodstream, and metastasize to bones, tissues, and organs far from the original malignancy. Malignant primary bone tumors are characterized as either bone cancers which originate in the hard material of the bone or soft-tissue sarcomas which begin in blood vessels, nerves, or tissues containing muscles, fat, or fiber.

Types of bone tumors

Osteogenic sarcoma, or osteosarcoma, is the most common form of bone **cancer**, accounts for 6 percent of all instances of the disease, and for about 5 percent of all cancers that occur in children. Nine hundred new cases of osteosarcoma are diagnosed in the United States every year. The disease usually affects teenagers and is almost twice as common in boys as in girls.

Osteosarcomas, which grow very rapidly, can develop in any bone but most often occur along the edge or on the end of one of the fast-growing long bones that support the arms and legs. About 80 percent of all osteosarcomas develop in the parts of the upper and lower leg nearest the knee (the distal femur or in the proximal tibia). The next likely location for an osteosarcoma is the bone of the upper arm closest to the shoulder (the proximal humerus).

Ewing's sarcoma is the second most common form of childhood bone cancer. Accounting for fewer than 5 percent of bone tumors in children, Ewing's sarcoma usually begins in the soft tissue (the marrow) inside bones of the leg, hips, ribs, and arms. It rapidly infiltrates the lungs and may metastasize to bones in other parts of the body.

More than 80 percent of patients who have Ewing's sarcoma are white, and the disease most frequently affects children between the ages five and nine and young adults between ages 20 and 30. About 27 percent of all cases of Ewing's sarcoma occur in children under the age of ten, and 64 percent occur in adolescents between the ages of ten and 20.

Chondrosarcomas are cancerous bone tumors that most often appear in middle age. Usually originating in strong connective tissue (cartilage) in ribs or leg or hip bones, chondrosarcomas grow slowly. They rarely spread to the lungs. It takes years for a chondrosarcoma to metastasize to other parts of the body, and some of these tumors never spread.

Parosteal osteogenic sarcomas, fibrosarcomas, and chordomas are rare. Parosteal osteosarcomas generally involve both the bone and the membrane that covers it. Fibrosarcomas originate in the ends of the bones in the arm or leg and then spread to soft tissue. Chordomas develop on the skull or spinal cord.

Osteochondromas, which usually develop between the ages of ten and 20, are the most common noncancerous primary bone tumors. Giant cell tumors generally develop in a section of the thigh bone near the knee. Giant cell tumors are originally benign but sometimes become malignant.

Causes and symptoms

The cause of bone cancer is unknown, but the tendency to develop it may be inherited. Children who have bone tumors are often tall for their age, and the disease seems to be associated with growth spurts that occur during childhood and **adolescence**. Injuries can make the presence of tumors more apparent but do not cause them.

A bone that has been broken or exposed to high doses of radiation used to treat other cancers is more likely than other bones to develop osteosarcoma. A history of noncancerous bone disease also increases bone-cancer risk.

The amount of radiation in diagnostic x rays poses little or no danger of bone-cancer development, but children who have a **family** history of the most common childhood cancer of the eye (**retinoblastoma**) or who have inherited rare cancer syndromes have a greater-than-average risk of developing bone cancer. Exposure to chemicals found in some paints and dyes can slightly raise the risk.

Both benign and malignant bone tumors can distort and weaken bone and cause **pain**, but benign tumors are generally painless and asymptomatic.

It is sometimes possible to feel a lump or mass, but pain in the affected area is the most common early symptom of bone cancer. Pain is not constant in the initial stages of the disease, but it is aggravated by activity and may be worse at night. If the tumor is located on a leg bone, the patient may limp. Swelling and weakness of the limb may not be noticed until weeks after the pain begins.

Other symptoms of bone cancer include:

- a bone that breaks for no apparent reason
- difficulty moving the affected part of the body
- fatigue
- fever
- a lump on the trunk, an arm or leg, or another bone
- persistent, unexplained back pain
- weight loss

Diagnosis

Physical examination and routine x rays may yield enough evidence to diagnose benign bone tumors, but removal of tumor tissue for microscopic analysis (biopsy) is the only sure way to rule out malignancy.

A needle biopsy involves using a fine, thin needle to remove small bits of tumor, or a thick needle to extract tissue samples from the innermost part (the core) of the growth. An excisional biopsy is the surgical removal of a small, accessible tumor. An incisional biopsy is performed on tumors too large or inaccessible to be completely removed. The surgeon performing an incisional biopsy cuts into the patient's skin and removes a portion of the exposed tumor. Performed under local or general anesthetic, biopsy reveals whether a tumor is benign or malignant and identifies the type of cancer cells the malignant tumor contains.

Bone cancer is usually diagnosed about three months after symptoms first appear, and 20 percent of malignant tumors have metastasized to the lungs or other parts of the body by that time.

Imaging techniques

The following procedures are used, in conjunction with biopsy, to diagnose bone cancer:

- Bone x rays usually provide a clear image of osteosarcomas.
- Computerized axial tomography (CAT scan), a specialized x ray that uses a rotating beam to obtain detailed information about an abnormality and its physical relationship to other parts of the body, can differentiate between osteosarcomas and other types of bone tumors, illustrate how tumor cells have infiltrated other tissues, and help surgeons decide which portion of a growth would be best to biopsy. Because more than four of every five malignant bone tumors metastasize to the lungs, a CAT scan of the chest is performed to see if these organs have been affected. Chest and abdominal CAT scans are used to determine whether Ewing's sarcoma has spread to the lungs, liver, or lymph nodes.
- Magnetic resonance imaging (MRI), a specialized scan that relies on radio waves and powerful magnets to reflect energy patterns created by tissue abnormalities and specific diseases, provides more detailed information than does a CAT scan about tumors and marrow cavities of the bone and can sometimes detect clusters of cancerous cells that have separated from the original tumor. This valuable information helps surgeons select the most appropriate approach for treatment.
- Radionuclide bone scans involve injecting a small amount of radioactive material into a vein. Primary tumors or cells that have metastasized absorb the radioactive material and show up as dark spots on the scan.

Cytogenic and molecular genetic studies, which assess the structure and composition of chromosomes and genes, may also be used to diagnose osteosarcoma. These tests can sometimes indicate what form of treatment is most appropriate.

Laboratory studies

A complete blood count (CBC) reveals abnormalities in the blood and may indicate whether bone marrow has been affected. A blood test that measures levels of the enzyme lactate dehydrogenase (LDH) can help predict the likelihood of a specific patient's survival.

Immunohistochemistry involves adding special antibodies and chemicals or stains to tumor samples. This technique is effective in identifying cells that are found

in Ewing's sarcoma but are not present in other malignant tumors.

Reverse transcription polymerase chain reaction (RTPCR) relies on chemical analysis of the substance in the body that transmits genetic information (RNA) to evaluate the effectiveness of cancer therapies, identify mutations consistent with the presence of Ewing's sarcoma, and reveal cancer that recurs after treatment has been completed.

Staging

Once bone cancer has been diagnosed, the tumor is staged. This process indicates how far the tumor has spread from its original location. The stage of a tumor suggests which form of treatment is most appropriate and predicts how the condition will probably respond to therapy.

An osteosarcoma may be localized or metastatic. A localized osteosarcoma has not spread beyond the bone where it arose or beyond nearby muscles, tendons, and other tissues. A metastatic osteosarcoma has spread to the lungs, to bones not directly connected to the bone in which the tumor originated, or to other tissues or organs.

Treatment

In the 1960s, amputation was the only treatment for bone cancer. Between then and the early 2000s **chemotherapy** drugs and innovative surgical techniques improved survival with intact limbs. Because osteosarcoma is so rare, patients should consider undergoing treatment at a major cancer center staffed by specialists familiar with the disease.

A treatment plan for bone cancer, developed after the tumor has been diagnosed and staged, may include the following:

- Amputation may be the only therapeutic option for large tumors involving nerves or blood vessels that have not responded to chemotherapy. MRI scans indicate how much of the diseased limb must be removed, and surgery is planned to create a cuff, formed of muscles and skin, around the amputated bone. Following surgery, an artificial (prosthetic) leg is fitted over the cuff. A patient who actively participates in the rehabilitation process may be walking independently as soon as three months after the amputation.
- Chemotherapy is usually administered in addition to surgery, to kill cancer cells that have separated from the original tumor and spread to other parts of the body. Although chemotherapy can increase the likelihood of later development of another form of cancer, the American Cancer Society maintains that the need for chemotherapeutic bone-cancer treatment is much greater than the potential risk.
- Surgery, coordinated with diagnostic biopsy, enhances the probability that limb-salvage surgery can be used to remove the cancer while preserving nearby blood vessels and bones. A metal rod or bone graft is used to replace the area of bone removed, and subsequent surgery may be needed to repair or replace rods that become loose or break. Patients who have undergone limb-salvage surgery need intensive rehabilitation. It may take as long as a year for a patient to regain full use of a leg following limb-salvage surgery, and patients who have this operation may eventually have to undergo amputation.
- Radiation therapy is used often to treat Ewing's sarcoma.
- Rotationoplasty, sometimes performed after a leg amputation, involves attaching the lower leg and foot to the thigh bone, so that the ankle replaces the knee. A prosthetic is later added to make the leg as long as it should be. Prosthetic devices are not used to lengthen limbs that remain functional after amputation to remove osteosarcomas located on the upper arm. When an osteosarcoma develops in the jawbone, the entire lower jaw is removed. Bones from other parts of the body are later grafted on remaining bone to create a new jaw.

Follow-up treatments

After a patient completes the final course of chemotherapy, CAT scans, bone scans, x rays, and other diagnostic tests may be repeated to determine if any traces of tumor remain. If none is found, treatment is discontinued, but patients are advised to see their oncologist and orthopedic surgeon every two or three months for the following year. X rays of the chest and affected bone are taken every four months. An annual echocardiogram is recommended to evaluate any adverse effect chemotherapy may have had on the heart, and CT scans are performed every six months.

Patients who have received treatment for Ewing's sarcoma are examined often—at gradually lengthening intervals—after completing therapy. Accurate growth measurements are taken during each visit and blood is drawn to be tested for side effects of treatment. X rays, CT scans, bone scans, and other imaging studies are generally performed every three months during the first year. If no evidence of tumor growth or recurrence is indicated, these tests are performed less frequently in the following years.

A specimen of a femur bone indicating the cancerous growth around the knee. Osteosarcoma is the most common primary cancer of the bone. *(Photo Researchers, Inc.)*

Some benign bone tumors shrink or disappear without treatment. However, regular examinations are recommended to determine whether these tumors have changed in any way.

Prognosis

Benign brain tumors rarely recur, but sarcomas can reappear after treatment was believed to have eliminated every cell.

Likelihood of long-term survival depends on the type and location of the tumor, how much the tumor has metastasized, and on what organs, bones, or tissues have been affected.

More than 85 percent of patients survive for more than five years after complete surgical removal of low-grade osteosarcomas (tumors that arise in mature tissue and contain a small number of cancerous cells). About 25 to 30 percent of patients diagnosed with high-grade osteosarcomas (tumors that develop in immature tissue and contain a large number of cancer cells) die of the disease.

Two-thirds of all children diagnosed with Ewing's sarcoma live for more than five years after the disease is detected. The outlook is most favorable for children under the age of ten, and least favorable in patients whose cancer is not diagnosed until after it has metastasized: fewer than three of every ten of these patients remain alive five years later. More than 80 percent of patients whose Ewing's sarcoma is confined to a small area and surgically removed live, for at least five years. Postsurgical radiation and chemotherapy add years to their lives. More than 70 percent of patients live five years or more with a small Ewing's sarcoma that cannot be removed, but only three out of five patients with large, unremovable tumors survive that long.

Prevention

There is no known way to prevent bone cancer.

Parental concerns

Careful attention to a child's diet can be very helpful for patients with cancer. This can be difficult when the cancer and/or the treatments are affecting the appetite, however. Whole foods, including grains, beans, fresh fruits and vegetables, and high quality fats, should be emphasized in the diet, while processed foods should be

avoided. Increased consumption of fish, especially cold-water fish such as salmon, mackerel, halibut, and tuna, provides a good source of omega-3 fatty acids. Nutritional supplements can build strength and help maintain it during and following chemotherapy, radiation, or surgery.

Guided imagery and relaxation techniques can be helpful for children undergoing difficult treatments. Support groups for the child and the family can be very helpful and can give provide an important emotional outlet for the child, the parents, and the siblings.

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Savant syndrome

Definition

Savant syndrome occurs when a person with below normal **intelligence** displays a special talent or ability in a specific area.

Description

Children who display savant syndrome have traditionally been referred to as idiot, retarded, or autistic savants. The negative connotations of the term "idiot" have led to the disuse of idiot savant. Because the syndrome is often associated with **autism**, the term autistic savant is more frequently heard. The first known description of a person displaying savant syndrome occurred in a German psychology journal in 1751. The term savant was first used in 1887 by J. Langdon Down (the doctor for whom **Down syndrome** is named).

Demographics

About half of all children with savant syndrome are autistic. Approximately 10 percent of all children with autism have savant syndrome. The rate increases to 25 percent of children with autism who have an IQ over 35. (Many autistic children have lower IQs.) About three times as many boys as girls have savant syndrome. This may be because more boys than girls are affected with autism. Less than 1 percent of the non-autistic population, including those with **mental retardation** and other developmental disorders, have savant syndrome.

Causes and symptoms

The causes of savant syndrome were as of 2004 not known. Some researchers hypothesize that it is caused by a change in a gene or genes, and others believe that it is caused by some kind of damage to the left hemisphere of the brain with compensation for this injury occurring in the right hemisphere. The reasons for the syndrome are not at all clear, however, and more research needs to be done.

Children with savant syndrome have an exceptional talent or skill in a particular area, such as the ability to process mathematical calculations at a phenomenal speed. Savant skills occur in a number of different areas, including music, visual arts, and mathematics. Experts believe that the most common skill demonstrated by savants is extraordinary memory. Children with savant syndrome may be able to memorize extensive amounts of data in such areas as **sports** statistics, population figures, and historical or biographical data. One particular skill common to those with savant syndrome is the ability to calculate what day of the week a particular date fell on or will fall on.

Diagnosis

Savant syndrome is diagnosed when a child's ability in one area is exceptionally higher than would be expected given his or her IQ or general level of functioning.

KEY TERMS

Autism—A developmental disability that appears early in life, in which normal brain development is disrupted and social and communication skills are retarded, sometimes severely.

Treatment

Savant syndrome is not known to have any drawbacks, so it does not have to be treated itself. The underlying disorders that usually accompany savant syndrome need to be treated, and it is believed that making use of the special talent of the child with savant syndrome may help treat the child's underlying developmental disorders.

Prognosis

The special skill associated with savant syndrome in a specific child is usually present for life. There has been at least one report of the skill being lost when progress was gained in other areas, but this appears to be very rare. In general, if the level of the skill changes it improves as the skill is practiced.

Prevention

There is no known way to prevent savant syndrome.

Parental concerns

Children with savant syndrome have a very special skill that can be nurtured. These children may respond better to treatments for any underlying disorder that make use in some way of the childs special underlying interest and talent.

See also Autism.

Resources

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Tish Davidson, A.M.

Scabies

Definition

Scabies is a relatively contagious infection caused by a tiny mite called *Sarcoptes scabiei*.

Description

Scabies is caused by a tiny insect about 0.3 mm long called a mite. When a human comes in contact with the female mite, the mite burrows under the skin, laying eggs along the line of its burrow. These eggs hatch, and the resulting offspring rise to the surface of the skin, mate, and repeat the cycle either within the skin of the original host or within the skin of its next victim.

The intense **itching** almost always caused by scabies is due to a reaction within the skin to the feces of the mite. The first time someone is infected with scabies, he or she may not notice any itching for a number of weeks (four to six weeks). With subsequent infections, the itchiness begins within hours of picking up the first mite.

Demographics

Prevalence rates are not clear; some studies suggest that between 6 and 27 percent of the population have scabies at any one time. Scabies is more common among schoolchildren and individuals living in crowded conditions.

Causes and symptoms

Scabies is most common among people who live in overcrowded conditions and whose ability to practice good hygiene is limited. Scabies can be passed between people by close skin contact. Although the mites can only live away from human skin for about three days, sharing clothing or bedclothes can pass scabies among **family** members or close contacts. In May 2002, the Centers for Disease Control (CDC) included scabies in

its updated guidelines for the treatment of **sexually transmitted diseases**.

The itching (pruritus) from scabies is worse after a hot shower and at night. Burrows are seen as winding, slightly raised gray lines along the skin. The female mite may be seen at one end of the burrow, as a tiny pearl-like bump underneath the skin. Because of the intense itching, burrows may be obscured by scratch marks left by the patient. The most common locations for burrows are the sides of the fingers, between the fingers, the top of the wrists, around the elbows and armpits, around the nipples of the breasts in women, in the genitalia of men, around the waist (beltline), and on the lower part of the buttocks. Babies may have burrows on the soles of their feet, palms of their hands, and faces.

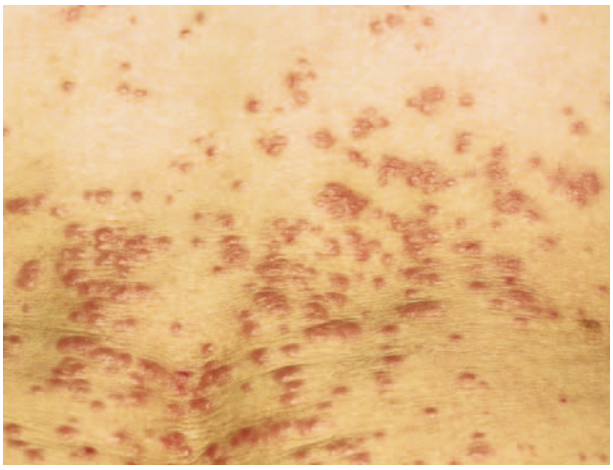
Scratching seems to serve some purpose in scabies, as the mites are apparently often inadvertently removed. Most infestations with scabies are caused by no more than 15 mites altogether.

Infestation with huge numbers of mites (on the order of thousands to millions) occurs when an individual does not scratch or when an individual has a weakened immune system. These patients include the elderly; those who live in institutions; the mentally retarded or physically infirm; those who have other diseases which affect the amount of sensation they have in their skin (leprosy or syringomyelia); leukemia or diabetes sufferers; those taking medications which lower their immune response (**cancer chemotherapy** or immunosuppressant drugs given after organ transplantation); or people with other diseases which lower their immune response (such as acquired **immunodeficiency** syndrome or **AIDS**). This form of scabies, with its major infestation, is referred to as crusted scabies or Norwegian scabies. Infected patients have thickened, crusty areas all over their bodies, including over the scalp. Their skin is scaly. Their fingernails may be thickened and horny.

Diagnosis

Diagnosis can be made simply by observing the characteristic burrows of the mites causing scabies. A sterilized needle can be used to explore the pearly bump at the end of a burrow, remove its contents, and place it on a slide to be examined. The mite itself may then be identified under a microscope.

Occasionally, a type of mite carried on dogs (*Sarcoptes scabiei var. canis*) may infect humans. These mites cannot survive for very long on humans, and so the infection is very light.



Close-up view of a scabies skin infection. *(© Dr. P Marazzi/ Photo Researchers, Inc.)*

Treatment

Several types of lotions (usually containing 5% permethrin) can be applied to the body and left on for 12 to 24 hours. One topical application is usually sufficient, although the scabicide may be reapplied after a week if mites remain. Preparations containing lindane are no longer recommended for treating scabies because of the potential for damage to the nervous system. Itching can be lessened by the use of calamine lotion or antihistamine medications.

In addition to topical medications, the doctor may prescribe oral ivermectin, a drug that was originally developed for veterinary practice as a broad-spectrum antiparasite agent. Studies done in humans, however, have found that ivermectin is as safe and effective as topical medications for treating scabies. A study published in 2003 reported that ivermectin is safe for people in high-risk categories, including those with compromised immune systems.

Prognosis

The prognosis for complete recovery from scabies infestation is excellent. In patients with weak immune systems, the biggest danger is that the areas of skin involved with scabies will become secondarily infected with bacteria.

Prevention

Good hygiene is essential in the prevention of scabies. When a member of a household is diagnosed with

KEY TERMS

Mite—An insect parasite belonging to the order Acarina. The organism that causes scabies is a mite.

Pruritus—The symptom of itching or an uncontrollable sensation leading to the urge to scratch.

Topical—Not ingested; applied to the outside of the body, for example to the skin, eye, or mouth.

scabies, all that person's recently worn clothing and bedding should be washed in very hot water.

Parental concerns

One of the biggest concerns among family members of an individual with scabies is its ready transmissibility. Care should be taken to avoid sharing bedding, towels, and clothing with an infected family member. Some healthcare providers recommend that all family members be treated with a scabicide, whether or not scabies is evident. Linens of all family members should be washed in the hottest water possible to avoid cross-contamination.

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Scarlatina *see* **Scarlet fever**

Scarlet fever

Definition

Scarlet fever is a rash that complicates a bacterial throat infection called **strep throat**.

Description

Scarlet fever, also known as scarlatina, gets its name from the fact that the patient's skin, especially on the cheeks, is flushed. The disease primarily affects children. A **sore throat** and a raised, sandpaper-like rash over much of the body are accompanied by fever and sluggishness (lethargy). The fever usually subsides within a few days, and recovery is complete by two weeks. After the fever is gone, the skin on the face and body flakes; the skin on the palms of the hands and soles of the feet peels more dramatically. Treatment for scarlet fever is intended to offset the possibility of serious complications such as **rheumatic fever** (a heart disease) or kidney inflammation (glomerulonephritis) can develop.

Scarlet fever is highly contagious and is spread by sneezing, coughing, or direct contact. The incubation period is three to five days, with symptoms usually beginning on the second day of the disease and lasting from four to ten days.

Early in the twentieth century, severe scarlet fever epidemics were common. In the early 2000s, the disease is rare. **Antibiotics** have helped, and it is possible that the strain of bacteria that causes scarlet fever has become weaker with time.

Demographics

Scarlet fever primarily affects children between the ages of five and 15 years. Approximately 10 percent of all children who have strep throat develop the characteristic scarlet fever rash.

Causes and symptoms

Scarlet fever is caused by group A streptococcal bacteria (*S. pyogenes*), highly toxic microbes that can also cause strep throat, wound or skin infections, **pneumonia**, and serious kidney infections. The group A streptococci are hemolytic bacteria, which means that they have the ability to break red blood cells. The strain of streptococcus that causes scarlet fever, unlike the one that causes most strep throats, produces an erythrogenic toxin, which causes the skin to flush.

The main symptoms and signs of scarlet fever are fever, lethargy, sore throat, and a bumpy rash that blanches under pressure. The rash appears first on the upper chest and spreads to the neck, abdomen, legs, arms, and in folds of skin such as under the arm or groin. In scarlet fever, the skin around the mouth tends to be pale, while the cheeks are flushed. The patient usually has a "strawberry tongue," in which inflamed bumps on the tongue rise above a bright red coating. Finally, dark red lines (called Pastia's lines) may appear in the creases of skin folds.

Diagnosis

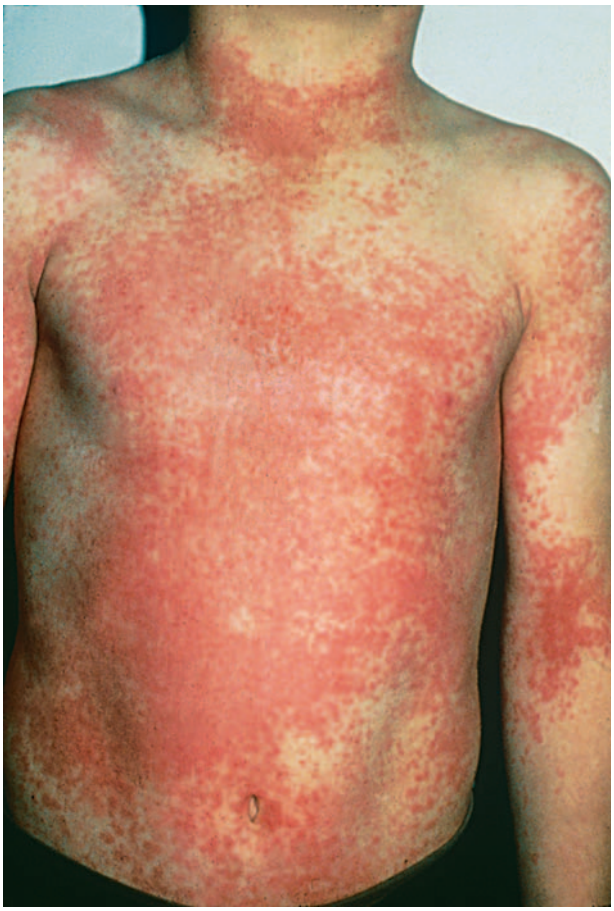
Cases of scarlet fever are usually diagnosed and treated by pediatricians or **family** medicine practitioners. The chief diagnostic signs of scarlet fever are the characteristic rash, which spares the palms and soles of the feet, and the presence of a strawberry tongue in children. Strawberry tongue is rarely seen in adults.

The doctor will take note of the signs and symptoms to eliminate the possibility of other diseases. For example, scarlet fever can be distinguished from **measles**, a viral infection that is also associated with a fever and rash, by the quality of the rash, the presence of a sore throat in scarlet fever, and the absence of the severe eye inflammation and severe runny nose that usually accompany measles.

Treatment

Although scarlet fever often clears up spontaneously within a few days, antibiotic treatment with either oral or injectable penicillin is usually recommended to reduce the severity of symptoms, prevent complications, and prevent spread to others. Antibiotic treatment shortens the course of the illness in small children but may not do so in adolescents or adults. Nevertheless, treatment with antibiotics is important to prevent complications.

One benzathine penicillin injection is required for treatment. But since penicillin injections are painful, oral penicillin may be preferable. If the patient is unable to tolerate penicillin, alternative antibiotics such as erythromycin or clindamycin may be used. However, the entire course of antibiotics, usually ten days, needs to be followed for the therapy to be effective. Because symptoms subside quickly, there is a temptation to stop therapy prematurely. It is important to take all of the pills in order to kill the bacteria. Not completing the course of therapy increases the risk of developing rheumatic fever and kidney inflammation.



Scarlet fever is characterized by a sandpaper-like rash on reddened skin. *(© Biophoto Associates/Photo Researchers, Inc.)*

Bed rest is not necessary, nor is isolation of the patient. **Acetaminophen** may be given for fever or relief of **pain**.

Prognosis

If treated promptly with antibiotics, full recovery is expected. Once a patient has had scarlet fever, the person develops immunity and cannot develop it again.

Prevention

Avoiding exposure to children who have the disease helps prevent the spread of scarlet fever.

Parental concerns

The most important thing to do for children with scarlet fever is to carefully and completely follow the

healthcare provider's instructions for administering a course of antibiotics.

See also Strep throat.

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Scheie syndrome *see* **Mucopolysaccharidoses**

Schizophrenia

Definition

Schizophrenia is a mental illness characterized by disordered thinking, delusions, hallucinations, emotional disturbance, and withdrawal from reality.

Description

Some experts view schizophrenia as a group of related illnesses with similar characteristics. Although the term, coined in 1911 by Swiss psychologist Eugene Bleuler (1857–1939), is associated with the idea of a "split" mind, the disorder is different from a "split personality" (dissociative identity disorder), with which it is frequently confused. In the United States, schizophrenics occupy more hospital beds than patients suffering from **cancer**, heart disease, or diabetes. At any given time, they account for up to half the beds in long-term care facilities and 40 percent of the treatment days.

Demographics

The incidence of childhood schizophrenia is thought to be one in 10,000 births. In comparison, the incidence among adolescents and adults is approximately one in 100. The condition occurs with equal frequency in males and females (although the onset of symptoms is usually earlier in males). At least 2.5 million Americans are thought to be afflicted with schizophrenia, with an estimated 100,000 to 200,000 new cases every year. Schizophrenia is commonly thought to disproportionately affect people in the lowest socioeconomic groups, although some people claim that socially disadvantaged persons with schizophrenia are only more visible than their more privileged counterparts, not more numerous.

Causes and symptoms

While the exact cause of schizophrenia is not known, it is believed to be caused by a combination of physiological and environmental factors. Studies have shown that there is clearly a hereditary component to the disorder. **Family** members of schizophrenics are ten times more prone to schizophrenia than the general population, and identical **twins** of schizophrenics have a 46 percent likelihood of having the illness themselves. Relatives of schizophrenics also have a higher incidence of other milder psychological disorders with some of the same symptoms as schizophrenia, such as suspicion, communication problems, and eccentric behavior.

In the years following World War II (1939–45), many doctors blamed schizophrenia on bad parenting. In the latter twentieth century, however, advanced neurological research strengthened the case for a physiological basis for the disease. It has been discovered that the brains of schizophrenics have certain features in common, including smaller volume, reduced blood flow to certain areas, and enlargement of the ventricles (cavities filled with fluid that are found at the brain's center). Much attention has focused on the connection between schizophrenia and neurotransmitters, the chemicals that transmit nerve impulses within the brain. One such chemical, dopamine, has been found to play an especially important role in the disease. Additional research has concentrated on how and when the brain abnormalities that characterize the disorder develop. Some are believed to originate prenatally for a variety of reasons, such as trauma, viral infections, **malnutrition** during pregnancy, or Rh sensitivity (a reaction caused when the mother lacks a certain blood protein called Rh that the baby has). Environmental factors associated with schizophrenia include birth complications, viral infections during infancy, and head injuries in childhood. While the notion of child-rearing practices causing schizophrenia has

been largely discredited, there is evidence that certain family dynamics do contribute to the likelihood of relapse in persons who already have shown symptoms of the disease.

Researchers have found correlations between childhood behavior and the onset of schizophrenia in adulthood. A 30-year longitudinal research project studied over 4,000 people born within a single week in 1946 in order to document any unusual developmental patterns observed in those children who later became schizophrenic. It was found that a disproportionate number of them learned to sit, stand, and walk late. They were also twice as likely as their peers to have **speech disorders** at the age of six and to have played alone when they were young. Home movies have enabled other researchers to collect information about the childhood characteristics of adult schizophrenics. One study found that the routine physical movements of these children tended to be slightly abnormal in ways that most parents would not suspect were associated with a major mental illness and that the children also tended to show **fear** and anger to an unusual degree.

The initial symptoms of schizophrenia usually occur between the ages of 16 and 30, with some variation depending on the type. Disorganized schizophrenia tends to begin early, usually in **adolescence** or young adulthood, while paranoid schizophrenia tends to start later, usually after the age of 25 or 30. The onset of schizophrenia before the age of 13 is rare and is associated with more serious symptoms. The onset of acute symptoms is referred to as the first psychotic break or break from reality. In general, the earlier the onset of symptoms, the more severe the illness is. Before the disease becomes full-blown, schizophrenics may go through a period called the prodromal stage, lasting about a year, when they experience behavioral changes that precede and are less dramatic than those of the acute stage. These may include social withdrawal, trouble concentrating or sleeping, neglect of personal grooming and hygiene, and eccentric behavior.

The prodromal stage is followed by the acute phase of the disease, which usually requires medical intervention. During this stage, three-fourths of schizophrenics experience delusions, illogical and bizarre beliefs that are held despite objections. An example of a delusion is the belief that the afflicted person is under the control of a sinister force located in the sewer system that dictates his every move and thought. Hallucinations are another common symptom of acute schizophrenia. These may be auditory (hearing voices) or tactile (feeling as though worms are crawling over one's skin). The acute phase of schizophrenia is also characterized by incoherent thinking, rambling or discontinuous speech, use of nonsense words, and odd physical behavior, including grimacing, pacing, and unusual postures. Persons in the grip of acute schizophrenia may also become violent, although often this violence is directed at themselves: it is estimated that 15 to 20 percent of schizophrenics commit **suicide** out of despair over their condition or because the voices they hear "tell" them to do so, and up to 35 percent attempt to take their own lives or seriously consider doing so. In addition, about 25 to 50 percent of people with schizophrenia abuse drugs or alcohol. As the positive symptoms of the acute phase subside, they may give way to what is called residual schizophrenia. Symptoms include flat or inappropriate emotions, an inability to experience pleasure (anhedonia), lack of motivation, reduced attention span, lack of interest in one's surroundings, and social withdrawal.

When to call the doctor

Parents should contact a healthcare professional if their child begins to have auditory or visual hallucinations, has a sudden change in behavior, shows signs of suicide ideation, or exhibits other symptoms of schizophrenia.

Diagnosis

Schizophrenia is generally divided into four types. The most prevalent, found in some 40 percent of affected persons, is paranoid schizophrenia, characterized by delusions and hallucinations centering on persecution, and by feelings of jealousy and grandiosity. Other possible symptoms include argumentativeness, anger, and violence. Catatonic schizophrenia is known primarily for its catatonic state, in which persons retain fixed and sometimes bizarre positions for extended periods of time without moving or speaking. Catatonic schizophrenics may also experience periods of restless movement. In disorganized (hebephrenic) schizophrenia, the patient is incoherent, with flat or inappropriate emotions, disorganized behavior, and bizarre, stereotyped movements and grimaces. Catatonic and disorganized schizophrenia affect far fewer people than paranoid schizophrenia. Most schizophrenics not diagnosed as paranoid schizophrenics fall into the large category of undifferentiated schizophrenia (the fourth type), which consists of variations of the disorder that do not correspond to the criteria of the other three types. Generally, symptoms of any type of schizophrenia must be present for six months before a diagnosis can be made.

Childhood schizophrenia has been known to appear as early as five years of age. Occurring primarily in males, it is characterized by the same symptoms as adult

schizophrenia. Diagnosis of schizophrenia in children can be difficult because delusions and hallucinations may be mistaken for childhood fantasies. Other signs of schizophrenia in children include moodiness, problems relating to others, attention difficulties, and difficulty dealing with change. In many cases, children are improperly diagnosed with the disease; one study found as many as 95 percent of children initially diagnosed with childhood-onset schizophrenia did not meet the diagnostic criteria.

It is important for schizophrenia to be diagnosed as early as possible. The longer the symptoms last, the less well afflicted individuals respond to treatment.

Treatment

Even when treated, schizophrenia interferes with normal development in children and adolescents and makes new learning difficult.

Schizophrenia has historically been very difficult to treat, usually requiring **hospitalization** during its acute stage. In the late 1900s, antipsychotic drugs became the most important component of treatment. These can control delusions and hallucinations, improve thought coherence, and, if taken on a long-term maintenance basis, prevent relapses. However, antipsychotic drugs do not work for all schizophrenics, and their use has been complicated by side effects, such as akathisia (motor restlessness), dystonia (rigidity of the neck muscles), and tardive dyskinesia (uncontrollable repeated movements of the tongue and the muscles of the face and neck). In addition, many schizophrenics resist taking medication, some because of the side effects, others because they may feel better and mistakenly decide they do not need the drugs anymore, or because being dependent on medication in order to function makes them feel bad about themselves. The tendency of schizophrenics to discontinue medication is very harmful. Each time a schizophrenic goes off medication, the symptoms of the disease return with greater severity, and the effectiveness of the drugs is reduced.

Low doses of antipsychotic medication have been used successfully with children and adolescents, especially when administered shortly after the onset of symptoms. Their rate of effectiveness in children between the ages of five and 12 has been found to be as high as 80 percent. Until about 1990, the drugs most often prescribed for schizophrenia were neuroleptics such as Haldol, Prolixin, Thorazine, and Mellaril. A major breakthrough in the treatment of schizophrenia occurred in 1990 with the introduction of the drug clozapine to the U.S. market. Clozapine, which affects the neurotransmitters in the brain (specifically serotonin and dopamine), has been dramatically successful in relieving symptoms of schizophrenia, especially in patients in whom other medications have not been effective. However, even clozapine does not work for all patients. In addition, about 1 percent of those who take it develop agranulocytosis, a potentially fatal blood disease, within the first year of use, and all patients on clozapine must be monitored regularly for this side effect. (Clozapine was first developed in the mid twentieth century but could not be introduced until it became possible to screen for this disorder.) The screening itself is expensive, creating another problem for those using the drug. Risperidone, a subsequent and safer medication that offers benefits similar to those of clozapine, was introduced in 1994 and is as of the early 2000s the most frequently prescribed antipsychotic medication in the United States. Olanzapine, another in the subsequent generation of schizophrenia drugs, received FDA approval in the fall of 1996, and more medications are under development. Electroconvulsive therapy (ECT, also called electric shock treatments) has been utilized to relieve symptoms of catatonia and depression in schizophrenics, especially in cases where medication is not effective.

Although medication is an important part of treatment, psychotherapy can also play an important role in helping schizophrenics manage **anxiety** and deal with interpersonal relationships, and treatment for the disorder usually consists of a combination of medication, therapy, and various types of rehabilitation. **Family therapy** has worked well for many patients, educating both patients and their families about the nature of schizophrenia and helping them in their cooperative effort to cope with the disorder.

Alternative treatment

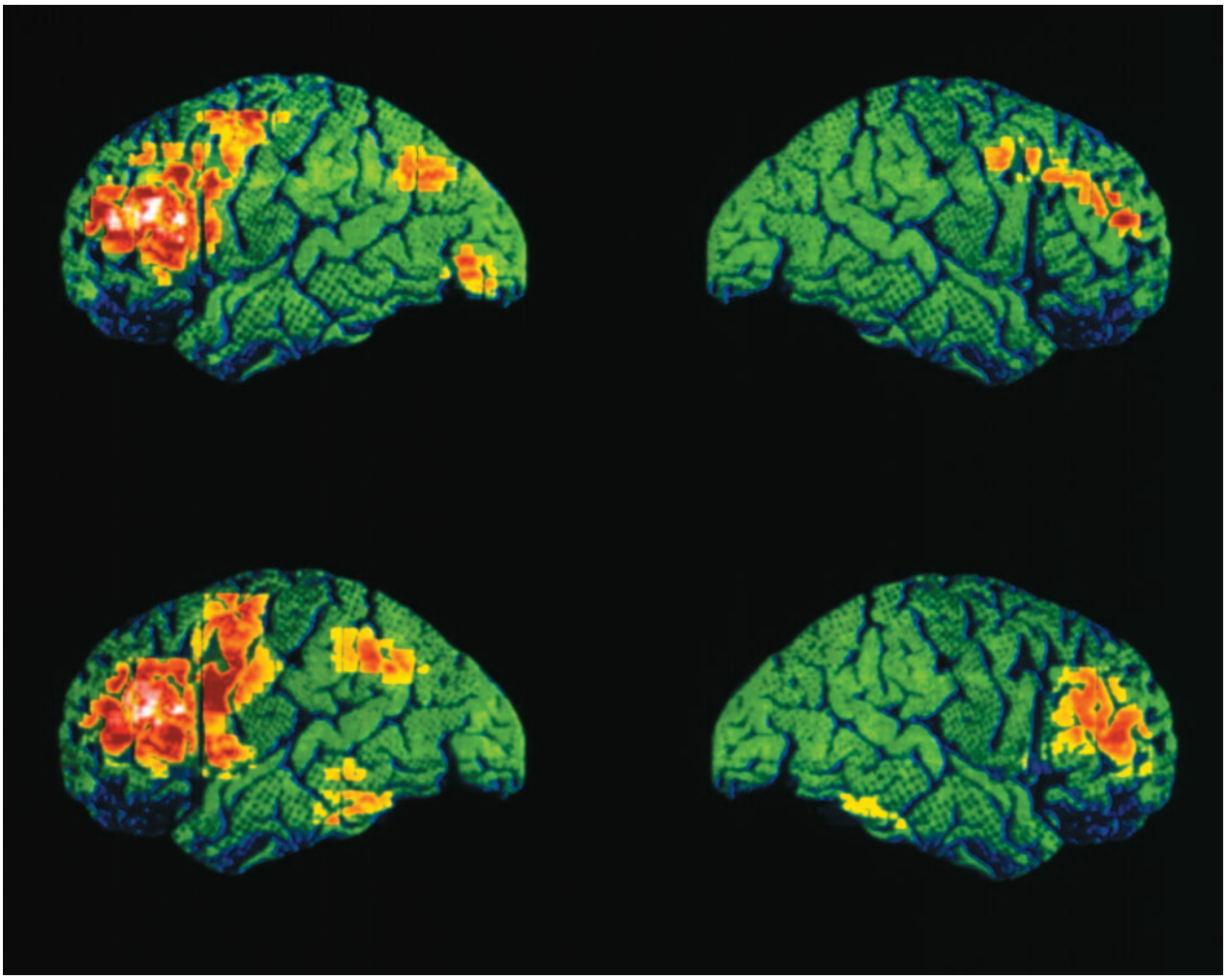
Some of the alternative treatments that have been used with varying success to treat children with schizophrenia include biofeedback, acupressure, chiropractic work, massage, and herbal drops.

Nutritional concerns

Some families have reported a benefit to making adjustments to or supplementing the diet of a child with schizophrenia, including reducing the amount of processed sugar consumed and supplementing with **vitamins** and **minerals** such as copper, zinc, **folic acid**, etc.

Prognosis

With the aid of antipsychotic medication to control delusions and hallucinations, about 70 percent of schizo-



Colored positron emission tomography (PET) brain scans of a schizophrenic, bottom, and normal patient, top. *(© Wellcome Dept. of Cognitive Neurology/Science Photo Library. Photo Researchers, Inc.)*

phrenics are able to function in society. Over the long term, about one-third of patients experience recovery or remission. Children afflicted with schizophrenia have a poorer prognosis than that of adults.

Prevention

There is no proven way to prevent onset of schizophrenia. Researchers have investigated the possibility of treating schizophrenia during the prodromal stage or even before symptoms start (such as when the likelihood of hereditary transmission is high). Other areas of research include the links between schizophrenia and family stress, drug use, and exposure to certain infectious agents.

Parental concerns

Parents play a key role in the everyday treatment and management of schizophrenia. The affected child should be closely monitored to ensure he or she is taking all prescribed medications. Working with the child's school teachers to formulate a day-to-day schedule can help maintain consistency for the child and address specific developmental delays. Parents should be educated on the signs of relapse and of adverse reactions to the medication, and encourage children in remission to self-report any possible signs of relapse.

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KEY TERMS

Neurotransmitters—Chemicals in the brain that transmit nerve impulses.

Ventricles—Four cavities within the brain that produce and maintain the cerebrospinal fluid that cushions and protects the brain and spinal cord.

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Stephanie Dionne Sherk

School phobia/school refusal

Definition

The term school phobia was first used in 1941 to identify children who fail to attend school because attendance causes emotional distress and **anxiety**. In Great Britain and as of the early 2000s in the United States, the term school refusal is preferred.

Description

School phobia is a complex syndrome that can be influenced by the child's **temperament**, the situation at school, and the **family** situation. Current thinking defines school phobia or school refusal as an anxiety disorder related to **separation anxiety**. Children refuse to attend school because doing so causes uncomfortable feelings, stress, anxiety, or panic. Many children develop physical symptoms, such as **dizziness**, stomachache, or **headache**, when they are made to go to school. School avoidance is a milder form of refusal to attend school. With school avoidance, the child usually tries to avoid a particular situation, such as taking a test or changing clothes for physical education, rather than avoiding the school environment altogether.

School refusal usually develops after a child has been home from school for an illness or vacation. It may also follow a stressful family event, such as **divorce**, parental illness or injury, death of a relative, or a move to a new school. Usually refusal to attend school develops gradually, with children putting up increasingly intense resistance to going to school as time passes. Psychiatrists believe that in young children, the motivating factor often is a desire to stay with the parent or caregiver rather than to avoid an unpleasant situation at school. In older children, or if school refusal comes on suddenly, it may be related to avoiding a distressing situation at school such as bullying, teasing, severe teacher criticism, or it may follow a humiliating event such as throwing up in class. The longer a child stays out of school, the more difficult it is for that child to return.

School refusal is not the same as **truancy**. Children who are school refusers suffer anxiety and physical symptoms when they go to school. They may have temper **tantrums** over going to school or become depressed. They may threaten to harm themselves if made to go to school. School refusers usually work to get their parent's permission to stay home. If allowed to stay home, they usually stay in the house or near the parent or caregiver. The child is willing to do make-up school work at home, so long as he or she does not have to go to school.

Children who are truants are not anxious about school; they simply do not want to be there. They try to hide their absence from their parents and have no interest in make-up schoolwork or meeting academic expectations. Unlike school phobia, truancy often occurs with other antisocial behaviors such as shoplifting, **lying**, and drug and alcohol use.

Demographics

Boys and girls refuse to attend school at the same rates. School phobia is highest in children ages five to seven and 11 to 14. These ages correspond with starting school, and transitioning through middle school or junior high school, both unusually stressful periods. Estimates suggest that about 4.5 percent of children ages 7 to 11 and 1.3 percent of children age 14 to 16 are school refusers. School phobia is an international problem, with an estimated rate of 2.4 percent of all school-age children worldwide refusing to attend classes.

Children who are more likely to become school refusers share certain characteristics. These include:

- reluctance to stay in a room alone or **fear** of the dark
- clinging attachment to parents or caregivers
- excessive worry that something dreadful will happen at home while they are at school
- difficulties sleeping or frequent **nightmares** about separation
- homesickness when away at places other than at school, or an excessive need to stay in touch with the parent or caregiver while away

Causes and symptoms

There appears to be a genetic component to all anxiety disorders, including school phobia. Children whose parents have anxiety disorders have a higher rate of anxiety disorders than children whose parents do not have these disorders. School phobia is often associated with other anxiety disorders such as agoraphobia or other mental health disorders such as depression. Some experts theorize that another possible cause of school refusal is traumatic and prolonged separation from the primary caregiver in early childhood.

Family functioning affects school refusal. Stressful events or a dysfunctional family can cause children to feel compelled to stay home. Young children are more likely to refuse to separate from their parent or caregiver because they fear something catastrophic will happen to the adult while they are at school. Older children may refuse to leave a parent who is ill or who has a substance abuse problem, in effect trying to cope for the parent. They may also be afraid of some specific aspect of school, such as riding the bus or eating in the cafeteria.

It is not uncommon for middle and high school students to become school refusers because they are afraid of violence either at school or on the way to school, are afraid of failing academically, have been repeatedly bullied or humiliated at school, feel they have no friends at school, or are excluded.

Children who refuse to attend school usually try to win a parent's permission to stay home, although some simply refuse to leave the house. Genuine physical symptoms are common and include dizziness, headaches, **nausea**, **vomiting**, **diarrhea**, shaking or trembling, fast heart rate, chest pains, and back, joint or stomach pains. These symptoms usually improve once the child is allowed to stay home. Behavioral symptoms include temper tantrums, crying, angry outbursts, and threats to hurt themselves (**self-mutilation**).

When to call the doctor

Parents with a child who is avoiding or refusing school should call their pediatrician and arrange to have physical symptoms evaluated. If no reason for physical symptoms such as abdominal **pain** can be found, the pediatrician should make a referral to a child or adolescent psychiatrist who can evaluate the child for a range of behavioral problems including social phobia, depression, **conduct disorder**, and post-traumatic stress syndrome.

Diagnosis

The most effective form of treatment is a combination of behavioral and cognitive therapy for an average period of six months. Behavioral therapy involves teaching both parents and children strategies for overcoming certain stressful behaviors such as separation and may involve desensitization by gradual exposure to the stressful event. Cognitive therapy teaches children to redirect their thoughts and actions into a more flexible and assertive pattern. **Family therapy** may also be used to help resolve family issues that may be affecting the child.

Depending on the diagnosis, children may also be treated with drugs to help alleviate depression, panic and anxiety, or other mental health disorders. In October 2003 the United States Food and Drug Administration issued an advisory indicating that children being treated with selective serotonin re-uptake inhibitor **antidepressants** (SSRIs) for major depressive illness may be at higher risk for committing **suicide**. A similar warning was issued in the United Kingdom. Parents and

physicians must weigh the benefits and risks of prescribing these medications for children on an individual basis.

Treatment

Diagnosis is made on the basis of family history, the absence of causes for physical symptoms such as heart palpitations, vomiting, or dizziness, and the results of a battery of **psychological tests**. Psychological evaluation varies with other findings and the age of the child but usually includes several assessments for anxiety and a behavioral checklist that evaluates the child's behavior at home and school.

Prognosis

The combination of cognitive and behavioral therapy appears to produce the most successful treatment results. In one study, more than 80 percent of children receiving this combination of therapies were attending school normally one year after treatment. Underlying conditions that might affect recovery from school phobia include **Tourette syndrome**, attention deficit disorder (ADD), depression, bipolar mental illness, panic disorder, or other anxiety disorders and **phobias**.

Prevention

Little can be done to prevent school refusal. However, parents can give their children appropriate opportunities to separate from them during the toddler and **preschool** years by exposing them to activities such as preschool, playgroups, **babysitters**, and daycare.

With older children, parents can step in to stop bullying behavior or remove their child from the bullying or humiliating situation as soon as it starts.

Parental concerns

Many parents recognize that their child is genuinely distressed by attending school and unwittingly encourage school refusal by allowing their child to stay home. However, the longer the child is at home, the harder it is to return to school. Parents need to make the school aware of their child's difficulties and take a firm stand in working with the school to resolve any issues of **safety** or bullying that may be preventing their child from experiencing a full education.

See also Separation anxiety.

KEY TERMS

Agoraphobia—Abnormal anxiety regarding public places or situations from which the person may wish to flee or in which he or she would be helpless in the event of a panic attack.

Selective serotonin reuptake inhibitors (SSRIs)—A class of antidepressants that work by blocking the reabsorption of serotonin in the brain, thus raising the levels of serotonin. SSRIs include fluoxetine (Prozac), sertraline (Zoloft), and paroxetine (Paxil).

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Tish Davidson A.M.

SCID *see* **Severe combined immunodeficiency**

Scoliosis

Definition

Scoliosis is a side-to-side curvature of the spine.

Description

When viewed from the rear, the spine usually appears perfectly straight. Scoliosis is a lateral (side-to-side) curve in the spine, usually combined with a rotation of the vertebrae. (The lateral curvature of scoliosis should not be confused with the normal set of front-to-back spinal curves visible from the side.) While a small degree of lateral curvature does not cause any medical problems, larger curves can cause postural imbalance and lead to muscle fatigue and **pain**. More severe scoliosis can interfere with breathing and lead to arthritis of the spine (spondylosis).

Demographics

Approximately 10 percent of all adolescents have some degree of scoliosis, although fewer than 1 percent have curves that require medical attention beyond monitoring. Scoliosis is found in both boys and girls, but a girl's spinal curve is much more likely to progress than a boy's. Girls require scoliosis treatment about five times more often than boys. The reason for these differences as of 2004 was not known.

Causes and symptoms

Four out of five cases of scoliosis are idiopathic, meaning the cause is unknown. While idiopathic scoliosis tends to run in families, no specific genes responsible for the condition have been identified. Children with idiopathic scoliosis appear to be otherwise entirely healthy and have not had any bone or joint disease early in life. Scoliosis is not caused by poor posture, diet, or carrying a heavy book bag on one shoulder.

Idiopathic scoliosis is further classified according to age of onset:

- Infantile: Curvature appears before age three. This type is quite rare in the United States but is more common in Europe.
- Juvenile: Curvature appears between ages three and ten. This type may be equivalent to the adolescent type, except for the age of onset.
- Adolescent: Curvature usually appears between ages of ten and 13, near the beginning of **puberty**. This is the most common type of idiopathic scoliosis.
- Adult: Curvature begins after physical maturation is completed.

Causes are known for three other types of scoliosis:

- Congenital scoliosis is due to abnormal formation of the bones of the spine and is often associated with other organ defects.
- Neuromuscular scoliosis is due to loss of control of the nerves or muscles that support the spine. The most common causes of this type of scoliosis are **cerebral palsy** and **muscular dystrophy**.
- Degenerative scoliosis may be caused by breaking down of the discs that separate the vertebrae or by arthritis in the joints that link them.

Scoliosis causes a noticeable asymmetry in the torso when viewed from the front or back. The first sign of scoliosis is often seen when a child is wearing a bathing suit or underwear. A child may appear to be standing with one shoulder higher than the other or to have a tilt in the waistline. One shoulder blade may appear more prominent than the other due to rotation. In girls, one breast may appear higher than the other or larger if rotation pushes one side forward.

Curve progression is greatest near the adolescent growth spurt. Scoliosis that begins early is more likely to progress significantly than scoliosis that begins later in puberty.

When to call the doctor

If the parent notices that a child's posture is abnormal, if when the child stands one hip appears to be higher than the other, if one shoulder blade appears to be sticking out, or the child appears to lean regularly to one side, the doctor should be notified. If the child is screened at school and the screener reports a suspicion of scoliosis, a doctor should be seen to follow up on this suspicion.

Diagnosis

Diagnosis for scoliosis is done by an orthopedist. A complete medical history is taken, including questions about **family** history of scoliosis. The physical examination includes determination of pubertal development in adolescents, a neurological exam (which may reveal a neuromuscular cause), and measurements of trunk asymmetry. Examination of the trunk is done while the patient is standing, bending over, and lying down and involves both visual inspection and use of a simple mechanical device called a scoliometer.

If a curve is detected, one or more x rays will usually be taken to define the curve or curves more precisely. An x ray is also used to document spinal maturity, any pelvic tilt or hip asymmetry, and the location, extent, and degree of curvature. The curve is defined in terms of where it begins and ends, in which direction it bends, and

by an angle measure known as the Cobb angle. The Cobb angle is found by taking an x ray of the spine. Lines are then projected out parallel to the vertebrae at the top and bottom of the curve. Then perpendicular lines are projected from these lines and the angle at which the lines intersect is measured. These angles are referred to when the angle of the curvature is discussed. To properly track the progress of scoliosis, it is important to project from the same points of the spine each time a measurement is made; otherwise, there is a risk of getting misleading measurements.

Occasionally, **magnetic resonance imaging** (MRI) is used as a diagnostic tool, primarily to look more closely at the condition of the spinal cord and nerve roots extending from it if neurological problems are suspected.

Treatment

Treatment decisions for scoliosis are based on the degree of curvature, the likelihood of significant progression, and the presence of pain, if any.

Curves less than 20 degrees are not usually treated, except by regular follow-up for children who are still growing. Watchful waiting is usually all that is required in adolescents with curves of 20 to 30 degrees as long as there is no pain.

For children or adolescents whose curves progress to 30 degrees and who have a year or more of growth left, bracing may be required. Bracing cannot correct curvature but may be effective in halting or slowing progression.

Two styles of braces are used for daytime wear. The Milwaukee brace consists of metal uprights attached to pads at the hips, rib cage, and neck. The other kind of brace is the underarm brace, which uses rigid plastic to encircle the lower rib cage, abdomen, and hips. Both these brace types hold the spine in a vertical position. Because it can be worn out of sight beneath clothing, the underarm brace is better tolerated and often leads to better compliance. A third style, the Charleston bending brace, is used at night to bend the spine in the opposite direction. Braces are often prescribed to be worn for 22 to 23 hours per day, though some clinicians allow or encourage removal of the brace for **exercise**.

Bracing may be appropriate for scoliosis due to some types of neuromuscular disease, including **spinal muscular atrophy**, before growth is finished. Duchenne muscular dystrophy is not treated by bracing. Surgery is likely to be required.

Surgery is usually the option of last resort in cases of scoliosis. Surgery for idiopathic scoliosis is usually recommended if one of the following conditions is present:

- The curve has progressed despite bracing.
- The curve is greater than 40 to 50 degrees before growth has stopped in an adolescent.
- There is significant pain.

Orthopedic surgery for neuromuscular scoliosis is often done earlier. The goals of surgery are to correct the deformity as much as possible, to prevent further deformity, and to eliminate pain as much as possible. Surgery can usually correct 40 to 50 percent of the curve, and sometimes as much as 80 percent. Surgery cannot always completely remove pain.

The surgical procedure for scoliosis is called spinal fusion, because the goal is to straighten the spine as much as possible and then to fuse the vertebrae together to prevent further curvature. To achieve fusion, the involved vertebra are first exposed and then scraped to promote regrowth. Bone chips are usually used to splint together the vertebrae to increase the likelihood of fusion. To maintain the proper spinal posture before fusion occurs, metal rods are inserted alongside the spine and are attached to the vertebrae by hooks, screws, or wires. Fusion of the spine makes it rigid and resistant to further curvature. The metal rods are no longer needed once fusion is complete but are rarely removed unless their presence leads to complications.

Spinal fusion leaves the involved portion of the spine permanently stiff and inflexible. While this leads to some loss of normal motion, most functional activities are not strongly affected, unless the very lowest portion of the spine (the lumbar region) is fused. Normal mobility, exercise, and even contact **sports** are usually all possible after spinal fusion. Full recovery takes approximately six months. Physical therapy is part of standard treatment as well.

Alternative treatment

Numerous alternative therapies have been touted to provide relief and help for individuals with scoliosis, but none has been proven beneficial in clinical trials. These include massage and electrical stimulation. In addition, alternatives such as rolfing or chiropractic manipulation of soft tissue to improve alignment may provide improved flexibility, stronger muscles, and pain relief but cannot prevent or correct the curvature of the spine or its progression.

Although important for general health and strength, exercise has not been shown to prevent or slow the development of scoliosis. It may help relieve pain from scolio-

sis by helping to maintain range of motion. Aquatic exercise, in particular, can increase flexibility and improve posture, balance, coordination, and range of motion. Because it decreases joint compression, it can lessen the pain caused by scoliosis or surgery.

Good **nutrition** is also important for general health, but no specific dietary regimen has been shown to control scoliosis development. In particular, dietary calcium levels do not influence scoliosis progression.

Chiropractic treatment may relieve pain, but it cannot halt scoliosis development and should not be a substitute for conventional treatment of progressing scoliosis. Acupuncture and acupressure may also help reduce pain and discomfort, but these treatments cannot halt scoliosis development either.

Prognosis

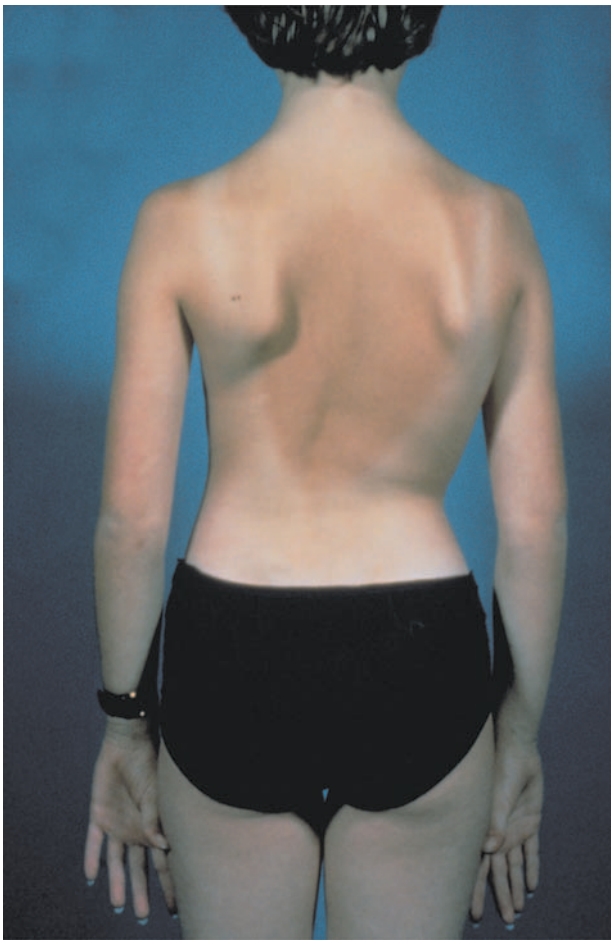
The prognosis for a child with scoliosis depends on many factors, including the age at which scoliosis begins and the treatment received. More importantly, mostly unknown individual factors affect the likelihood of progression and the severity of the curve. Most cases of mild adolescent idiopathic scoliosis need no treatment and do not progress. Untreated severe scoliosis often leads to spondylosis and may impair breathing. Degenerative arthritis of the spine, sciatica, and severe physical deformities can also result if severe scoliosis is left untreated. Finally, scoliosis can also poorly affect the individual's **self-esteem** and cause serious emotional problems.

Prevention

There is no known way to prevent the development of scoliosis. Progression of scoliosis may be prevented through bracing or surgery. More than 30 states have screening programs in schools for adolescent scoliosis, usually conducted by trained school nurses or physical education teachers. These programs can help to catch scoliosis early, so that treatment can begin and progression can often be halted or slowed.

Parental concerns

Children with scoliosis often have a negative self-image associated with irregular posture or having to wear a brace. This problem is being combated with new braces that can be worn under the clothing and are more discreet than traditional braces. Scoliosis can be life threatening if it is not treated and progresses to a point at which breathing is impaired. This is very rare, however.



This patient suffers from scoliosis, or curvature of the spine. *(Custom Medical Stock Photo Inc.)*

Scoliosis should be watched carefully by a physician for signs of worsening, but it usually does not progress to the point at which treatment is needed.

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KEY TERMS

Cobb angle—A measure of the curvature of scoliosis, determined by measurements made on x rays.

Rolfing—A holistic system of bodywork that uses deep manipulation of the body's soft tissue to realign and rebalance the body's myofacial (connective) structure. It is used to improve posture, relieve chronic pain, and reduce stress.

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Scoliosis Research Society. 55 East Wells St. Suite 1100. Milwaukee, WI 53202–3823. Web site: <www.srs.org>.

Tish Davidson, A.M.
Liz Meszaros

Scrapes *see* **Wounds**

Seasonal allergies *see* **Allergic rhinitis**

Seborrheic dermatitis

Definition

Seborrheic **dermatitis** is a common inflammatory disease of the scalp and skin characterized by scaly lesions usually on the scalp, hairline, face and body. In infants, it is sometimes called cradle cap.

Description

Seborrheic dermatitis appears as red, inflamed skin covered by greasy or dry scales that may be white, yellowish, or gray. It can affect the scalp, eyebrows, forehead, face, folds around the nose and ears, the chest, armpits, and groin. In infants it appears most commonly on the scalp and is called cradle cap. Dandruff is a mild form of seborrheic dermatitis and appear as fine white scales without red skin or inflammation. Dandruff can also be caused by other skin conditions, especially in children.

Seborrheic dermatitis is a common, mild disease of newborns. The red, scaly rash can spread to the forehead, behind the ears, and in the creases of the neck and armpits. The rash is not itchy and usually does not bother babies. Occasionally babies also develop this skin disease in the diaper area. When seborrheic dermatitis occurs in the diaper area, it is often accompanied by a yeast infection. When yeast is present, the rash is itchy and uncomfortable. Seborrheic dermatitis usually disappears by the end of the first year and does not reappear until **puberty**.

Transmission

Seborrheic dermatitis is not an infection and is not transmitted from individual to individual.

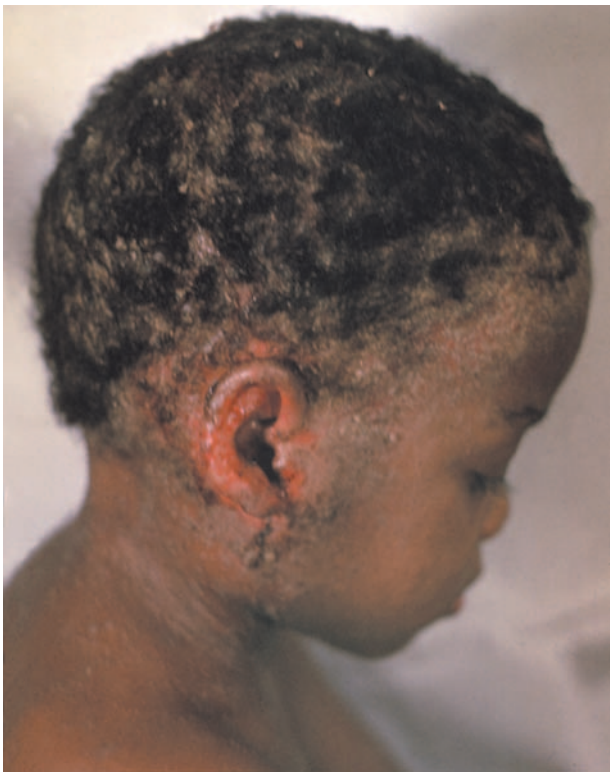
Demographics

Seborrheic dermatitis is a very common among newborns. It usually appears the first six weeks of life and rarely after the age of nine to 12 months. It affects babies of all races and both genders. Seborrheic dermatitis can reappear at puberty and into adulthood.

Causes and symptoms

As of 2004 the cause of seborrheic dermatitis was not clear. However, it is not an infection or an allergy, it is not contagious, and it is not caused by poor hygiene. Seborrheic refers to the sebaceous, or oil producing, glands of the skin. It appears that in pregnancy, hormone changes in the mother may cause these glands to produce too much oil. When this happens, scales develop in the area where the oil glands are most dense. Seborrheic dermatitis may also be linked to genetic factors.

Babies exhibit a characteristic non-itchy greasy red scaly rash or dry whitish or grayish scales on the scalp and possibly on other areas.



This young boy is afflicted with seborrheic dermatitis. *(Custom Medical Stock Photo Inc.)*

KEY TERMS

Cortisone—Glucocorticoid produced by the adrenal cortex in response to stress. Cortisone is a steroid with anti-inflammatory and immunosuppressive properties.

Dermatitis—Inflammation of the skin.

Salicylic acid—An agent prescribed to treat a variety of skin disorders, such as acne, dandruff, psoriasis, seborrheic dermatitis, calluses, corns, and warts.

Sebaceous—Related to the glands of the skin that produce an oily substance called sebum.

When to call the doctor

If the rash does not improve after regular washings with baby shampoo or if the rash spreads and becomes red and itchy, especially in the diaper area, the doctor should be consulted.

Diagnosis

Diagnosis is made on visual inspection of the rash.

Treatment

Frequent washing of the scalp with a mild baby shampoo followed by brushing with a soft brush to remove scales usually clears up cradle cap. In stubborn cases, a special shampoo containing sulfur and salicylic acid can be used. This treatment should be done only after consultation with a pediatrician, since this shampoo may be irritating to babies. Sometimes an ointment containing cortisone, an anti-inflammatory medication, is prescribed. If the seborrheic dermatitis is complicated by a yeast infection, an ointment containing anti-yeast medications such as nystatin is applied to the infected area three or four times daily.

Alternative treatment

Parents may rub mineral oil into their child's scalp to soften and loosen the scales, but the oil should be brushed or shampooed out and not left to accumulate.

Prognosis

Seborrheic dermatitis normally resolves without difficulty, usually by the age of six months and almost always by the end of the first year. The rash does not leave scars.

Prevention

Seborrheic dermatitis cannot be prevented from developing, although it may be controlled through frequent hair washings with a mild baby shampoo.

Parental concerns

Parents are often concerned that the rash will leave a scar on their baby's skin. However, scarring does not occur.

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Security objects

Definition

Security objects are items, usually soft and easily held or carried, that offer a young child comfort. They also are referred to as transitional objects, substitute objects, cuddlies, soothers, "loveys," and security blankets.

Description

Security objects or transitional objects are items that help a young child make the emotional transition from dependence to independence. Attachment to an object often develops toward the end of the first year, although attachment to pacifiers happens earlier. Strong attachment to blankets peaks at 18 and 24 months, stays near this high level through 39 months, and then diminishes steadily. By five and a half, fewer than to 8 percent of children are attached to blankets. Other objects of attachment can be articles of clothing, cloth diapers, soft **toys**, or almost anything with a soft, pleasing texture.

In the 1940s, attachment to a special object was regarded as a childhood problem reflecting an unhealthy relationship between the mother and the child. Gradually this attitude began changing as researchers concluded that the child's attachment to a security object is normal and even desirable. Nevertheless, throughout the 1970s, but progressively less in the 1980s and 1990s, a stigma remained attached to children who, like Linus in the "Peanuts" comic strip, hugged a blanket or other security object in times of stress. The subsequently discredited stereotype was that these children were overly anxious and insecure. As a result, the security object was often taken away from the child, sometimes forcibly, just when it could have been beneficial. As of 2004 research indicated that there is no justification for such drastic actions. Evidence does not find children abnormal or overanxious just because they demonstrate an attachment to a security object. Blanket-attached children appear to be neither more nor less maladjusted or insecure than other children.

Although several theories exist about the role of security objects in development, it is not clear from any of these theories why some children engage in comfort habits with security objects while others do not. Child-rearing practices are frequently seen as contributing factors, and evidence suggests that the security of a child's attachment to its mother does predict how a security object will be used in new or stressful situations.

Despite thinking in the early 2000s that attachment to transitional objects is normal and almost universal, this attachment is actually culture-specific. For instance, in the United States, about 60 percent of children have at least a mild degree of attachment to a soft, inanimate object at some time during their life, and 32 percent exhibit strong attachment. The incidence of attachments to soft objects in the Netherlands, New Zealand, and Sweden is comparable to that in the United States. Korean children have substantially fewer attachments to blankets (18%) than do American children, but Korean-born children living in the United States display an intermediate percentage (34%). Only 5 percent of rural Italian children have transitional objects, compared to 31 percent of native Italian children living in Rome. However, only 16 percent of children living in London have a special security object. In the United States, attachments to various security objects are generally regarded as normal throughout the first five years of life.

Attachment to a security object can be beneficial to a child. The security object may serve as a substitute for the parent in his or her absence and may assist separation from the mother or father by providing the comfort of something familiar. At bedtime, it can soothe and facilitate **sleep**. Studies have also shown that during a routine third-year pediatric examination, children allowed security objects were less distressed than children undergoing the medical evaluation without their security object. The comfort provided by a blanket in new situations has even been shown to enhance children's learning.

Common problems

Parents often become frustrated with their child's need for a security object as it falls to the parent to keep track of the object and soothe the child if it is misplaced or lost. Occasionally daycare centers forbid the child to bring anything from home, including a security object, causing unnecessary stress for the child.

Parental concerns

Parents worry that as the security object becomes dirty, it will spread germs. Other concerns are related to specific objects, such as pacifiers, which may cause dental deformity or objects that, due to their size, shape, or composition, are awkward or undesirable as "loveys." In

Child using his stuffed toy as a security object. *(© Gilbert Patrick/Corbis.)*

these cases, it may be possible, with patience, to substitute one security object for another.

When to call the doctor

Attachment to a security object is normal and should be outgrown without intervention by age five, although 8 percent of children still remain attached to blankets after this age.

See also Separation anxiety.

Resources

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Richard Passman, Ph.D.

Seizure disorder

Definition

A seizure is a sudden disruption of the brain's normal electrical activity accompanied by altered consciousness and/or other neurological and behavioral manifestations. Epilepsy is a disorder of the brain characterized by recurrent seizures that may include repetitive muscle jerking called convulsions.

Description

There are more than 20 different seizure disorders, although epilepsy is the most familiar. Most seizures are benign, but a seizure that lasts a long time can lead to status epilepticus, a life-threatening condition characterized by continuous seizures, sustained loss of consciousness, and respiratory distress. In addition, non-convulsive epilepsy can impair physical coordination, vision, and other senses. Undiagnosed seizures can lead to conditions that are more serious and more difficult to manage. Ten percent of Americans have a seizure at some time in their lives.

Generalized seizures

A generalized seizure occurs when electrical abnormalities exist throughout the brain. A generalized tonic-clonic (grand-mal) seizure typically begins with a loud cry before the individual having the seizure loses consciousness and falls to the ground. The muscles become rigid for about 30 seconds during the tonic phase of the seizure and alternately contract and relax during the clonic phase, which lasts 30 to 60 seconds. The skin sometimes acquires a bluish tint, and the person may bite

the tongue, lose bowel or bladder control, or have trouble breathing.

A grand mal seizure lasts two to five minutes, and the person may be confused or have trouble talking after regaining consciousness (post-ictal state). The individual may complain of head or muscle aches or weakness in the arms or legs before falling into a deep **sleep**.

Primary generalized seizures

A primary generalized seizure occurs when electrical discharges begin in both halves (hemispheres) of the brain at the same time. Primary generalized seizures are more likely to be major motor attacks than to be absence seizures. Motor attacks cause parts of the body to jerk repeatedly. A motor attack usually lasts less than an hour and may last only a few minutes.

Absence seizures

Absence (petit mal) seizures generally begin at about the age of four and stop by the time the child becomes an adolescent. Absence seizures usually begin with a brief loss of consciousness and last 15 to 20 seconds. An individual having a petit mal seizure becomes very quiet and may blink, stare blankly, roll the eyes, or move the lips. When a petit mal seizure ends, individual resumes whatever he or she was doing before the seizure began and does not remember the seizure. The individual may not realize that anything unusual has happened. Untreated, petit mal seizures can recur as many as 100 times a day and may progress to grand mal seizures.

Myoclonic seizures

Myoclonic seizures are characterized by brief, involuntary spasms of the tongue or muscles of the face, arms, or legs. Myoclonic seizures are most apt to occur when waking after a night's sleep.

A Jacksonian seizure is a partial seizure characterized by tingling, stiffening, or jerking of an arm or leg. Loss of consciousness is rare. The seizure may progress in characteristic fashion along the limb.

Limp posture and a brief period of unconsciousness are features of akinetic seizures. These occur in young children. Akinetic seizures, which cause the child to fall, also are called drop attacks.

Partial seizures

Simple partial seizures do not spread from the focal area of the brain where they arise. Symptoms are determined by the part of the brain affected. The individual usually remains conscious during the seizure and can later describe it in detail. In 2003, it was reported that people who experience partial seizures are twice as likely to have sleep disturbances as people their same age and gender who do not have seizures.

Complex partial seizures

A distinctive smell, taste, or other unusual sensation (aura) may signal the start of a complex partial seizure. Complex partial seizures start as simple partial seizures but move beyond the focal area of the brain and cause loss of consciousness. Complex partial seizures can become major motor seizures. Although individuals having a complex partial seizure may not seem to be unconscious, they do not know what is happening and may behave inappropriately. They will not remember the seizure but may seem confused or intoxicated for a few minutes after it ends.

Demographics

One in ten Americans has a seizure during their lifetime, and at least 200,000 Americans have at least one seizure a month. Epilepsy affects 2.5 million Americans of all ages, and of those, 25 percent of all cases develop before the age of five. Some 181,000 new cases are diagnosed annually and 45,000 of them are children under the age of 15. Though the incidence rate for children is in the early 2000s trending down, epilepsy remains a significant problem for many children.

In all people, the risk of developing epilepsy is approximately 1 percent. However, certain groups are at higher risk. The expectations of the onset of epilepsy in these populations are as follows:

- children with **mental retardation**: 10%
- children with **cerebral palsy**: 10%
- children with both cerebral palsy and mental retardation: 50%
- children of mothers with epilepsy: 8.7%
- children of fathers with epilepsy: 2.4%

In addition, males are somewhat more likely to develop epilepsy than females, and African-Americans are more likely to develop it than Caucasians. The incidence of epilepsy is greater in those who are socioeconomically disadvantaged.

Causes and symptoms

The cause of 70 percent of new cases of epilepsy is unknown (idiopathic). Epilepsy sometimes is the result of trauma at birth. Such neonatal causes include insuffi-

cient oxygen to the brain, **head injury**, heavy bleeding, incompatibility between a woman's blood and the blood of her baby, or infection immediately before, after, or at the time of birth.

Other causes of epilepsy include the following:

- head trauma resulting from a car accident, gunshot wound, or other injury
- alcoholism
- brain abscess or inflammation of membranes covering the brain or spinal cord
- phenylketonuria (PKU) or other inherited disorders or genetic factors
- infectious diseases such as **measles**, **mumps**, and diphtheria
- degenerative disease
- lead **poisoning**, mercury poisoning, **carbon monoxide poisoning**, or ingestion of other poisonous substances

Status epilepticus, a condition in which an individual suffers from continuous seizures and may have trouble breathing, can be caused by the following factors:

- suddenly discontinuing anti-seizure medication
- hypoxic or metabolic encephalopathy (brain disease resulting from lack of oxygen or malfunctioning of other physical or chemical processes)
- acute head injury
- blood infection caused by inflammation of the brain or the membranes that cover it

Symptoms

Different types of seizures have different symptoms. Generalized epileptic seizures occur when electrical abnormalities exist throughout the brain. Partial seizures do not involve the entire brain, although a partial seizure may spread to other parts of the brain and cause a generalized seizure. Some people who have epilepsy have more than one type of seizure.

Motor attacks cause parts of the body to jerk repeatedly. Sensory seizures cause **numbness** or tingling in one area. The sensation may move along one side of the body or the back before subsiding.

Visual seizures, which affect the area of the brain that controls sight, cause people to see things that are not there. Auditory seizures affect the part of the brain that controls hearing and cause the individual to imagine voices, music, and other sounds. Other types of seizures can cause confusion, upset stomach, or emotional distress.

When to call the doctor

Parents should call the doctor or local emergency number the first time a child has a seizure. For children who have been diagnosed with epilepsy, the doctor should give guidelines about when to call. However, the following situations merit emergency attention:

- a longer seizure than the child usually has or an unusual number of seizures
- seizures that recur repeatedly in the course of a few minutes
- consciousness not regained between seizures
- occurrence of new neurological symptoms
- occurrence of side effects from medication, which could include drowsiness and rash for most anticonvulsants (Specific possible side effects should be reviewed for each medication with the physician and/or pharmacist.)

Diagnosis

Personal and **family** medical history, description of seizure activity, and physical and neurological examinations help primary care physicians, neurologists, and epileptologists diagnose this disorder. Doctors rule out conditions that cause symptoms that resemble seizure disorders, including small strokes (transient ischemic attacks, or TIAs), fainting (syncope), pseudoseizures, and sleep attacks (**narcolepsy**).

Neuropsychological testing uncovers learning or memory problems. Neuroimaging provides views of brain areas involved in seizure activity.

The **electroencephalogram** (EEG) is the main test used to diagnose epilepsy. EEGs use electrodes placed on or within the skull to record the brain's electrical activity and pinpoint the exact location of abnormal discharges.

Other tests used to diagnose seizure disorders include:

- Magnetic resonance imaging (MRI), which provides clear, detailed images of the brain. Functional MRI (fMRI), performed while the patient does various tasks, can measure shifts in electrical intensity and blood flow and indicate which brain region each activity affects.
- Positron emission tomography (PET) and single photon emission tomography (SPECT) monitor blood flow

and chemical activity in the brain area being tested. PET and SPECT are very effective in locating the brain region where metabolic changes take place between seizures.

- Urine and blood lab tests can screen for electrolyte disturbances and possible metabolic disorders.

Treatment

Seizure disorders in children are usually treated with anticonvulsant drugs. Doctors attempt to use a single drug for this purpose, but more than one may be required. Medications are prescribed based on the seizure type. Even when the drugs suppress seizures, they should not be discontinued without a doctor's advice. Most individuals require at least several years of treatment.

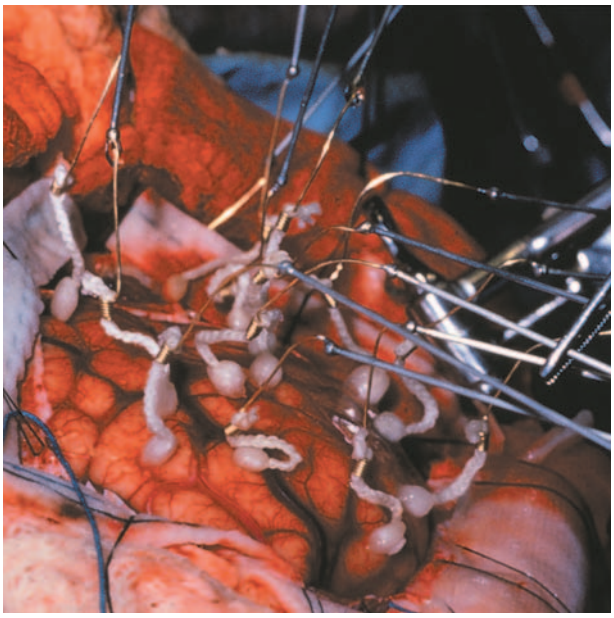
If medication is not successful in preventing seizures, surgery, a ketogenic diet, or vagus nerve stimulation (VNS) may be tried. Brain surgery can be useful in certain cases to remove small groups of cells causing the problem.

The ketogenic diet is a high fat, low carbohydrate, limited calorie diet that forces the child's body to burn fat instead of glucose derived from carbohydrates. Burning fat produces chemicals called ketones. One out of three children who begins the diet becomes free or almost free from seizures, while another third improve, and the final third show no improvement. This diet, which is usually begun in the hospital, is extremely rigorous and must be monitored by a doctor and dietician.

The United States Food and Drug Administration (FDA) has approved the use of vagus nerve stimulation (VNS) in patients over the age of 16 who have intractable partial seizures. This non-surgical procedure uses a pacemaker-like device implanted under the skin in the upper left chest, to provide intermittent stimulation to the vagus nerve. Stretching from the side of the neck into the brain, the vagus nerve affects swallowing, speech, breathing.

Prognosis

Prognosis depends on the type of seizures, the ability to control them with medication, the age of the individual, and the underlying cause of the seizures. Seventy percent of individuals with epilepsy can be expected to go into remission, which is defined as five or more years without seizures while on medication. Three-fourths of those who are seizure free for two to five years while on medication can have the medication reduced or eliminated. However, in 10 percent of new epilepsy cases, the seizures are not controlled by medication.



This patient's brain is exposed during surgery in order for surgeons to remove the mass responsible for epileptic seizures. *(Custom Medical Stock Photo Inc.)*

Prevention

There is no known way to prevent the onset of seizure disorders, but seizures may be controlled and sometimes prevented by the use of medication. Up to 80 percent of those with seizure disorder can have their seizures substantially or completed controlled, allowing them to live normal or close to normal lives.

Parental concerns

Seizure disorders are long-term illnesses, with the added problem of being public. Besides the difficulty of controlling medication and possibly diet, the parents of a child with a seizure disorder must sometimes deal with the public visibility of seizure episode. Parents should be supportive of the child and make sure the child does not consider himself to blame for the seizures.

Siblings are also affected by a child with a seizure disorder. Siblings may feel neglected by parents who focus on care for one child. They may also feel responsible for their brother or sister getting the disease, and they may worry about having seizures themselves. Siblings should be assured that seizure disorders are not contagious. They should be given appropriate information both for themselves and for friends who might be present during seizures.

KEY TERMS

Epileptologist—A physician who specializes in the treatment of epilepsy.

Glucose—A simple sugar that serves as the body's main source of energy.

Phenylketonuria (PKU)—A rare, inherited, metabolic disorder in which the enzyme necessary to break down and use phenylalanine, an amino acid necessary for normal growth and development, is lacking. As a result, phenylalanine builds up in the body causing mental retardation and other neurological problems.

Some parents worry that stress might bring on a seizure and are therefore unwilling to **discipline** a child with a seizure disorder and might give in to the usual childish demands. Although stress can be a factor, parents should consult with their doctor on the level of risk and methods of discipline that can be effective for their child.

Teenagers have special concerns. In many states, those who have not been seizure-free for a certain time are not allowed to drive, which affects a teen's mobility and social life. Having seizures in front of friends can be embarrassing. Parents should resist being excessively overprotective of their teenager and should consult with their physician as to which activities are safe for their child to pursue.

Some physicians recommend avoidance of swimming in children with epilepsy. Nearly all practitioners would advise against unaccompanied swimming in persons with seizure disorders. Avoidance of exposure to flashing lights or other triggers might be necessary in some persons with seizure disorders.

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Seizure medication *see* **Antiepileptics**

Self-esteem

Definition

Considered an important component of emotional health, self-esteem encompasses both self-confidence and self-acceptance. It is the way individuals perceive themselves and their self-value.

Description

Self-esteem is the way individuals think and feel about themselves and how well they do things that are important to them. In children, self-esteem is shaped by what they think and feel about themselves. Their self-esteem is highest when they see themselves as approximating their "ideal" self, the person they would like to be. Children who have high self-esteem have an easier time handling conflicts, resisting negative pressures, and making friends. They laugh and smile more and have a generally optimistic view of the world and their life.

Children with low self-esteem have a difficult time dealing with problems, are overly self-critical, and can become passive, withdrawn, and depressed. They may hesitate to try new things, may speak negatively about themselves, are easily frustrated, and often see temporary problems as permanent conditions. They are pessimistic about themselves and their life.

Self-esteem comes from different sources for children at different stages of development. The development of self-esteem in young children is heavily influenced by parental attitudes and behavior. Supportive parental behavior, including encouragement and praise for accomplishments, as well as the child's internalization of the parents' own attitudes toward success and failure, are the most powerful factors in the development of self-esteem in early childhood. As children get older their experiences outside the home, in school, and with peers, become increasingly important in determining their self-esteem.

Schools can influence their students' self-esteem through the attitudes they foster toward competition and diversity and their recognition of achievement in academics, **sports**, and the arts. By middle childhood, friendships have assumed a pivotal role in a child's life. Studies have shown that school-age youngsters spend more time with their friends than they spend doing homework, watching television, or playing alone. In addition, the amount of time in which they interact with their parents is greatly reduced from when they were younger. At this stage, social acceptance by a child's peer group plays a major role in developing and maintaining self-esteem.

The physical and emotional changes that take place in **adolescence**, especially early adolescence, present new challenges to a child's self-esteem. Boys whose growth spurt comes late compare themselves with peers who have matured early and seem more athletic, masculine, and confident. In contrast, early physical maturation can be embarrassing for girls, who may feel gawky and self-conscious in their newly developed bodies. Both boys and girls expend inordinate amounts of time and energy on personal grooming, spending long periods of time in the bathroom trying to achieve a certain kind of look. Fitting in with their peers becomes more important than ever to their self-esteem, and, in later adolescence, relationships with the opposite sex (or sometimes the same sex) can become a major source of confidence or insecurity. Up to a certain point, adolescents need to gain a sense of competence by making and learning from their own mistakes and by being held accountable for their own actions.

Peer acceptance and relationships are important to children's social and emotional development and to their development of self-esteem. Peer acceptance, especially friendships, provides a wide range of learning and development opportunities for children. These include companionship, recreation, social skills, participating in group problem solving, and managing competition and conflict. They also allow for self-exploration, emotional growth, and moral and ethical development.

There are several factors that influence self-esteem. These include the following:

- Age: Self-esteem tends to grow steadily until middle school when the transition of moving from the familiar environment of elementary school to a new setting confronts children with new demands. Self-esteem either continues to grow after this period or begins to decrease.
- Gender: Girls tend to be more susceptible to having low self-esteem than boys, perhaps because of increased social pressure that emphasizes appearance more than **intelligence** or athletic ability.
- Socioeconomic status: Researchers have found that children from higher-income families usually have a better sense of self-esteem in the mid- to late-adolescence years.
- Body image: Especially true for teens but also important for younger children, body image is evaluated within the context of media images from television, movies, and advertising that often portray girls as thin, beautiful, and with perfect complexion. Boys are portrayed as muscular, very good looking, and tall. Girls who are overweight and boys who are thin or short often have low self-esteem because they compare themselves against these cultural and narrow standards.

Infancy

Infants start building self-esteem as soon as they are born. Their self-esteem is first built by having their basic needs met, including the need for love, comfort, and closeness. They gradually learn that they are loved as the people who care for them consistently treat them gently, kindly, comfort them when they cry, and show them attention. How their parents or primary caregivers treat them sets the stage for later development of self-esteem. Parents who give their babies love and attention teach the infants that they are important, safe, and secure.

Toddlerhood

During toddlerhood, children still have not developed a clear understanding of self-esteem or self-identity. Each time they learn a new skill they add to their sense of their ability and their comprehension of who they are. Toddlers learn about themselves by learning what they look like, what they can do, and where they belong. They find it difficult to share since they are just starting to learn who they are and what is theirs.

Toddlers see themselves through the eyes of their parents, **family**, or primary caregivers. If their parents show them love and treat them as special, toddlers will develop self-esteem. Toddlers who feel unloved find it more difficult to develop a sense of self-worth.

Preschool

By the age of three, children have a clearer understanding of who they are and how they fit into the world they know. They have begun learning about their bodies and that, within limits, they are able to think and make decisions on their own. They can handle time away from their parents or primary caregivers because they feel safe on their own or with other children and adults. They develop their self-esteem in mostly physical ways, by comparing their appearance to that of other children, such as height, size, agility, and abilities.

Preschoolers learn self-esteem in stages through developing their senses of trust, independence, and initiative. During this age, parents can help foster the child's self-esteem by teaching problem-solving skills, involving them in tasks that give them a sense of accomplishment, asking for and listening to their opinions, and introducing them to social settings, especially with their peers. Young children learn self-esteem through what they can do and what their parents think of them.

School age

A critical point in a child's development of self-esteem occurs when they start school. Many children's self-esteem falls when they have to cope with adults and peers in a new situation with rules that may be new and strange. In the early school-age years, self-esteem is about how well children manage learning tasks in school and how they perform in sports. It also depends on their physical appearance and characteristics and their ability to make friends with other children their own age.

Stresses at home, such as parents arguing a lot, and problems at school, such as difficult lessons, being bullied, or not having friends, can have a negative impact on a child's self-esteem. Children with overly developed self-esteem may tend to be **bullies**, while children with lower self-esteem may become the victims of bullies. Parents can help children develop an inner sense of self-control, which comes from having experience in making decisions.

Teenagers' self-esteem is often affected by the physical and hormonal changes they experience, especially during **puberty**. Teens undergo major changes in their lives and their self-esteem can often become fragile. They are usually extremely concerned about how they look and how they are perceived and accepted by their peers. Teens who set goals in their lives have higher self-esteem than those who do not. High self-esteem is also directly related to teens who have a very supportive family.

Body image is a major component in teenagers' self-esteem, and they are very concerned about how their peers see them. Teens who have high self-esteem like the way they look and accept themselves the way they are. Teens with low self-esteem usually have a poor body image and think they are too fat, not pretty enough, or not muscular enough. There are some physical features that teens cannot change, but accepting themselves as they are without undue self-criticism is challenging. If there are characteristics that cause low self-esteem but can be changed, teens may be able to set reasonable goals for making change. For example, if teens think they are overweight, they should first verify their perception with a healthcare provider. If they are actually overweight, they can set goals to lose weight by eating nutritiously and exercising regularly.

The "Teens Health" section of the Web site Kids Health (available online at <www.kidshealth.org>) offers the following advice for teens to improve self-esteem: "When you hear negative comments coming from within, tell yourself to stop. Your inner critic can be retrained. Try exercises like giving yourself three compliments every day. While you're at it, every evening list three things in your day that really gave you pleasure. It can be anything from the way the sun felt on your face, the sound of your favorite band, or the way someone laughed at your jokes. By focusing on the good things you do and the positive aspects of your life, you can change how you feel about yourself."

Parents can enhance teenagers' self-esteem by asking for their help or advice and listening to their opinions.

Common problems

Numerous studies have linked low self-esteem to a wide range of problems, including poor school achievement, criminal and violent behavior; being the victim of bullying; teenage pregnancy; **smoking** and the use of alcohol and other drugs; dropping out of school; depression; and thoughts of **suicide**, suicide attempts, and suicide. Also, children and teens who have low self-esteem have more physical health problems than those with higher self-esteem.

Parental concerns

Every child and teen has low self-esteem at some time in his or her life. Criticism from parents or others can make children with low self-esteem feel worse. Children can also develop low self-esteem if parents or others press them to reach unrealistic goals. Parents should be concerned when a child's low self-esteem interferes with his or her daily activities or causes depression. Some common signs of low-self esteem in children and teens are as follows:

- feeling they must always please other people
- general feelings of not liking themselves
- feelings of unhappiness most of the time
- feeling that their problems are not normal and that they to blame for their problems
- needing constant validation or approval
- not making friends easily or having no friends
- needing to prove that they are better than others

When to call the doctor

Sometimes a lack of self-esteem is too much for a child to handle alone. Parents may need to seek professional psychological help for children suffering from low self-esteem when the child is depressed or shows an inability to create friendships. Help may also be needed for adolescents whose lack of self-esteem is expressed in negative behaviors, such as criminal activities, gang affiliation, smoking, and alcohol and other drug dependency. If the child talks about or threatens suicide, professional help should be sought immediately.

Tips for raising your child's self-esteem

1. Be a role model for high self-esteem. If you have a positive attitude, chances are your children will have one too.
2. Have realistic expectations. Unreasonable goals will set your child up for feelings of failure.
3. Respect your child's individuality. Their accomplishments should be praised even if they are not in your area of interest, or if their level of academic success, for instance, is generally lower than a sibling's.
4. Praise your child's efforts, even if they are ultimately unsuccessful. Making a great effort should be rewarded, even he or she did not come in "first."
5. Be careful when correcting your child's behavior. Constructive criticism is much more useful than pinning your child with a label like "lazy" or "stupid."

SOURCE: McKesson Health Solutions, http://www.med.umich.edu/1libr/pa/pa-esteemup_pep.htm, 2004.

(Table by GGS Information Services.)

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KEY TERMS

Peer acceptance—The degree to which a child or adolescent is socially accepted by peers, usually of about the same age; the level of peer popularity.

Primary caregiver—A person who is responsible for the primary care and upbringing of a child.

Puberty—The point in development when the ability to reproduce begins. The gonads begin to function and secondary sexual characteristics begin to appear.

Self-identity—The awareness that an individual has of being unique.

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Ken R. Wells

Self-mutilation

Definition

Self-mutilation, also called self-harm, self-injury or cutting, is the intentional destruction of tissue or alteration of the body done without the conscious wish to commit **suicide**, usually in an attempt to relieve tension.

Description

Self-mutilation has become an increasing problem among adolescents since the 1990s. Cutting one's skin with razors or knives is the most common pattern of self-mutilation. Other forms of self-harm include biting, hitting, or bruising oneself; picking or pulling at skin or hair; burning oneself with cigarettes, or amputating parts of the body. Self-mutilation can be episodic (infrequent) or repetitive. Episodic self-harm can progress to repetitive self-harm after as few as five or as many as 20 episodes.

Professional thinking about self-mutilation has evolved over the past 20 years. Before the 1990s, self-mutilation was often identified as a failed suicide attempt. This concept is no longer accepted. As of 2004 self-mutilation was not a specific diagnosis recognized by the American Psychiatric Association. Instead, it is recognized as a feature of other psychiatric disorders. Some researchers dispute this designation and feel self-mutilation should be a separate diagnosis. Self-mutilation should not be confused with current fads for **tattoos** and body **piercing**. In some cases, however, it may be difficult to distinguish between an interest in these fads and the first indications of a disorder.

Demographics

It is estimated that one in every 100 individuals in the United States, or more than 2.5 million people, are self-mutilators. Girls are four times more likely to engage in self-harm than boys, with girls between the ages of 16 and 25 at highest risk, although many girls begin cutting in middle school (ages 12 or 13). At risk individuals also include those who have underlying psychiatric disorders. Up to half of individuals who are self-mutilators were sexually abused as children.

Causes and symptoms

It is not entirely clear why some individuals mutilate themselves. However, self-injury appears to give these people an immediate release from almost unbearable tension caused by **anxiety**, anger, or sadness. Some researchers ascribe this response to the release of certain chemicals in the body in response to **pain**. Like other addictive behaviors, self-mutilation gradually takes more and more destruction to achieve release. Some researchers separate self-mutilators into several groups, based on their psychological condition, motivation for harming themselves, and degree to which they practice self-injury.

The most common form of self-mutilation, and the one usually seen in adolescents, is impulsive self-mutilation consisting of superficial skin cutting and burning. Psychiatrists generally believe that this is a maladaptive form of self-help or self-preservation and is done to achieve release from almost unbearable psychic tension and to give the individual a feeling of control. It is often a feature of psychiatric disorders including the following:

- borderline personality disorder
- antisocial personality disorder
- dissociative disorders
- anorexia or bulimia

- post traumatic stress syndrome
- substance abuse
- depression

Compulsive self-mutilation consists of repetitive hair pulling (**trichotillomania**), nail biting, and skin picking. It is often found in individuals with **obsessive-compulsive disorder** (OCD). Major self-mutilation is the least frequent form of self-harm. It involves infrequent episodes of destruction of large amounts of tissue, for example self-castration or self-amputation. Major self-mutilation occurs most often with psychotic or highly intoxicated individuals and occasionally with institutionalized mentally retarded individuals. It is also occasionally associated with **autism**, **Tourette syndrome**, and **schizophrenia**.

The symptoms of self-mutilation typically include wearing long-sleeved or baggy clothing, even in hot weather, and an unusual need for privacy. Self-mutilators are often hesitant to change their clothes or undress around others. In most cases the individual also shows signs of depression.

When to call the doctor

Parents and caregivers should consult a psychiatrist or psychotherapist with professional expertise in self-mutilation as soon as the behavior is discovered. Adolescents rarely do not outgrow this behavior. If left untreated, it can continue into adulthood.

Diagnosis

Self-mutilation is usually diagnosed by a psychiatrist or psychotherapist upon referral from a **family** member, physician, nurse, or social worker who has noticed scars, **bruises**, or other physical evidence of self-injury.

Treatment

Individuals who mutilate themselves should seek treatment from a therapist with some specialized training and experience with this behavior. Most self-mutilators are treated as outpatients, although some specialized inpatient programs for self-mutilators exist. A number of different treatment approaches are used with self-mutilators, including individual therapy, **family therapy**, and group therapy. Therapies focus on teaching self-awareness, alternate coping skills, behavior modification, and improved **communication skills**. Techniques may include journaling, music and art therapy, and role-playing.

Self-inflicted lacerations on the arms of a teenage girl. *(Photo Researchers, Inc.)*

Underlying psychological disorders are also treated with medication and/or psychotherapy. Although there are no medications specifically for self-mutilation, **antidepressants** are often given, particularly if the patient meets the diagnostic criteria for a depressive disorder. However, in October 2003, the United States Food and Drug Administration issued an advisory indicating that children being treated with selective serotonin re-uptake inhibitor antidepressants (SSRIs) for major depressive illness may be at higher risk for committing suicide. A similar warning was issued in the United Kingdom. Parents and physicians must weigh the benefits and risks of prescribing these medications for children on an individual basis.

Alternative treatment

Mindfulness training, which is a form of meditation, has been used to teach self-mutilators to observe and identify their feelings in order to have some control over them.

Prognosis

The prognosis depends on the presence and severity of other emotional disorders and history of sexual abuse and/or suicide attempts. In general, teenagers without a history of abuse or other disorders have a good prognosis. Patients diagnosed with borderline personality disorder and/or a history of attempted suicide are considered to have the worst prognosis.

KEY TERMS

Borderline personality disorder (BPD)—A pattern of behavior characterized by impulsive acts, intense but chaotic relationships with others, identity problems, and emotional instability.

Dissociative disorders—A group of mental disorders in which dissociation is a prominent symptom. Patients with dissociative disorders have a high rate of self-mutilation.

Selective serotonin reuptake inhibitors (SSRIs)—A class of antidepressants that work by blocking the reabsorption of serotonin in the brain, thus raising the levels of serotonin. SSRIs include fluoxetine (Prozac), sertraline (Zoloft), and paroxetine (Paxil).

Prevention

Some society-wide factors that influence self-mutilation, such as the sexual abuse of children and media portrayals of cutting, are difficult to change. Parents should emphasize self-respect and respect for one's body. In general, young people who have learned to express themselves in words or through art and other creative activities are less likely to deal with painful feelings by injuring their bodies.

Parental concerns

Parents should be alert to Internet sites and movies that promote and/or glorify self-mutilation as a way to cope with problems. Experts feel that some children may be induced to try this behavior as a coping mechanism because of the way it is presented in these media.

See also Depressive disorders.

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American Psychiatric Association. 1400 K Street, NW, Washington, DC 20005. Web site: <www.psych.org>.

Focus Adolescent Services. Web site: <www.focusas.com>.

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Separation anxiety

Definition

Separation **anxiety** is distress or agitation resulting from separation or **fear** of separation from a parent or caregiver to whom a child is attached.

Description

Separation anxiety is a normal part of development. It emerges during the second half year in infants. Separation anxiety reflects a stage of brain development rather than the onset of problem behaviors. On the other hand, prolonged separation anxiety that develops in school age children is considered an anxiety disorder by the American Psychiatric Association.

Normal separation anxiety

Developmentally normal separation anxiety usually begins somewhere around eight to 10 months and peaks by 18 months, after which it gradually diminishes until by age three. Only occasional bouts of separation anxiety then occur when the child is faced with new situations, such as starting **preschool** or the addition of a new baby to the **family**.

Before about six months of age, infants show little special attachment to a particular caregiver and no distress at being left alone. At about eight months, babies begin to react by crying and fussing whenever their primary caregiver leaves the room. Bedtimes may become

a struggle, with the child refusing care from all but the primary caregiver and crying, fussing, and calling the caregiver when it is time to go to **sleep**. This unwillingness to be left alone can continue for many months.

When left with a babysitter, even a familiar individual that the infant formerly accepted, the child may scream and cry to exhaustion. This is likely to be due to a combination of separation anxiety and **stranger anxiety**, which arise at about the same time and has similar origins.

Separation anxiety is thought to develop because as babies mature mentally, they begin to recognize their caregivers as unique individuals. However, infants lack the mental capacity to understand that the caregiver still exists when out of sight. To the infant, once the caregiver cannot be seen, she is gone forever (lack of object permanence). This inability to project beyond what is immediately visible, coupled with the newly formed attachment to the caregiver, causes distress that is usually expressed by crying. Although this is a difficult stage for parents, the fact that a child fusses when the preferred caregiver leaves is a sign of healthy **bonding** and normal development. With experience and increased mental maturity, the child will eventually understand that he is not being abandoned permanently and that the caregiver will return.

Although separation anxiety is normal in infants and toddlers, cultural practices have an impact on the timing of its emergence and its extent. Babies who remain in constant contact with their mothers may develop separation anxiety earlier and possibly for more intense and prolonged periods than infants frequently cared for by a variety of different caregivers.

Separation anxiety disorder

Separation anxiety disorder occurs when older children refuse to leave a parent or other caregiver to whom they have become attached. Often separation anxiety disorder begins around age six or seven at a time when it can interfere with school attendance. School phobia can be a type of separation anxiety disorder.

Children with separation anxiety disorder repeatedly show at least three of the following behaviors at a developmentally inappropriate age:

- excessive distress at leaving home or leaving the primary caregiver, or even distress in anticipation of leaving
- excessive worry that something catastrophic will happen at home or to the caregiver while the child is away
- extreme fear that something will happen to them, such as getting lost or kidnapped, that will prevent their return to the caregiver
- unwillingness to be alone, even in familiar settings
- nightmares about separation from home and loved ones
- inability to stay at a friend's house overnight or go away to camp due to worry about what is happening at home
- physical complaints such as stomach pains, **dizziness**, headaches, or **vomiting** when faced with separation from home or caregiver
- refusal to attend school not related to events at school such as bullying or academic failure
- attachment to home or caregiver that interferes with social life and school attendance

Unlike developmentally normal separation anxiety, children do not outgrow separation anxiety disorder. This disorder is usually treated with a combination of behavioral and cognitive therapy. Behavioral therapy involves teaching parents and children strategies for overcoming stressful separation and may involve desensitization by gradual exposure to longer and longer periods apart. Cognitive therapy teaches children to redirect their thoughts and actions into a more flexible and assertive pattern. **Family therapy** may also be used to help resolve family issues that may be negatively affecting the child.

Separation anxiety disorder sometimes occurs in conjunction with other psychiatric disorders, such as pervasive developmental disorder, **schizophrenia**, other anxiety or panic disorders, and major depression. Depending on the diagnosis, children may also be treated with drugs to help alleviate these disorders. However, the use of **antidepressants** in minors is currently under review. In October 2003, the United States Food and Drug Administration issued an advisory indicating that children being treated with selective serotonin re-uptake inhibitor antidepressants (SSRIs) for major depressive illness may be at higher risk for committing **suicide**. A similar warning was issued in the United Kingdom. Parents and physicians must weigh the benefits and risks of prescribing these medications for children on an individual basis.

Common problems

Parents are frequently frustrated by the intensity of their child's separation anxiety while an infant and toddler and believe that something is wrong with their child rather than accepting this natural stage of development. In school-age children, refusal to attend school due to

KEY TERMS

Selective serotonin reuptake inhibitors (SSRIs)—A class of antidepressants that work by blocking the reabsorption of serotonin in the brain, thus raising the levels of serotonin. SSRIs include fluoxetine (Prozac), sertraline (Zoloft), and paroxetine (Paxil).

separation anxiety disorder is common. This can lead to academic failure and difficulty in making friends and developing relationships outside the home.

Parental concerns

Parents are distressed and concerned when their child is distressed. However, since anxiety disorders have an inherited component, in some families a parent will also have an anxiety disorder. The parent's anxieties can add to the child's concerns about separating, worsening the separation anxiety. In this case, family therapy as well as individual therapy for the parent and child may be appropriate.

When to call the doctor

Parents should call the doctor when a child in kindergarten or older shows extreme reluctance to separate from the parent to the point where it interferes with the child's normal life and social development. After a physical examination, a psychological evaluation that includes several assessments for anxiety and a behavioral checklist that evaluates the child's behavior at home and school should be done by a psychologist or psychiatrist with experience in separation anxiety.

See also Stranger anxiety; School phobia/school refusal.

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Serum hepatitis *see* **Hepatitis B**

Severe combined immunodeficiency

Definition

Severe combined **immunodeficiency** (SCID) is the most serious primary or congenital human immunodeficiency disorder. It is a group of congenital (present from birth) disorders in which the immune system does not work properly. Children with SCID are vulnerable to recurrent severe infections, retarded growth, and early death.

Description

The immune system is composed of elements that are needed for the body to fight infections by recognizing disease agents and attacking them. It includes many classes of T-lymphocytes (white blood cells that detect foreign proteins called antigens). It also includes B cells, which are the only cells in the body that make antibodies. Natural killer (NK) cells are cells that destroy infected cells. In children with SCID, the immune system does not function properly because T, B, and NK cells are either absent or defective. When the immune system does not function correctly, the child is left open to repeated severe diseases and infections.

Several different immune system disorders are grouped under SCID. These include the following:

- X-linked: The most common form of SCID accounts for about half of all cases. Because this is an X-linked condition, it occurs only in boys. Children with

X-linked SCID have low T-cell and natural killer (NK) cell levels but elevated B-cell levels.

- Adenosine deaminase deficiency (ADA): About 20 percent of SCID cases are of this type. ADA deficiency leads to low levels of B and T cells in the child's immune system.
- Janus Kinase 3 (Jak3) deficiency: This form of SCID accounts for about 6 percent of cases. There are very low levels of T and NK cells, or they are not present at all. There is an elevated level of B cells. In this form of SCID the lymphocyte or white blood evaluation is identical to X-linked SCID but is autosomal recessive and, therefore, occurs in girls and boys.

Demographics

The rate of SCID is not perfectly documented. It is estimated that it occurs in between one in 50,000 and one in 500,000 infants. It is about three times more common in boys than in girls.

Causes and symptoms

SCID is an inherited disorder. In all forms of SCID, B and T cells are non-functioning. They may or may not be present in various forms of SCID, but they are always non-functioning. In some forms of SCID, NK cells are also absent or non-funtioning.

For the first few months after birth, a infant with SCID is often protected by antibodies acquired before birth from the mother's blood. As early as three months of age, however, the SCID child begins to suffer from mouth infections (thrush), chronic **diarrhea**, **otitis media**, and pulmonary infections, including pneumocystis **pneumonia**. The child loses weight, becomes very weak, and if untreated eventually dies from an opportunistic infection.

When to call the doctor

If a child has unusual infections, unusually severe infections, infections with unusual organisms, or unusual complications of usual infections, a doctor should be consulted to evaluate for possible immune deficiency. This is particularly important if there is a **family** history of immune deficiency.

Diagnosis

The first screening test for SCID is a white blood cell count with a count of the lymphocytes (differential) because in most forms of SCID the lymphocyte count will be very low. Blood tests can then be done to test for the numbers of B, T, and NK type lymphocytes. If the numbers of all of these cell types are normal and SCID is still suspected, more specialized tests can be done to test the lymphocyte cell functions. Rarely there are children with SCID who have normal lymphocyte numbers and nonfunctioning cells.

Treatment

Patients with SCID should be treated aggressively with **antibiotics** for any infection, and intravenous immunoglobulin should be given to replace the antibiotics the children cannot make, but these treatments cannot cure the disorder. Bone marrow transplants are as of 2004 regarded as one of the few effective standard treatments for most types of SCID. For those children with ADA deficiency, ADA infusions are the accepted treatment of choice. Up to 95 percent of children who are treated with bone marrow transplants, especially those who are treated before three months of age, survive.

Investigational treatments

As an example of gene therapy for SCID children with ADA deficiency, the child receives periodic infusions of his or her own T cells corrected with a gene for ADA that has been implanted in an activated virus. This should allow these cells to function normally. Other types of SCID have been treated with gene therapy, but these procedures have been put on hold due to serious complications (malignancies). Researchers are as of 2004 also investigating treating SCID in the yet unborn fetus, which has been done successfully a few times.

Prognosis

There is no cure for SCID. Nearly all untreated patients die before age two, most before one year of age. Children who are treated with bone marrow transplants have a much better prognosis.

Prevention

There is no known way to prevent SCID. Genetic counseling is recommended for parents of a child with SCID who are considering having more children and for potential parents who have a family history of the disease and believe they may be carriers.

Parental concerns

Without prompt treatment SCID is nearly always fatal. Treatment can be very successful if done early, preferably within the first three months of life. Research is

KEY TERMS

Adenosine deaminase (ADA)—An enzyme that is lacking in a specific type of severe combined immunodeficiency disease (SCID). Children with an ADA deficiency have low levels of both B and T cells.

Antigen—A substance (usually a protein) identified as foreign by the body's immune system, triggering the release of antibodies as part of the body's immune response.

B cell—A type of white blood cell derived from bone marrow. B cells are sometimes called B lymphocytes. They secrete antibodies and have a number of other complex functions within the human immune system.

Congenital—Present at birth.

Gene therapy—An experimental treatment for certain genetic disorders in which a abnormal gene is replaced with the normal copy. Also called somatic-cell gene therapy.

Lymphocyte—A type of white blood cell that participates in the immune response. The two main groups are the B cells that have antibody molecules on their surface and T cells that destroy antigens.

T cell—A type of white blood cell that is produced in the bone marrow and matured in the thymus gland. It helps to regulate the immune system's response to infections or malignancy.

Thrush—An infection of the mouth, caused by the yeast *Candida albicans* and characterized by a whitish growth and ulcers.

continuing into in utero treatment options, and some in utero treatments have been successfully carried out, so fetal screening may be helpful if there is a possibility that the child has SCID.

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Sexual abuse *see* **Rape and sexual assault**

Sexually transmitted diseases

Definition

Sexually transmitted diseases (STDs) are viral and bacterial infections passed from one person to another through sexual contact.

Description

Adolescence is a time of opportunities and risk when many health behaviors are established. Although many of these behaviors are health-promoting, some are health-compromising, resulting in increasingly high rates of adolescent morbidity and mortality. For example, initiation of sexual intercourse and experimentation with alcohol and drugs are normative adolescent behaviors. However, these behaviors often result in negative health outcomes such as the acquisition of STDs. As a consequence of STDs, many adolescents experience serious health problems that often alter the course of their adult lives, including infertility, difficult pregnancy, genital and cervical **cancer**, neonatal transmission of infections, and **AIDS** (acquired **immunodeficiency** syndrome).

Examples of STDs with high prevalence among sexually-active adolescents include:

- Gonorrhea: Caused by the bacteria *Neisseria gonorrhoeae*, gonorrhea infects the reproductive tract of women, causing pelvic inflammatory disease (PID), a major cause of infertility. The bacteria are found in vaginal secretions and semen.
- Chlamydia: The bacteria that causes chlamydia, *Chlamydia trachomatis*, trigger inflammation of the reproductive tract, leading to PID in women and epididymitis (inflammation of the epididymis) in men.

- Syphilis: *Treponema pallidum* is the bacteria that causes syphilis. The course of syphilis is broken down into four distinct segments: primary syphilis, occurring within a few weeks or months of initial exposure; secondary syphilis, occurring generally between six weeks and six months of initial exposure; latent syphilis, an asymptomatic period which may stretch for years; and late syphilis, the most serious stage. If left untreated, syphilis can infect a number of organ systems and cause serious complications.
- Herpes simplex virus: Two different types of HSV (HSV-1 and HSV-2) cause lesions on the genitals, although HSV-2 is associated with the majority of cases. (HSV-1 is most commonly associated with oral lesions, or "cold sores.")
- Human papillomavirus (HPV): HPV causes condylomata acuminata, more commonly known as venereal **warts** or genital warts. The warts may affect any of the external and internal genital organs in men and women.
- Human immunodeficiency virus (HIV). HIV is the causative agent of acquired immune deficiency syndrome (AIDS), a potentially fatal condition in which the immune system fails and the individual becomes prone to frequent and unusual infections.

Transmission

The mode of transmission varies among the different sexually transmitted diseases. Some bacteria or virus are found in vaginal secretions or semen (e.g. HIV and gonorrhea), while others are shed from the skin of and around the genitals (e.g. HSV and HPV). Infection typically occurs during sexual intercourse or when the genitals come into close contact. Infection may also occur during oral sex, such as transmission of HSV from an oral lesion to the genitals or vice versa, or transmission of HIV from genital secretions through a cut in the mouth. STDs may be transmitted during nonconsensual sex acts such as **rape** or molestation.

The transmission of many STDs is more efficient from men to women than from women to men. For example, with just one unprotected sexual encounter with an infected partner, a woman is twice as likely as a man to acquire gonorrhea or chlamydia. In addition, different STDs have different rates of transmissibility. For example, with one exposure of unprotected sexual intercourse, a woman has a 1 percent chance of acquiring HIV, a 30 percent chance of acquiring herpes, and 50 percent chance of contracting gonorrhea if her partner is infected.

Demographics

STDs among sexually experienced adolescents occur at alarmingly high rates. One-fourth of the estimated 12 million new cases reported annually occur among adolescents between 15 and 19 years of age. Moreover, since many STDs are asymptomatic, they are often undiagnosed and untreated, thus increasing their potential for proliferation among adolescents.

Gonorrhea and chlamydia, the most prevalent bacterial STDs, disproportionately affect adolescents. The rates of gonorrhea in adolescents ages 15 to 19 years declined between 1990 and 2004, but in the early 2000s they continue to be higher than rates for any five-year age group between 20 and 44 years, particularly among women and African Americans.

Numerous prevalence studies for chlamydia have shown rates to be highest among adolescents and young adults under 25 years of age, many of whom are minorities. Rates of chlamydia reported by gender indicate that women, overall, have higher rates than men due in large part to increased efforts in screening women for asymptomatic chlamydial infections. The low rates of chlamydia for men suggest that the sexual partners of women diagnosed with chlamydia are not being diagnosed or treated. Chlamydia has been detected in more than 10 percent of sexually experienced women during screening.

While rates of syphilis declined between 1990 and 2004, the disease continues to be an important cause of sexually transmitted infection. The rate of syphilis infection among adolescents ages 15 to 19 is 1.3 per 100,000 population for males and 2.2 per 100,000 population for females. For comparison, the syphilis rates among males 20 to 24 is 5.5 per 100,000, and among females of the same age, 3.3 per 100,000.

HSV and HPS occur at alarming rates among sexually experienced adolescents. Studies indicate that one in six Americans is infected with HSV-2, reflecting a ninefold increase between 1975 and 2005. Prevalence of HSV-2 in adolescents and young adults varies by the demographic and behavioral characteristics of the populations studied as well as the diagnostic methods used. As of the early 2000s approximately 4 percent of Caucasians and 17 percent of African Americans are infected with HSV-2 by the end of their teenage years. One study of young pregnant women of low income status found an HSV-2 infection rate of 11 percent in women 15 to 19 years of age and 22 percent in women 25 to 29 years of age.

In 2002, there were 4,785 reported cases of AIDS among teenagers between the ages of 13 and 19, more

than double the 1994 figures. Most adolescents with AIDS were infected as a result of high risk sexual and substance use behaviors. Among adolescents ages 13 to 19 years infected with HIV, 49 percent are male and 51 percent are female. Studies also indicate that African-American and Latino teens are overrepresented among persons with AIDS relative to their proportion in the population. Although these epidemiological statistics on AIDS in the United States provide a descriptive overview of the prevalence and patterns of HIV exposure in adolescents, the extent of asymptomatic **HIV infection** remains largely unknown.

Causes and symptoms

The chance for adolescents of getting and transmitting STDs is affected by complex interrelationships between key factors (sociodemographic, biologic, psychosocial, and behavioral). For example, many STD-related risk markers (e.g. age, gender, race/ethnicity) correlate with more fundamental determinants of risk status (e.g., access to health care, living in communities with high prevalence of STDs) to influence adolescents' risk for STDs.

Developmental factors such as pubertal timing, **self-esteem**, and peer affiliation may also increase their risk of exposure to STDs. An assessment of these interrelationships is critical to preventing and controlling STDs in adolescents. Moreover, since behavior is the common means by which STDs occur, an important first step in fighting STDs is to understand the prevalence and patterns of risk behaviors as well as the psychosocial context in which these behaviors occur.

Behavioral factors

Although biologic factors play an important role in the transmission of STDs, it is also the health-risking behaviors of adolescents that place them at increased risk for exposure to STDs. Behavioral risk factors include the age of sexual activity, number of sexual partners, use of contraceptives, and use of alcohol and drugs.

SEXUAL ACTIVITY Early initiation of sexual intercourse has been associated with high-risk sexual activities, including ineffective use of contraceptives, multiple sex partners over a short period of time, high-risk sex partners, and acquisition of STDs and their consequences of cervical cancer and dysplasia. The average age of first sexual intercourse is between 16 and 17 years for adolescent men and between the age of 17 and 18 years for adolescent women, and has been found to be as young as age 12 in some high-risk populations. Research on adolescents' decision to initiate sexual intercourse indicates an interaction between biological and social factors. However, much remains unknown about the interactions between hormones, behavior, and social factors.

The Youth Risk Behavior Surveillance System (YRBSS), a self-reported survey of a national representative sample of high school students in grades nine to 12, indicated that in 2003, 46.7 percent of the students reported having had sex. By grade level, the rates were 32.8 percent for ninth grade, 44.1 percent for tenth grade, 53.2 percent for eleventh grade, and 61.6 percent for twelfth grade. Approximately 7.4 percent of students reported having sex for the first time before age 13. Prevalence rates of sexual experience differed by race/ethnicity and gender. African-American students were significantly more likely (73.8% of males and 60.9% of females) than Caucasian (40.5% of males and 43.0% of females) and Hispanic (56.8% of males and 46.4% of females) students to have engaged in sexual intercourse. Moreover, data from the National Survey of Family Growth (NSFG), a large-scale national survey of women ages 15 to 44 years, reveal that family income is associated with adolescents' protection against HIV and many other STDs; adolescents from poor and low-income families are more likely to report an earlier age of sexual experience than their counterparts from higher income families.

In addition to early sexual activity, many adolescents have multiple sex partners within a short period of time in a pattern of serial monogamy which also increases their risk of acquiring STD for two important reasons: it increases the likelihood of being exposed to a sexually transmitted pathogen, and it may reflect poor choices of sexual partners. Among the sexually experienced high school students responding to the YRBSS, 14.4 percent reported having four or more sex partners. Multiple sex partners were noted more frequently among African-American students (41.7% of males and 16.3% of females), compared to Hispanic (20.5% of males and 11.2% of females) and Caucasian (11.5% of males and 10.1% of females) students.

Involuntary sexual intercourse such as rape and sexual abuse may occur more commonly among adolescents, especially younger adolescent women, and often pose a potential risk for acquisition of STDs. A study on the effects of **child abuse** (i.e., incest, extra-familial sexual abuse, and physical abuse) on adolescent males showed a strong association between abuse and a number of risk-taking behaviors, such as forcing female sexual partners into having sexual intercourse and drinking alcohol prior to sexual intercourse. Moreover, when sexual intercourse is intermittent, as it is with most sexually

experienced adolescents, the adolescents are less likely to take proper measures to safeguard against STDs.

CONTRACEPTIVE USE Sexually experienced adolescents are also at risk for STDs because of their patterns of contraceptive use, especially their use of barrier-method contraceptives. Some data indicate that adolescents do not use effective methods to reduce their risk of STDs or unintended pregnancies. Sexual abstinence is the only sure method of eliminating risk for STDs. When used consistently and correctly, however, condoms offer the best protection against acquisition of STDs, including HIV. Even when condoms are used improperly they reduce the risk of acquiring infections by 50 percent.

The overall reported use of contraceptives, particularly condoms, has increased among adolescents between 1994 and 2004. Data from the 2003 YRBSS reveal that 63.0 percent of the students who reported sexual activity in the three months prior to the survey also reported using condoms during their last sexual encounter; this behavior was more common among males of virtually all ages and racial/ethnic groups. In contrast, 20.6 percent of adolescent women ages 15 to 19 years reported use of birth control pills. It appears that while the use of **oral contraceptives** provides some protection against the development of gonococcal and nongonococcal forms of PID, it may increase the risk of chlamydial endocervical infections, and provides no protection against most STDs.

Differences in the types and patterns of contraceptive use by race/ethnicity, age, and socioeconomic status have also been noted. Also, adolescent women of higher income are more likely than young women of lower income to use oral contraceptives. These factors are related to access and use of medical services for reproductive health care. Thus, providing all sexually experienced adolescents with reproductive health counseling and education about the importance of consistently and correctly using barrier-method contraceptives such as condoms may play a crucial role in reducing their risk of acquiring and transmitting STDs.

ALCOHOL AND OTHER DRUG USE Use of alcohol and other drugs is prevalent among adolescents and thus poses a significant threat to their health. About 40 percent of high school youth responding to the YRBSS have used marijuana at least once with 22.4 percent of these students reporting use of this substance within 30 days before the survey. Cocaine was used at least once by 8.7 percent of the students and by 4.1 percent within 30 days of the survey. The substance of choice, however, is alcohol: 74.9 percent of students had at least one drink at some point in time and nearly half (44.9%) consumed alcohol in the 30 days prior to the survey. Among the current alcohol users, 28.3 percent had five or more drinks on at least one occasion, suggesting that a sizeable proportion of the students are periodic heavy drinkers. Grade, age, and gender differences were noted for lifetime and current use of alcohol and other illicit substances. In general, students in higher grade levels (grades 11 and 12) and males were more likely to use all substances. Racial/ethnic differences in use of substances were also found. Heavy use of alcohol was most prevalent among Caucasian and Hispanic males and females, while marijuana use was most common among African-American and Hispanic males.

Although these data strongly suggest that adolescents are at increased risk for social and physical morbidities, and even premature mortality because of their use of alcohol and other illicit substances, they underrepresent the actual prevalence of substance use among all adolescents. Teens who have dropped out or who are repeatedly absent from school and those who are homeless or otherwise disenfranchised are not represented by the reported data; many of these teens are potentially at higher risk for STDs because of their substance use behavior.

Substance use prior to sexual intercourse is likely to be related to a number of risk-taking behaviors: sexual intercourse with a casual acquaintance, lack of communication about use of condoms or previous sexual experiences, and no use of condoms. This association remained significant regardless of demographic factors, sexual experience, and dispositional factors such as adventure and thrill seeking. It appears that early intervention to prevent the use and abuse of alcohol and other substances may significantly decrease their risk of acquiring STDs.

Psychosocial factors

One study of college students examined the relationship between sexual behavior, substance use, and specific constructs from social cognitive theory (i.e., perceptions of self-efficacy, vulnerability to HIV risk, social norms, negative outcome expectancies of condoms, and knowledge of HIV risk and prevention). The results indicate that although young men expected more negative outcomes of **condom** use and were more likely to have sexual intercourse under the influence of alcohol and other drugs, young women reported perceptions of higher self-efficacy to practice safer sex. The study further revealed that perceptions of higher self-efficacy to engage in safer sexual behaviors, perceptions of fewer negative outcomes of condom use, and less frequent alcohol and drug use with sexual intercourse were the best predictors of safer sexual behaviors.

Evaluating STD risk

The information, motivation, and behavioral skills (IMB) model is one method of evaluating risk for STDs. This model posits that information, motivation, and behavior are the primary determinants of AIDS-related preventive behavior. Specifically, the model asserts that information regarding the transmission of HIV and information concerning specific methods of preventing HIV (e.g., condom use, decreasing the number of partners) are necessary prerequisites of reducing risk behaviors.

Motivation to change risk behaviors is another determinant of prevention and affects whether a person acts on his or her knowledge of the transmission and prevention of HIV. The IMB contends that motivation to engage in prevention behaviors is a function of one's attitudes toward the behavior and of subjective norms regarding prevention behaviors. Other critical factors which are hypothesized to influence motivation to engage in prevention behaviors are perceived vulnerability to acquiring HIV, perceived costs and benefits of engaging in prevention behaviors, intention to engage in prevention behaviors regarding HIV, as well as characteristics of the sex partner and/or the sexual relationship (e.g. primary vs. secondary partner).

Behavioral skills for engaging in specific prevention behaviors are a third determinant of prevention; it affects whether a knowledgeable, highly motivated person will be able to change his or her behavior to prevent HIV. Important skills required to engage in prevention behaviors include the ability to effectively communicate with one's sex partner about safer sex, refusal to engage in unsafe sexual practices, proper use of barrier-method contraceptives, and the ability to exit a situation when prevention behaviors are not possible. In addition, individuals who are able to practice prevention skills are presumed to have a strong belief in their ability to practice these prevention behavioral skills. Overall, the IMB asserts that information and motivation trigger behavioral skills to affect the initiation and maintenance of HIV prevention behaviors.

Symptoms of common STDs

The symptoms of some STDs may seriously affect an infected individual's quality of life or eventually become fatal, while others are so mild as to go undetected. The symptoms of some of the more prevalent STDs include:

- Gonorrhea: The most common symptoms among infected adolescent girls are vaginal discharge, bleeding between menstrual cycles, and painful urination. Among adolescent boys, common symptoms are burning or painful urination and pus-like discharge from the penis. Many infections, however, remain asymptomatic in both females (32%) and males (2%). Symptoms are similar among young children who have contracted gonorrhea from a sexual abuser.
- Chlamydia: Symptoms of chlamydia are similar to those of gonorrhea and sometimes difficult to differentiate clinically. Chlamydial infections are more likely to be asymptomatic than gonorrheal infections and thus are of longer duration on average.
- Syphilis: In primarily syphilis, the characteristic symptom is the appearance of a chancre (painless ulcer) at the site of initial exposure (e.g. external genitalia, lips, tongue, nipples, or fingers). In some cases, the infected individual will experience swollen lymph glands. In secondary syphilis, the infection becomes systemic and the individual experiences symptoms such as **fever**, **headache**, **sore throat**, rash, and swollen glands. During latent syphilis, symptoms go unnoticed. During the late stage of syphilis, the infection has spread to organ systems and may cause blindness, signs of damage to the nervous system and heart, and skin lesions.
- Herpes simplex virus: The symptoms of genital herpes include burning and **itching** of the genital area, blisters or sores on the genitals, discharge from the vagina or penis, and/or flu-like symptoms such as headache and fever.
- Human papillomavirus (HPV): The warty growths of HPV can appear on the external or internal reproductive organs of males and females but are commonly found on the labia minora and the opening to the vagina in females and the penis in males. They may be small and few or combine to form larger growths.
- Human immunodeficiency virus (HIV): Some persons who are newly infected with HIV have rash, fever, enlarged lymph nodes, and a flu-like illness sometimes called HIV seroconversion syndrome. This initial syndrome passes without intervention, and later symptoms, when T-cells become depleted, include weight loss, chronic **cough**, fever, fatigue, chronic **diarrhea**, swollen glands, white spots on the tongue and inside of the mouth, and dark blotches on the skin or in the mouth.

When to call the doctor

If a child or adolescent develops any of the symptoms of STDs, he or she should be evaluated for possible infection. Routine pelvic exams are recommended for all sexually active females and all females over the age of 18.

Diagnosis

A history of sexual activity is collected from all individuals at increased risk of contracting an STD, including adolescents who admit to being sexually active or who are pregnant or have undergone therapeutic abortion, adolescents or children with symptoms indicative of infection with an STD, and adolescents or children suspected of being victims of sexual abuse or rape. The healthcare provider will take a complete medical history and perform a thorough physical examination. Depending on the STD in question, additional tests may be performed such as blood work, Papanicolaou (pap) smear, rectal swabs, or biopsy.

Treatment

The treatment of sexually transmitted diseases varies according to the diagnosed infection. Gonorrhea, chlamydia, and syphilis are curable in most cases with **antibiotics**, although antibiotic-resistant strains do exist. As viruses, HSV, HPV, and HIV are treatable but not curable. The frequency and duration of HSV lesions can be reduced with antiviral therapy, including acyclovir (Zovirax), famciclovir (Famvir), and valacyclovir (Valtrex). Common methods to reduce genital warts include application of a topical cream called imiquimod (Aldara), cryotherapy (freezing of the wart), elecrosurgery (applying an electrical current to the wart), and surgical removal. The course of HIV infection can be slowed with a number of different kinds of drugs, including reverse transcriptase inhibitors, protease inhibitors, nonnucleoside reverse transcriptase inhibitors, and fusion inhibitors.

Alternative treatment

A number of different alternative therapies may be pursued to treat STDs, such as the use of herbs, homeopathy, acupuncture, and nutritional supplements, although minimal research has been done to establish their efficacy.

Nutritional concerns

In some cases, supplementation with specific nutrients may enhance immunity and minimize outbreaks. Examples are vitamin C (to boost the immune system), zinc (to reduce the frequency of HSV outbreaks), aloe (a possible antiviral), lemon balm (to speed healing), and licorice (with anti-inflammatory and antiviral effects).

Prognosis

Most STDs have excellent prognoses and respond well to treatment. While HSV and HPS are not curable, outbreaks can be managed and infection generally has little effect on quality of life. HIV, however, is a potentially fatal disease which can be treated but not cured.

Prevention

The prevalence data on STDs, HIV, and AIDS in adolescents indicate that younger women, gay and bisexual teens, and poor, urban and racial/ethnic minority young people have higher rates of STDs and HIV relative to their peers. Primary prevention of initial STD infections through prevention and risk reduction programs are essential for stemming the tide of these sexually acquired diseases. Moreover, secondary prevention through screening at risk adolescents for asymptomatic STD infections and effectively treating the index case and his or her sexual contact(s) are the most effective means of eliminating long-term medical and psychosocial consequences from STDs.

Prevention of high risk sexual, contraceptive, and substance use behaviors through cognitive-behavioral skills training and prevention and risk reduction counseling programs is a key strategy for decreasing the high incidence of STDs in adolescents. Prevention and risk reduction strategies should be developed and implemented in settings in which most adolescents can be reached, including schools or community-based programs in which there are multiple opportunities to intervene with adolescents or clinical settings where one-to-one risk reduction counseling can occur and actual risk can be assessed.

Cognitive-behavioral skills building interventions

In order to prevent new STD infections, adolescents must not only be informed about the risk and prevention of STDs, they must also have skills to resist **peer pressure**, negotiate the use of condoms, and project the future consequences of their behaviors. In addition, prevention of STDs in adolescents requires that they have the necessary means, resources, and social support to develop self-regulative skills and self-efficacy to effectively reduce their risk of disease transmission. Such cognitive-behavioral skills building programs have been shown to be effective in developing skills, delaying the onset of sexual activity, and changing high risk behaviors associated with pregnancy, STDs, and HIV infection. Moreover, cognitive-behavioral skills building programs should be immediate, sustained, and cost-effective. Specifically, these programs should be

designed to increase knowledge about the prevention and transmission of STDs and their consequences; formulate realistic attitudes and perceptions about personal susceptibility to acquiring infections; enhance self-efficacy and self-motivation; monitor and regulate STD-related risk behaviors; address the role of social peer norms; and develop appropriate decision-making, problem-solving, and **communication skills**.

Prevention and risk reduction counseling

Counseling strategies to prevent and reduce the risk of STDs should be conducted in a confidential and nonjudgmental manner that is both developmental and culturally appropriate for the adolescent. Counseling should focus on a number of key elements such as maintenance and support of healthy sexual behaviors (e.g. delaying initiation of sexual intercourse, limiting the number of sexual partners), use of barrier-method contraceptives (e.g. condoms, diaphragms, spermicide), routine medical care and advice (e.g. seeking medical care if the adolescent has participated in high-risk behavior), compliance with treatment recommendations (e.g. taking all medications as directed), and encouraging sex partners to seek medical care. Adolescents should also be informed about the myths and misconceptions of acquiring STDs. Moreover, adolescents should receive anticipatory guidance to assist them in defining appropriate options and alternatives to engaging in high-risk behaviors.

Parental concerns

Parents should be encouraged to talk to their children about sexually transmitted diseases and the risks of sexual activity. By asking preteens or teenagers questions about what they knows about STDs or by using cues from television shows or newspaper articles, parents can help make their children more comfortable talking about sex and the risks of infection, thereby opening the lines of communication. It is important that adolescents be provided accurate information, even if they already have some knowledge on the topic. Research has shown teens are not more likely to have sex if they are informed about safe sex practices, but they are more likely to practice safer sex.

KEY TERMS

Opportunistic infection—An infection that is normally mild in a healthy individual, but which takes advantage of an ill person's weakened immune system to move into the body, grow, spread, and cause serious illness.

Pap test—A screening test for precancerous and cancerous cells on the cervix. This simple test is done during a routine pelvic exam and involves scraping cells from the cervix. These cells are then stained and examined under a microscope. Also known as the Papanicolaou test.

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Shaken baby syndrome

Definition

Shaken baby syndrome (SBS) is a collective term for the internal head injuries a baby or young child sustains from being violently shaken.

Description

Shaken baby syndrome was first described in medical literature in 1972. Physicians earlier labeled these injuries as accidental, but as more about **child abuse** became known, more cases of this syndrome were properly diagnosed.

Demographics

Every year, nearly 50,000 children in the United States are forcefully shaken by their caretakers. More than 60 percent of these children are boys. Nearly 2,000 children die every year as a result of being shaken. The victims are on average six to eight months old, but may be as old as five years or as young as a few days.

Men are more likely than women to shake a child; typically, these men are in their early 20s and are the baby's father or the mother's boyfriend. Women who inflict SBS are more likely to be **babysitters** or child care providers than the baby's mother. The shaking may occur as a response of frustration to the baby's inconsolable crying or as an action of routine abuse.

Causes and symptoms

Infants and small children are especially vulnerable to SBS because their neck muscles are still too weak to adequately support their disproportionately large heads, and their young brain tissue and blood vessels are extremely fragile. When an infant is vigorously shaken by the arms, legs, shoulders, or chest, the whiplash motion repeatedly jars the baby's brain with tremendous force, causing internal damage and bleeding. While there may be no obvious external signs of injury following shaking, the child may suffer internally from brain bleeding and bruising (called subdural hemorrhage and hematoma); brain swelling and damage (called cerebral edema); **mental retardation**; blindness, hearing loss, paralysis, speech impairment, and learning disabilities; and death.

Physicians may have difficulty initially diagnosing SBS because there are usually few witnesses to give a reliable account of the events leading to the trauma, few if any external injuries, and, upon close examination, the physical findings may not agree with the account given. A shaken baby may present one or more signs, including **vomiting**; difficulty breathing, sucking, swallowing, or making sounds; seizures; and altered consciousness.

When to call the doctor

A physician should be called when a baby exhibits one or more of the following: vomiting; difficulty breathing, sucking, swallowing, or making sounds; seizures; and altered consciousness. An unresponsive child should never be put to bed, but must be taken to a hospital for immediate care.

Diagnosis

To diagnose SBS, physicians look for at least one of three classic conditions: bleeding at the back of one or both eyes (retinal hemorrhage), **subdural hematoma**, or cerebral edema. The diagnosis is confirmed by the results of either a **computed tomography** scan (CT scan) or **magnetic resonance imaging** (MRI).

Treatment

Appropriate treatment is determined by the type and severity of the trauma. Physicians may medically manage both internal and external injuries. Behavioral and educational impairments as a result of the injuries require the attention of additional specialists. Children with SBS may need physical therapy, speech therapy, vision therapy, and **special education** services.

Prognosis

Unfortunately, children who receive violent shaking have a poor prognosis for complete recovery. Those who do not die may experience permanent blindness, mental retardation, seizure disorders, or loss of motor control.

Prevention

Shaken baby syndrome is preventable with public education. Adults must be actively taught that shaking a child is never acceptable and can cause severe injury or death.

Parental concerns

When the frustration from an incessantly crying baby becomes too much, caregivers should have a strategy for coping that does not harm the baby. The first step is to place the baby in a crib or playpen and leave the room in order to calm down. Counting to 10 and taking deep breaths may help. A friend or relative may be called to come over and assist. A calm adult may then resume trying to comfort the baby. A warm bottle, a dry diaper, soft music, a bath, or a ride in a swing, stroller, or car may be offered to soothe a crying child. Crying may also indicate **pain** or illness, such as from abdominal cramps or an earache. If the crying persists, the child should be seen by a physician.

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KEY TERMS

Cerebral edema—The collection of fluid in the brain, causing tissue to swell.

Hematoma—A localized collection of blood, often clotted, in body tissue or an organ, usually due to a break or tear in the wall of blood vessel.

Hemorrhage—Severe, massive bleeding that is difficult to control. The bleeding may be internal or external.

Retinal hemorrhage—Bleeding of the retina, a key structure in vision located at the back of the eye.

Subdural hematoma—A localized accumulation of blood, sometimes mixed with spinal fluid, in the space between the middle (arachnoid) and outer (dura mater) membranes covering the brain. It is caused by an injury to the head that tears blood vessels.

Shigellosis

Definition

Shigellosis is an infection of the intestinal tract by a group of bacteria called *Shigella*.

Description

Shigellosis is a well-known cause of traveler's **diarrhea** and illness throughout the world. The major symptoms of shigellosis are diarrhea, abdominal cramps, **fever**, and severe fluid loss (**dehydration**). The bacteria causing shigellosis is named after Shiga, a Japanese researcher, who discovered the organism in 1897. Four different groups of *Shigella* can affect humans; of these, *S. dysenteriae* generally produces the most severe attacks, and *S. sonnei* the mildest. Shigella are extremely infectious bacteria, and ingestion of just 10 organisms is enough to cause dysentery. The most serious form of the disease is called dysentery, which is characterized by severe watery (and often blood- and mucus-streaked) diarrhea, abdominal cramping, rectal **pain**, and fever. *Shigella* is only one of several organisms that can cause dysentery, but the term bacillary dysentery is usually another name for shigellosis.

Demographics

Shigella accounts for 10 to 20 percent of all cases of diarrhea worldwide, and in any given year infects over 140 million persons and kills 600,000, mostly children and the elderly.

Most deaths are in less-developed or developing countries, but even in the United States, shigellosis can be a dangerous and potentially deadly disease. Poor hygiene, overcrowding, and improper storage of food are leading causes of infection. Shigellosis is often passed within families when young children are not yet toilet-trained and hand washing is poorly done. The following statistics show the marked difference in the frequency of cases between developed and less-developed countries: in the United States, about 30,000 individuals are hit by the disease each year or about 10 cases per 100,000 population. By contrast, infection in some areas of South America is 1,000 times more frequent. Shigellosis is most common in children below the age of five years and occurs less often in adults over 20.

Causes and symptoms

Shigella share several of the characteristics of a group of bacteria that inhabit the intestinal tract. E. coli, another cause of food-borne illness, can be mistaken for *Shigella* both by physicians and the laboratory. Careful testing is needed to assure proper diagnosis and treatment.

Shigella are very resistant to the acid produced by the stomach, and this allows them to easily pass through the gastrointestinal tract and infect the colon (large intestine). The result is a colitis that produces multiple ulcers, which can bleed. *Shigella* also produce a number of toxins (Shiga toxin and others) that increase the amount of fluid secretion by the intestinal tract. This fluid secretion is a major cause of the diarrhea symptoms.

Shigella infection spreads through food or water contaminated by human waste. Sources of transmission are as follows:

- contaminated milk, ice cream, vegetables, and other foods which often cause epidemics
- household contacts (40% of adults and 20% of children develop infection from such a source)
- poor hygiene and overcrowded living conditions
- daycare centers
- sexual practices which lead to oral-anal contact, directly or indirectly

Symptoms can be limited to mild diarrhea or develop into dysentery. Dehydration results from the large fluid losses due to diarrhea, **vomiting**, and fever. Inability to eat or drink worsens the situation.

In developed countries, most infections are of the less severe type and are often due to *S. sonnei*. The period between infection and symptoms (incubation period) varies from one to seven days. Shigellosis can last from a few days to several weeks, with the average duration seven days.

Complications

Areas outside the intestine can be involved, including the following:

- nervous system (irritation of the meninges or **meningitis**, **encephalitis**, and seizures)
- kidneys (producing hemolytic uremic syndrome or HUS which leads to kidney failure)
- joints (leading to an unusual form of arthritis called Reiter's syndrome)
- skin (rash)

One of the most serious complications of this disease is HUS, which involves the kidney. The main findings are kidney failure and damage to red blood cells. As many as 15 percent of patients die from this complication, and half the survivors develop chronic kidney failure, requiring dialysis.

Another life-threatening condition is toxic megacolon. Severe inflammation causes the colon to dilate or stretch, and the thin colon wall may eventually tear. Certain medications (particularly those that diminish intest-

inal contractions) may increase this risk, but this interaction is unclear. Clues to this diagnosis include sudden decrease in diarrhea, swelling of the abdomen, and worsening abdominal pain.

Diagnosis

Shigellosis is one of the many causes of acute diarrhea. Culture (growing the bacteria in the laboratory) of freshly obtained diarrhea fluid is the only way to be certain of the diagnosis. But even this is not always positive, especially if the patient is already on **antibiotics**. *Shigella* are identified by their appearance under the microscope, along with various chemical tests. These studies take several days.

Treatment

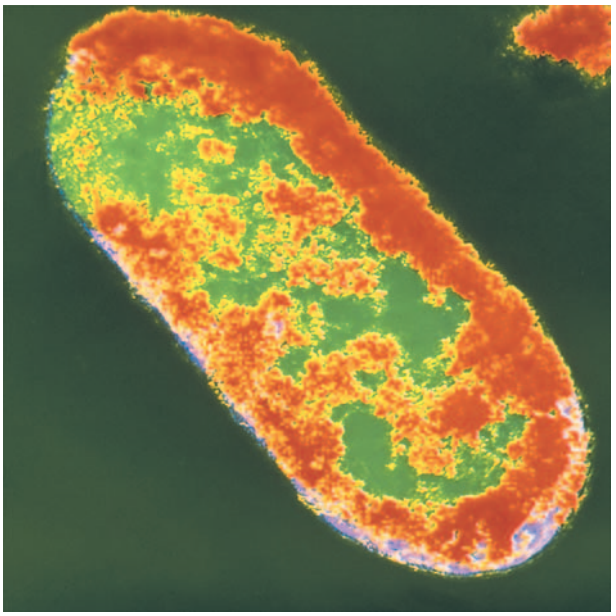
The first aim of treatment is to keep up **nutrition** and avoid dehydration. Ideally, a physician should be consulted before starting any treatment. Antibiotics may not be necessary, except for the more severe infections. Many cases resolve before the diagnosis is established by culture. Medications that control diarrhea by slowing intestinal contractions can cause problems and should be avoided by patients with bloody diarrhea or fever, especially if antibiotics have not been started.

Rehydration

The World Health Organization (WHO) has developed guidelines for a standard solution taken by mouth and prepared from ingredients readily available at home. This oral rehydration solution (ORS) includes salt, baking powder, sugar, orange juice, and water. Commercial preparations, such as Pedialyte, are also available. In many patients with mild symptoms, this is the only treatment needed. Severe dehydration usually requires intravenous fluid replacement.

Antibiotics

Patients who have very mild cases of shigellosis may improve without any antibiotic therapy; therefore, these drugs are indicated only for treatment of moderate or severe disease, as found in the tropics. Choice of antibiotic is based on the type of bacteria found in the geographical area and on laboratory results. Recommended antibiotics include ampicillin, sulfa derivatives such as trimethoprim-sulfamethoxazole (TMP-SMX) sold as Bactrim, or fluoroquinolones, such as Ciprofloxacin.



A transmission electron microscopy (TEM) scan of *Shigella*, a genus of aerobic bacteria that causes dysentery in humans and animals. *(Custom Medical Stock Photo Inc.)*

Prognosis

Many patients with mild infections need no specific treatment and recover completely. In those with severe infections, antibiotics decrease the length of symptoms and the number of days bacteria appear in the feces. In rare cases, an individual may fail to clear the bacteria from the intestinal tract; the result is a persistent carrier state. This may be more frequent in patients with acquired immune deficiency syndrome (**AIDS**). Antibiotics are about 90 percent effective in eliminating these chronic infections.

In patients who have suffered particularly severe attacks, some degree of cramping and diarrhea can last for several weeks. This is usually due to damage to the intestinal tract, which requires some time to heal. Since antibiotics can also produce a form of colitis, which may cause persistent or recurrent symptoms.

Prevention

Shigellosis is an extremely contagious disease; good hand washing techniques (especially after toileting young children or changing diapers) and proper precautions in food handling help in avoiding the spread of infection. Children in daycare centers need to be reminded about hand washing during an outbreak to minimize contagion. Shigellosis in schools or daycare

KEY TERMS

Antibiotics—Drugs that are designed to kill or inhibit the growth of the bacteria that cause infections.

Antimotility drug—A medication, such as loperamide (Imodium), dephenoxylate (Lomotil), or medications containing codeine or narcotics that decrease the ability of the intestine to contract.

Carrier state—The continued presence of an organism (bacteria, virus, or parasite) in the body that does not cause symptoms, but is able to be transmitted and infect other persons.

Colitis—Inflammation of the colon (large intestine).

Dialysis—A process of filtering and removing waste products from the bloodstream, it is used as a treatment for patients whose kidneys do not function properly. Two main types are hemodialysis and peritoneal dialysis. In hemodialysis, the blood flows out of the body into a machine that filters out the waste products and routes the cleansed blood back into the body. In peritoneal dialysis, the cleansing occurs inside the body. Dialysis fluid is injected into the peritoneal cavity and wastes are filtered through the peritoneum, the thin membrane that surrounds the abdominal organs.

Dysentery—A disease marked by frequent watery bowel movements, often with blood and mucus, and characterized by pain, urgency to have a bowel movement, fever, and dehydration.

Fluoroquinolones—A relatively new group of antibiotics used to treat infections with many gram-negative bacteria, such as *Shigella*. One drawback is that they should not be used in children under 17 years of age, because of possible effect on bone or cartilage growth.

Food-borne illness—A disease that is transmitted by eating or handling contaminated food.

Meninges—The three-layer membranous covering of the brain and spinal cord, composed of the dura mater, arachnoid, and pia mater. It provides protection for the brain and spinal cord, as well as housing many blood vessels and participating in the appropriate flow of cerebrospinal fluid.

Oral rehydration solution (ORS)—A liquid preparation of electrolytes and glucose developed by the World Health Organization that can decrease fluid loss in persons with diarrhea. Originally developed to be prepared with materials available in the home, commercial preparations have recently come into use.

Stool—The solid waste that is left after food is digested. Stool forms in the intestines and passes out of the body through the anus.

Traveler's diarrhea—An illness due to infection from a bacteria or parasite that occurs in persons traveling to areas where there is a high frequency of the illness. The disease is usually spread by contaminated food or water.

settings almost always disappears when holiday breaks occur, which severs the chain of transmission.

Traveler's diarrhea (TD)

Shigella accounts for about 10 percent of diarrhea illness in travelers to Mexico, South America, and the tropics. Most cases of TD are more of a nuisance than a life-threatening disease.

In some cases, though, aside from ruining vacation plans, these infections can interrupt business conference schedules and, in the worst instances, lead to a life-threatening illness. Therefore, researchers have tried to find a safe, yet effective, way of preventing TD. Of course the best prevention is to follow closely the rules outlined by the WHO and other groups regarding eating fresh fruits, vegetables, and other foods.

One safe and effective method of preventing TD is the use of large doses of Pepto Bismol. Tablets are easier for use during travel; usage must start a few days before departure. Patients should be aware that bismuth turns bowel movements black.

Antibiotics have also proven highly effective in preventing TD. They can also produce significant side effects. Therefore, a physician should be consulted before use. Like Pepto Bismol, antibiotics need to be started before beginning travel.

Parental concerns

Parents of children suffering from shigellosis need to follow closely their healthcare provider's directions for preventing dehydration. Excellent hand washing is crucial to prevent the spread of the infection throughout

family members. This is particularly important while helping to toilet train a child or while changing diapers.

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Shock, anaphylactic *see* **Anaphylaxis**

Shyness

Definition

Shyness is a psychological state that causes a person to feel discomfort in social situations in ways that interfere with enjoyment or that cause avoidance of social contacts altogether.

Description

Shyness can vary from mild feelings to moderately uncomfortable in social circumstances to debilitating levels of **anxiety** that interfere in children with the process of socialization (social withdrawal). Shyness is a personality trait that affects a child's **temperament**. Some infants are born shy and more sensitive. Some of them are quiet when new people enter a room. A shy baby might sink his head into his mother's shoulder, while a baby who is outgoing might smile or squeal with delight when someone new visits. Some children may feel shy in certain situations, like when meeting new people. Other children may learn to be shy because of experiences in school or at home. As of 2004, research tended to distinguish shyness from introversion. Introverts simply prefer solitary to social activities but do not **fear** social encounters as shy people do, while extroverts prefer social to solitary activities.

Evidence suggests a genetic component to shyness. Studies on the biological basis of shyness have shown that shyness in adults can often be traced as far back as the age of three. A Harvard study of two-year olds showed that, even at that age, widely different personality types can be recognized: roughly 25 percent of children are bold, sociable, and spontaneous regardless of the novelty of the situation, while 20 percent are shy and restrained in new situations. The remaining 55 percent of newborns fall between the extremes of shyness and boldness. These two basic temperaments were also recognized in studies examining infants as young as four months old. As children grow, their shy temperament tends to display itself in predictable ways: for example, in **play** groups at age seven, shy children play by themselves, while more outgoing children seek to play together in groups. Evidence of a genetic predisposition for shyness is found in parents and grandparents of shy infants who report childhood shyness more often than relatives of children who are not shy. Further evidence for a congenital link to shyness is found in studies that show that identical **twins** (who have identical genes) are more likely to be shy than fraternal twins (who are no more alike than other siblings).

Research shows, however, that 25 percent of the time genetic predisposition to shyness does not develop into shyness. Some researchers believe that a shy temperament may require environmental triggers, such as insecurity of attachment in the form of difficult relationships with parents, **family** conflict or chaos, frequent criticism, a dominating older sibling, or a stressful school environment.

Research has also identified a strong cultural link to shyness. In the United States, shyness surveys typically show that shyness is highest among Asian Americans and lowest among Jewish Americans. Using culturally sensitive adaptations of the Stanford Shyness Inventory, researchers in eight countries administered the inventory to groups of 18 to 21 year olds. Results showed that a large proportion of participants in all cultures reported experiencing shyness to a considerable degree—from 31 percent in Israel to 57 percent in Japan and 55 percent in Taiwan. In Mexico, Germany, India, and Canada, shyness levels were close to the U.S figure of 40 percent. In all countries, shyness is perceived as more negative than positive, with 60 percent or more considering shyness to be a problem. There is no gender difference in reported shyness, but males tend to conceal their shyness because it is considered a feminine trait in most countries. For example, in Mexico, males report shyness less often than females do.

Common problems

When shyness is intense, it can often lead to social anxiety disorder or to avoidant personality disorder, both characterized by the avoidance of interpersonal contacts accompanied by significant fears of embarrassment in social interaction. According to the most recent statistics, provided by the National Co-morbidity Survey—carried out in 1994—approximately 40 percent of Americans consistently report since the early 1970s that they are shy to the extent of considering it a problem in their lives. Subsequent research showed that the percentage of problem-related shyness gradually increased during the 1990s to nearly 50 percent. The National Co-morbidity Survey results were also indicative of a lifetime prevalence of social anxiety of 13.3 percent, making it the third most prevalent psychiatric disorder in the United States.

Excessive shyness usually leads to social withdrawal. If it is based on social fear and anxiety despite a desire to interact socially—such as in children who are unhappy because they are unable to make friends, it is called "conflicted shyness"; if it is based on the lack of a strong motivation to engage in social interaction, it is called "social disinterest." Both types are detected at an early age. The major behavioral components of excessive shyness in children are as follows:

- difficulty talking, stammering, **stuttering**, blushing, shaking, sweating hands when around other people
- difficulty thinking of things to say to people
- absence of outgoing mannerisms such as good eye contact or an easy smile
- reluctance to play with other kids, to go to school, to visit relatives and neighbors

Parental concerns

Parents may worry about if their shy infant, child, or teenager has a socialization problem. Parents should know that a child who seems mildly anxious or quiet at certain times may be shy. The best thing they can do is to help the child feel comfortable about being shy, by explaining that shyness can be a normal part of growing up. Teasing or being critical can make a shy child even more afraid to be around people. Sometimes, just encouraging a quiet child to play with others helps the child overcome shy feelings. Although many children who are shy remain shy all of their lives, many others overcome it in time as they develop social skills. Many children overcome shyness by themselves, some through associating with younger children, which allows them to display leadership behavior, still others through contact with other sociable children. Nothing assists in overcoming shyness more than experiencing social successes, as when a child takes the initial risk of engaging in some social activity that is rewarded, for example, in successfully developing friendships.

The use of **video games**, CD-ROM games, Web surfing, and other computer-related marvels all interfere with the time required to seek out direct contact with others for fun and friendship. Increasingly, social time is being replaced with the anonymous exchange of information within an externally imposed medium that effectively promotes shyness in young people. While some shy children may benefit from using the anonymity and structural control features of cyberspace, the danger is that for many others virtual on-line reality may become a substitute for the reality of close human relationships. Many parents are concerned because their young children prefer "chat time" on their computers more than actually talking face to face with other children, so these children may not socialize as much in the homes of neighbors and friends.

When to call the doctor

When a child is excessively shy, when shyness is persistent and results in high levels of anxiety in social settings and leads to social withdrawal and parents feel that their child is unhappy being shy, they should seek professional help. It is very important to determine if there is a social anxiety disorder, and if so, what treatment can best help the child overcome shyness. Child and adolescent psychiatrists are trained to help parents sort out whether their child's level of shyness is normal introversion or indicative of a disorder.

See also Parent-child relationships; Personality development; Personality disorders.

Resources

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KEY TERMS

Avoidant personality disorder—Chronic and long-standing fear of negative evaluation and tendency to avoid interpersonal situations without a guarantee of acceptance and support, accompanied by significant fears of embarrassment and shame in social interaction.

Extroversion—A personal preference for socially engaging activities and settings.

Extrovert—A person who is outgoing and performs well socially.

Introversion—A personal preference for solitary, non-social activities and settings.

Personality—The organized pattern of behaviors and attitudes that makes a human being distinctive. Personality is formed by the ongoing interaction of temperament, character, and environment.

Social anxiety disorder—Persistent avoidance and/or discomfort in social situations that significantly interferes with functioning.

Social withdrawal—Avoidance of social contacts.

Socialization—The process by which new members of a social group are integrated in the group.

Temperament—A person's natural disposition or inborn combination of mental and emotional traits.

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American Psychological Association (APA). 750 First Street, NE, Washington, DC 20002–4242. Web site: <www.apa.org>.

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Sibling rivalry

Definition

Sibling rivalry is antagonism between brothers and/or sisters that results in physical fighting, verbal hostility, teasing, or bullying.

Description

Psychologists believe that sibling rivalry comes from competition for parental attention, love, and approval. The amount of conflict depends on the perception of parents about the role of each child in the **family**, the personalities of the parents and children, the number and spacing of children in the family, outside resources available to the children, and parental beliefs about child

rearing, including their attitudes toward gender, **birth order**, and competition. Sibling rivalry is also affected by the presence in the family of a special needs child, **divorce** or other family trauma, and ethnic and cultural attitudes toward family relationships.

Studies suggest that sister/sister relationships are the least competitive and hostile while brother/brother relationships, especially when brothers are close in age, are the most hostile and competitive. However, this is a generalization that does not apply in many specific instances. Some psychologists believe that moderate levels of sibling rivalry can help children learn to share, compromise, and negotiate with others.

Infancy

The birth of a new baby in the family often creates jealousy and distress for older children. Not only does a new baby increase the number children that must share parental attention, newborns are inordinately time consuming, leaving older children to feel they have been displaced and abandoned. Mothers often are exhausted and sometimes depressed after the birth of a child. While in the hospital and immediately after the birth, they may withdraw from their older children to care for the newborn, leaving day-to-day care taking of the other children to friends, relatives, or hired caregivers. Friends and other family members tend to focus on the newborn, further displacing older children. If the new baby is born with special needs, the time and energy spent focusing on the new sibling may be quite extensive.

Toddlers may react to a new baby by reverting to younger behaviors in an attempt to gain parental attention. For example, a toddler who is toilet trained may start having accidents in his or her pants. Verbal toddlers may express their disgust with the new sibling by asking, "Isn't it time to send him back?" Others may pinch and poke the new baby. Older children may become more difficult, temperamental, and uncooperative, as they see their role in the family changing. Although responses like these are, within reason, normal, they challenge parents and create conflict within the family.

Parents can help their other children prepare for the arrival of a new sibling by reading books to them about babies and involving them in preparations for their new sibling. After the child is born, in a two-parent family, the father can step in and spend extra time with older siblings, taking some of the pressure off the relationship between the mother and her older children. Many children feel more connected to their new sibling if they are given some specific age-appropriate task that helps to care for the baby.

Toddlerhood

Toddlers are active, curious people who are beginning to explore both their physical and social world. As noted above, they may respond to the birth of a new sibling by reverting to more childish behaviors. Toddlers are developing a sense of themselves as individuals and pushing the limits of their physical abilities. This testing and accompanying frustration often manifests itself in **tantrums** and other socially unacceptable behaviors such as an unwillingness to share **toys**. Since toddlers usually lack the ability to perceive the needs and desires of others and do not have the verbal capacity to express their emotions or abstract thoughts, sibling rivalry at this age usually takes the form of physical aggression.

Toddlers who are working out social boundaries may take toys from others or refuse to share or take turns. They may go through a stage of wanting whatever a sibling has, even if the moment they get it, they no longer want it. This can be a normal, if not socially desirable, stage of development. However, it creates friction with older siblings that often degenerates into kicking, hitting, punching, pinching, and even biting.

Parents need to intervene when sibling rivalry becomes physical. Younger toddlers can sometimes be distracted, but older ones need to be separated and given a break from each other. Many experts recommend punishing both children rather than becoming involved in trying to figure out who was "right" and who was "wrong."

Preschool

Preschool children are more verbal than toddlers, and much of their hostility toward siblings takes the form of name calling, verbal abuse, and teasing. Parents need to set limits on what is acceptable. Another source of sibling conflict at this age is the preschooler's desire to be part of his older siblings friendships. Although it may be easier for parents to tell their older child to include the younger one, this often intensifies the older child's hostility toward the younger one. Parents should be alert to the need to protect each child's personal possessions and friendships.

School age

Sibling rivalry can and often does continue into adulthood. By the time children reach school age, the level of sibling rivalry is affected by family attitude toward competition, ethnic and cultural attitudes, comparisons of siblings by teachers and coaches, the family's expectations for each child in the family, and their method of applying "fairness" in their relationships with

their children. Hostility can take the form of physical or verbal fighting, invading each other's privacy, or destroying each other's possessions.

At this age, children often begin to carve out their own area in which to excel. One child may concentrate on soccer while another concentrates on music and a third on schoolwork. This differentiation can help reduce competition and sibling rivalry. Parents can reduce the level of sibling rivalry by supporting each child's interest with an equal investment of enthusiasm and time. At this age, the approval and support of individuals outside the family also plays a role in reducing sibling rivalry. Resentment and hostility can be increased when parents insist that all children in the family do the same activities all the time, always include each other in their **play** and friendships, and put older children in charge of younger ones for long periods on a regular basis.

Common problems

The presence of a special needs child who dominates the parents' attention can add to increased resentment and rivalry. Other common problems include assuming that the older child is always at fault in sibling fights, giving each child a label such as "the smart one" or "the wild one," that suggest one child is "good" and another is "bad," reinforcing cultural attitudes that place a higher value on sons than daughters and on first children rather than later children, and overprotecting younger children or children who are perceived as weaker than their siblings. Single parents may face an especially difficult time balancing the needs of their children in ways that reduce rivalry simply because they are the only adult in the family.

Parental concerns

Parents often worry about preferring one child over another and being fair to all their children. In reality, parents may love their children equally but find at different times in their development some of their children are more likeable and easier to get along with than others. Parents can help reduce sibling rivalry by following these steps:

- working to see each child as a unique individual with his or her own strengths and weaknesses
- spending some one-on-one time with each child every week
- encouraging children to develop their own interests and friends independent of the interests and friends of their siblings
- limiting the amount of care giving expected of older siblings for younger ones
- setting and enforcing firm rules about name calling, teasing, and physical aggression in the family
- praising cooperative behavior
- insisting that each child's personal possessions and privacy are respected by the other children in the family

When to call the doctor

Parents may wish to consult their pediatrician, a child and adolescent psychologist, or a family therapist if any of the following occurs:

- Serious attempts at reducing sibling rivalry have failed.
- Siblings physically harm each other.
- Siblings constantly tease and belittle or bully each other.
- One or more children in the family seem to have other behavioral problems at home or school.
- Siblings gang up on one child in the family.
- The level of sibling rivalry appears to be destructive to any member of the family.

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Tish Davidson, A.M.

Sickle cell anemia

Definition

Sickle cell anemia, also called sickle cell disease (SS disease), is an inherited condition caused by having abnormal hemoglobin, the protein that carries oxygen in the blood. People with sickle cell anaemia have sickle hemoglobin (HbS) which is different from the normal hemoglobin (HbA).

Description

Children with sickle cell anemia produce two abnormal hemoglobin proteins (inheriting one from each parent), which makes their red blood cells easily destructible while giving them a sickle-like shape. Since the red blood cells do not have a normal shape, their circulation in the small blood vessels is impaired as well as the function of the abnormal hemoglobin (HbS) which can no longer carry oxygen with maximum efficiency.

Transmission

Sickle cell anemia is usually inherited from parents who are carriers, who have the sickle cell trait—a milder form of sickle cell anemia, or one abnormal hemoglobin.

Demographics

Sickle cell anemia and sickle cell trait are found mainly in people whose families come from Africa, the Caribbean, the Eastern Mediterranean, Middle East, and Asia. In the United States, sickle cell anemia affects some 72,000 people. The families of most of the people affected come from Africa. The disease occurs in about one in every 600 African-American births and in one in every 1,000 to 1,400 Hispanic-American births. Some 2 million Americans carry the sickle cell trait and about one in 12 African Americans have the trait.

Causes and symptoms

Sickle cell anemia is caused by an error in the gene that signals the body how to make hemoglobin. The defective gene tells the body to make the abnormal hemoglobin HbS instead of the normal HbA, and this results in deformed red blood cells. The error in the hemoglobin gene is due to a genetic mutation that occurred many thousands of years ago in people living in Africa, the Mediterranean basin, the Middle East, and India. A deadly form of malaria was very common at that time, and research has shown that in areas where malaria was endemic, children who inherited one HbS gene and who, therefore, carried the sickle cell trait, had a survival advantage because, unlike the children who had normal HbA genes, they survived malaria. They grew up, had their own children, and passed on the gene for HbS.

Symptoms or complications associated with sickle cell anemia usually start after the age of four to six months and can include all or some of the following:

- anemia, caused by low amounts of red blood cells in the bloodstream, resulting in insufficient oxygen delivery to tissues and organs
- vaso-occlusive **pain**, meaning severe episodes of pain in the arms, legs, or back, due to impaired blood circulation in the blood vessels
- chest pain and **fever** with coughing
- dactylitis, or hand-foot syndrome, with painful swelling of the bones in the hands or feet of young children
- aplastic crises, during which the body stops making new red blood cells causing severe anemia usually following an infection with the parovirus B19, which causes fifth disease
- priapism, a painful and prolonged erection
- **stroke**, usually causing sudden weakness of one side of the body
- acute splenic sequestration, with pooling of blood causing a sudden enlargement of the spleen
- jaundice, a yellowing of the skin and white of the eyes
- frequent infection with certain bacteria, particularly pneumococcus and salmonella, which are due to autoinfarction of the spleen or death of the spleen due to poor blood flow

When to call the doctor

Children suffering from sickle anemia have episodes during which they suddenly become unwell or complain of severe abdominal or chest pain, **headache**, stiffness of the neck or drowsiness. Parents should know that a child having a sickle cell crisis requires urgent hospital treatment. They should also call a doctor if the child has a temperature above 101°F (38.5°C).

Diagnosis

The diagnosis of sickle cell anemia is established during the newborn screen testing that is performed in the nursery at time of birth. For children who are not tested, an electrophoresis test of the blood can detect the abnormal hemoglobin of sickle cell anemia. This test

measures the speed at which a molecule moves in a gel and can detect abnormal hemoglobin HbS.

Treatment

Treatment usually includes frequent monitoring of red blood counts, **antibiotics** for infections, transfusions for aplastic crises and splenic sequestration when required, and oxygen as well as respiratory support for chest syndrome. Some patients with severe symptoms receive regular blood transfusions to prevent crises and/or other complications such as stroke and organ damage.

Children with sickle cell disorders are at risk of developing severe infections, and penicillin is usually prescribed to prevent dangerous pneumococcal infections.

Sickle cell pain can be managed with a variety of measures including the following:

- warmth, to increase the blood flow, by massaging and rubbing and by heat from hot water bottles and deep heat creams
- bandaging to support the painful region
- rest
- getting the child to relax, by deep breathing exercises and distracting the attention, and by other psychological methods
- use of pain-killing medicines (**analgesics**)

Analgesics should only be given as recommended by the treating physician. The gentlest analgesic usually prescribed is paracetamol, given three times a day (62.5 mgm under 12 months; 125 mgm 1–4 years; 250 mgm 4–10 years; 500 mgm 10–14 years; and 1 gm 15 years upwards). The next gentlest is codeine phosphate, given four times a day, at 1–2 mgm for every kilogram of body weight.

Bone marrow transplantation has been shown to provide a cure for severely affected children with sickle cell disease, but the procedure is not entirely without risk. In addition, the marrow must come from a healthy matched sibling donor and only about 18 percent of children with sickle cell anemia are likely to have a matched sibling.

Alternative treatment

Research contributed a great deal about sickle cell anemia from 1970 to the early 2000s concerning what causes it, how it affects the patient, and how to treat it. Scientists were as of 2004 starting to be successful at developing drugs that prevent the symptoms of sickle cell anemia and procedures that they hope should eventually provide a cure. Drug research is focused on identifying drugs, such as hydroxyurea, that can increase the level of fetal hemoglobin in the blood. Fetal hemoglobin is a form of hemoglobin that all humans produce before birth, but most stop producing it after birth. It has been observed that some children with sickle cell anemia continue to produce large amounts of fetal hemoglobin after birth, and studies have shown that these children have less severe cases of the disease. Fetal hemoglobin seems to prevent "sickling" of red cells, and cells containing fetal hemoglobin tend to survive longer in the bloodstream. Butyrate, a substance widely used as a food additive, was also being investigated as of 2004 as an agent that may increase fetal hemoglobin production.

Nutritional concerns

Thirst and **dehydration** caused by not drinking enough, even if thirst is not felt, are known to trigger sickle pain. Parents should accordingly monitor fluid intake closely.

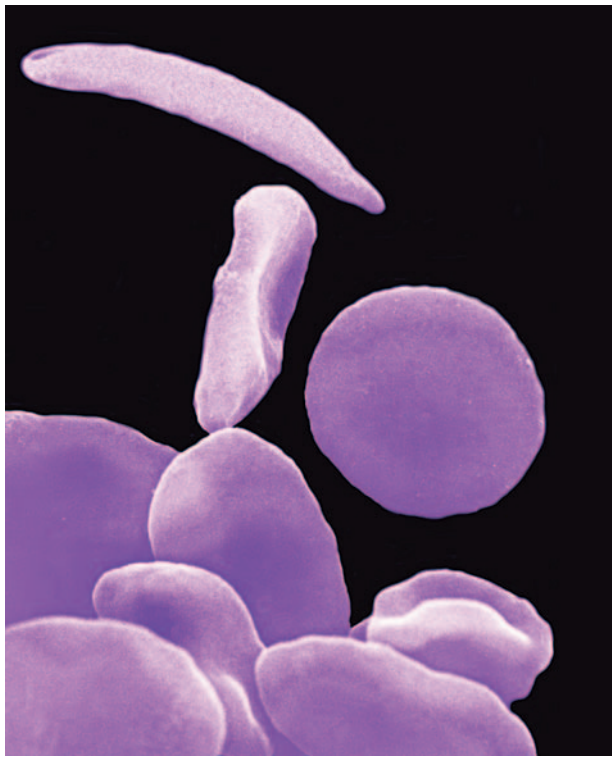
Children with sickle cell anemia are anemic to various degrees. Most of the time they feel quite well, but if the anemia gets worse, they may feel very tired. **Folic acid**, a vitamin found in fruit and vegetables, supports making blood and anemic children especially need it to prevent them from becoming run down.

Prognosis

Sickle cell anemia is an inherited disease and lasts a lifetime.

Prevention

Both sickle cell trait and sickle cell anemia are inherited. Therefore, parents can pass it to their offspring. It is important for parents to get tested. If one partner has sickle cell trait and the other does not, their children each have a 50 percent chance of having the sickle cell trait, and a 50 percent chance of having normal hemoglobin. If one parent has sickle cell trait it is extremely important that the other parent be tested. If both parents have sickle cell trait, there is a 25 percent chance that the child will have normal hemoglobin, a 50 percent chance that the child will have the sickle cell trait, and a 25 percent chance the child will have sickle cell disease. If both parents have sickle cell trait and want to know whether the unborn child has sickle cell anemia, testing can be performed as early as the tenth week of pregnancy. If the results are normal, the parents can be reassured. If the tests show that the baby will be affected, the parents can be better prepared and they can make an informed decision concerning the pregnancy.



Normal red blood cells are smooth and round. In sickle cell anemia, the red blood cells become shaped like sickles or crescents. *(© Dr. Gopal Murti/Photo Researchers, Inc.)*

Parental concerns

Parents should be aware that children with sickle cell anemia are also at increased risk of infection, especially from the *Streptococcus pneumonia* and *H. influenzae* bacteria.

Sickle cell anemia does not affect **intelligence**. Children with sickle cell disorders can almost always attend school and participate fully in normal activities. The child's teacher and the school principal should know of the diagnosis and understand the limitations sickle cell anemia can impose on a child, for instance the need for frequent drinks and easy access to the bathroom, and the triggering of pain by over-exertion or cold.

See also Anemias.

KEY TERMS

Acute splenic sequestration—Retention of blood in the spleen.

Anemia—A condition in which there is an abnormally low number of red blood cells in the bloodstream. It may be due to loss of blood, an increase in red blood cell destruction, or a decrease in red blood cell production. Major symptoms are paleness, shortness of breath, unusually fast or strong heart beats, and tiredness.

Aplastic anemia—A disorder in which the bone marrow greatly decreases or stops production of blood cells.

Bone marrow—The spongy tissue inside the large bones in the body that is responsible for making the red blood cells, most white blood cells, and platelets.

Dactylitis—Inflammation of the hands or feet.

Endemic—Natural to or characteristic of a particular place, population, or climate.

Hemoglobin—An iron-containing pigment of red blood cells composed of four amino acid chains (alpha, beta, gamma, delta) that delivers oxygen from the lungs to the cells of the body and carries carbon dioxide from the cells to the lungs.

Priapism—A painful, abnormally prolonged penile erection.

Sickle cell trait—Condition that occurs in people who have one of two possible genes responsible for the abnormal hemoglobin of sickle cell anemia. People with this trait may suffer milder symptoms of sickle cell anemia or may have no symptoms. Some scientists believe that the trait actually provides an advantage in tropical environments because the slightly altered shape of the blood cells cause a person to be more resistant to malaria.

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Monique Laberge, Ph.D.

SIDS *see* **Sudden infant death syndrome**

Single-parent families

Definition

Single-parent families are families with children under age 18 headed by a parent who is widowed or divorced and not remarried, or by a parent who has never married.

Description

One out of every two children in the United States will live in a single-parent **family** at some time before they reach age 18. According the United States Census Bureau, in 2002 about 20 million children lived in a household with only their mother or their father. This is more than one-fourth of all children in the United States.

Since 1950, the number of one-parent families has increased substantially. In 1970, about 11 percent of children lived in single-parent families. During the 1970s, **divorce** became much more common, and the number of families headed by one parent increased rapidly. The number peaked in the 1980s and then declined slightly in the 1990s. By 1996, 31 percent of children lived in single-parent families. In 2002, the number was 28 percent. Many other children have lived in single-parent families for a time before their biological parent remarried, when they moved into a two-parent family with one biological parent and one step parent.

The reasons for single-parent families have also changed. In the mid-twentieth century, most single-parent families came about because of the death of a spouse. In the 1970s and 1980s, most single-parent families were the result of divorce. In the early 2000s, more and more single parents have never married. Many of these single parents live with an adult partner, sometimes even the unmarried father of their child. These families are counted by the Census Bureau as single-parent families, although two adults are present. Still other families are counted as single-parent families if the parents are married, but one is away for an extended period, for example, on military deployment.

The most common type of single-parent family is one that consists of a mother and her biological children. In 2002, 16.5 million or 23 percent of all children were living with their single mother. This group included 48 percent of all African-American children, 16 percent of all non-Hispanic white children, 13 percent of Asian/Pacific Islander children, and 25 percent of children of Hispanic origin. However, these numbers do not give a true picture of household organization, because 11 percent of all children were actually living in homes where their mother was sharing a home with an adult to whom she was not married. This group includes 14 percent of white children, 6 percent of African-American children, 11 percent of Asian/Pacific Islander, and 12 percent of Hispanic children.

Households headed by a single father increased substantially after the early 1980s, reflecting society's changing attitudes about the role of fathers in child rearing. In 1970, only 1 percent of children lived with a single father. In 2002, about 5 percent of children under age 18 lived with their single fathers. Single fathers, however, are much more likely to be divorced than never married and much more likely than single mothers to be sharing a home with an adult to whom they are not married. For example, 33 percent of Caucasian children lived with fathers who were unmarried but cohabiting with another adult. The rate was 29 percent for African-American children, 30 percent for Asian/Pacific Islanders, and 46 percent for children of Hispanic origin. It is clear that not all single-parent families are the same and that within different ethnic and racial groups, the number and type of single-parent families varies considerably.

Adoption by single individuals has also soared. In 1970 only 0.5 to 4 percent of adoptive parents were single. In the 1980s this rate increased from 8 to 34 percent. According the United States Department of Health and

Single parent and her children spending time together. *(© Rick Gomez/Corbis.)*

Human Services, 33 percent of children adopted from **foster care** are adopted by single parents.

Common problems

Single-parent families face special challenges. One of these is economic. In 2002, twice as many single-parent families earned less than $30,000 per year compared to families with two parents present. At the opposite end of the spectrum, 39 percent of two-parent families earned more than $75,000 compared to 6 percent of single-mother families and 11 percent of single-father families. Single-parent families are challenged in other ways. Children living with single fathers were the least likely of all children to have health insurance coverage.

Social scientists have found that children growing up in single-parent families are disadvantaged in other ways when compared to a two-biological-parent families. Many of these problems are directly related to the poor economic condition of single-parent families, not just to parenting style. These children are at risk for the following:

- lower levels of educational achievement
- twice as likely to drop out of school
- more likely to become teen parents
- more conflict with their parent(s)
- less supervised by adults
- more likely to become truants
- more frequently abuse drugs and alcohol
- more high-risk sexual behavior
- more likely to join a gang
- twice as likely to go to jail
- four times as likely to need help for emotional and behavioral problems
- more likely to participate in violent crime
- more likely to commit suicide
- twice as likely to get divorced in adulthood

Studies have also found that children who live in a two-parent family where one parent is abusive or has a high level of **antisocial behavior** do not do as well as children whose parents divorce if the child then lives in a single-parent family with the nonabusive parent.

It is important to remember that every single-parent family is different. Children who are living with a widowed mother will have a home life that is different from children with divorced parents or those whose parents were never married. Children of divorced parents will have a wide range of relationships with their parents and parents' partners depending on custody arrangements and the commitment of the non-custodial parent to maintaining a relationship with the child. Despite the fact that children from single-parent families often face a tougher time economically and emotionally than children from two-biological-parent families, children from single-parent families can grow up doing well in school and maintaining healthy behaviors and relationships.

Parental concerns

Being a single parent can be hard and lonely. There is often no other adult with whom to share decision-making, **discipline**, and financial responsibilities. The full burden of finding responsible childcare, earning a living, and parenting falls on one individual. However, the lack of a second parent often has a less negative impact on children than family instability, lack of structure, and inconsistent enforcement of parental standards. Single parents may want to follow these steps in order to create positive experiences for their children:

- Find stable, safe child care.
- Establish a home routine and stick to it.
- Apply rules and discipline clearly and consistently.
- Allow the child to be a child and not ask him or her to solve adult problems.
- Get to know the important people (teachers, coaches, friends) in the child's life.
- Answer questions about the other parent calmly and honestly.
- Avoid behavior that causes the child to feel pressed to choose between divorced parents.
- Explain financial limitations honestly.

When to get help

If parents feel their child is out of control and is not responding to their parenting, they need to get help from the child's school, social service agencies, and mental health professionals. If they feel their own life is spiraling downward and falling apart, they can seek help from many organizations that provide social, emotional, financial, and legal support for single-parent families.

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Parents without Partners. 1650 South Dixie Highway, Suite 510, Boca Raton, Florida 33431 Web site: <www.parentswithoutpartners.org>.

Single and Custodial Fathers Network Inc. Web site: <http://scfn.org>.

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Tish Davidson, A.M.

Sinusitis

Definition

Sinusitis is an inflammation of the sinuses, which are airspaces within the bones of the face. Sinusitis is most often due to an infection within these spaces.

Description

The sinuses are paired air pockets located within the bones of the face. There are:

- the frontal sinuses, located above the eyes, in the center region of each eyebrow
- the maxillary sinuses, located within the cheekbones, just to either side of the nose

- the ethmoid sinuses, located between the eyes, just behind the bridge of the nose
- the sphenoid sinuses, located just behind the ethmoid sinuses, and behind the eyes.

The sinuses are connected with the nose. They are lined with the same kind of skin found elsewhere within the respiratory tract. This skin has tiny little hairs projecting from it called cilia. The cilia beat constantly to help move the mucus produced in the sinuses into the respiratory tract. The beating cilia sweeping the mucus along the respiratory tract helps to clear the respiratory tract of any debris or of any organisms that may be present. When the lining of the sinuses is at all swollen, the swelling interferes with the normal flow of mucus. Trapped mucus can then fill the sinuses, causing an uncomfortable sensation of pressure and providing an excellent environment for the growth of infection-causing bacteria.

Demographics

It is estimated that about 37 million Americans are affected by sinusitis each year. Having a cold increases the chance of getting sinusitis. Immune system disorders also increase this likelihood. Children with **asthma** are also considered more likely to be affected by sinusitis.

Causes and symptoms

Sinusitis is usually due to an infection, although swelling from **allergies** can mimic the symptoms of pressure, **pain**, and congestion, and allergies can set the stage for a bacterial infection. Bacteria are the most common cause of sinus infection. *Streptococcus pneumoniae* causes about 33 percent of all cases, while *Haemophilus influenzae* causes about 25 percent of all cases. Sinusitis in children may be caused by *Moraxella catarrhalis* (20%). In people with weakened immune systems (including patients with diabetes, acquired **immunodeficiency** syndrome or **AIDS**, and patients who are taking medications that lower their immune resistance, such as **cancer** and transplant patients), sinusitis may be caused by fungi such as *Aspergillus*, *Candida*, or *Mucorales*.

Acute sinusitis usually follows some type of upper respiratory tract infection or cold. Instead of ending, the cold seems to linger on, with constant or even worsening congestion. Drainage from the nose often changes from a clear color to a thicker, yellowish-green. The individual may develop a **fever**. **Headache** and pain over the affected sinuses may occur, as well as a feeling of pressure that may worsen when the patient bends over. There may be pain in the jaw or teeth. Some children, in particular, get upset stomachs from the infected drainage going down the back of their throats and being swallowed. Some patients develop a **cough**.

Chronic sinusitis occurs when the problem has existed for at least three months. There is rarely a fever with chronic sinusitis. Sinus pain and pressure are frequent, as is nasal congestion. Because of the swelling in the sinuses, they may not be able to drain out the nose. Drainage, therefore, drips constantly down the back of the throat, resulting in a continuously **sore throat** and bad breath.

When to call the doctor

If the child is displaying the signs of sinusitis for more than a few days, the doctor should be contacted. If a cold seems to be getting better and then gets worse again, it may have developed into sinusitis. Likewise, colds that linger beyond a week may indicate sinusitis, and the doctor should be called.

Diagnosis

Diagnosis is sometimes tricky, because the symptoms so often resemble those of an uncomplicated cold. However, sinusitis should be strongly suspected when a cold lingers beyond about a week's time.

Medical practitioners have differing levels of trust in certain basic examinations commonly conducted in the office. For example, tapping over the sinuses may cause pain in patients with sinusitis, but it may not. A procedure called "sinus transillumination" may or may not also be helpful. Using a flashlight pressed up against the skin of the cheek, the practitioner will look in the patient's open mouth. When the sinuses are full of air (under normal conditions), the light will project through the sinus and will be visible on the roof of the mouth as a lit-up, reddened area. When the sinuses are full of mucus, the light will be stopped. While this simple test can be helpful, it is certainly not a perfect way to diagnose or rule out the diagnosis of sinusitis.

X-ray pictures and CT scans of the sinuses are helpful for both acute and chronic sinusitis. People with chronic sinusitis should also be checked for allergies, and they may need a procedure called nasal endoscopy where a very slender lighted fiber optic tube is placed in the nose in order for the doctor to see if any kind of anatomic obstruction is causing the illness. For example, the septum (the cartilage which separates the two nasal cavities from each other) may be slightly displaced (a deviated septum). This condition can result in chronic obstruction, setting the person up for the development of an infection.

Treatment

Antibiotic medications are used to treat acute sinusitis. Suitable **antibiotics** include sulfa drugs, amoxicillin, and a variety of cephalosporins. These medications are usually given for about two weeks but may be given for even longer periods. **Decongestants** or the short-term use of decongestant nose sprays can be useful. **Acetaminophen** (Tylenol) and ibuprofen (Motrin, Advil) can decrease the pain and headache associated with sinusitis. Also, running a humidifier can prevent mucus within the nasal passages from drying out uncomfortably and can help soothe any accompanying sore throat or cough.

Chronic sinusitis is often treated initially with antibiotics. Steroid nasal sprays may be used to decrease swelling in the nasal passages. If an anatomic reason is found for chronic sinusitis, it may need to be corrected with surgery. If a surgical procedure is necessary, samples are usually taken at the same time to allow identification of any organisms present which may be causing infection.

Fungal sinusitis requires surgery to clean out the sinuses. Then, a relatively long course of a very strong antifungal medication called amphotericin B is given through a needle in the vein (intravenously).

Alternative treatment

Some practitioners believe that chronic sinusitis is associated with **food allergies**. These doctors would suggest an elimination/challenge diet to identify and eliminate allergenic foods. While linking chronic sinusitis to food is widely considered controversial, many practitioners link the problem to aero-allergies. Irrigating the sinuses with a salt-water solution is thus recommended for sinusitis and allergies, in order to clear the nasal passages of mucus. Another solution for nasal lavage (washing) uses powdered goldenseal (*Hydrastis canadensis*). Other herbal treatments, taken internally, include a mixture made of eyebright (*Euphrasia officinalis*), goldenseal, yarrow (*Achillea millefolium*), and horseradish, or, when infection is present, a mixture made of echinacea (*Echinacea* spp.), wild indigo, and poke root (*Phytolacca decandra-Americana*).

Homeopathic practitioners find a number of remedies useful for treating sinusitis. Among those they recommend are: *Arsenicum album*, *Kalium bichromium*, *Nux vomica*, *Mercurius iodatus*, and *Silica*.

Acupuncture has been used to treat sinusitis, as have a variety of dietary supplements, including **vitamins** A, C, and E, and the mineral zinc. Contrast hydrotherapy (hot and cold compresses, alternating three minutes hot, 30 seconds cold, repeated three times always ending with cold) applied directly over the sinuses can relieve pressure and enhance healing. A direct inhalation of essential oils (two drops of oil to two cups of water) using thyme, rosemary, and lavender can help open the sinuses and kill bacteria that cause infection.

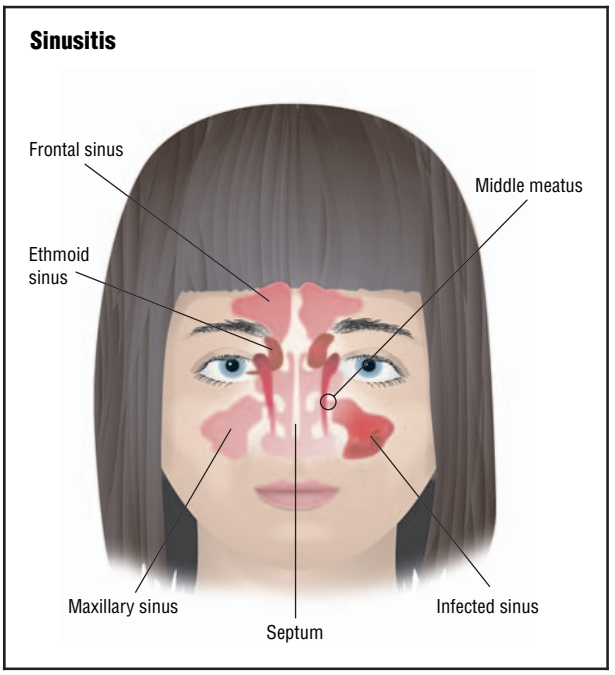

Illustration of an infected left maxillary sinus, which radiates pain and pressure to the surrounding sinus areas. *(Illustration by GGS Information Services.)*

Prognosis

Prognosis for sinus infections is usually excellent, although some individuals may find that they are particularly prone to contracting such infections after a cold. Fungal sinusitis, however, has a relatively high death rate.

Prevention

Prevention involves the usual standards of good hygiene to cut down on the number of colds an individual catches. Avoiding exposure to cigarette smoke, identifying and treating allergies, and avoiding deep dives in swimming pools may help prevent sinus infections. During the winter, it is a good idea to use a humidifier. Humidifiers should be adequately and frequently cleaned with bleach or comparable cleanser to avoid mold which can be aerosolized and then exacerbate existing allergies. Dry nasal passages may crack, allowing bacteria to enter. When allergies are diagnosed, a

KEY TERMS

Cilia—Tiny hairlike projections on certain cells within the body. Cilia produce lashing or whipping movements to direct or cause motion of substances or fluids within the body. Within the respiratory tract, the cilia act to move mucus along, in an effort to continually flush out and clean the respiratory tract.

Sinus—A tubular channel or cavity connecting one body part with another or with the outside. Often refers to one of the air-filled cavities surrounding the eyes and nose that are lined with mucus-producing membranes. They cleanse the nose, add resonance to the voice, and partially determine the structure of the face.

number of nasal sprays are available to assist in preventing inflammation within the nasal passageways, thus allowing the normal flow of mucus.

Parental concerns

Sinusitis can usually be treated successfully with antibiotics. It can, however, be very dangerous or even fatal if left untreated it becomes progressively worse such that the infection spreads.

See also Allergic rhinitis.

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Rosalyn Carson-DeWitt, MD

Skeletal development

Definition

Skeletal development refers to the development of the human skeletal system from the early days of pregnancy until the bones have reached full development in late **puberty**.

Description

The early development of the skeletal system begins in the third week after conception with the formation of the notochord (a rod-like structure along the back of the embryo that later becomes the spine, spinal cord, and brain), followed in the fourth week by the first signs of arms and legs. Between the fifth and eighth weeks, the limbs (first the arms, hands, and fingers, followed by the legs, feet, and toes) begin to extend and take on a definite shape.

By the end of the fifth week, the embryo has doubled in size and has grown a tail-like structure that becomes the coccyx (lowermost tip of the backbone). By the seventh week the embryo is about 2 cm (1 in) long and

facial features are visible. At this stage, the 206 bones of the human body are all set down, in surprisingly adult form. However, the process of osteogenesis (development of bone) has not progressed to the point where the bones are "bony." Ossification (the process whereby tissue becomes bone) of most bony nuclei of the long bones and round bones does not complete until after birth.

The major types of human bones are:

- long (e.g. the arm and leg bones)
- short (e.g. the small bones in the wrists and ankles)
- flat (e.g. the bones of the skull or the ribs)
- irregular (e.g. vertebrae)

Long, short, and irregular bones develop by endochondral ossification, where cartilage is replaced by bone. Flat bones develop by intramembranous ossification, where bone develops within sheets of connective tissue. Compact cortical bone, representing about 80 percent of the mature skeleton, supports the body, and features extra thickness at the midpoint in long bones to prevent the bones from bending. Cancellous bone, whose porous structure with small cavities resembles sponge, predominates in the pelvis and the 33 vertebrae from the neck to the tailbone.

Bone growth is more complicated than simple elongation or simple enlargement. Most long bones add width on the outside by a process referred to as subperiosteal apposition (layers added to those already existing), while losing bone on the inside by endosteal resorption (breaking down and reabsorbing material at the center of a mass). At the same time, long bones gain in length by adding to the epiphyseal plate (the surface at the end of the bone). As they elongate, bones of this type go through a process called remodeling during which they change in outer shape as well. Conversely, the individual bones of the skull grow by circumferential apposition (adding layers at the circumference), while gaining in thickness by adding layers (apposition) at the surface with simultaneous resorption at the inner surface. By this process, the skull expands and becomes thicker while allowing for more brain space within.

Linear growth of the long bones occurs by a different process. At birth, long bones have more than one ossification center (regions from which bone growth starts). These grow during childhood until the ends of the bone (epiphyseal plates) become fused with the shaft of the bone (the diaphysis). This process is stimulated by the hormones produced by the testes and ovaries, which provide the developmental signal that the linear growth of the long bones should reach completion or full development. Both round and flat bones of the skeleton are capable of continued growth throughout life.

Ossification centers and their development

The many ossification centers of the body—hand, foot, knee, elbow, and pelvis, for example—are not visible by radiography (x rays) until they begin to mineralize or ossify, even though they are actually present long before such mineralization begins. The age at appearance of individual ossification centers then becomes a useful measure of skeletal development and especially in the form of "bone age" assessments of the hand, foot, or knee. Such assessments, made by taking a series of radiographs and comparing them against appropriate standards, are both highly reliable and useful estimates of the stage of physical development. Bone age assessments are, therefore, used in pediatric evaluation, especially when **malnutrition**, malabsorption, food intolerance, or endocrinopathies (such as hypopituitarism or **hypothyroidism**) are suspected. Bone age assessments also have forensic application, such as estimating the chronological age of a cadaver. In addition, they can provide data for making age assessments for children whose birth date in unknown or for whom a birth certificate does not exist or is suspected of being inaccurate. Families adopting infants or children from countries in which there has been socioeconomic stress may find bone age **assessment** helpful in establishing the chronological age their adopted child has attained.

The normal variability of skeletal age is about 10 percent of attained chronological age. Thus, some chronological 12-year-olds may be assessed as 14 years of age in terms of skeletal development, while others may be assessed as ten. Bone age is useful in projecting final stature; research has shown that it is more meaningful in making such projections than chronological age alone.

Factors affecting bone growth and remodeling

Girls mature earlier than boys, grow for a shorter time, and ultimately have shorter overall bone lengths by about 7 percent. Adolescent girls are, in general, shorter-legged than adolescent boys; this proportional difference is also reflected in the hand and foot skeletons as well. Thus, even at comparable stature, females are shorter-legged and shorter-handed than boys; in addition, girls' bones are more gracile (narrower) than boys and are, therefore, more affected by adult bone loss.

There are major genetically determined differences in relative growth rates of individual bones, in both length and width. Bone widths in general parallel differences in muscle mass and overall frame size. The sequence of ossification of the bones also differs signifi-

cantly from child to child, and the different sequences are controlled by genetics. Differences in growth patterns even among siblings confirm this genetic component. There are also major population differences in skeletal proportions and bone sizes and ratios. Children and adults of African ancestry have relatively longer bones in their hands and feet; the same bones in children and adults of Japanese, Korean, and Chinese ancestry are relatively shorter.

Common problems

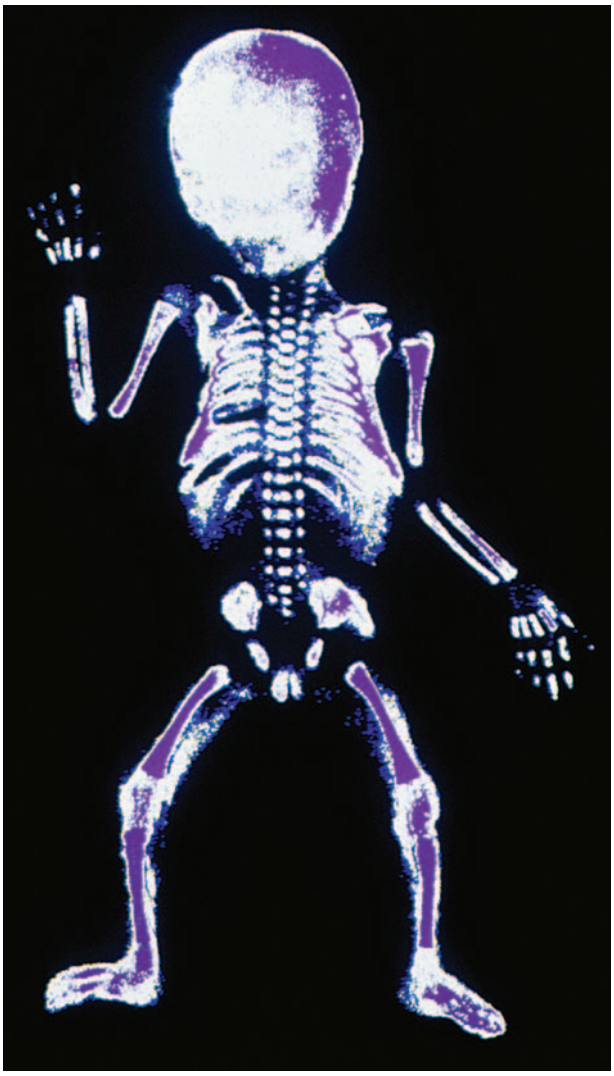
In some cases, abnormalities in skeletal development are caused by nutritional deficiencies that may or may not be reversible. Other disorders are congenital and caused by genetic abnormalities. Steroids used for chronic inflammatory illnesses can lead to thinning of bone in adults (osteoporosis) and to slower bone growth in long bones and therefore shorter status in children.

Problems relating to nutritional status

During childhood, bones are growing rapidly. Bone growth is fueled by a positive energy balance, created by a well-balanced diet and healthy living environment. Even in circumstances of severe malnutrition, there may be some formation of new bone; it will, however, occur while bone formed earlier is deteriorating. During protein malnutrition, bone growth is largely halted, and existing bone is cannibalized by the body as a source of protein. Bone growth may also be limited by **vitamin D deficiency**, resulting in a condition called rickets (osteomalacia), which leads to soft and/or deformed bones and is caused by an inability to absorb calcium due to lack of vitamin D. Treatment and prevention involves sun exposure and vitamin D supplementation.

With growing concern about adult osteoporosis, it is important to realize that the mass of skeleton built during childhood and into early adulthood constitutes bone banked against inevitable later withdrawals. For this reason there is much interest in the proposition that a calcium intake over 1,500 mg per day may build a greater skeletal mass. This proposition, however, in the early 2000s favored in the United States, is met with great skepticism in the United Kingdom. Moreover, calcium intakes during childhood are far greater in the United States than in most countries, but there is no particular evidence that the adult North American skeleton has a greater bone density (bone mass divided by bone volume).

Bone growth, bone remodeling, and the timing of skeletal maturation are all profoundly affected by nutritional status throughout the growing period. In bone remodeling, complex chemical signals prompt cells called osteoclasts to break down and remove (resorb) old bone, and others called osteoblasts to deposit new bone. Many elements influence bone remodeling, including whether the bone is weight-bearing, vitamin D intake, growth factors, and production of various hormones, including estrogen, thyroid, parathyroid, and calcitonin. Thus, poorly nourished boys and girls may be delayed in linear bone growth, diminished in all bone widths, later in the appearance of ossification centers, and delayed in epiphyseal union (completion of long-bone growth). Children living in poverty worldwide may exhibit evidence of smaller amounts of incremental growth of all long bones and vertebrae, and delay in epiphyseal union. By contrast, obese boys and girls evidence greater



X ray showing the skeleton of a newborn. Gaps between bones indicate cartilage, which will develop into bone tissue as the child ages. *(© Howard Sochurek/Corbis.)*

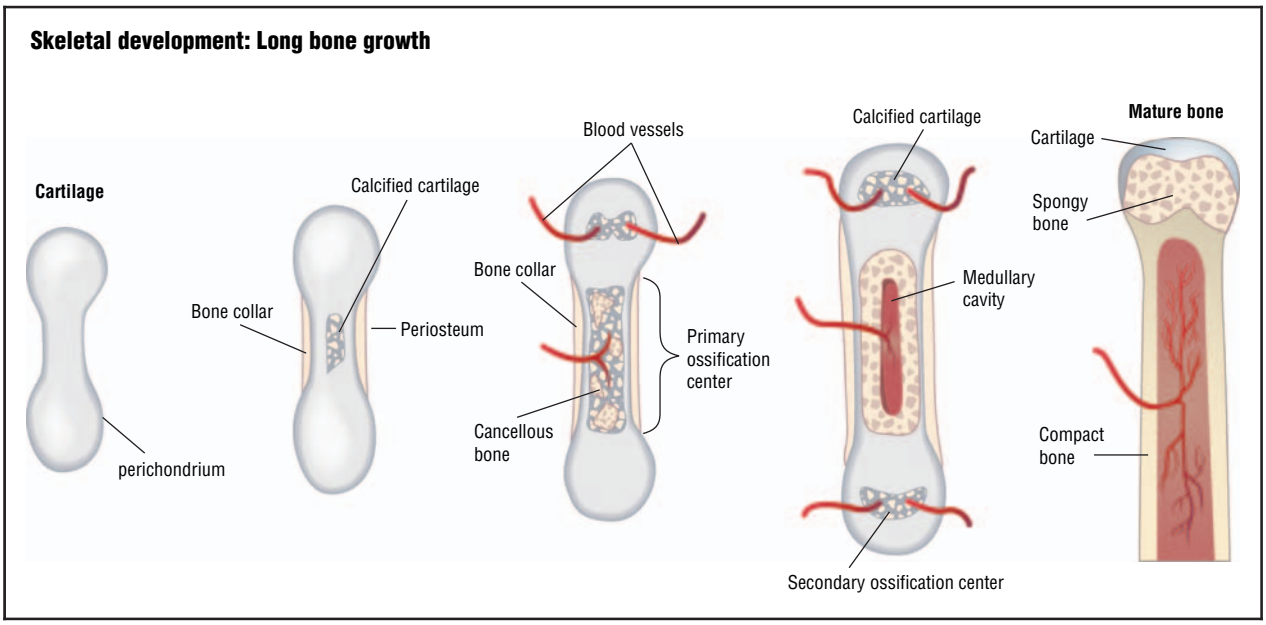


Illustration depicting the stages of long bone growth, showing the process of cartilage calcifying and becoming mature, compact bone. *(Illustration by GGS Information Services.)*

growth in bone lengths and widths, earlier appearance of ossification centers, and earlier completion of epiphyseal union. In other words, obese children grow more, though for a shorter time period, because of their elevated caloric and nutrient intake.

Bone growth is not only affected by simple caloric malnutrition due to inadequate food intake, but also if there is protein deficiency (protein-calorie malnutrition or Kwasiorkor). In protein-calorie malnutrition, lower rates of bone formation may be exceeded by higher rates of bone loss. Thus children and adolescents with protein-calorie malnutrition may show a marked thinning of the outer walls of tubular bones, and an increased incidence of bone **fractures** as a result. Excessive bone loss in protein-calorie malnutrition is also common in juvenile and adolescent cases of **anorexia nervosa**. Individuals with this condition show diminished bone density (bone mass/bone width). There is some evidence that high levels of sodium in the diet of girls ages eight to 13 can significantly increase calcium lost. This effect is particularly powerful in girls whose calcium intake is less than the recommended 1,500 mg—the amount in five glasses of milk—per day.

Congenital skeletal disorders

In some cases, skeletal abnormalities are inherited from one or both parents or occur as a result of genetic mutation. Examples of different congenital disorders of the skeletal system include:

- Achondroplasia: This form of **dwarfism** is characterized by short stature, abnormal body proportion (limbs are shorter than normal while the torso remains of average size), and facial deformities. As a genetic disease, there is no cure. Adult height is generally less than 1.2 m (4 ft).
- Giantism: This condition is characterized by excessive bone growth and is caused by too much growth hormone being produced before puberty (hyperpituitarism). Giantism is treated by inhibiting the production of pituitary hormones.
- Hypopituitarism: Caused by insufficient production of growth hormone, this condition leads to growth failure and delayed skeletal maturation. It can be treated (if diagnosed at an early enough age) with hormone replacement therapy. Hypopituitarism can also be caused by damage to the pituitary gland.
- Osteogenesis imperfecta: Also called brittle bone disease, this condition is characterized by fragile bones that are prone to fracturing. It is a genetic condition that cannot be cured; treatment may involve restricting activity to minimize stress on bones and joints.
- Osteopetrosis: The congenital form of osteopetrosis is rare and involves the formation of overly dense but fragile bones that can lead to frequent fractures, blindness, deafness, and strokes. The more severe form of congenital osteopetrosis is usually fatal within the first ten years of life unless successfully treated with a bone marrow transplant.

KEY TERMS

Cartilage—A tough, elastic connective tissue found in the joints, outer ear, nose, larynx, and other parts of the body.

Endochondral ossification—The process by which cartilage is converted into bone.

Endosteal resorption—The process by which bones are thinned from the inside.

Intramembranous ossification—The process by which bone tissue is formed within sheets of connective tissue.

Osteoporosis—Literally meaning "porous bones," this condition occurs when bones lose an excessive amount of their protein and mineral content, particularly calcium. Over time, bone mass and strength are reduced leading to increased risk of fractures.

Subperiosteal apposition—The process by which bones are made thicker from the outside.

Parental concerns

The question of long-bone growth and completion is of particular concern to parents of children whose growth falls at the outside edges of the normal range. In some cases long-bone growth may be accelerated by growth hormone administration if given by age nine, without speeding the timing of completion of long-bone growth. By encouraging the development of the three key elements of self esteem—acceptance, competence, and purpose—parents can help a child with skeletal abnormalities develop positive body image and confidence in his or her abilities.

When to call the doctor

A healthcare provider should be contacted if a child exhibits symptoms of skeletal or growth abnormalities, such as abnormally short or tall height for age, frequent bone fractures, bony growths, or bone or joint **pain**.

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Skin allergy test *see* **Allergy tests**

Skin rash *see* **Rashes**

Skin tag *see* **Polydactyly and syndactyly**

Sleep

Definition

Sleep is a biological imperative critical to the maintenance of mental and physical health. It is a state of lessened consciousness and decreased physical activity during which the organism slows down and repairs itself. The sleep cycle involves two distinct phases that alternate cyclically from light sleep to deep then deeper and deepest sleep throughout the sleep period. There are two main phases of sleep.

- rapid eye movement (REM) sleep, during which dreaming occurs
- non-rapid eye movement (NREM) or slow-wave sleep (SWS)

Description

The timing and progression of the sleep cycle and the total amount of nightly sleep required for optimal

health varies from infancy to adulthood, depending on developmental stage and **temperament**. Children, particularly infants, require the most sleep during a 24-hour period. The natural sleep-wake cycle, governed by an internal "biological clock," tends toward a 25-hour day. It is affected by the relative balance of light and darkness in the environment. As darkness approaches, the hormone melatonin is secreted by the pineal gland and signals the brain that it is time to sleep.

NREM deep sleep

Sleep begins in stage one of the sleep phase known as NREM, or non-rapid eye movement, sleep. NREM sleep has four stages: light sleep, deeper sleep, and two stages of deepest sleep. Stage one is the "drifting off" period of light sleep in the transition between wakefulness and sleep and comprises about 5 percent of the entire sleep period. Stage two sleep involves a change in brain-wave patterns and increased resistance to arousal and accounts for 45–55 percent of total sleep time. Stages three and four are the deepest levels of sleep and occur only in the first third of the sleep period. NREM stage four sleep usually takes up 12 to 15 percent of total sleep time. Sleep terrors, sleep walking, and bedwetting episodes generally occur within stage four sleep or during partial arousals from this sleep stage.

It typically takes about 90 minutes to cycle through the four deepening stages of NREM sleep before onset of the second phase of sleep known as REM or dream sleep.

REM dream sleep

Rapid eye movement (REM) sleep is qualitatively different from NREM sleep. REM sleep is characterized by extensive central nervous system (CNS) activity with an increase in brain metabolism accompanied by the vivid imagery of dreams. During REM sleep the body is nearly paralyzed, a condition called "atonic," that serves to inhibit the dreamer from physical movement during active dreaming.

"Waking and dreaming are two states of consciousness, with differences that depend on chemistry," according to J. Allan Hobson, professor of psychiatry at Harvard Medical School. Physical activity and thought are suppressed in sleep, but the brain nonetheless remains active "processing information, consolidating and revising memory, and learning newly acquired skills." The brain self-activates, radically changing its chemical climate from wakefulness to sleep states.

REM sleep is also known as "paradoxical sleep" because muscle activity is suppressed even as the CNS registers intense brain activity and spontaneous rapid eye movements can be observed. Brain-wave monitoring of REM sleep with an electroencephalograph (EEG) reveals a low-voltage, fast-frequency, non-alpha wave record. Beyond infancy, REM sleep comprises 20–25 percent of the entire sleep period. This sleep phase is concerned with memory and the consolidation of new information.

Infancy

Newborn infants usually sleep for brief periods at a time around the clock, with the total of day and nighttime sleep roughly equal. A newborn's total sleep need is from 16 to 18 hours in every 24-hour period. Newborns spend approximately 50 percent of their sleep period in the REM phase. Infants are most easily awakened during this phase of sleep that is accompanied by yawning, squirming, and quiet vocalizations.

Infants move through REM and non-REM sleep stages in a 90 minute cycle, and they rise to a near-waking state every three to four hours, more often in breast-fed infants. By about six months of age, babies usually will sleep through the night for 12 or more hours and will continue to nap several times throughout the day.

Researchers conducting a 2004 survey for the National Sleep Foundation discovered that children in every age group fail to meet even the low-end requirements for adequate sleep. By the third month of life, a child's sleep requirement is about 14 to 15 out of every 24 hours, a need that continues until about 11 months of age. However, research indicates that children age three months to 11 months sleep only 12.7 hours on average.

Toddlerhood

Toddlers are far more physically active than infants, and their sleeping behavior and the timing of sleep cycles reflects their maturing brains. A toddler will spend only about 30 percent of her sleep time in REM dream sleep. Toddlers on average require 12 to 14 hours of sleep and may no longer need an afternoon nap to meet this sleep requirement. But research shows that children in the one to three-year-old range may actually average only about 11.7 hours of sleep.

Preschool

Children in this age group tend to be more troubled with **nightmares** and **night terrors** than younger children. They may resist going to bed at night because of **fear** of the dark or of some monster lurking under the bed. Parental reassurance and comfort and the addition of a night light may alleviate some of these concerns.

Preschool children may also feel anxiety around the issue of **toilet training** and bedwetting.

School age

School-age children require from eight to 10 hours of sleep nightly. Adequate sleep is especially important as school children's lives become busier and stress levels rise. Sleep disruptions such as nightmares tend to increase with this age group as the child has more life experiences and anxieties to process. Parents should also monitor the child's use of caffeinated beverages which can cause sleep difficulties and add to the overall loss of adequate sleep.

Adolescents require at least 10 hours of nightly sleep. This is a busy time when many teens' lifestyles include school, work, **sports**, and other **extracurricular activities**, as well as socializing with peers. This increase in activity, together with early-morning school schedules, leaves little time for adequate sleep. Various psychological disorders also may trouble the adolescent, particularly anxiety and depression. Parents should pay attention to a young teen who shows sudden changes in eating habits, loss of interest in usual activities, and other behavioral clues that may indicate onset of depression.

Common problems

According to the "2004 Sleep in America Poll" published by the National Sleep Foundation, 69 percent of children younger than age 10 experience problems with sleep that may occur as often as several times a week. Sleep disruptions in children are usually a normal symptom of central nervous system development. In older children sleep disruptions may increase and intensify due to external stressors in the home or school environment. Sleep difficulties can also be a sign of physical or mental health problems. They are often present in children with attention-deficit/hyperactivity disorder (AD/HD) and in children who have experienced physical, psychological, or sexual abuse.

Childhood sleep problems and parasomnias include:

- Bedwetting: A common sleep problem characterized by involuntary urination during sleep. This is a routine occurrence in children up to five years of age. Bedwetting is also called "nocturnal enuresis."
- Nightmares: A common parasomnia characterized by dreams with frightening psychological content, a feeling of imminent physical danger, and a sensation of being trapped or suffocated. Nightmares occur during REM, or dream-time, sleep and trigger a partial or full awakening. The word "mare" in Old English means "demon."
- Insomnia: Difficulty falling asleep and remaining asleep, or early-morning awakenings. Insomnia may be short-term, due to stress or physical or psychological problems, or may be due to the lack of a healthy bedtime routine.
- Night terrors: A common childhood sleep disruption characterized by an abrupt arousal from stage 4 sleep within the first hour of the sleep period. The child may sit bolt upright in acute terror, screaming inconsolably. Night terrors are a confusional arousal resulting from immature sleep patterns with an intense activation of the flight or fight emotion. They occur in the deepest stage of slow-wave non-REM sleep. Night terrors are also called "pavor nocturnus."
- Sleep apnea: A serious and potentially life-threatening sleep disruption characterized by brief interruptions of airflow during sleep and frequent partial arousals throughout the night. Sleep apnea is less common than other sleep disturbances, occurring in about 2 percent of children.
- Sleep bruxism: A sleep disturbance characterized by grinding the teeth or clenching of the jaws during sleep. Sleep bruxism is common among children of all ages. This sleep problem usually subsides over time.
- Sleep rocking and head banging: A sleep disturbance characterized by rhythmical movements of the body during sleep. Rhythmical movements may be observed in children as young as six months. More dramatic movements, involving head banging and rocking, occur in as many as 60 percent of nine-month-old children. These sleep disturbances tend to decrease with age, appearing in only about 5 percent of children over two years of age.
- Sleep walking: A sleep disturbance characterized by a partial-arousal involving walking about for a few steps, or for much longer distances, with a glassy, trance-like appearance to the eyes. Sleepwalking occurs in the deepest stages of slow-wave, non-REM sleep within the first few hours of sleep onset. Researchers have found that as many as 15–30 percent of children experience at least one sleepwalking episode. Sleepwalking can be triggered by external stimuli, such as an abrupt noise, or by moving a sleeping child to a standing position. This sleep disturbance tends to run in families. Sleepwalking is also called "somnambulism."

Losing sleep

All children need regular and adequate sleep to assure optimal mental and physical health. Sleeping patterns developed in infancy usually persist into adulthood. It is important that parents help the child to establish a

healthy bedtime routine that will assure adequate sleep time, minimize bedtime struggles, and help to reduce the occurrence of common childhood sleep problems.

As reported by Steven Reinberg, research by Maria M. Wong of the University of Michigan, published in 2004 in the journal *Alcoholism: Clinical and Experimental Research,* cautions parents to pay more attention to their children's sleep habits. "Sleep problems are a risk factor for alcohol and drug problems," Wong concluded from data obtained in the first study to link alcohol and drug use with **sleep disorders** in early childhood. The study obtained sleep data from 257 boys ages three to five years and followed them until they were 12–14 years old. Almost half of the children in the study who experienced childhood sleep problems began using alcohol and drugs by the time they were 14 years old.

In many households, electronic distractions interfere with the establishment of a regular bedtime routine that would help a child to settle down and prepare for restful sleep. Calming-down activities, such as being read to by a parent, have been replaced with electronic stimulation resulting in less sleep time.

As reported in *Manchester Online*, Luci Wiggs, a research fellow at Oxford University, is co-author of a 2004 poll of more than 1,000 parents with children four to 10 years of age. She found that 67 percent of these children had a television, computer, or game machine in their bedroom. These stimulating diversions, which she calls "digital distractions," resulted in a cumulative sleep deficit for at least one fifth of the children surveyed that may "compromise children's physical health, academic achievements, and mental health."

Children who consume **caffeine** throughout the day, in soda or iced tea beverages, also lose the sleep required for optimal health and cognitive functioning. A survey by the National Sleep Foundation released in 2004 found that 26 percent of children ages three and older drink at least one caffeinated beverage a day and suffer a loss of about 3.5 hours of sleep each week.

Parental concerns

Parents are on a journey of discovery with each child whose temperament, biology, and sleep habits result in a unique sleep-wake pattern. It can be frustrating when children's sleep habits do not conform to the household schedule. Helping the child develop good sleep habits in childhood takes time and parental attention, but it will have beneficial results throughout life. An understanding of the changing patterns of the typical sleep-wake cycle in children will help alleviate any unfounded concerns. Maintaining a sleep diary for each child will provide the parent with baseline information in assessing the nature and severity of childhood sleep problems. Observant parents will come to recognize unusual sleep disruptions or those that persist or intensify.

When to call the doctor

Developmental changes throughout childhood bring differences in the sleep-wake cycle and in the type and frequency of parasomnias that may interrupt sleep. Medical consultation to rule out illness, infection, or injury is prudent if the child's sleep problems prevent adequate sleep and result in an ongoing sleep deficit. As reported by *News-Medical in Child Health News*, children's sleep problems should be taken seriously as they may be a "'marker' for predicting later risk of early adolescent substance use." In the same article, University of Michigan psychiatry professor Kirk Brower, who has studied "the interplay of alcohol and sleep in adults," stressed that "The finding does not mean there's a cause-and-effect relationship."

Consultation with a child psychologist may be helpful if frightening dreams intensify and become more frequent as this may indicate a particular problem or life circumstance that needs to be changed or one that the child may need extra help working through.

Most childhood sleep disturbances will diminish over time as the brain matures and a regular sleep-wake cycle is established. Parental guidance is crucial to development of healthy sleep habits in children.

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Biological clock—A synonym for the body's circadian rhythm, the natural biological variations that occur over the course of a day.

Parasomnia—A type of sleep disorder characterized by abnormal changes in behavior or body functions during sleep, specific stages of sleep, or the transition from sleeping to waking.

Suprachiasmatic nuclei (SCN)—SCN is that part of the brain that functions as a person's "biological clock" to regulate many body rhythms. The SCN is located on top of the main junction of nerve fibers that connects to the eyes.

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Clare Hanrahan

Sleep apnea, infant *see* **Apnea of infancy**

Sleep disorders

Definition

Sleep disorders are a group of syndromes characterized by disturbance in the individual's amount of sleep, quality or timing of sleep, or in behaviors or physiological conditions associated with sleep.

Description

Although sleep is a basic behavior in animals as well as humans, researchers still do not completely understand all of its functions in maintaining health. Since 1975, however, laboratory studies on human volunteers have yielded information about the different types of sleep. Researchers have learned about the cyclical patterns of different types of sleep and their relationships to breathing, heart rate, brain waves, and other physical functions. These measurements are obtained by a technique called polysomnography. There are about 70 different sleep disorders. To qualify for the diagnosis of sleep disorder, the condition must be a persistent problem, cause the patient significant emotional distress, and interfere with his or her social, academic, or occupational functioning.

There are five stages of human sleep. Four stages have non-rapid eye movement (NREM) sleep, with unique brain wave patterns and physical changes occurring. Dreaming occurs in the fifth stage, during rapid eye movement (REM) sleep.

- Stage one NREM sleep. This stage occurs while a child is falling asleep. It represents about 5 percent of sleep time.

- Stage two NREM sleep. In this stage, (the beginning of "true" sleep), the child's **electroencephalogram** (EEG) will show distinctive waveforms called sleep spindles and K complexes. About 50 percent of sleep time is stage two NREM sleep.
- Stages three and four NREM sleep. Also called delta or slow wave sleep, these are the deepest levels of human sleep and represent 10 to 20 percent of sleep time. They usually occur during the first 30 to 50 percent of the sleeping period.
- REM sleep. REM sleep accounts for 20 to 25 percent of total sleep time. It usually begins about 90 minutes after the child falls asleep. It alternates with NREM sleep about every hour and a half throughout the night. REM periods increase in length over the course of the night.

Sleep cycles vary with a person's age. Children and adolescents have longer periods of stage three and stage four NREM sleep than do middle aged or elderly adults. Because of this difference, the doctor needs to consider the individual's age when evaluating a sleep disorder. Total REM sleep also declines with age.

The average length of nighttime sleep varies among individuals. Most people sleep between seven and nine hours a night. This population average appears to be constant throughout the world. In temperate climates, however, people often notice that sleep time varies with the seasons. It is not unusual for people in North America and Europe to sleep about 40 minutes longer per night during the winter. Infants can regularly sleep up to 16 hours a day. The total amount of sleep declines as the infant gets older. Teenagers may actually need more sleep than slightly younger children and often sleep nine or more hours a day.

Sleep disorders are classified based on what causes them. Primary sleep disorders are distinguished as those that are not caused by other mental disorders, prescription medications, substance abuse, or medical conditions. The two major categories of primary sleep disorders are the dyssomnias and the parasomnias.

Dyssomnias

Dyssomnias are primary sleep disorders in which the patient suffers from changes in the amount, restfulness, and timing of sleep. The most important dyssomnia is primary insomnia, which is defined as difficulty that lasts for at least one month in falling asleep or remaining asleep. Primary insomnia can be caused by many things, including a traumatic event related to sleep or bedtime, and it is often associated with increased physical or psychological arousal at night. Children who experience primary insomnia may develop **anxiety** related to not being able to sleep. The child may come to associate all sleep-related things (their bed, bedtime, etc.) with frustration, making the problem worse. The child may then becomes more stressed about not sleeping.

Hypersomnia is a condition marked by excessive sleepiness during normal waking hours. The individual has either lengthy episodes of daytime sleep or episodes of daytime sleep on a daily basis even though he or she is sleeping normally at night. In some cases, people with primary hypersomnia have difficulty waking in the morning and may appear confused or angry. This condition is sometimes called sleep drunkenness and is more common in males.

The number of people with primary hypersomnia is unknown, although 5 to 10 percent of patients in sleep disorder clinics have the disorder. Primary hypersomnia usually affects young adults between the ages of 15 and 30.

Kleine-Levin syndrome is a recurrent form of hypersomnia that usually starts in late teen years. Doctors do not know the cause of this syndrome. It is marked by excessive drowsiness and for short spells, maybe two to three days, the person sleeps 18 to 20 hours per day, overeats, and is highly irritable. Males are three or four times more likely than females to have the syndrome.

PARASOMNIAS Parasomnias are primary sleep disorders in which the individual's behavior is affected by specific sleep stages or transitions between sleeping and waking. They are sometimes described as disorders of physiological arousal during sleep.

Nightmare disorder is a parasomnia in which the child is repeatedly awakened from sleep by frightening dreams and is fully alert on awakening. The actual rate of occurrence of nightmare disorder is unknown. Approximately 10 to 50 percent of children between three and five years old have **nightmares**, as do many older children. The nightmares occur during REM sleep, usually in the second half of the night. The child is usually able to remember the content of the nightmare and may be afraid to go back to sleep. More females than males have this disorder, but it is not known whether the sex difference reflects a difference in occurrence or a difference in reporting. Nightmare disorder is most likely to occur in children under severe or traumatic stress.

Sleep terror disorder is a parasomnia in which the child awakens screaming or crying. The child also has physical signs of arousal, like sweating and shaking. Sleep terror is sometimes referred to as pavor nocturnus. Unlike nightmares, sleep terrors typically occur in stage three or stage four NREM sleep during the first third of

the night. The child may be confused or disoriented for several minutes and cannot recall the content of the dream. He or she may fall asleep again and not remember the episode the next morning. Sleep terror disorder is most common in children four to 12 years old and is usually outgrown in **adolescence**. It affects about 3 percent of children. In children, more males than females have the disorder.

Sleepwalking disorder, which is sometimes called **somnambulism**, occurs when the child is capable of complex movements during sleep, including walking. Like sleep terror disorder, sleepwalking occurs during stage three and stage four NREM sleep during the first part of the night. If the child is awakened during a sleepwalking episode, he or she may be disoriented and have no memory of the behavior. In addition to walking around, individuals with sleepwalking disorder have been reported to eat, use the bathroom, unlock doors, or talk to others. It is estimated that 10 to 30 percent of children have at least one episode of sleepwalking. However, only 1 to 5 percent meet the criteria for sleepwalking disorder. The disorder is most common in children eight to 12 years old.

Demographics

In the United States, 20 to 25 percent of children have some kind of sleep problem. Nightmares are believed to occur in about 30 percent of children, usually in younger children. Sleepwalking occurs more than once in about 25 to 30 percent of children. The most common age group to experience sleepwalking is children under 10. Insomnia is reported to occur in approximately 23 percent of children. Many other sleep disorders occur less frequently but are still a problem for many children.

Causes and symptoms

The causes of sleep disorders vary depending on the disorder. Many times, stress, anxiety, or other factors are found to be the cause. Often the underlying cause of the sleep disorder is never found.

The most important symptoms of sleep disorders are insomnia and sleepiness during waking hours. Insomnia is by far the more common of the two symptoms. It covers a number of different patterns of sleep disturbance. These patterns include inability to fall asleep at bedtime, repeated awakening during the night, and/or inability to go back to sleep once awakened.

When to call the doctor

If a child does not seem to be getting enough sleep at night or the child wakes frequently or seems tired frequently during the day, it may be helpful to consult a doctor.

Diagnosis

Diagnosis of sleep disorders usually requires a psychological history as well as a medical history. Physical examinations are not usually revealing. The patient's sex and age are useful starting points in assessing the problem. The doctor may also talk to other **family** members in order to obtain information about the patient's symptoms. The family's observations are particularly important for evaluating sleepwalking, kicking in bed, snoring loudly, or other behaviors that the patient cannot remember.

Psychological testing

The doctor may use **psychological tests** or inventories to evaluate insomnia because it is frequently associated with mood or affective disorders. The **Minnesota Multiphasic Personality Inventory** (MMPI), the Millon Clinical Multiaxial Inventory (MCMI), the Beck Depression Inventory, and the Zung Depression Scale are the tests most commonly used in evaluating this symptom.

Laboratory studies

If the doctor is considering breathing-related sleep disorders, myoclonus, or **narcolepsy** as possible diagnoses, he or she may ask the patient to be tested in a sleep laboratory or at home with portable instruments.

POLYSOMNOGRAPHY Polysomnography can be used to help diagnose sleep disorders as well as conduct research into sleep. In some cases the patient is tested in a special sleep laboratory. The advantage of this testing is the availability and expertise of trained technologists, but it is expensive. Since 2001, however, portable equipment is available for home recording of certain specific physiological functions.

MULTIPLE SLEEP LATENCY TEST (MSLT) The multiple sleep latency test (MSLT) is frequently used to measure the severity of the patient's daytime sleepiness. The test measures sleep latency (the speed with which the patient falls asleep) during a series of planned naps during the day. The test also measures the amount of REM sleep that occurs. Two or more episodes of REM sleep under these conditions indicates narcolepsy. This test can also be used to help diagnose primary hypersomnia.

REPEATED TEST OF SUSTAINED WAKEFULNESS (RTSW) The repeated test of sustained wakefulness (RTSW) measures sleep latency by challenging the patient's ability to stay awake. In the RTSW, the patient is placed in a quiet room with dim lighting and is asked to stay awake. As with the MSLT, the testing pattern is repeated at intervals during the day.

Treatment

Treatment for a sleep disorder depends on what is causing the disorder. For example, if major depression is the cause of insomnia, then treatment of the depression with **antidepressants** or psychological counseling should resolve the insomnia. The use of antidepressants in minors is a matter of debate. In October 2003, the United States Food and Drug Administration issued an advisory indicating that children being treated with selective serotonin re-uptake inhibitor antidepressants (SSRIs) for major depressive illness may be at higher risk for committing **suicide**. A similar warning was issued in the United Kingdom. Parents and physicians must weigh the benefits and risks of prescribing these medications for children on an individual basis.

Medications

Medications for sleep disorders are generally not recommended for use by children. In most cases medications are the treatment of last resort. If children with sleep terror disorder or sleepwalking are treated with medication, then they may be given benzodiazepines because this type of medication suppresses stage three and stage four NREM sleep.

Psychotherapy

Psychotherapy is recommended for patients with sleep disorders associated with other mental disorders. In many cases the patient's scores on the Beck or Zung inventories will suggest the appropriate direction of treatment.

Sleep preparation

Children with sleep disorders such as insomnia may benefit from a regular pattern of pre-bedtime rituals designed to help the child relax and prepare for bed. Fluid intake should usually be limited in the hours before bed to reduce the need to get out of bed and use the toilet. Children should generally not be given **caffeine** in the evening, as it may make it harder for them to fall asleep. Children with nightmare disorder may benefit from limits on television or movies. Violent scenes or frightening science fiction stories appear to influence the frequency and intensity of children's nightmares.

Alternative treatment

Some alternative approaches may be effective in treating insomnia caused by anxiety or emotional stress. For some people, meditation practice, breathing exercises, and **yoga** can break the vicious cycle of sleeplessness, worry about inability to sleep, and further sleeplessness. Yoga can help some people to relax muscular tension in a direct fashion. The breathing exercises and meditation can keep some patients from obsessing about sleep.

Homeopathic practitioners recommend that people with chronic insomnia see a professional homeopath. They do, however, prescribe specific remedies for at-home treatment of temporary insomnia: *Nux vomica* for alcohol or substance-related insomnia, *Ignatia* for insomnia caused by grief, *Arsenicum* for insomnia caused by **fear** or anxiety, and *Passiflora* for insomnia related to mental stress.

Melatonin has also been used as an alternative treatment for sleep disorders. Melatonin is produced in the body by the pineal gland at the base of the brain. This substance is thought to be related to the body's circadian rhythms.

Practitioners of traditional Chinese medicine usually treat insomnia as a symptom of excess yang energy. Cinnabar is recommended for chronic nightmares. Either magnetic magnetite or "dragon bones" is recommended for insomnia associated with hysteria or fear. If the insomnia appears to be associated with excess yang energy arising from the liver, the practitioner will give the patient oyster shells. Acupuncture treatments can help bring about balance and facilitate sleep.

Dietary changes such as eliminating stimulant foods (coffee, cola, chocolate) and late-night meals or snacks can be effective in treating some sleep disorders. Nutritional supplementation with magnesium, as well as botanical medicines that calm the nervous system, can also be helpful. Among the botanical remedies that may be effective for sleep disorders are valerian (*Valeriana officinalis*), passionflower (*Passiflora incarnata*), and skullcap (*Scutellaria lateriflora*).

Prognosis

The prognosis depends on the specific disorder. Children usually outgrow sleep disorders. Patients with Kleine-Levin syndrome usually get better around age 40. The prognosis for sleep disorders related to many other

KEY TERMS

Apnea—The temporary absence of breathing. Sleep apnea consists of repeated episodes of temporary suspension of breathing during sleep.

Cataplexy—A symptom of narcolepsy in which there is a sudden episode of muscle weakness triggered by emotions. The muscle weakness may cause the person's knees to buckle, or the head to drop. In severe cases, the patient may become paralyzed for a few seconds to minutes.

Circadian rhythm—Any body rhythm that recurs in 24-hour cycles. The sleep-wake cycle is an example of a circadian rhythm.

Dyssomnia—A primary sleep disorder in which the patient suffers from changes in the quantity, quality, or timing of sleep.

Electroencephalogram (EEG)—A record of the tiny electrical impulses produced by the brain's activity picked up by electrodes placed on the scalp. By measuring characteristic wave patterns, the EEG can help diagnose certain conditions of the brain.

Hypersomnia—An abnormal increase of 25% or more in time spent sleeping. Individuals with hypersomnia usually have excessive daytime sleepiness.

Hypnotics—A class of drugs that are used as a sedatives and sleep aids.

Hypopnea—Shallow or excessively slow breathing usually caused by partial closure of the upper airway during sleep, leading to disruption of sleep.

Insomnia—A sleep disorder characterized by inability either to fall asleep or to stay asleep.

Jet lag—A temporary disruption of the body's sleep-wake rhythm following high-speed air travel across several time zones. Jet lag is most severe in people who have crossed eight or more time zones in 24 hours.

Kleine-Levin syndrome—A disorder that occurs primarily in young males, three or four times a year. The syndrome is marked by episodes of hypersomnia, hypersexual behavior, and excessive eating.

Narcolepsy—A life-long sleep disorder marked by four symptoms: sudden brief sleep attacks, cataplexy (a sudden loss of muscle tone usually lasting up to 30 minutes), temporary paralysis, and hallucinations. The hallucinations are associated with falling asleep or the transition from sleeping to waking.

Nocturnal myoclonus—A disorder in which the patient is awakened repeatedly during the night by cramps or twitches in the calf muscles. Also sometimes called periodic limb movement disorder.

Non-rapid eye movement (NREM) sleep—A type of sleep that differs from rapid eye movement (REM) sleep. The four stages of NREM sleep account for 75–80% of total sleeping time.

Parasomnia—A type of sleep disorder characterized by abnormal changes in behavior or body functions during sleep, specific stages of sleep, or the transition from sleeping to waking.

Pavor nocturnus—Another name for sleep terror disorder.

Polysomnography—An overnight series tests designed to evaluate a patient's basic physiological processes during sleep. Polysomnography generally includes monitoring of the patient's airflow through the nose and mouth, blood pressure, electrocardiographic activity, blood oxygen level, brain wave pattern, eye movement, and the movement of respiratory muscles and limbs

Primary sleep disorder—A sleep disorder that cannot be attributed to a medical condition, another mental disorder, or prescription medications or other substances.

Rapid eye movement (REM) latency—The amount of time it takes for the first onset of REM sleep after a person falls asleep.

Rapid eye movement (REM) sleep—A phase of sleep during which the person's eyes move rapidly beneath the lids. It accounts for 20-25% of sleep time. Dreaming occurs during REM sleep.

Restless legs syndrome (RLS)—A disorder in which the patient experiences crawling, aching, or other disagreeable sensations in the calves that can be relieved by movement. RLS is a frequent cause of difficulty falling asleep at night.

Sedative—A medication that has a calming effect and may be used to treat nervousness or restlessness. Sometimes used as a synonym for hypnotic.

Sleep latency—The amount of time that it takes to fall asleep. Sleep latency is measured in minutes and is important in diagnosing depression.

Somnambulism—Another term for sleepwalking.

conditions depends on successful treatment of the underlying problem. The prognosis for primary sleep disorders is affected by many things, including the patient's age, sex, occupation, personality characteristics, family circumstances, neighborhood environment, and similar factors.

Prevention

There is no known way to prevent sleep disorders, although having a good, regular, sleep schedule with a nighttime ritual intended to reduce stress may help.

Parental concerns

Children who do not get enough sleep, or do not get good quality sleep, may seem irritable or uncooperative during the day. Lack of sleep reduces the ability to concentrate and decreases mental functioning, so children who are not getting enough good sleep at night may have poor concentration skills and poor academic performance.

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Small-for-gestational-age infant *see* **Intrauterine growth retardation**

Smoke inhalation

Definition

Smoke inhalation is breathing in the harmful gases, vapors, and particulate matter contained in smoke.

Description

Smoke inhalation typically occurs in victims or firefighters caught in structural fires. However, cigarette **smoking** also causes similar damage on a smaller scale over a longer period of time. People who are trapped in fires may suffer from smoke inhalation independent of receiving skin **burns**; however, the incidence of smoke inhalation increases with the percentage of total body surface area burned. Smoke inhalation contributes to the total number of fire-related deaths each year for several reasons: the damage is serious; its diagnosis is not always easy because as of 2004 there were no sensitive diagnostic tests; and people may not show symptoms until 24 to 48 hours after the event.

Demographics

According to the National Safety Council, 3,900 people died from exposure to fire, flame, and smoke in the United States in 2001, the most recent year as of 2004 for which data were available. Smoke inhalation accounts for the majority of deaths in home fires. Children under age 11 and adults over age 70 are most vulnerable to the effects of smoke inhalation.

Causes and symptoms

The harmful materials given off by combustion injure the airways and lungs in three ways: heat damage, tissue irritation, and oxygen starvation of tissues (asphyxiation). Signs of heat damage are singed nasal hairs, burns around and inside the nose and mouth, and internal swelling of the throat. Tissue irritation of the throat and lungs may appear as noisy breathing, coughing, hoarseness, black or gray spittle, and fluid in the lungs. Asphyxiation is apparent from shortness of breath and blue-gray or cherry-red skin color. In some cases, the person may not be conscious or breathing.

When to call the doctor

A doctor should be called whenever smoke is inhaled for more than a few minutes or whenever the inhaled smoke and fumes are known to contain toxic substances.

Diagnosis

In addition to looking for the signs of heat damage, tissue irritation, and asphyxiation, the physician will assess the individual's breathing by the respiratory rate (number of breaths per minute) and motion of the chest as the lungs inflate and deflate. The person's circulation

is also evaluated by the pulse rate (number of heartbeats per minute) and blood pressure. Blood tests will indicate the levels of oxygen and byproducts of poisonous gases. Chest x rays are too insensitive to show damage to delicate respiratory tissues but can show fluid in the lungs (pulmonary edema).

The physician may perform a bronchoscopy, a visual examination in which the airways and lungs are seen through a fiber optic tube inserted down the person's windpipe (trachea). Other **pulmonary function tests** may be performed to measure how efficiently the lungs are working.

Treatment

Treatment varies with the severity of the damage caused. The primary focus of treatment is to maintain an open airway and provide an adequate level of oxygen. If the airway is open and stable, the individual may be given high-flow humidified 100 percent oxygen by mask. If swelling of the airway tissues is closing off the airway, the person may require the insertion of an endotracheal tube to artificially maintain an open airway.

Oxygen is often the only medication necessary. However, people who have a **cough** with wheezing (bronchospasm), indicating that the bronchial airways are narrowed or blocked, may be given a bronchodilator to relax the muscles and increase ventilation. There are also antidotes for specific poisonous gases in the blood; dosage is dependent upon the level indicated by blood tests. **Antibiotics** are not given until sputum and blood cultures confirm the presence of a bacterial infection.

In institutions where it is available, hyperbaric oxygen therapy may be used to treat smoke inhalation, resulting in severe carbon monoxide or cyanide **poisoning**. This treatment requires a special chamber in which the person receives pure oxygen at three times the normal atmospheric pressure, thus receiving more oxygen faster to overcome loss of consciousness, altered mental state, cardiovascular dysfunction, pulmonary edema, and severe neurological damage.

Botanical medicine can help to maintain open airways and heal damaged mucous membranes. It can also help support the entire respiratory system. Acupuncture and homeopathic treatment can provide support to the whole person who has suffered a traumatic injury such as smoke inhalation.

Prognosis

Although the outcome depends of the severity of the smoke inhalation and the severity of any accompanying burns or other injuries, with prompt medical treatment, the prognosis for recovery is good. However, some people may experience chronic pulmonary problems following smoke inhalation, and those with **asthma** or other chronic respiratory conditions prior to smoke inhalation may find their original conditions have been aggravated by the inhalation injury.

Prevention

Smoke inhalation is best avoided by preventing structural fires. Doing so involves inspection of wiring; safe use and storage of flammable liquids; and maintenance of clean, well-ventilated chimneys, wood stoves, and space heaters. Properly placed and working smoke detectors in combination with rapid evacuation plans minimize a person's exposure to smoke in the event of a fire. When escaping a burning building, a person should move close to the floor where there the air is cooler and clearer to breathe because hot air rises, carrying gases and particulate matter upward. Finally, firefighters should always wear proper protective gear.

Parental concerns

Parents should monitor their homes to make sure they provide a safe environment for everyone, including their children. They should also monitor **play** and recreational activities to limit exposure to smoke or toxic fumes. Parents should regularly check smoke detectors and change batteries every six months. In addition, families should have a fire escape plan, including a designated meeting area away from the house. This plan should be practiced periodically.

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KEY TERMS

Asphyxiation—Oxygen starvation of tissues. Chemicals such as carbon monoxide prevent the blood from carrying sufficient oxygen to the brain and other organs. As a result, the person may lose consciousness, stop breathing, and die without artificial respiration (assisted breathing) and other means of elevating the blood oxygen level.

Hyperbaric oxygen therapy—Medical treatment in which oxygen is administered in specially designed chambers, under pressures greater than that of the atmosphere, in order to treat specific medical conditions, such as carbon monoxide poisoning, smoke inhalation, and certain bacterial infections.

Pulmonary—Referring to the lungs and respiratory system.

Pulmonary edema—An accumulation of fluid in the tissue of the lungs.

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American College of Emergency Physicians. PO Box 619911, Dallas, TX 75261–9911. Web site: <www.acep.org/>.

American College of Occupational and Environmental Medicine. 55 West Seegers Rd., Arlington Heights, IL 60005. Web site: <www.acoem.org/>.

American College of Osteopathic Emergency Physicians. 142 E. Ontario St., Suite 550, Chicago, IL 60611. Web site: <www.acoep.org/>.

American College of Physicians. 190 N Independence Mall West, Philadelphia, PA 19106–1572. Web site: <http://www.acponline.org/>.

American Lung Association. 1740 Broadway, New York, NY 10019. Web site: <www.lungusa.org/diseases/lungtb.html>.

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Smoking

Definition

Smoking is the inhalation of the smoke of burning tobacco that is used mostly in three forms: cigarettes, pipes, and cigars.

Description

Casual smoking is the act of smoking only occasionally, usually in a social situation or to relieve stress. A smoking habit is a physical **addiction** to tobacco products. Many health experts as of 2004 regarded habitual smoking as a psychological addiction, one with serious health consequences. Nicotine, the active ingredient in tobacco, is inhaled into the lungs, where most of it stays. The rest passes into the bloodstream, reaching the brain in about 10 seconds and dispersing throughout the body in about 20 seconds.

Depending on the circumstances and the amount consumed, nicotine can act as either a stimulant or tranquilizer. This dual role explains why some people report that smoking gives them energy and stimulates their mental activity, while others note that smoking relieves **anxiety** and relaxes them. The initial effect results in part from the drug's stimulation of the adrenal glands and resulting release of epinephrine into the blood. Epinephrine causes several physiological changes: it temporarily narrows the arteries, raises the blood pressure, raises the levels of fat in the blood, and increases the heart rate and flow of blood from the heart. Some researchers think epinephrine contributes to smokers' increased risk of high blood pressure.

Nicotine, by itself, increases the risk of heart disease. However, when a person smokes, he or she is ingesting a lot more than nicotine. Smoke from a cigarette, pipe, or cigar is made up of many additional toxic chemicals, including tar and carbon monoxide. Tar is a

sticky substance that forms as deposits in the lungs, causing lung **cancer** and respiratory distress. Carbon monoxide limits the amount of oxygen that the red blood cells can convey throughout the body. Nicotine may also damage the inner walls of the arteries, which allows fat to build up in them.

Besides tar, nicotine, and carbon monoxide, tobacco smoke contains 4,000 different chemicals. More than 200 of these chemicals are known to be toxic. Nonsmokers who are exposed to tobacco smoke also take in these toxic chemicals. They inhale the smoke exhaled by the smoker as well as the more toxic sidestream smoke—the smoke from the end of the burning cigarette, cigar, or pipe.

The harmful effects of teenage smoking are both short-term and long-term. During **adolescence**, smoking interferes with ongoing lung growth and development, preventing the attainment of full lung function. Teenagers who smoke are less fit than their nonsmoking peers and more apt to experience shortness of breath, **dizziness**, coughing, and excess phlegm in their lungs. They are also more vulnerable to colds, flu, **pneumonia**, and other respiratory problems. Smoking for even a short time can produce a chronic smoker's **cough**. In addition to respiratory problems and a diminished level of overall well-being in adolescence, teenage smoking is also responsible for health problems in adulthood.

It is estimated that one third of the teenagers who start smoking each year eventually die of diseases related to tobacco use, diseases that will shorten their lives by an average of 12–15 years. Cigarette smoking is a major risk factor for cardiovascular disease, including coronary heart disease, atherosclerosis (hardening of the arteries), and **stroke**. Reports by the surgeon general link teenage smoking to cardiovascular disease in both adolescents and adults. The same reports cite evidence that the length of time a person has smoked has a greater impact on the risk of developing lung cancer and other smoking-related cancers than the number of cigarettes smoked; in other words, starting to smoke at an early age is an even greater health risk than being a heavy smoker.

Demographics

The National Survey on Drug Use and Health (NSDUH) is conducted annually by the Substance Abuse and Mental Health Services Administration (SAMHSA) of the U.S. Department of Health and Human Services. The study found that an estimated 70.8 million Americans reported current (past month) use of a tobacco product in 2003. This is 29.8 percent of the population aged 12 or older, similar to the rate in 2002 (30.4%). Young adults aged 18–25 reported the highest rate of past month cigarette use (40.2%), similar to the rate among young adults in 2002. An estimated 35.7 million Americans aged twelve or older in 2003 were classified as nicotine dependent in the past month because of their cigarette use (15% of the total population), about the same as for 2002.

Young adults aged 18 to 25 had the highest rate of current use of cigarettes (40.2%), similar to the rate in 2002. Past month cigarette use rates among youths in 2002 and 2003 were 13 percent and 12.2 percent, respectively, not a statistically significant change. However, there were significant declines in past year (from 20.3% to 19%) and lifetime (from 33.3% to 31%) cigarette use among youths aged 12 to 17 between 2002 and 2003. Among persons aged twelve or older, a higher proportion of males than females smoked cigarettes in the past month in 2003 (28.1% versus 23%). Among youths aged 12 to 17, however, girls (12.5%) were as likely as boys (11.9%) to smoke in the past month. There was no change in cigarette use among boys aged 12 to 17 between 2002 and 2003. However, among girls, cigarette use decreased from 13.6 percent in 2002 to 12.5 percent in 2003.

Causes and symptoms

No one starts smoking to become addicted to nicotine. It is not known how much nicotine may be consumed before the body becomes addicted. However, once smoking becomes a habit, the smoker faces a lifetime of health risks associated with one of the strongest addictions known to humans.

Smoking risks

Smoking is recognized as the leading preventable cause of death, causing or contributing to the deaths of approximately 430,700 Americans each year. Anyone with a smoking habit has an increased chance of cancer (lung, cervical, and other types); respiratory diseases (emphysema, **asthma**, and chronic **bronchitis**); and cardiovascular disease (heart attack, high blood pressure, stroke, and atherosclerosis). The risk of stroke is especially high in women who take birth control pills.

Smoking can damage fertility, making it harder to conceive, and it can interfere with the growth of the fetus during pregnancy. It accounts for an estimated 14 percent of premature births and 10 percent of infant deaths. There is some evidence that smoking may cause impotence in men. Because smoking affects so many of the body's systems, smokers often have vitamin deficiencies and suffer oxidative damage caused by free radicals.

Free radicals are molecules that steal electrons from other molecules, turning the other molecules into free radicals and destabilizing the molecules in the body's cells.

Studies reveal that the more a person smokes, the more likely he is to sustain illnesses such as cancer, chronic bronchitis, and emphysema. But even smokers who indulge in the habit only occasionally are more prone to these diseases. Some brands of cigarettes are advertised as low tar, but no cigarette is truly safe. If a smoker switches to a low-tar cigarette, he is likely to inhale longer and more deeply to get the chemicals his body craves. A smoker has to quit the habit entirely in order to improve his health and decrease the chance of disease.

Though some people believe chewing tobacco is safer, it also carries health risks. People who chew tobacco have an increased risk of heart disease and mouth and throat cancer. Pipe and cigar smokers have increased health risks as well, even though these smokers generally do not inhale as deeply as cigarette smokers do. These groups have not been studied as extensively as cigarette smokers, but there is evidence that they may be at a slightly lower risk of cardiovascular problems but a higher risk of cancer and various types of circulatory conditions. Some research reveals that passive smokers, or those who unavoidably breathe in second-hand tobacco smoke, have an increased chance of many health problems such as lung cancer, asthma, and **sudden infant death syndrome** in babies.

Smokers' symptoms

Smokers are likely to exhibit a variety of symptoms that reveal the damage caused by smoking. A nagging morning cough may be one sign of a tobacco habit. Other symptoms include shortness of breath, wheezing, and frequent occurrences of respiratory illness, such as bronchitis. Smoking also increases fatigue and decreases the smoker's sense of smell and taste. Smokers are more likely to develop poor circulation, with cold hands and feet, and premature wrinkles.

Sometimes the illnesses that result from smoking come with little warning. For instance, coronary artery disease may exhibit few or no symptoms. At other times, there will be warning signs, such as bloody discharge from a woman's vagina, a sign of cancer of the cervix. Another warning sign is a hacking cough, worse than the usual smoker's cough, that brings up phlegm or blood, a sign of lung cancer.

Withdrawal symptoms

A smoker who tries to quit may expect one or more of these withdrawal symptoms: **nausea**, **constipation** or **diarrhea**, drowsiness, loss of concentration, insomnia, **headache**, nausea, and irritability.

When to call the doctor

Smokers should seek medical help if they want to quit smoking but are unable to do so, or if they exhibit signs of any of the illnesses associated with long-term tobacco use. Persons who are frequently around smokers should seek medical advice if they show any of the symptoms associated with illnesses caused by smoking since second-hand smoke can be more damaging to health than first-hand smoke.

Diagnosis

It is not easy to quit smoking. That is why it may be wise for smokers to turn to their physician for help. For the greatest success in quitting and to help with the withdrawal symptoms, smokers should talk over a treatment plan with their doctor or alternative practitioner. They should have a general physical examination to gauge their general health and uncover any deficiencies. They should also have a thorough evaluation for some of the serious diseases that smoking can cause.

Research shows that most smokers who want to quit benefit from the support of other people. It helps to quit with a friend or to join a group such as those organized by the American Cancer Society. These groups provide support and teach behavior modification methods that can help the smoker quit. The smoker's physician can often refer him to such groups.

Other alternatives to help with the withdrawal symptoms include nicotine replacement therapy in the form of gum, patches, nasal sprays, and oral inhalers. These are available by prescription or over the counter. A physician can provide advice on how to use them. They slowly release a small amount of nicotine into the bloodstream, satisfying the smoker's physical craving. Over time, the amount of gum the smoker chews is decreased and the amount of time between applying the patches is increased. This process helps wean the smoker from nicotine slowly. However, if the smoker smokes while taking a nicotine replacement, a nicotine overdose may occur.

The drug buproprion hydrochloride has shown some success in helping smokers quit. This drug contains no nicotine and was originally developed as an antidepres-

sant. It is not known exactly how buproprion works to suppress the desire for nicotine.

Alternative treatment

There are a wide range of alternative treatments that can help a smoker quit the habit, including hypnotherapy, herbs, acupuncture, and meditation. For example, a controlled trial demonstrated that self-massage can help smokers crave less intensely, smoke fewer cigarettes, and in some cases completely give them up.

Prognosis

Research on smoking shows that 80 percent of all smokers desire to quit. But smoking is so addictive that fewer than 20 percent of the people who try ever successfully break the habit. Still, many people attempt to quit smoking over and over again, despite the difficulties—the cravings and withdrawal symptoms, such as irritability and restlessness.

For those who do quit, it is well worth the effort. The good news is that once a smoker quits the health effects are immediate and dramatic. After the first day, oxygen and carbon monoxide levels in the blood return to normal. At two days, nerve endings begin to grow back and the senses of taste and smell revive. Within two weeks to three months, circulation and breathing improve. After one year of not smoking, the risk of heart disease is reduced by 50 percent. After 15 years of abstinence, the risks of health problems from smoking virtually vanish. A smoker who quits for good often feels a lot better too, with less fatigue and fewer respiratory illnesses.

Prevention

How do smokers give up their cigarettes for good and never go back to them again? Here are a few tips from the experts:

- People should tell their friends and neighbors that they are quitting. Doing so helps make quitting a matter of pride.
- They should chew sugarless gum or eat sugar-free hard candy to redirect the oral fixation that comes with smoking and to prevent weight gain.
- They should eat as much as they want, but only low-calorie foods and drinks. They should drink plenty of water, which may help with the feelings of tension and restlessness that quitting can bring. After eight weeks, they will lose their craving for tobacco, so it is safe then to return to their usual eating habits.
- They should stay away from situations that prompt smoking, avoiding other people who smoke and dining in the nonsmoking section of restaurants.

Parental concerns

Parents and guardians need to be aware of the power they have to influence the development of their kids throughout the pre-teen and teenage years. Adolescence brings a new and dramatic stage to **family** life. The changes that are required are not just the teen's to make; parents need to change their relationship with their teenager. It is best if parents are proactive about the challenges of this life cycle stage, particularly those that pertain to the possibility of experimenting with and using tobacco. Parents should not be afraid to talk directly to their kids about smoking, even if they have had problems with smoking themselves. Parents should give clear, no-use messages about smoking and its negative consequences on health. It is important for kids and teens to understand that the rules and expectations set by parents are based on parental love and concern for their well-being. Parents should also be actively involved and demonstrate interest in their teen's friends and social activities. Spending quality time with teens and setting good examples are essential. Even if tobacco use already exists in the teen's life, parents and families can still have a positive influence on their teen's behavior.

Resources

BOOKS

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Haugen, Hayley Mitchell. *Teen Smoking*. San Diego, CA: Greenhaven Press, 2004.

Shipley, Robert H. *Stop Smoking Kit: Quit Smart Stop Smoking Guide, Hypnosis Quit Smoking CD, and Cigarette Substitute*. Durham, NC: QuitSmart Stop Smoking Resources, Inc., 2004.

PERIODICALS

Brook, Judith S., et al. "Tobacco Use and Health in Young Adulthood." *Journal of Genetic Psychology* (September 2004): 310–23.

Frieden, Joyce. "Peer Pressure Likely to Prompt Tobacco Use: Behavior Predictors Studied." *Family Practice News* (June 15, 2004): 66.

McCollum, Sean. "Up in Smoke: Smoking Harms Your Health and Empties Your Wallet. How Much Does This Lethal Habit Cost? Do the Math and Find Out." *Scholastic Choices* (February-March 2004): 16–20.

KEY TERMS

Antioxidant—Any substance that reduces the damage caused by oxidation, such as the harm caused by free radicals.

Chronic bronchitis—A smoking-related respiratory illness in which the membranes that line the bronchi, or the lung's air passages, narrow over time. Symptoms include a morning cough that brings up phlegm, breathlessness, and wheezing.

Emphysema—A chronic respiratory disease that involves the destruction of air sac walls to form abnormally large air sacs that have reduced gas exchange ability and that tend to retain air within the lungs. Symptoms include labored breathing, the inability to forcefully blow air out of the lungs, and an increased susceptibility to respiratory tract infections. Emphysema is usually caused by smoking.

Epinephrine—A hormone produced by the adrenal medulla. It is important in the response to stress and partially regulates heart rate and metabolism. It is also called adrenaline.

Flavonoid—A food chemical that helps to limit oxidative damage to the body's cells, and protects against heart disease and cancer.

Free radical—An unstable molecule that causes oxidative damage by stealing electrons from surrounding molecules, thereby disrupting activity in the body's cells.

Nicotine—A colorless, oily chemical found in tobacco that makes people physically dependent on smoking. It is poisonous in large doses.

Nicotine replacement therapy—A method of weaning a smoker away from both nicotine and the oral fixation that accompanies a smoking habit by giving the smoker smaller and smaller doses of nicotine in the form of a patch or gum.

Secondhand smoke—A mixture of the smoke given off by the burning end of a cigarette, pipe, or cigar and the smoke exhaled from the lungs of smokers.

Sidestream smoke—The smoke that is emitted from the burning end of a cigarette or cigar, or that comes from the end of a pipe. Along with exhaled smoke, it is a constituent of second-hand smoke.

"Reports: Fewer U.S., Canadian Youth are Lighting Up." *Tobacco Retailer* (August 2004): 7–8.

"Tobacco Use among Middle and High School Students—United States, 2002." *Morbidity and Mortality Weekly Report* (November 14, 2003): 1096–98.

ORGANIZATIONS

Campaign for Tobacco-Free Kids. 1400 Eye Street, Suite 1200, Washington DC 20005. Web site: <www.tobaccofreekids.org>.

Youth Anti-Tobacco Collaborative. 1469 Park Ave., San Jose, CA 95128. Web site: <www.notbuyinit.org>.

WEB SITES

"Kids against Tobacco Smoke." *Roy Castle Lung Cancer Foundation.* Available online at <www.roycastle.org/kats/about.htm> (accessed November 3, 2004).

"Stand Up. Speak Out against Tobacco." Available online at <www.standonline.org> (accessed November 3, 2004).

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Social competence

Definition

Social competence is the condition of possessing the social, emotional, and intellectual skills and behaviors needed to succeed as a member of society.

Description

Social competence refers to the social, emotional, and cognitive skills and behaviors that children need for successful social adaptation. Despite this simple definition, social competence is an elusive concept, because the skills and behaviors required for healthy social development vary with the age of the child and with the demands of particular situations. A socially competent **preschool** child behaves differently from a socially competent adolescent. Conversely, the same behaviors (e.g., aggression, **shyness**) have different implications for social adaptation depending on the age of the child and the particulars of the social context.

A child's social competence depends upon a number of factors including the child's social skills, social

awareness, and self-confidence. The term social skills describes the child's knowledge of and ability to use a variety of social behaviors that are appropriate to a given interpersonal situation and that are pleasing to others in each situation. The capacity to inhibit egocentric, impulsive, or negative social behavior is also a reflection of a child's social skills. The term emotional **intelligence** refers to the child's ability to understand the emotions of others, perceive subtle social cues, "read" complex social situations, and demonstrate insight about others' motivations and goals. Children who have a wide repertoire of social skills and who are socially aware and perceptive are likely to be socially competent.

Social competence is the broader term used to describe a child's social effectiveness. It defines a child's ability to establish and maintain high quality and mutually satisfying relationships and to avoid negative treatment or victimization from others. In addition to social skills and emotional intelligence, factors such as the child's self-confidence or social **anxiety** can affect his or her social competence. Social competence can also be affected by the social context and the extent to which there is a good match between the child's skills, interests, and abilities and those of peers. For example, a quiet and studious boy may appear socially incompetent in a peer group full of raucous athletes but may do fine socially if a more complementary peer group can be found for him, such as children who share his interests in quiet games or computers.

Importance of social competence

Parents are the primary source of social and emotional support for children during the first years of life, but in later years peers begin to play a significant role in a child's social-emotional development. Increasingly with age, peers rather than parents become preferred companions, providing important sources of entertainment and support. In the context of peer interactions, young children engage in fantasy play that allows them to assume different roles, learn to take another person's perspective, and develop an understanding of the social rules and conventions of their culture. In addition, relationships with peers typically involve more give-and-take than relationships with adults and thus provide an opportunity for the development of social competencies such as cooperation and negotiation.

During **adolescence**, peer relations become particularly important for children. A key developmental task of adolescence is the formation of an identity or sense of the kind of person one is and the kind of person one wants to be. Adolescents try on different social roles as they interact with peers, and peers serve as a social stepping stone as adolescents move away from their emotional dependence upon their parents and toward autonomous functioning as an adult. In many ways, then, childhood peer relations serve as training grounds for future interpersonal relations, providing children with opportunities to learn about reciprocity and intimacy. These skills are associated with effective interpersonal relations in adult life, including relations with co-workers and with romantic partners.

When children experience serious difficulties in peer relations, the development of social competencies may be threatened. Rejection or victimization by peers may become a source of significant stress to children, contributing to feelings of loneliness and low **self-esteem**. In addition, peer rejection can escalate in a negative developmental spiral. That is, when children with poor social skills become rejected, they are often excluded from positive interactions with peers that are critical for learning social skills. Rejected children typically have fewer options in terms of play partners and friends than do accepted children. Observations of rejected children have revealed that they spend more time playing alone and interacting in smaller groups than their more popular peers. In addition, the companions of rejected children tend to be younger or more unpopular than the companions of accepted children. Exclusion from a normal peer group can deprive rejected children of opportunities to develop adaptive social behaviors. Hence, the social competence deficits of rejected children may increase over time, along with feelings of social anxiety and inadequacy.

Social competence deficits and peer rejection

Many children experience difficulties getting along with peers at some point during their youth. Sometimes these problems are short-lived and for some children the effects of being left out or teased by classmates are transitory. For other children, however, being ignored or rejected by peers may be a lasting problem that has lifelong consequences, such as a dislike for school, poor self-esteem, social withdrawal, and difficulties with adult relationships.

Considerable research has been undertaken to try to understand why some children experience serious and long-lasting difficulties in the area of peer relations. To explore factors leading to peer difficulties, researchers typically employ the sociometric method to identify children who are or are not successful with peers. In this method, children in a classroom or a group are asked to list the children they like most and those whom they like least. Children who receive many positive ("like most") nominations and few negative ("like least") nominations

are classified as "popular." Those who receive few positive and few negative nominations are designated "neglected," and those who receive few positive and many negative nominations are classified as "rejected."

Evidence compiled from studies using child interviews, direct observations, and teacher ratings all suggest that popular children exhibit high levels of social competence. They are friendly and cooperative and engage readily in conversation. Peers describe them as helpful, nice, understanding, attractive, and good at games. Popular and socially competent children are able to consider the perspectives of others, can sustain their attention to the play task, and are able to remain self-controlled in situations involving conflict. They are agreeable and have good problem-solving skills. Socially competent children are also sensitive to the nuances of "play etiquette." They enter a group using diplomatic strategies, such as commenting upon the ongoing activity and asking permission to join in. They uphold standards of equity and show good sportsmanship, making them good companions and enjoyable play partners.

Children who have problems making friends, those who are either "neglected" or "rejected" by their peers, often show deficits in social skills. One of the most common reasons for friendship problems is behavior that annoys other children. Children, like adults, do not like behavior that is bossy, self-centered, or disruptive. It is simply not fun to play with someone who does not share or does not follow the rules. Sometimes children who have learning problems or attention problems can have trouble making friends, because they find it hard to understand and follow the rules of games. Children who get angry easily and lose their temper when things do not go their way can also have a hard time getting along with others. Children who are rejected by peers often have difficulties focusing their attention and controlling their behavior. They may show high rates of noncompliance, interference with others, or aggression (teasing or fighting). Peers often describe rejected classmates as disruptive, short-tempered, unattractive, and likely to brag, to start fights, and to get in trouble with the teacher.

Not all aggressive children are rejected by their peers. Children are particularly likely to become rejected if they show a wide range of conduct problems, including disruptive, hyperactive, and disagreeable behaviors in addition to physical aggression. Socially competent children who are aggressive tend to use aggression in a way that is accepted by peers (e.g., fighting back when provoked), whereas the aggressive acts of rejected children include **tantrums**, verbal insults, cheating, or tattling. In addition, aggressive children are more likely to be rejected if they are hyperactive, immature, and lacking in positive social skills.

Children can also have friendship problems because they are very shy and feel uncomfortable and unsure of themselves around others. Sometimes children are ignored or teased by classmates because there is something "different" about them that sets them apart from other children. When children are shy in the classroom and ignored by children, becoming classified as "neglected," it does not necessarily indicate deficits in social competence. Many neglected children have friendships outside the classroom setting, and their neglected status is simply a reflection of their quiet attitude and low profile in the classroom.

Developmentally, peer neglect is not a very stable classification, and many neglected children develop more confidence as they move into classrooms with more familiar or more compatible peers. However, some shy children are highly anxious socially and uncomfortable around peers in many situations. Shy, passive children who are actively disliked and rejected by classmates often become teased and victimized. These children often do have deficits in core areas of social competence that have a negative impact on their social development. For example, many are emotionally dependent on adults and immature in their social behavior. They may be inattentive, moody, depressed, or emotionally volatile, making it difficult for them to sustain positive play interactions with others.

The long-term consequences of sustained peer rejection can be quite serious. Often, deficits in social competence and peer rejection coincide with other emotional and behavioral problems, including attention deficits, aggression, and depression. The importance of social competence and satisfying social relations is life-long. Studies of adults have revealed that friendship is a critical source of social support that protects against the negative effects of life stress. People with few friends are at elevated risk for depression and anxiety.

Childhood peer rejection predicts a variety of difficulties in later life, including school problems, mental health disorders, and **antisocial behavior**. In fact, in one study, peer rejection proved to be a more sensitive predictor of later mental health problems than school records, achievement, intelligence quotient (IQ) scores, or teacher ratings.

It appears, then, that positive peer relations play an important role in supporting the process of healthy social and emotional development. Problematic peer relations are associated with both present and future maladjustment of children and warrant serious attention from par-

ents and professionals working with children. When assessing the possible factors contributing to a child's social difficulties and when planning remedial interventions, it is important to understand developmental processes associated with social competence and peer relations.

Developmental changes and social competence

The key markers of social competence listed in the previous section are consistent across the developmental periods of the preschool years, middle childhood, and adolescence. Across these developmental periods, prosocial skills (friendly, cooperative, helpful behaviors) and self-control skills (anger management, negotiation skills, problem-solving skills) are key facets of social competence. In addition, however, developmental changes occur in the structure and quality of peer interactions that affect the complexity of skills contributing to social competence. That is, as children grow, their preferences for play change, and the thinking skills and language skills that provide a foundation for social competence also change. Hence, the kinds of interactions that children have with peers change qualitatively and quantitatively with development.

Preschool

During the preschool years, social competence involves the ability to separate from parents and engage with peers in shared play activities, particularly fantasy play. As preschool children are just learning to coordinate their social behavior, their interactions are often short and marked by frequent squabbles, and friendships are less stable than at later developmental stages. In addition, physical rough-and-tumble play is common, particularly among boys. During the preschool and early grade school years, children are primarily focused on group acceptance and having companions with whom they can play.

School age

By grade school, children begin to develop an interest in **sports**, structured board games, and group games with complex sets of rules. Being able to understand and follow game rules and being able to handle competition in appropriate ways (e.g., being a good sport) become important skills for social competence. Children play primarily in same-sex groups of friends and expect more stability in their friendships. Loyalty and dependability become important qualities of good friends.

During the middle to late grade school years, children begin to distinguish "regular" friends from "best" friends. The establishment of close, best friendships is an important developmental milestone. That is, in addition to gaining acceptance from a group of peers, one of the hallmarks of social competence is the ability to form and maintain satisfying close friendships.

During the preadolescent and early adolescent years, communication (including sending notes, calling on the phone, and "hanging out") becomes a major focus for peer interactions. Increasingly, social competence involves the willingness and ability to share thoughts and feelings with one another, especially for girls. When adolescent friends squabble, their conflicts typically center on issues such as gossiping, disclosing secrets, or loyalty and perceived betrayal. It is at this stage that friends and romantic partners consistently rival parents as the primary sources of intimacy and social support.

Many of the positive characteristics that promote popularity (such as cooperativeness, friendliness, and consideration for others) also assist children in developing and maintaining friendships. Friendships emerge when children share similar activities and interests and, in addition, when they develop a positive and mutual bond between them. Group acceptance and close friendships follow different timetables and serve different developmental functions, with the need for group acceptance emerging during the early grade school years and filling a need for belonging and the need for close friends emerging in preadolescence to meet newfound needs for affection, alliance, and intimacy outside the **family**. Key features of close friendships are reciprocity and similarity, mutual intimacy, and social support.

Common problems

Many children who are rejected by peers have lower self-esteem, feel lonelier, and are more dissatisfied with their social situations than are average or popular children. These feelings can cause them to give up and avoid social situations, which can in turn exacerbate their peer problems. Interestingly, not all rejected children feel badly about their social difficulties. Studies have shown that aggressive-rejected children, who tend to blame outside factors for their peer problems, are less likely to express distress than withdrawn-rejected children, who often attribute their problems to themselves.

Assessing social competence

There is an important difference between not being "popular" and having friendship problems. Some children are outgoing and have many friends. Other children are quite content with just a good friend or two. Either one of these friendship patterns is healthy. Distinguishing normal friendship problems from problem peer

relations that signal serious deficits in social competence is an important goal of **assessment**. There are several key signs that a child's peer difficulties may be more serious and long-lasting rather than temporary. First, the nature of the child's social behavior is important. If children behave aggressively with peers, act bossy and domineering, or are disruptive and impulsive at school, they are more likely to have long-lasting peer difficulties than are children who are simply shy. Children who display aggressive or disruptive behavior often have many discouraging experiences at school, including **discipline** problems and learning difficulties, as well as poor peer relations. School adjustment can be a downhill slide for these children as teachers may get discouraged and peers may be angered by their behaviors. Peers may attempt to "get back" at these children by teasing, which only increases the frustrations and helplessness experienced by aggressive, disruptive children.

Second, children who are actively disliked, teased, or ostracized by peers are at more risk than children who are simply ignored. It is not necessary for a child to be popular in order for that child to gain the advantages of peer support. When children are ignored by peers and are neither disliked nor liked, teachers and parents can take steps to foster friendship development and peer support. When children are actively disliked by peers and the victims of teasing or ostracism, the task is harder for parents and teachers and the likelihood of the child reestablishing positive peer relations without help decreases.

Third, the stability and timing of peer problems should be considered. It is not unusual for children to experience short-term social difficulties when they are moving into new peer situations, such as a new school or a new classroom. Peer problems may also emerge if children are distressed about other changes in their lives, such as a reaction to parental conflict or the birth of a sibling. When peer problems emerge at a time that corresponds to other family or situational changes, they may serve as signals to let parents and teachers know that the child needs extra support at that time. When peer problems have been stable and have existed for a long time, more extensive intervention focused on improving peer relations may be needed.

A variety of methods are available for the assessment of social competence. When choosing a particular assessment strategy, it is important to consider the nature of a particular child's problem. Some children have difficulty with all types of social relationships, while others do well in their neighborhoods or in one-on-one friendships but experience problems with the peer group at school. When problems occur in the school setting, teachers and other school personnel who have opportunities to see children interacting in several peer group situations (such as the classroom, playground, and lunchroom) are often the best first step in assessment. Teachers can often provide information about how children treat and are treated by peers and can also offer opinions about how typical or unusual a child's peer problems are relative to others of the same age. Teacher assessments can include behavioral checklists and rating scales and direct observations of specific social behaviors.

Similarly, parents can provide information about children's social competence. Parents can help to identify problem behaviors such as aggression, withdrawal, and noncompliance that may interfere with social skills. In addition, parents are usually more aware than teachers of their children's social activities outside of school, such as their participation in sports, clubs, or hobbies.

Because they do not have access to the full range of situations in which children interact, however, teachers and parents may not always be the best source of information on children's peer problems. In some cases, it is most helpful to get information directly from peers themselves. One method of obtaining such information is the use of sociometric ratings and nominations. With these procedures, all of the children in a classroom are asked to rate how much they like to play with or spend time with each of their classmates. In addition, they nominate specific peers whom they particularly like or dislike, and they may be asked to identify peers who exhibit particular behavioral characteristics (e.g., nice, aggressive, shy, etc.). The sociometric method, although cumbersome to administer, identifies children who are popular, rejected, and neglected by their peers more accurately than parent or teacher reports and provides useful information about the reasons for peer dislike.

A third approach to assessment of social competence involves children's self-reports. Although input from parents, teachers, and peers can provide valuable insight into children's social behavior and their status within the peer group, information regarding children's thoughts, feelings, and perceptions of their social situations can be obtained only by asking the children themselves. Depending upon the age of the child, information about social competence can be obtained through the use of questionnaires and rating scales that measure children's self-perceptions of their peer relations, the use of stories and hypothetical social situations to elicit information about the child's social reasoning, or simply talking with children to determine their perspectives on their social situations.

Because children may have different experiences in different kinds of peer settings and because no one particular method of assessment is entirely reliable or complete, it is desirable to use a variety of sources when

attempting to assess children's social competence. Teacher, parent, peer, and self-reports may yield distinct but complementary information, so by gathering multiple perspectives a more complete picture of a child's social strengths and weaknesses can be obtained.

Interventions to promote social competence

Different strategies may be needed to help children develop social competencies and establish positive peer relations depending on the age of the child and the type of peer problem being experienced. Different children have different needs when it comes to helping them get along better with others and making friends. The age of the child, the kinds of behaviors that are part of the problem, and the reasons for the friendship problem may all affect the helping strategy.

One strategy involves social skill training. Observations have revealed that children who are well liked by peers typically show helpful, courteous, and considerate behavior. The purpose of social skill training is to help unpopular children learn to treat their peers in positive ways. The specific skills taught in different programs vary depending upon the age and type of child involved. Commonly taught skills include helping, sharing, and cooperation. Often children are taught how to enter a group, how to be a good group participant, how to be a fair player (e.g., following rules, taking turns), and how to have a conversation with peers. The skills might also include anger management, negotiation, and conflict resolution skills. Problem-solving skills (e.g., identifying the problem, considering alternative solutions, choosing a solution, and making a plan) are often included in social skill training programs. Sometimes social skill training is done individually with children, but often it is done in a small group. A particular skill concept is discussed, and children may watch a short film or hear a story that illustrates the usefulness of the skill. They then have the opportunity to practice the skill during activities or role-plays with other children in the group. A trained group leader helps guide the children in their use of the skill and provides support and positive feedback to help children become more natural and spontaneous in socially skillful behavior.

Another intervention strategy focuses on helping children who are having trouble getting along with others because of angry, aggressive, or bossy behavior. It can be difficult to suppress aggressive and disruptive behaviors in peer settings for several reasons. For one thing, these behaviors often "work" in the sense that they can be instrumental in achieving desired goals. By complaining loudly, hitting, or otherwise using force or noise, children may be able to get access to a toy they want, or they may be able to get peers to stop doing something obnoxious to them. In this type of situation, an adult's expressed disapproval may suppress the behavior, but the behavior is likely to emerge again in situations where an adult supervisor is not present. Often contracts and point systems are used to suppress **aggressive behavior** and bossiness; however, positive skill training must be used in conjunction with behavior management in order to provide the child with alternative skills to use in situations requiring negotiations with peers. Often parents are included in programs to help children develop better anger management skills and to help children reduce fighting. Trained counselors, educators, or psychologists work with parents to help them find positive discipline strategies and positive **communication skills** to promote child anger management and conflict resolution skills.

A third helping strategy focuses on finding a good social "niche" for the child. Large, unstructured peer group settings (such as recess) are particularly difficult situations for many of the children who have peer problems. These children need a structured, smaller peer interaction setting in which an adult's support is available to guide positive peer interaction. Finding a good social niche for some children can be a difficult task, but an important one. Sometimes a teacher can organize cooperative learning groups that help an isolated child make friends in the classroom. Sometimes parents can help by inviting potential friends over to play or by getting their child involved in a social activity outside of school that is rewarding (such as a church group, a sports group, or a scouting club). Providing positive opportunities for friendship development is important, as it provides children with an appropriate and positive learning environment for the development of social competence.

Parental concerns

Because the family is the primary setting for social development, there are a number of ways in which family interaction patterns may help or hinder the development of children's social competence. Some researchers have speculated that the origins of social competence can be found in infancy, in the quality of the parent-child attachment relationship. Studies have shown that babies whose parents are consistent and sensitive in their responses to distress are less irritable, less anxious, and better emotionally regulated. By contrast, parents who are inconsistent and insensitive to their infants' signals are more likely to have anxious, irritable babies who are difficult to soothe. These children may learn both to model their parents' insensitivity and to rely on intrusive, demanding behavior of their own in order to get attention. If they then generalize these socially incompetent behaviors to their peer interactions, peer rejection may result.

As children get older, family interaction styles and the ways in which parents discipline may play a primary role in the development of noncompliant or aggressive behaviors in children. In families where parents are extremely demanding and use inconsistent, harsh, and punitive discipline strategies, family interaction patterns are frequently characterized by escalation and conflict, and children often exhibit behavior problems. When children generalize the aggressive and oppositional behavior that they have learned at home to their interactions with peers, other children often reject them. Indeed, research has revealed that aggressive behavior is the common link between harsh, inconsistent discipline and rejection by peers.

By contrast, parents of popular children are typically more positive and less demanding with their children than parents of unpopular children. In addition, parents of popular children set a good example by modeling appropriate social interactions and assist their children by arranging opportunities for peer interaction, carefully supervising these experiences, and providing helpful feedback about conflict resolution and making friends.

Child characteristics and social competence

In addition to family interaction patterns and various aspects of the parent-child relationship, children's own thoughts, feelings, and attitudes may influence their social behavior. Research has revealed that many rejected children make impulsive, inaccurate, and incomplete judgments about how to behave in social situations and are lacking in social problem-solving skills. They may make numerous errors in processing social information, including misinterpretation of other people's motives and behavior, setting social goals for themselves that are unrealistic or inappropriate and making poor decisions about their own conduct in social situations. For example, aggressive children are more likely to interpret an accidental push or bump from a peer as intentionally hostile and respond accordingly. Similarly, socially incompetent children are often more interested in "getting even" with peers for injustices than they are in finding positive solutions to social problems and expect that aggressive, coercive strategies will lead to desired outcomes.

When to call the doctor

If the child has significant problems with social competence, especially those which may be caused by an underlying disorder such as anxiety, a doctor or mental health professional should be consulted.

See also Peer acceptance.

Emotional and social development

Age	Activity
Two months	Smiles at person's face. Shows happiness and distress. May be soothed by rocking.
Three months	Smiles when spoken to. Coos or squeals with pleasure.
Four months	Enjoys being cuddled. Recognizes parents and distinguishes them from strangers. Recognizes patterns of feeding, bathing, and dressing. Laughs aloud.
Six months	Smiles and "talks" to mirror image. Sticks out tongue in imitation. May start to show fear of strangers and protest separation from mother or other primary care giver. Enjoys playing peek-a-boo.
Seven months	Responds to name. Tries to engage a person by coughing or making other noise.
Eight months	Responds to "no."
Ten months	May pull on clothing of caregiver to attract attention. Waves bye-bye and plays pat-a-cake. Helps with dressing by holding out arm or leg.
Twelve months	Repeats an action that evoked laughter from adults. May kiss on request, or kiss mirror image. Tends to be shy. Gives and takes objects.
Fifteen months	Asks for object by pointing. Shows affection for familiar people and objects. Shows dependency on primary caregiver. Negativism begins.
Eighteen months	Does the opposite of what is requested. May have temper tantrums.
Two years	Tends to be jealous of own toys and attention of parents. Engages in parallel play with other children. Negativism increases.
Two and a half years	Negativism peaks. Shows fear of separation. Can hit or thrash about when angry. Able to play tricks and pretend.
Three years	Has a more easy-going nature and greater sense of identity. Shows jealousy of same-sex parent and attachment for opposite-sex one. Begins to have imaginary fears of the dark or getting hurt. Engages in cooperative play.
Four years	More sure of self. Often negative and can be defiant. Tests limits. Enjoys cooperative play and group games.
Five years	More stable and secure. Likes to follow rules and enjoys some responsibility. Enjoys organized play and table games requiring taking turns and following rules.

SOURCE: *Miller-Keane Encyclopedia and Dictionary of Medicine, Nursing, and Allied Health, 5th ed.* and Child Development Institute, http://www.childdevelopmentinfo.com.

(Table by GGS Information Services.)

Resources

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KEY TERMS

Cognitive skills—Skills required to perform higher cognitive processes, such as knowing, learning, thinking, and judging.

Emotional intelligence—The ability to perceive and interpret the emotions of others.

Social skills—The knowledge of and ability to use a variety of social behaviors that are appropriate to interact positively with other people.

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Janet Welsh, Ph.D.
Karen Bierman, Ph.D.

Soft tissue cancers *see* **Sarcomas**

Somnambulism

Definition

Somnambulism is also known as sleepwalking. It is a common disorder among children that involves getting out of bed and moving about while still asleep.

Description

Somnambulism is similar to *pavor nocturnus* (**night terrors**) in that it occurs during the non-dreaming stage of **sleep**, usually within an hour or two of going to bed. The sleepwalking child feels an intense need to take action and may appear alert, purposeful, or anxious as he or she moves about. For many years, people believed that it was dangerous to awaken a sleepwalker, but there is no basis for this view. There is, however, little reason to awaken a sleepwalking child, and it may be impossible to do so. Episodes of sleepwalking may be signs of a child's heightened **anxiety** about something.

Demographics

Somnambulism, or sleepwalking, affects an estimated 15 percent of children in their early school years. It decreases in frequency with increasing age. It is very uncommon among adults.

Causes and symptoms

The root cause of sleepwalking is not known. Anxiety and stress are the most commonly given reasons for sleepwalking.

If sleepwalking is common among **family** members, it is more likely that the child may respond to even slight increases in anxiety with sleepwalking behavior.

When to call the doctor

A doctor or other health care provider should be called when episodes of sleepwalking cannot be comfortably managed in the home.

Diagnosis

A diagnosis of somnambulism is made by observation and history. There are no laboratory tests. An **electroencephalogram** may be used as a part of an analysis in a sleep laboratory, but this is the exception rather than the rule.

Treatment

Sleepwalking children should be gently guided back to bed. They will usually be cooperative in this effort.

Prognosis

The prognosis for sleepwalking is good. Most children experience a few episodes of somnambulism and then simply stop, often when a source of stress or anxiety

is removed. Sleepwalking rarely affects persons outside of one's own family circle.

Prevention

There is no known way to prevent episodes of sleepwalking.

Nutritional concerns

There is no known link between sleepwalking and **nutrition**.

Parental concerns

Parents should give careful consideration to events and environmental changes that may have triggered the onset of sleepwalking. Potential hazards that may injure children should be removed from their sleeping areas.

Resources

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Carney, Paul R. et al. *Clinical Sleep Disorders*. Philadelphia: Lippincott Williams & Wilkins, 2004.

Hertz, Grett J. J. et al. *Olie's Bedtime Walk*. Long Island City, NY: Star Bright Books, 2002.

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Lecendreux, M., et al. "HLA and genetic susceptibility to sleepwalking." *Molecular Psychiatry* 8, no. 1 (2003): 114–17.

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KEY TERMS

Electroencephalogram (EEG)—A record of the tiny electrical impulses produced by the brain's activity picked up by electrodes placed on the scalp. By measuring characteristic wave patterns, the EEG can help diagnose certain conditions of the brain.

ORGANIZATIONS

American Academy of Family Physicians. 11400 Tomahawk Creek Parkway, Leawood, KS 66211-2672. (913) 906-6000. fp@aafp.org. <www.aafp.org>

American Academy of Pediatrics. 141 Northwest Point Boulevard, Elk Grove Village, IL 60007-1098. (847) 434-4000, Fax: (847) 434-8000. kidsdoc@aap.org. <www.aap.org/default.htm>

American Academy of Sleep Medicine. 6301 Bandel Road NW, Suite 101, Rochester, MN 55901. (507) 287-6006. Fax: (507) 287-6008. info@aasmnet.org. <www.asda.org>

American College of Physicians, 190 N. Independence Mall West, Philadelphia, PA 19106-1572. (800) 523-1546, x2600 or (215) 351-2600. <www.acponline.org>

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"Sleep Disorders and Sleep Problems in Childhood." *American Academy of Family Physicians*. Available online at <www.aafp.org/afp/20010115/277.html>.

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Sore throat

Definition

Sore throat is a painful inflammation of the mucous membranes lining the pharynx.

Description

Sore throat is also called pharyngitis. It is a symptom of many conditions, but is most often associated with colds or **influenza**. Sore throat may be caused by either viral or bacterial infections or environmental conditions. Most sore throats heal without complications, but they should not be ignored, as some develop into serious illnesses.

Sore throats can be either acute or chronic. Acute sore throats are more common than chronic sore throats. They appear suddenly and last from three to about seven days. A chronic sore throat lasts much longer and is a symptom of an unresolved underlying condition or disease, such as a sinus infection.

Transmission

The way in which a sore throat is transmitted depends on the agent causing the sore throat. Viral and bacterial sore throats are usually passed in the same way as the **common cold**: sneezing, coughing, sharing drinking glasses or silverware, or in any other way germ particles can easily move from one person to another. Some sore throats are caused by environmental factors or **allergies**. These sore throats cannot be passed from one person to another.

Demographics

Almost everyone gets a sore throat at one time or another, although children in child care or grade school have them more often than adolescents and adults. Sore throats are most common during the winter months when upper respiratory infections (colds) are more frequent.

About 10 percent of children who go to the doctor each year have pharyngitis. Forty percent of the time that children are taken to the doctor with a sore throat, the sore throat is diagnosed as viral. An antibiotic cannot help to cure a virus; a virus has to be left to run its course.

In about 30 percent of the cases for which children are taken to the doctor, bacteria are found to be responsible for the sore throat. Many of these bacterial sore throats are cases of **strep throat**. Sore throats caused by bacteria can be successfully treated with **antibiotics**. In about 40 percent of these cases of pharyngitis, it is never clear what caused the sore throat. In these cases it is possible that the virus or bacteria was not identified, or that other factors such as environment or post-nasal drip may have been responsible.

Causes and symptoms

Sore throats have many different causes, and may or may not be accompanied by cold symptoms, **fever**, or swollen lymph glands. Proper treatment depends on understanding the cause of the sore throat.

Viral sore throat

Viruses cause most sore throats. Cold and flu viruses are the main culprits. These viruses cause an inflammation in the throat and occasionally the tonsils (**tonsillitis**). Cold symptoms usually accompany a viral sore throat. These can include a runny nose, **cough**, congestion, hoarseness, **conjunctivitis**, and fever. The level of throat **pain** varies from uncomfortable to excruciating, when it is painful for the patient to eat, breathe, swallow, or speak.

Another group of viruses that causes sore throat are the adenoviruses. These may also cause infections of the lungs and ears. In addition to a sore throat, symptoms that accompany an adenovirus infection include cough, runny nose, white bumps on the tonsils and throat, mild **diarrhea**, **vomiting**, and a rash. The sore throat lasts about one week.

A third type of virus that can cause severe sore throat is the coxsackie virus. It can cause a disease called herpangina. Although anyone can get herpangina, it is most common in children up to age 10 and is more prevalent in the summer or early autumn. Herpangina is sometimes called summer sore throat.

Three to six days after being exposed to the coxsackie virus, an infected person develops a sudden sore throat that is accompanied by a substantial fever, usually between 102–104°F (38.9–40°C). Tiny grayish-white blisters form on the throat and in the mouth. These fester and become small ulcers. Throat pain is often severe, interfering with swallowing. Children may become dehydrated if they are reluctant to eat or drink because of the pain. In addition, children with herpangina may vomit, have abdominal pain, and generally feel very ill.

One other common cause of a viral sore throat is mononucleosis. Mononucleosis occurs when the Epstein-Barr virus infects one specific type of lymphocyte. The infection spreads to the lymphatic system, respiratory system, liver, spleen, and throat. Symptoms appear 30–50 days after exposure.

Mononucleosis, sometimes called the kissing disease, is extremely common. It is estimated that by the age of 35–40, 80–95 percent of Americans will have had mononucleosis. Often, symptoms are mild, especially in young children, and are diagnosed as a cold. Since symp-

toms are more severe in adolescents and adults, more cases are diagnosed as mononucleosis in this age group. One of the main symptoms of mononucleosis is a severe sore throat.

Although a runny nose and cough are much more likely to accompany a sore throat caused by a virus than one caused by a bacteria, there is no absolute way to tell what is causing the sore throat without a laboratory test.

Bacterial sore throat

Fewer sore throats are caused by bacteria than are caused by viruses. The most common bacterial sore throat results from an infection by group A *Streptococcus*. This type of infection is commonly called strep throat. Anyone can get strep throat, but it is most common in school age children.

Noninfectious sore throat

Not all sore throats are caused by infection. Postnasal drip can irritate the throat and make it sore. It can be caused by hay fever and other allergies that irritate the sinuses. Environmental and other conditions, such as breathing secondhand smoke, breathing polluted air or chemical fumes, or swallowing substances that burn or scratch the throat can also cause pharyngitis. Dry air, like that in airplanes or from forced hot air furnaces, can make the throat sore. Children who breathe through their mouths at night because of nasal congestion often get sore throats that improve as the day progresses. Sore throat caused by environmental conditions is not contagious.

When to call the doctor

If the child has had a sore throat and fever for more than 24 hours, a doctor should be contacted so a strep test can be performed. Identifying and treating strep throat within about a week is vital to preventing **rheumatic fever**. If the child has had a sore throat, even without fever, for more than 48 hours, the doctor should be consulted. If the child has trouble swallowing or breathing, or is drooling excessively (in small children), emergency medical attention should be sought immediately.

Diagnosis

It is easy for people to tell if they have a sore throat, but difficult to know what has caused it without laboratory tests. Most sore throats are minor and heal without any complications. A small number of bacterial sore throats do develop into serious diseases. Because of this, it is advisable to see a doctor if a sore throat lasts more than a few days or is accompanied by fever, **nausea**, or abdominal pain.

Diagnosis of a sore throat by a doctor begins with a physical examination of the throat and chest. The doctor will also look for signs of other illness, such as a sinus infection or **bronchitis**. Since both bacterial and viral sore throat are contagious and pass easily from person to person, the doctor will seek information about whether the patient has been around other people with flu, sore throat, colds, or strep throat. If it appears that the patient may have strep throat, the doctor will do laboratory tests.

If mononucleosis is suspected, the doctor may do a mono spot test to look for antibodies indicating the presence of the Epstein-Barr virus. The strep test is inexpensive, takes only a few minutes, and can be done in a physician's office. An inexpensive blood test can also determine the presence of antibodies to the mononucleosis virus.

Treatment

Effective treatment varies depending on the cause of the sore throat. Viral sore throats are best left to run their course without drug treatment, because antibiotics have no effect on a viral sore throat. They do not shorten the length of the illness, nor do they lessen the symptoms.

Sore throat caused by streptococci or another bacteria must be treated with antibiotics. Penicillin is the preferred medication, although other antibiotics are also effective if the child is allergic to penicillin. Oral penicillin must be taken for 10 days. Patients need to take the entire amount of antibiotic prescribed, even after symptoms of the sore throat improve. If it is unlikely that the parent will be able to ensure that the child will take the full course of antibiotics, a one-time injection of antibiotics can be administered instead. Cessation of the antibiotic early can lead to a return of the sore throat.

Because a virus causes mononucleosis, there is no specific drug treatment available. Rest, a healthy diet, plenty of fluids, limiting heavy **exercise** and competitive **sports**, and treatment of aches with **acetaminophen** (Datril, Tylenol, Panadol) or ibuprofen (Advil, Nuprin, Motrin, Medipren) will help the illness pass. Nearly 90 percent of mononucleosis infections are mild. The infected person does not normally get the disease again.

In the case of chronic sore throat, it is necessary to treat the underlying disease to heal the sore throat. If a sore throat is caused by environmental factors, the aggravating stimulus should be eliminated from the sufferer's environment.

Home care for sore throat

Regardless of the cause of a sore throat, there are some home care steps that people can take to ease their discomfort. These include:

- taking acetaminophen or ibuprofen for pain (aspirin should not be given to children because of its association with increased risk for **Reye's syndrome**, a serious disease)
- gargling with warm double strength tea or warm salt water made by adding 1 tsp of salt to 8 oz (237 ml) of water
- drinking plenty of fluids, but avoiding acid juices such as orange juice, which can irritate the throat (sucking on popsicles is a good way to get fluids into children)
- eating soft, nutritious foods like noodle soup and avoiding spicy foods
- resting until the fever is gone, then resuming strenuous activities gradually
- using a room humidifier to make sore throat sufferers more comfortable
- using antiseptic lozenges and sprays with caution, as they may aggravate the sore throat rather than improve it

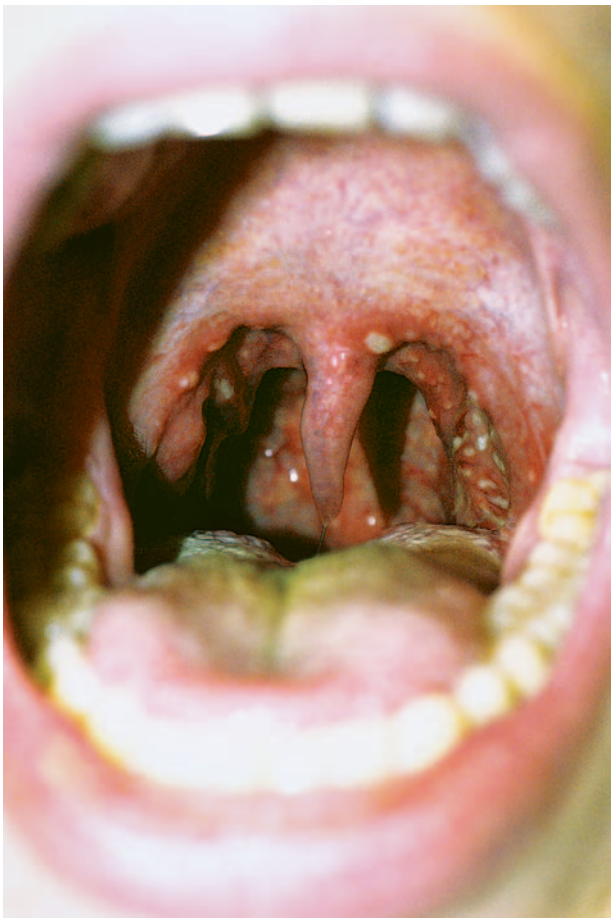
Alternative treatment

Alternative treatment focuses on easing the symptoms of sore throat using herbs and botanical medicines.

- Aromatherapists recommend inhaling the fragrances of the essential oils of lavender (*Lavandula officinalis*), thyme (*Thymus vulgaris*), eucalyptus (*Eucalyptus globulus*), sage (*Salvia officinalis*), and sandalwood.
- Ayurvedic practitioners suggest gargling with a mixture of water, salt, and tumeric (*Curcuma longa*) powder or astringents such as alum, sumac, sage, and bayberry (*Myrica* spp.).
- Herbalists recommend taking osha root (*Ligusticum porteri*) internally for infection or drinking ginger (*Zingiber officinale*) or slippery elm (*Ulmus fulva*) tea for pain.
- Homeopaths may treat sore throats with superdilute solutions of *Lachesis*, *Belladonna*, *Phytolacca*, or yellow jasmine (*Gelsemium*).

Nutritional concerns

Nutritional recommendations include zinc lozenges every two hours along with vitamin C with bioflavonoids, vitamin A, and beta-carotene supplements. Although it may hurt to swallow, it is very important that the child does not become dehydrated. Sucking on popsicles or drinking warm broth can help. If the child shows any signs of **dehydration** he or she should be taken to the doctor.



Sore throat caused by a viral infection. *(© Scott Camazine/ Photo Researchers, Inc.)*

Prognosis

Sore throat caused by a viral infection generally clears up on its own within one week with no complications. The exception is mononucleosis. Ninety percent of cases of mononucleosis clear up without medical intervention or complications, so long as dehydration does not occur. In young children, the symptoms may last only a week, but in adolescents the symptoms usually last longer. In all age groups, fatigue and weakness may continue for up to six weeks after other symptoms disappear.

In rare cases of mononucleosis, breathing may be obstructed because of swollen tonsils, adenoids, and

KEY TERMS

Antigen—A substance (usually a protein) identified as foreign by the body's immune system, triggering the release of antibodies as part of the body's immune response.

Lymphocyte—A type of white blood cell that participates in the immune response. The two main groups are the B cells that have antibody molecules on their surface and T cells that destroy antigens.

Pharynx—The throat, a tubular structure that lies between the mouth and the esophagus.

lymph glands. If this happens, the individual should seek emergency medical care immediately.

Patients with bacterial sore throat begin feeling better about 24 hours after starting antibiotics. Untreated strep throat has the potential to cause **scarlet fever**, kidney damage, or rheumatic fever. Scarlet fever causes a rash and can cause high fever and convulsions. Rheumatic fever causes inflammation of the heart and damage to the heart valves. Taking antibiotics within the first week of a strep infection will prevent these complications. People with strep throat remain contagious until they have taken antibiotics for 24 hours.

Prevention

There is no way to prevent a sore throat; however, the risk of getting one or passing one on to another person can be minimized by:

- washing hands well and frequently
- avoiding close contact with someone who has a sore throat
- not sharing food and eating utensils with anyone
- staying out of polluted air

Parental concerns

Viral sore throats usually resolve themselves fairly quickly although they may be very uncomfortable. If the child has a fever and sore throat for more than 24 hours it may be a sign of a bacterial infection and the child should be taken to the doctor. Prompt treatment with antibiotics for strep throat is important because it can prevent rheumatic fever, a serious disease that can cause damage to the heart.

See also Common cold; Mononucleosis.

Resources

PERIODICALS

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Tish Davidson, A.M.

Spastic colon *see* **Irritable bowel syndrome**

Spasticity

Definition

Spasticity is an abnormal increase in muscle tone. It may be associated with involuntary **muscle spasms**, sustained muscle contractions (dystonia), and exaggerated deep tendon reflexes that make movement difficult or uncontrollable. Although it most commonly affects the legs and arms, spasticity can affect any part of the body including the trunk, neck, eyelids, face, or vocal cords.

Description

Spastic muscles are resistant to the normal stretching that occurs during use and may remain contracted for long periods. Spasticity may not be present all the time and varies based on initial muscle tone; length of responding muscle groups; and the person's position, posture, and state of relaxation. Spasticity may increase with **anxiety**, emotions, **pain**, or sensory stimulation. It may worsen with movement of the involved muscles. Spasticity may be aggravated by temperature extremes, humidity, skin problems such as a pressure ulcer or ingrown toenail, bladder or bowel problems, infections, and sometimes tight clothing.

The severity of spasticity ranges from slight muscle stiffness to spasms that come and go, to permanent contracture. Spasticity-induced contracture can be treated with medications. If muscle contractures are not treated,

fixed contracture can occur, leaving the muscle permanently shortened.

Severe spasticity can interfere with a child's normal functioning, motor and speech development, and/or comfort. Spasticity can be painful, especially if joints are pulled into abnormal positions or if range of motion is limited.

Simple activities of daily living (such as walking, eating, dressing, and bathing) may become time-consuming and difficult for both the child and caregiver. When spasticity limits activity for long periods, it can cause additional medical problems such as **sleep** disturbances, pressure sores, and **pneumonia**.

Demographics

The exact incidence of spasticity is not known. Estimates report spasticity may affect over 500,000 Americans and over 12 million people world-wide.

Causes and symptoms

Causes

Spasticity occurs when certain nerve signals do not reach the muscles because of injury or disease that affects parts of the brain or spinal cord. With spasticity, muscles receive improper nerve signals, causing them to contract, while the brain is unable to communicate with the motor nerves to stop the muscles from contracting.

Common neurological conditions associated with spasticity include **cerebral palsy**, brain injury or trauma, severe **head injury**, **stroke**, multiple sclerosis, **spinal cord injury**, and some metabolic diseases.

Symptoms

Spasticity is characterized by increased muscle tone (hypertonicity) and by muscle stiffness.

Symptoms associated with spasticity include the following:

- increased muscle stretch reflexes
- involuntary contraction and relaxation of muscles (spasms)
- prolonged muscle contractions (dystonia)
- rapid, repetitive jerky motions (clonus)
- exaggerated deep tendon jerks or reflexes
- involuntary crossing of the legs (also called scissoring reflex)
- abnormal posture or abnormal positioning of the shoulder, arm, wrist, or finger
- increased resistance to movement of certain muscle groups

When to call the doctor

If a child has any of the following symptoms, the parent or caregiver should call the child's doctor:

- worsening spasticity
- apparent development of muscle contractures
- worsening overall health

Diagnosis

A diagnosis of spasticity is often made with the diagnosis of cerebral palsy or following a brain or spinal cord injury. A multi-disciplinary team may be consulted to provide an accurate diagnosis of spasticity so the proper treatment can be planned.

The diagnosis of spasticity includes:

- review of personal and **family** medical history
- review of current medications
- review of other health problems
- physical examination
- diagnostic tests

The medical history helps the physician evaluate the presence of other conditions or disorders that might contribute to or cause the spasticity. Records of previous diagnoses, surgeries, and treatments are reviewed. The child's family medical history is evaluated to determine if there is a history of muscular or neurological disorders.

Questions about the child's medical history may include:

- When were the symptoms first noticed?
- How long have the symptoms lasted?
- Are the symptoms always present?
- What muscles are affected?
- What makes the symptoms improve?
- What specific treatments or techniques have been tried?
- What makes the symptoms worse?
- Do certain activities, emotions, or events seem to aggravate the symptoms?
- Are other symptoms present?

- Is the spasticity preventing function or independence?

The doctor will review the presence of other health problems such as swallowing function, bowel and bladder function, and learning difficulties.

The physical exam may include an evaluation of the child's motor reflexes including muscle tone, mobility, strength, balance and endurance; heart and lung function; cranial nerve function; and an examination of the child's abdomen, spine, throat, and ears. The child's height and weight and blood pressure also are checked and recorded.

To confirm the diagnosis of spasticity, the following tests can be performed to evaluate the child's arm and leg movements, muscular activity, range of motion, and ability to perform self-care activities:

- x rays of the spine and hips
- occupational and physical therapy evaluations to determine upper and lower extremity movement patterns and passive range of motion
- diagnostic blocks with local anesthetics to provide information on the effectiveness of potential treatments
- nerve conduction studies to evaluate muscle or nerve damage
- electromyogram (EMG or myogram) to detect abnormal muscle electrical activity.

Nerve conduction studies and an electromyogram (EMG) are usually performed together to provide a comprehensive assessment of nerve and muscle function. In both tests, the examiner uses a computer, monitor, amplifier, loudspeaker, stimulator and high-tech filters to see and hear how the muscles and nerves are responding during the test.

In the nerve conduction study, small electrodes are placed on the skin over the muscles to be examined. A stimulator delivers a very small electrical current (that does not cause damage to the body) through the electrodes, causing the nerves to fire. In the electromyogram, a very thin, sterilized needle is inserted into various muscles, usually those affected most by spasticity symptoms. The needle is attached by wires to a recording machine. The patient is asked to relax and contract the muscles being examined. The electrical signals produced by the nerves and muscles during these tests are measured and recorded by a computer and displayed as electrical waves on the monitor. The test results are interpreted by a specially trained physician.

Treatment

There is no cure for spasticity, but it can be managed with the appropriate treatment. Treatment options include physical and occupational therapy, medications, surgery, or a combination of these treatments. The goals of treatment are to increase the child's comfort, decrease pain, ease mobility, help with activities of daily living including hygiene, ease rehabilitation procedures, and prevent or decrease the risk of developing a joint contracture. The type of treatment recommended will depend upon the severity of the spasticity; the patient's overall health; the potential benefits, limitations, and side effects of the treatment; and the impact of the treatment on the child's quality of life.

In some cases, treatment is not recommended or desired, because it would actually interfere with the patient's current mobility and not improve function. For example, some people with multiple sclerosis who experience significant leg weakness find that spasticity makes their legs more rigid, helping them to stand, transfer to a chair or bed, or walk.

Clinicians should work with the child and the parents or caregivers to develop an individual treatment plan. Specific treatment goals will vary from one person to the next. Treatment should be provided by a pediatric neurologist and a multi-disciplinary team of specialists that may include a physiatrist, physical therapist, occupational therapist, gait and movement specialists, social worker, and surgical specialists as applicable, such as a pediatric orthopedic surgeon or pediatric neurosurgeon.

Physical and occupational therapy

Physical therapy includes stretching exercises, muscle group strengthening exercises, and range of motion exercises to prevent muscles from shortening (contracture), preserve flexibility and range of motion, and reduce the severity of symptoms. Exercises should be practiced daily, as recommended by the physical therapist. Prolonged stretching can lengthen muscles to help decrease spasticity. Strengthening exercises can restore the proper strength to muscles affected by spasticity. Aquatic therapy also may be recommended, since in water there is less stress on the body.

A physical therapist can instruct the patient on proper posture guidelines. Proper posture is critical, especially while sitting and sleeping, to maintain proper alignment of the hips and back. Balancing rest and **exercise** is also important.

Occupational therapy may include splints, casts, or braces on the affected arm or leg to enable proper limb positioning and maintain flexibility and range of motion.

It may also include training for proper limb positioning while seated in a wheelchair or lying in bed.

Physical and occupational therapists can provide guidelines on how to adapt the child's environment to ensure **safety** and comfort.

Other treatments

Brief application (about 10 minutes) of cold packs to spastic muscles may help ease pain and improve function for a short period of time.

Electrical stimulation may be used to reduce spasticity for a short period of time or to stimulate a weak muscle to counteract the action of a stronger, spastic muscle.

Biofeedback training may be used to teach the patient how to consciously reduce muscle tension. Biofeedback uses an electrical signal that indicates when a spastic muscle relaxes. The patient may be able to use biofeedback to learn how to consciously reduce muscle tension and possibly reduce spasticity. However, little research had been conducted as of 2004 to determine the effectiveness of biofeedback on reducing spasticity.

Medications

Medications to treat spasticity are taken by mouth, injected, or received through continuous delivery systems. These medications work by preventing nerves from signaling the muscles to contract, thereby preventing muscle contractions.

If treatment with a single medicine fails to effectively treat spasticity, a different medicine may be tried or an additional medicine may be prescribed. The most important medication guidelines include making sure the child takes the medicine exactly as prescribed and not discontinuing medication without first talking to the child's doctor, even if the medication does not seem to be working or is causing unwanted side effects.

ORAL MEDICATIONS The most commonly prescribed oral medication is baclofen (Lioresal). Baclofen is a muscle relaxant that works on nerves in the spinal cord to reduce spasticity. The benefits of baclofen include decreased stretch reflexes, improved passive range of motion, and reduced muscle spasms, pain, and tightness. Side effects include drowsiness and sedation, as well as weakness, decreased muscle tone, confusion, fatigue, **nausea**, and **dizziness**. Baclofen should not be taken with central nervous system depressants or alcohol.

Benzodiazepines, such as diazepam (Valium), clonazepam (Klonopin, Rivotril), and lorazepam (Ativan) reduce spasticity by acting on the central nervous system. The benefits of benzodiazepines include improved passive range of motion, less muscle overactivity, fewer painful spasms, and overall relaxation. These medications are often taken at night because they cause drowsiness. They are also taken at night to relieve muscle spasms that interrupt sleep. Side effects include unsteadiness, loss of strength, low blood pressure, gastrointestinal symptoms, memory problems, confusion, and behavioral problems.

Datrolene sodium (Dantrium) acts on the muscles to directly interfere with the chemistry of the muscle contraction. It is generally used when other medications are not effective. Benefits may include improved passive movement, decreased muscle tone, and reduced muscle spasms, tightness, and pain. Side effects include generalized weakness, including weakness of the respiratory muscles, as well as drowsiness, fatigue, **diarrhea**, and sensitivity to the sun. Liver problems may occur with this medication, so frequent lab tests are performed to evaluate liver function.

Tizanidine (Zanaflex) reduces spasticity by acting on the central nervous system. It does not usually cause reduced muscle strength. The most common side effect is sedation, and other side effects include low blood pressure, dry mouth, dizziness, and hallucinations. Liver problems may occur with this medication, so frequent lab tests are performed to evaluate liver function.

INJECTED MEDICATIONS Botulinum toxin type A (Botox, Dysport) or type B (Myobloc) is injected locally into the affected muscle group to relax the muscles. It works by preventing nerves from sending signals to the muscles that cause them to contract. Although the treatment takes one to two weeks to reach its full effectiveness, the beneficial effects last three to four months. Botulinum-toxin allows more normal limb positioning and improved mobility. In some patients, the injections also decrease pain. Injections may be used to make casting easier, ease the adjustment of a new brace, or delay surgery.

Botulinum toxin is made by the bacteria that cause **botulism**. However, the amount of botulinum toxin injected to treat spasticity is so small that it would not cause botulism poisoning. This treatment is very safe, and the injections can be given in a doctor's office without the use of sedation or anesthesia. Injections can be repeated but should be spaced apart from three to six months to avoid exceeding the recommended dose. Botulinum-toxin injections can be used in combination with oral medications or intrathecal baclofen to treat spasticity.

Botulinum-toxin injections are typically expensive and may not be covered by insurance. A Reimbursement Hotline established by Allergan, the manufacturer of Botox, is a resource for reimbursement questions: available online at <www.botox.com>. Elan, the manufacturer of Myobloc, also has resources available to answer questions about reimbursement: available online at <www.elan.com>.

Alcohol and phenol are injected in combination but are less commonly used to treat spasticity. The medications are injected directly onto nerves that supply spastic muscles to destroy them. The injections cut off the signals to those muscles, allowing them to relax. This treatment may be used to treat spasticity in larger muscle groups closer to the trunk, such as the thigh muscles. Although this treatment is generally less expensive than botulinum-toxin injections, there are more serious side effects.

Short-term medications such as lidocaine, a local anesthetic, can be used to assess the potential benefit of botulinum toxin or alcohol and phenol injections.

CONTINUOUS DELIVERY MEDICATIONS Baclofen usually is taken as an oral medication but also can be delivered directly into the spinal fluid when the oral medication does not effectively control symptoms. An intrathecal baclofen delivery system continuously releases prescribed amounts of baclofen in small doses directly into the spinal fluid via a small catheter and pump. This type of delivery system causes fewer and less severe side effects than the oral baclofen.

To determine the potential effectiveness of the system, an initial trial of the intraspinal therapy is conducted. During this trial, the medication is delivered into the spinal fluid via a lumbar puncture procedure. The medication usually reaches its peak effectiveness within four hours. If the patient responds favorably to the trial, the intrathecal system can be considered.

The intrathecal baclofen delivery system is placed by a neurosurgeon during a surgical procedure under local or general anesthesia. First, a catheter (thin, flexible tube) is inserted through a needle and guided into the spinal canal, close to where pain pathways enter the spinal cord. The other end of the catheter is tunneled under the skin to the abdomen where a pocket is created. There, the pump is implanted under the skin (epidermal area) through an incision in the abdomen. The baclofen pump is a round, titanium disc about one inch thick and about three inches in diameter. The pump is anchored to surrounding tissue and connected to the catheter. The incision is then closed.

The pump reservoir is filled with the prescribed amount of medication. Medication can be filled and refilled in the pump by inserting a needle through the skin into a filling port (called a diaphragm) in the center of the pump.

The medication is dispensed, either continuously or at certain intervals as determined by the doctor, via a tiny motor in the pump that moves the medication from the pump reservoir through the catheter. Baclofen flows freely in the spinal canal, affecting the nerves to control hyperactive muscles. The system contains a computer chip, so adjustments to the dose, rate, and timing of the medication can be made by the physician using an external programmer. The system also has an alarm to indicate when the reservoir needs to be refilled, the battery is low, or the pump is not delivering the medication. If the system does not appear to be effective in treating spasticity, it can be turned off and eventually removed.

Pump refills and medication adjustments are generally made once every two to three months after the initial dosage is established. The pump system lasts from three to five years, at which time the system needs to be replaced.

Surgery

Surgery is only recommended when all other treatments have been tried and have not effectively controlled the child's spasticity symptoms. Surgical options for chronic spasticity include selective dorsal rhizotomy and tendon release surgery.

Selective dorsal rhizotomy surgery, also called selective posterior rhizotomy, involves a surgical resection of part of the spinal nerve. By cutting the sensory nerve rootlets that cause the spasticity, muscle stiffness is decreased while other functions are maintained. Potential benefits of this surgical procedure include pain relief, reduced spasticity to improve walking or aid sitting in a wheelchair, increased ability to bend at the waist, and improved use of the hands. Sometimes, rhizotomy results in improved breathing and better control of the arms, legs, and head.

Orthopedic surgery for spasticity may be performed to correct a contracture. During contracture release surgery, the tendon of a contractured muscle is cut, the joint is repositioned to a more normal angle, and a cast is applied. Regrowth of the tendon to this new length occurs over several weeks following surgery. After the cast is removed, physical therapy can help strengthen the muscles and improve range of motion. This procedure is most commonly performed on the Achilles tendon but

may also be performed on the knees, hips, shoulders, elbows, and wrists.

Tendon transfer surgery is another technique to treat contractures. During this procedure, the tendon attached to a spastic muscle is cut and transferred to a different site, preventing the muscle from being pulled into an abnormal position.

The disadvantages of these orthopedic procedures are that they are irreversible and that they may need to be repeated.

Other orthopedic surgeries that may accompany contracture release surgery include osteotomy, in which a small wedge is removed from a bone to allow repositioning. A cast is applied while the bone heals in a more natural position. Osteotomy is more commonly performed on the bones in the hips or feet. Arthrodesis is a fusing of bones that normally move independently to limit the ability of a spastic muscle to pull the joint into an abnormal position. Arthrodesis is more commonly performed on the bones in the ankle.

Nutritional concerns

Dietary guidelines are individualized, based on the child's age, diagnosis, overall health, severity of disability, and level of functioning. Specific nutritional problems, such as swallowing or feeding difficulties, may be a concern in some patients and should be managed by a team of specialists including a speech therapist. Early identification, treatment, and correction of specific feeding problems will improve the health and nutritional status of the patient.

A well-balanced and carefully planned diet will help maintain general good health for people with spasticity. Specialists recommend that people with multiple sclerosis and other **movement disorders** adhere to the same low-fat, high-fiber diet that is recommended for the general population.

Children with spasticity may have different energy needs, depending on their condition. One study indicated that ambulatory and non-ambulatory adolescents with cerebral palsy had decreased energy needs compared with a control group of normal adolescents. Therefore, a child's specific calorie needs should be evaluated by a registered dietitian who can work with the parents to develop an individualized meal plan. The child's weight should be obtained once a week or at least once a month to determine if caloric intake is adequate.

A child's self-feeding skills can impact his or her health outcome. One study indicated that 90 percent of children with good to fair motor and feeding skills reached adulthood. In contrast, a lack of self-feeding skills was associated with a six-fold increase in mortality (rate of death).

Maintaining a healthy weight is important to prevent the development of chronic diseases such as diabetes, high blood pressure (**hypertension**), and heart disease.

Tube feedings may be required in some patients with **failure to thrive**, aspiration pneumonia, difficulty swallowing, or an inability to ingest adequate calories orally to maintain nutritional status or promote growth.

Alternative treatment

Alternative and complementary therapies include approaches that are considered to be outside the mainstream of traditional health care.

Techniques that reduce stress, such as **yoga**, Tai Chi, meditation, deep breathing exercises, guided imagery, and relaxation training, may be helpful to induce relaxation and manage spasticity. Acupuncture and biofeedback training also may help induce relaxation. Before learning or practicing any particular technique, it is important for the parent/caregiver and child to learn about the therapy, its safety and effectiveness, potential side effects, and the expertise and qualifications of the practitioner. Although some practices are beneficial, others may be harmful to certain patients.

Initial trials of cannabinoids, the active ingredient in marijuana, have shown promise in the treatment of muscle stiffness and limb straightening associated with multiple sclerosis. Further research is needed to determine the beneficial effects of marijuana-derived substances on neuromuscular symptoms associated with movement disorders. Researchers caution that **smoking** marijuana is dangerous, especially since there may be other harmful substances mixed in with the illegal drug.

Relaxation techniques and dietary supplements should not be used as a substitute for medical therapies prescribed by a doctor. Parents should discuss these alternative treatments with the child's doctor to determine the techniques and remedies that may be beneficial for the child.

Prognosis

There is no cure for spasticity, and it cannot be prevented. However, it can be well-managed with the proper combination of physical and occupational therapy, medication, and surgery. The long-term outlook for those with spasticity depends on the severity of the spasticity and the associated disorder.

KEY TERMS

Active motion (spontaneous)—Motions produced by the activity of a person. Active range of motion exercises are those that are performed by the patient without assistance.

Activities of daily living (ADL)—The activities performed during the course of a normal day, for example, eating, bathing, dressing, toileting, etc.

Anoxia—Lack of oxygen.

Central nervous system—Part of the nervous system consisting of the brain, cranial nerves, and spinal cord. The brain is the center of higher processes, such as thought and emotion and is responsible for the coordination and control of bodily activities and the interpretation of information from the senses. The cranial nerves and spinal cord link the brain to the peripheral nervous system, that is the nerves present in the rest of body.

Cerebral palsy—A nonprogressive movement disability caused by abnormal development of or damage to motor control centers of the brain.

Clonic—Referring to clonus, a series of muscle contractions and partial relaxations that alternate in some nervous diseases in the form of convulsive spasms.

Contraction—A tightening of the uterus during pregnancy. Contractions may or may not be painful and may or may not indicate labor.

Contracture—A tightening or shortening of muscles that prevents normal movement of the associated limb or other body part.

Dysphagia—Difficulty in swallowing.

Dystonia—Painful involuntary muscle cramps or spasms.

General anesthesia—Deep sleep induced by a combination of medicines that allows surgery to be performed.

Hyperactive reflexes—Reflexes that persist too long and may be too strong. For example, a hyperactive grasp reflex may cause the hand to stay clenched in a tight fist.

Hypertonia—Having excessive muscular tone or strength.

Local anesthesia—Pain-relieving medication used to numb an area while the patient remains awake. Also see general anesthesia.

Muscle spasm—Localized muscle contraction that occurs when the brain signals the muscle to contract.

Neurologist—A doctor who specializes in disorders of the nervous system, including the brain, spinal cord, and nerves.

Neurosurgeon—Physician who performs surgery on the nervous system.

Occupational therapist—A healthcare provider who specializes in adapting the physical environment to meet a patient's needs. An occupational therapist also assists patients and caregivers with activities of daily living and provide instructions on wheelchair use or other adaptive equipment.

Orthopedist—A doctor specializing in treatment of the musculoskeletal system.

Passive movement—Movement that occurs under the power of an outside source such as a clinician. There is no voluntary muscular contraction by the individual who is being passively moved.

Peripheral nerves—Nerves outside the brain and spinal cord that provide the link between the body and the central nervous system.

Physiatrist—A physician who specializes in physical medicine and rehabilitation.

Physical therapist—A healthcare provider who teaches patients how to perform therapeutic exercises to maintain maximum mobility and range of motion.

Pressure ulcer—Also known as a decubitus ulcer or bedsore, a pressure ulcer is an open wound that forms whenever prolonged pressure is applied to skin covering bony prominences of the body. Patients who are bedridden are at risk of developing pressure ulcers.

Range of motion (ROM)—The range of motion of a joint from full extension to full flexion (bending) measured in degrees like a circle.

Rigidity—A constant resistance to passive motion.

Scissoring—Involuntary crossing of the legs.

Stroke—Interruption of blood flow to a part of the brain with consequent brain damage. A stroke may be caused by a blood clot or by hemorrhage due to a burst blood vessel. Also known as a cerebrovascular accident.

Parental concerns

Parents should work closely with the child's therapists and doctors to create an effective treatment plan. It is important for parents to communicate their treatment goals with the healthcare team. Parents should take an active role in the child's exercise program and help the child practice the exercises as prescribed every day. Raising a child with a movement disorder can be challenging. There are several support groups available to provide information and assistance.

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National Center on Birth Defects and Developmental Disabilities, Centers for Disease Control. 4770 Buford Hwy., NE, Ste. F-35, Atlanta, GA 30341. Web site: <http://cdc.gov/ncbddd/dh>.

National Institute on Disability and Rehabilitation Research, Office of Special Education and Rehabilitative Services. U.S. Department of Education, 400 Maryland Ave., SW, Washington, DC 20202–7100. Web site: <www.ed.gov/about/offices/list/osers/nidrr/>.

National Institute of Neurological Disorders and Stroke (NINDS), National Institutes of Health. PO Box 5801, Bethesda, MD 20824. Web site: <www.ninds.nih.gov/about_ninds/>.

National Rehabilitation Information Center (NARIC). 4200 Forbes Blvd., Ste. 202, Lanham, MD 20700. Web site: <www.naric.com>.

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Angela M. Costello

Special education

Definition

Special education refers to a range of educational and social services provided by the public school system and other educational institutions to individuals with disabilities who are between three and 21 years of age.

Purpose

Special education is designed to ensure that students with disabilities are provided with an environment that allows them to be educated effectively. Disabilities that qualify for special education include physical disabilities, such as deafness or blindness; mental disabilities, such as Down's syndrome and **autism**; medical conditions, such as oxygen dependence or traumatic brain injury; learning deficits, such as **dyslexia**; and behavioral disorders, such as attention deficit hyperactivity disorder (ADHD) and conduct disorders.

In 1975, the Education for All Handicapped Children Act (EHCA, PL 94-142) mandated that states provide a "free and appropriate public education" (FAPE) to all students, including those with physical, mental, or behavioral disabilities. This special education must include a comprehensive screening and diagnosis by a multi-disciplinary team and the development of an annual Individualized Education Plan (IEP) for each student, outlining academic and behavioral goals, services to be provided, and methods of evaluation. The student's parents must consent to initial screening and must be invited to participate in all phases of the process.

In 1997, the Individuals with Disabilities Education Act (IDEA) expanded special education services by mandating that all children with disabilities—regardless of the type or severity of their disability—between the ages of three and 21 years are entitled to FAPE in the least restrictive environment. That is, children requiring special education must by educated with nondisabled children to the maximum extent possible in an appropriate program to meet their special needs. While the majority of children with disabilities are taught at least part-time in a general classroom setting, many children are segregated, most often due to a lack of staff and resources to support special needs students in general classrooms. This stipulation that special-needs children be educated in the least restrictive environment led to the practice of mainstreaming, which is the policy of placing special education students in regular classrooms as much as possible and using separate resource rooms where the students receive special tutoring, review, and instruction.

Although gifted and talented students are not usually considered candidates for special education and there is no federal mandate to support these students, exceptionally gifted children may also be entitled to receive special education services. Gifted children who are not identified and continue to be taught in a general classroom may develop behavioral issues due to boredom. Specially designed gifted education programs are available in many school districts. In addition, bilingual children may require special education services. Children whose native language is not English may not receive appropriate education due to their language barrier. Bilingual language support services should be provided.

Description

Special education can include a range of support services, depending on the special needs of the student. Support services may involve physical assistance and therapy, counseling and psychotherapy, modified learning environments and assistive learning devices, educational and psychological assessments, and behavioral modification techniques.

According to U.S. Department of Education statistics, approximately 600,000 children aged three to five years were served by special education services in 2001. In **preschool** children, the most prevalent disability was speech or language impairment. Approximately 5.8 million students aged six to 21 years were served by special education services in 2001. Common disabilities include specific learning disabilities (e.g., dyslexia), speech or language impairment, **mental retardation**, and emotional disturbance.

In order to qualify for special education a child must be diagnosed as having a disability and the disability must be found to "adversely affect educational performance" so as to require special services. Referral and evaluation for special education varies widely. For children with severe disabilities, a physician and the parents usually identify and refer the child to special education. Other disabilities or deficits in the child's developing physical and cognitive abilities may be identified by teacher and parent observation or revealed by academic or developmental tests. Most school districts have standardized programs to screen large numbers of children between kindergarten and third grade. Other disabilities may be subtle or compensated for, such as dyslexia, and may not be discovered until demands on the student increase in college. After referral, a meeting is held to determine whether the child should be assessed or evaluated to determine the type of disability he or she may have. Tests attempt to identify the cognitive (academic), social, or physical tasks that the child has difficulty performing and why the difficulty exists, i.e., what disability or disabilities are present. Tests may include: reading, writing, spelling, and math tests; psychological or **intelligence** tests; speech and language; vision and hearing tests; or an examination by a physician. Parents must consent to all testing, evaluation, and placement and can appeal most decisions if they disagree with the conclusions.

After disabilities and special needs are identified, an IEP is developed by school staff with input from the parents. The IEP development team is interdisciplinary and usually includes the special education teacher, another regular academic teacher, the parents, a school administrator, a school psychologist, and other school staff (e.g., nurse, coach, counselor). The IEP should be comprehensive and include the following:

- current performance measures based on multiple tests and **assessment** methods
- educational goals and objectives that define how problems will be addressed in the short and long term

Special education student receiving one-on-one instruction. *(© Richard T. Nowitz/Corbis.)*

- definition of how the child's progress will be measured on an ongoing basis
- disciplinary methods (especially for children with emotional and behavioral issues)
- an individualized healthcare plan (IHP) for students also requiring special medical attention or medications

IEPs vary widely in length and complexity according to the type of disability. More effective IEPs specifically outline the child's needs; are mutually agreed upon by parents, teachers, and counselors; support activities that are typical of other students in the same age-group; promote school and community membership, and clearly facilitate the student's long-range life goals. Often IEPs do not specifically address how progress is to be measured. An effective IEP clearly defines the types of tests and assessments that are to be given to measure the child's progress. Although subjective assessment by teachers can provide valuable insight, objective tests that specifically measure academic and other skills must be included in the IEP.

After the IEP is developed, the student is placed in the appropriate educational setting. Certified special education teachers deliver programs in separate classrooms using modified educational curricula and specially designed assistive education techniques. Children with physical disabilities are provided with any assistive learning technology or equipment they need to complete educational requirements. Examples of such technology include special computers for speech/hearing/language assistance, modified desks, and writing support devices. Specially trained support staff assist students mainstreamed in general classrooms. When the public school cannot provide the appropriate environment and resources to meet the educational needs of the student, it is obligated to find and pay for an alternative educational setting, such as a day program in a mental/behavioral health facility, **home schooling** with appropriate medical/mental health support, an **alternative school** dedicated to serving disabled children, or a private school with special education support services.

Parental concerns

Children with disabilities and their parents have certain legal rights, most importantly, the right to challenge any recommendation made by a school and its staff. Par-

ents who disagree with the school's educational program can hire legal representation, request formal and informal hearings (due process), and obtain additional evaluation from an independent consultant.

Children with emotional disturbances and related behavioral disorders have historically been unrecognized as being eligible for special education services. However, emotional problems can in fact act as a barrier to education. For children with emotional disturbances to qualify for special education, evidence from psychological testing and observation (by teachers or therapists) must demonstrate that the emotional issues significantly affect educational performance. Most public schools do not have the staff and resources to handle children with emotional disturbances, in addition to other children with disabilities. Many alternative schools exist for children with emotional disturbances and behavioral disorders who have average and above-average academic abilities. If the public school cannot adequately provide FAPE for such students, parents can seek legal representation to obtain funding from the public school for their child to attend an appropriate alternative school. Students with emotional disturbances and behavioral disorders should have mental health support services integrated with their IEP.

According to parents, 14 percent of students with disabilities in elementary and middle school had been expelled or suspended at some point in their school careers. And special needs children have a high drop-out rate—approximately 25 percent drop out of school and another 20 percent leave for other reasons. Emotionally disturbed students have the highest drop-out rate (35%), according to Department of Education statistics, while deaf-blind students have the lowest rate (4%). Graduation and employment rates for students with disabilities rose through the two decades that followed the passage of EHCA and IDEA and other disability legislation such as the Americans with Disabilities Act. Depending on the disability, as many as 45 to 70 percent of disabled adults may remain unemployed. However, some special needs students are quite successful. Students with learning disabilities and **speech disorders** have the lowest rates of unemployment, usually because they have participated in vocational education programs with a comprehensive vocational assessment, including assessment of independent living skills.

KEY TERMS

Alternative school—An educational setting designed to accommodate educational, behavioral, and/or medical needs of children and adolescents that cannot be adequately addressed in a traditional school environment.

Attention deficit hyperactivity disorder (ADHD)—A condition in which a person (usually a child) has an unusually high activity level and a short attention span. People with the disorder may act impulsively and may have learning and behavioral problems.

Dyslexia—A type of reading disorder often characterized by reversal of letters or words.

Individualized educational plan (IEP)—A detailed description of the educational goals, assessment methods, behavioral management plan, and educational performance of a student requiring special education services.

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Specific language impairment

Definition

Specific language impairment (SLI) describes a condition of markedly delayed **language development** in the absence of any apparent handicapping conditions.

Description

Many different terms have been used to describe the childhood disorder that is characterized by markedly delayed language development in the absence of any conditions such as deafness, **autism**, or **mental retardation** that would explain the delay. SLI is also sometimes called childhood dysphasia or developmental language disorder.

Demographics

Estimates of true SLI vary according to the age of identification. Some experts argue that as many as 10 percent of two-year-olds may have a specific language impairment, but by age three or four, that percentage drops considerably, presumably because some difficulties resolve themselves. The incidence in the general population is estimated at about 1 percent. SLI is more common in boys than girls.

Causes and symptoms

Children with SLI usually begin to talk at roughly the same age as normal children but are markedly slower in the progress they make. They seem to have particular problems with inflection and word forms, such as leaving off endings when forming verb tenses (for example, the -ed ending when forming the past tense). This problem can persist much longer than early childhood, often into the grade school years and beyond, where these children encounter difficulties in reading and writing. The child with SLI also often has difficulties learning language incidentally, that is, in picking up the meaning of a new word from context or generalizing a new syntactic form. This is in decided contrast to the normal child's case, where incidental learning and generalization are the hallmarks of language acquisition. Children with SLI are not cognitively impaired and are not withdrawn or socially aloof like an autistic child.

Very little is known about the cause or origin of specific language impairment, although evidence in the early 2000s is growing that the underlying condition may be a form of brain abnormality. Any such brain abnormality, however, is not readily apparent with existing diagnostic technologies. SLI children do not have clear brain lesions or marked anatomical differences from other children in either brain hemisphere. However, there is some indication that SLI can be passed down from parents to children. Research as of 2004 suggested a possible genetic link, although there are many problems in identifying such a gene. Sometimes the siblings of an affected child show milder forms of the difficulty, complicating the picture. One of the major stumbling blocks is the definition of the disorder, because children with SLI show many different kinds of symptoms which makes it hard to determine what the genetic cause of the disorder might be.

Some investigators have attributed the difficulties that children with SLI have to problems with speech sound perception, suggesting that inflection and word forms such as endings are hard for the child to perceive because those items are fleeting and unstressed in speech. It is not that the child is deaf in general but that he or she has a specific difficulty discriminating some speech sounds.

Other researchers have argued that this difficulty is not specific to speech but reflects a general perceptual difficulty with the processing of rapidly timed events, of which speech is the most taxing example. The left hemisphere of the brain seems to be specialized for processing rapid acoustic events, so perhaps the child with SLI has a unique difficulty in that part of the brain. Some researchers investigate children with SLI who speak different languages to see if any patterns emerge in the kinds of difficulties the children experience.

When to call the doctor

If a parent notices that a child is having problems with speech or is not achieving language milestones around the usual time, a doctor should be consulted.

Diagnosis

Early identification is very important for the success of interventions for SLI. The disorder is usually diagnosed by comparing a child's linguistic abilities to those that are expected for children of the same age. If the child is significantly behind his or her age peers in terms of language development, SLI is likely. One procedure for diagnosing children aged 24 to 36 months asks parents to complete a standardized questionnaire in which they check off the vocabulary the child knows and write down examples of the child's two-word sentences. If the child's vocabulary contains fewer than 50 words and the child does not use any two-word sentences, that is an indication of SLI or another language disorder.

Treatment

SLI is generally treated by intervention that focuses on helping the child with whatever specific language problems he or she is having. The child with SLI may become increasingly aware of his or her difficulties with language and may lose spontaneity and avoid speaking as he or she gets older. Intensive language intervention can allow these children to make considerable gains, with modeling of appropriate linguistic forms that the child is having difficulty with being especially effective.

Prognosis

The prognosis for children with SLI depends very heavily on the type and severity of the language problem experienced. Many language problems can be largely overcome, although some difficulties usually persist.

Prevention

There is no known way to prevent SLI.

Parental concerns

Children with SLI are often at risk for reduced performance in other areas of their lives because of their difficulty in mastering language. SLI can lead to decreased social interaction and decreased school performance.

KEY TERMS

Inflection—Variations in the pitch or tone of a voice.

Resources

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Speech disorders

Definition

Speech disorders are characterized by a difficulty in producing normal speech patterns.

Description

Children go through many stages of speech production while they are learning to communicate. What is normal in the speech of a child of one age may be a sign of a problem in an older child. Speech disorders include voice disorders (abnormalities in pitch, volume, vocal quality, resonance, or duration of sounds), articulation

disorders (problems producing speech sounds), and fluency disorders (impairment in the normal rate or rhythm of speech, such as **stuttering**.

Demographics

Speech disorders are common. More than a million children in the public schools' **special education** programs have been diagnosed with a speech disorder. One in 10 people in the United States is affected by a communication disorder (speech, language, or hearing disorders).

Causes and symptoms

The causes of most speech disorders are not known. Deafness and hearing loss are significant causes of speech delays and disorders. The symptoms of a speech disorder depend heavily on the age of the child. There are no symptoms of speech disorders that apply to all ages of children. Basic guidelines about what kind of speech is normal at what age can be helpful in determining if a child is missing significant speech milestones.

- Twelve months: By this time babies should respond nonverbally, have different types of cries, and may know one or a few simple words (e.g. "mama" or "dada"). At this age babies should coo and babble.
- Eighteen months: Children of this age should be increasing their vocabularies slowly and be able to produce five to 20 common words.
- Twenty-four months: At this point vocabulary building should begin to speed up. At this age children should be able to produce simple sentences made up of two words.
- Three years: Children should begin to be able to produce speech that is understood by those outside immediate caretakers. Sentences become longer and more complex, and vocabulary increases drastically.

When to call the doctor

If a child continuously misses speech milestones, or is significantly behind what is generally considered average for his or her age, a doctor should be consulted. If hearing loss is ever suspected, such as if a child only responds when the parent speaking is in eyesight, the doctor should be consulted without delay.

Diagnosis

A doctor will do a hearing test on the child to ensure that a hearing problem is not responsible for the speech delay. The doctor may interact with the child to determine linguistic competence. In addition, he or she will interview the parents or other caregivers or have them fill out a list indicating the child's verbal skills. The doctor will typically refer the child to a speech pathologist—a professional specializing in treating speech problems. The speech pathologist will work with the child, the child's **family**, and any other caregivers to develop a plan to help the child.

Treatment

Children with isolated speech disorders are often helped by articulation therapy, in which they practice repeating specific sounds, words, phrases, and sentences. For stuttering and other fluency disorders, a popular treatment method is fluency training, which develops coordination between speech and breathing, slows down the rate of speech, and develops the ability to prolong syllables. A child may practice saying a single word fluently and then gradually add more words, slowly increasing the amount and difficulty of speech that can be mastered without stuttering. The speaking situations can gradually be made more challenging as well, beginning with speaking alone to the pathologist and ending with speaking to a group of people.

Delayed auditory feedback (DAF), in which stutterers hear an echo of their own speech sounds, has also been effective in treating stuttering. When a speech problem is caused by serious or multiple disabilities, a neurodevelopmental approach, which inhibits certain reflexes to promote normal movement, is often preferred. Other techniques used in speech therapy include the motor-kinesthetic approach and biofeedback, which helps children know whether the sounds they are producing are faulty or correct. For children with severe communication disorders, speech pathologists can assist with alternate means of communication, such as manual signing and computer-synthesized speech.

Prognosis

When speech disorders are detected and treated early, the prognosis is generally very good. Many speech disorders that are not caused by other underlying problems resolve themselves, and most others can be resolved completely or nearly completely with prompt treatment. Stuttering resolves itself without treatment in about 50–80 percent of children.

KEY TERMS

Speech pathologist—An individual certified by the American Speech-Language-Hearing Association (ASHA) to treat speech disorders.

Prevention

There is no known way to prevent most speech disorders, although making sure that children have a language-rich environment is thought to help disorders related to lack of input.

Parental concerns

Speech disorders and significant speech delays can have a lasting negative impact on children. Children who have speech disorders may not want to communicate with their peers or even adults which may adversely affect their performance in school and social development.

See also Language delay; Language disorders.

Resources

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ORGANIZATIONS

American Speech-Language-Hearing Association. 10801 Rockville Pike, Rockville, MD 20852. (800) 638–8255. Web site: <www.asha.org>.

Tish Davidson

Spina bifida

Definition

Spina bifida is a birth abnormality in which the spine is malformed and lacks its usual protective skeletal and soft tissue coverings.

Description

Spina bifida may appear in the body midline anywhere from the neck to the buttocks. In its most severe form, termed spinal rachischisis, the entire spinal canal is open, exposing the spinal cord and nerves. More commonly, the abnormality appears as a localized mass on the back that is covered by skin or by the meninges, the three-layered membrane that envelops the spinal cord. Spina bifida is usually readily apparent at birth because of the malformation of the back and paralysis below the level of the abnormality.

Various forms of spina bifida are known as meningomyelocele, myelomeningocele, spina bifida aperta, open spina bifida, myelodysplasia, spinal dysraphism, spinal rachischisis, myelocele, and meningocele. The term meningocele is used when the spine malformation contains only the protective covering (meninges) of the spinal cord. The other terms indicate involvement of the spinal cord and nerves in the malformation. A related term, spina bifida occulta, indicates that one or more of the bony bodies in the spine are incompletely hardened, but that there is no abnormality of the spinal cord itself.

Demographics

Spina bifida occurs worldwide, but there has been a steady downward trend in occurrence rates since about 1940, particularly in regions of high prevalence. The highest prevalence rates, about one in 200 pregnancies, have been reported from certain northern provinces in China. Intermediate prevalence rates, about one in 1000 pregnancies, have been found in Central and South America. The lowest prevalence rates, less than one in 2,000 pregnancies, have been found in European countries. The highest regional prevalence in the United States of about one in 500 pregnancies has occurred in the Southeast.

Causes and symptoms

Spina bifida may occur as an isolated abnormality or in the company of other malformations. As an isolated abnormality, spina bifida is caused by the combination of genetic factors and environmental influences that bring about malformation of the spine and spinal column. The specific genes and environmental influences that contribute to the many-factored causes of spina bifida were not as of 2004 completely known. An insufficiency of **folic acid** is known to be one influential nutritional factor. Changes (mutations) in genes involving the metabolism of folic acid are believed to be significant genetic risk factors. The recurrence risk after the birth of an infant with isolated spina bifida is 3 to 5 percent.

Recurrence may be for spina bifida or another type of spinal abnormality.

Spina bifida may arise because of chromosome abnormalities, single gene mutations, or specific environmental insults such as maternal **diabetes mellitus** or prenatal exposure to certain anticonvulsant drugs. The recurrence risk varies with each of these specific causes.

In most cases, spina bifida is obvious at birth because of malformation of the spine. The spine may be completely open, exposing the spinal cord and nerves. More commonly, the spine abnormality appears as a mass on the back covered by membrane (meninges) or skin. Spina bifida may occur any where from the base of the skull to the buttocks. About 75 percent of abnormalities occur in the lower back (lumbar) region. In rare instances, the spinal cord malformation may occur internally, sometimes with a connection to the gastrointestinal tract.

In spina bifida, many complications arise, dependent in part on the level and severity of the spine malformation. As a rule, the nerves below the level of the abnormality develop in a faulty manner and fail to function, resulting in paralysis and loss of sensation below the level of the spine malformation. Since most abnormalities occur in the lumbar region, the lower limbs are paralyzed and lack sensation. Furthermore, the bowel and bladder have inadequate nerve connections, causing an inability to control bowel and bladder function. Most infants also develop hydrocephaly, an accumulation of excess fluid in the four cavities of the brain. At least one of every seven cases develops findings of Chiari II malformation, a condition in which the lower part of the brain is crowded and may be forced into the upper part of the spinal cavity.

There are a number of mild variant forms of spina bifida, including multiple vertebral abnormalities, skin dimples, tufts of hair, and localized areas of skin deficiency over the spine. Two variants, lipomeningocele and lipomyelomeningocele, typically occur in the lower back area (lumbar or sacral) of the spine. In these conditions, a tumor of fatty tissue becomes isolated among the nerves below the spinal cord, which may result in tethering of the spinal cord and complications similar to those with open spina bifida.

Diagnosis

Few disorders are to be confused with open spina bifida. The diagnosis is usually obvious based on the external findings at birth. Paralysis below the level of the abnormality and fluid on the brain (hydrocephaly) may contribute to the diagnosis. Other spine abnormalities such as congenital **scoliosis** and kyphosis, or soft tissue tumors overlying the spine, are not likely to have these accompanying findings. In cases in which there are no external findings, the diagnosis is more difficult and may not become evident until neurological abnormalities or hydrocephaly develop weeks, months, or years following birth.

Prenatal diagnosis may be made in most cases with ultrasound examination after 12 to 14 weeks of pregnancy. Many cases are also detected by the testing of the mother's blood for the level of alpha-fetoprotein at about 16 weeks of pregnancy. If the spine malformation is not skin covered, alpha-fetoprotein from the fetus's circulation may leak into the surrounding amniotic fluid, a small portion of which is absorbed into the mother's blood.

Treatment

Aggressive surgical and medical management have improved the survival and function of infants with spina bifida. Initial surgery may be carried out during the first days of life, in the hope of providing protection against injury and infection. Subsequent surgery is often necessary to protect against excessive curvature of the spine, and in the presence of hydrocephaly, to place an echanical shunt to decrease the pressure and amount of cerebrospinal fluid in the cavities of the brain. Because of weakness or paralysis below the level of the spine abnormality, most children require physical therapy, bracing, and other orthopedic assistance in order to be able to walk. A variety of approaches including periodic bladder catheterization, surgical diversion of urine, and **antibiotics** are used to protect urinary function.

Although most individuals with spina bifida have normal intellectual function, learning disabilities or **mental retardation** occur in a minority. This deficit may result, in part, from hydrocephaly and/or infections of the nervous system. Children so affected may benefit from early educational intervention, physical therapy, and occupational therapy. Counseling to improve self-image and lessen barriers to socialization becomes important in late childhood and **adolescence**.

Open fetal surgery has been performed for spina bifida during the last half of pregnancy. After direct closure of the spine malformation, the fetus is returned to the womb. By preventing chronic intrauterine exposure to mechanical and chemical trauma, **prenatal surgery** improves neurological function and leads to fewer complications after birth. Fetal surgery is considered experimental, and results have been mixed.

Prognosis

More than 80 percent of infants born with spina bifida survive with surgical and medical management. Although complications from paralysis, hydrocephaly, Chiari II malformation, and urinary tract deterioration threaten the well-being of the survivors, the outlook for normal intellectual function is good.

Prevention

Prevention of isolated spina bifida and other spinal abnormalities became possible in the 1980s and 1990s. The major prevention is through the use of folic acid, one of the B **vitamins**, for several months prior to and following conception. The Centers for Disease Control and Prevention (CDC) recommend the intake of 400 micrograms of synthetic folic acid every day for all women of childbearing years. For women who have had a previous child with spina bifida, the CDC recommends a daily intake of 4 milligrams of synthetic folic acid to help prevent a recurrence of spina bifida in future pregnancies.

Parental concerns

Caring for a child with spina bifida can be a daunting endeavor. Initially, parents may be overwhelmed with the medical decisions to be made and with the grief experienced after the birth of a special needs child. Many parents benefit from early and continuing involvement of an experienced social worker. There will be a multitude of medical decisions to be made. Children with spina bifida require a multidisciplinary team of healthcare providers, including surgeons, physicians, and therapists. Parents may find it helpful to designate a physician, usually the primary pediatrician, or an experienced rehabilitation counselor to act as an advocate for their child and to aid them in coordinating their child's treatment program.

Parental concerns may be two-fold, medical and emotional. Medical concerns include monitoring their child's condition after surgery. Children with spina bifida may have many surgical procedures throughout their lives. Post surgical complications are common but may often be avoided. Parents will be given care instructions after each surgery. Children with spina bifida face a multitude of heath issues such as monitoring bladder and bowel function, maintaining proper **nutrition**, preventing broken bones, promoting healthy growth and development, and encouraging activity and mobility. Many children with spina bifida have non-surgical treatments as well, such as positional aides to help the child sit and stand, physical therapy, and bracing and splints usually of the lower extremities.

Doctor examining a child with spinal bifida. *(© Annie Griffiths Belt/Corbis.)*

Parents of children with spina bifida experience an array of emotions, including grief, **fear**, **anxiety**, and stress. Spina bifida impacts not only the affected child but the entire **family**. Groups and networks of other families affected by spina bifida can provide valuable support. Parents may need to be active in ensuring that their child receives the early intervention and educational services available in their community. Each state has programs to encourage healthy development in children with special needs.

Finally, parents should remember that most children with spina bifida live productive and happy lives. For the most part, children with spina bifida have average or above-average **intelligence**. Many of these children can go on to higher education, have active careers, and live self-sufficiently. It is important for parents to encourage

KEY TERMS

Bracing—Using orthopedic devices to hold joints or limbs in place.

Chiari II anomaly—A structural abnormality of the lower portion of the brain (cerebellum and brainstem) associated with spina bifida. The lower structures of the brain are crowded and may be forced into the foramen magnum, the opening through which the brain and spinal cord are connected.

Fetus—In humans, the developing organism from the end of the eighth week to the moment of birth. Until the end of the eighth week the developing organism is called an embryo.

Hydrocephalus—An abnormal accumulation of cerebrospinal fluid within the brain. This accumulation can be harmful by pressing on brain structures, and damaging them.

Splint—A thin piece of rigid or flexible material that is used to restrain, support, or immobilize a part of the body while healing takes place.

strong self esteem in their child and to foster independent living skills.

Resources

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ORGANIZATIONS

March of Dimes Birth Defects Foundation. 1275 Mamaroneck Ave., White Plains, NY 10605. Web site: <www.modimes.org>.

National Birth Defects Prevention Network. Web site: <www.nbdpn.org>.

Shriners Hospitals for Children. International Shrine Headquarters, 2900 Rocky Point Dr., Tampa, FL 33607–1460. Web site: <www.shrinershq.org>.

Spina Bifida Association of America. 4590 MacArthur Blvd. NW, Suite 250, Washington, DC 20007–4226. Web site: <www.sbaa.org>.

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Spinal cord injury

Definition

Spinal cord injury (SCI) is damage to the spinal cord that results in a loss of function such as mobility or feeling. The spinal cord does not have to be severed in order for a loss of function to occur. In most SCI cases, the spinal cord is intact, but the damage to it results in loss of function.

Description

The spinal cord and the brain are the two components of the central nervous system (CNS). The spinal cord extends from the base of the brain, down the middle of the back, to the lower back, and it coordinates movement and sensation in the body. It contains nerve cells, supporting cells, and long nerve fibers (axons) that connect to the brain and carry signals downward from the brain along descending pathways and upward to the brain along ascending pathways. Axons are covered by sheaths of an insulating whitish substance called myelin, and the region in which they lie is accordingly called white matter. The nerve cells themselves, with long branches (dendrites) that receive signals from other nerve cells, make up the gray matter that lies in a butterfly-shaped region in the center of the spinal cord. Like the brain, the spinal cord is enclosed in three membranes (meninges). The innermost layer is called the pia mater, the middle layer is the arachnoid, and the dura mater is the tougher outer layer. The spinal cord consists of several segments along its length, with higher segments controlling movement and sensation in upper parts of the body and lower segments controlling the lower parts of the body. The segments in the neck (cervical region), referred to as C1 to C8, control signals to the neck, arms, and hands. Those in the thoracic or upper back region (T1 to T12) control signals to the torso and some parts of the arms. Those in the mid-back (upper lumbar region) just below the ribs (L1 to L5) control signals to the hips and legs. Finally, the sacral segments (S1 to S5) lie just below the lumbar segments in the mid-back and control signals to the groin, toes, and some parts of the legs.

The types of disability associated with SCI thus depend directly on the type and severity of the injury, the level of the cord at which the injury occurs, and the nerve fiber pathways that are damaged. Severe injury to the spinal cord causes paralysis and complete loss of sensation to the parts of the body controlled by the spinal cord segments below the point of injury. Spinal cord injuries also can lead to many complications, including

pressure sores and increased susceptibility to respiratory diseases.

Demographics

According to the National Institute of Neurological Disorders and **Stroke** (NINDS), accidents and violence cause an estimated 10,000 spinal cord injuries each year, and more than 200,000 Americans live day-to-day with the disabling effects of SCI. The incidence of spinal cord injuries peaks among people in their early 20s, with a small increase in the elderly population due to falls and degenerative diseases of the spine. SCI is an uncommon source of morbidity and mortality in children.

Causes and symptoms

According to the National Spinal Cord Injury Association (NSCIA), spinal cord injuries are caused in the United States by motor vehicle accidents (44%), acts of violence (24%), falls (22%), **sports** (8%), and other causes (2%) such as abscesses, tumors, **polio**, **spina bifida** and Friedrich's Ataxia, a rare inherited disorder. For infants, motor vehicle crash is the leading cause of SCI. Falls rank highest for ages two to nine years and sports for the 10 to 14 age group. The most common injury level for the five to 13 age group is the high cervical spine (C1-C4).

SCI symptoms usually appear immediately after the injury. However, symptoms can develop slowly, if an infection or tumor is gradually increasing pressure on the spinal cord. General symptoms are as follows:

- weakness, poor coordination or paralysis, particularly below the level of the injury
- **numbness**, tingling, or loss of sensation
- loss of bowel or bladder control
- **pain**

When to call the doctor

Immediate medical attention is required if a parent suspects a child may have injured his or her neck or back, or if a child has poor coordination or paralysis in any part of the body. Spinal cord injury is not always obvious: numbness or paralysis may result immediately after SCI or later on as swelling gradually occurs in or around the spinal cord. In either case, the time between injury and treatment is critical and can significantly influence the extent of complications and the level of recovery. Any child who has experienced significant trauma to the head, back, or neck should be medically evaluated for the possibility of SCI.

Diagnosis

The possibility of SCI is usually suspected in anyone with significant trauma to the head and/or neck. Physicians accordingly assume that such patients have a spine fracture until proven otherwise.

Diagnosis is established with the help of x-rays of the spine that allow doctors to determine the extent of the damage. The following imaging tests are also used: CT scan (**computed tomography**), MRI (**magnetic resonance imaging**), and myelogram (x ray after injection of dye into the spinal canal).

Treatment

A person suspected of having a spinal cord injury should not be moved and treatment of SCI begins with **immobilization**, commonly achieved by enclosing the cervical spine in a rigid collar and use of rigid backboards. Paramedics and other rescue workers receive extensive training in immobilizing the spine. Immobilization prevents further injuries to the cord at the scene of the injury and has helped reduce worsening of any neurological SCI injury. At the time of injury, treatment is focused on stabilizing the spine and relieving cord compression. Prompt steroid drug injections (within eight hours of the injury) are also used to minimize cell damage and improve the chance of recovery.

Surgery cannot reverse damage to the spinal cord but is often needed to stabilize the spine to prevent future pain or deformity. It may involve fusing together vertebrae or inserting metal pins; or removing bone chips, bullets, or other **foreign objects**; or draining fluid to relieve pressure. Long-term treatment of spinal cord injuries usually involves drug therapy, the use of neural prostheses, and rehabilitation. Complementary treatment includes **nutrition** management, psychological counseling, and careful monitoring by physicians.

Drug therapy

Effective drug therapy for spinal cord injury was demonstrated in 1990, when methylprednisolone, the first drug shown to improve recovery from spinal cord injury, was approved for standard use. Completely paralyzed patients given methylprednisolone recover an average of about 20 percent of their lost motor function, compared to only 8 percent recovery of function in untreated patients. Partially paralyzed patients recover an average of 75 percent of their function, compared to 59 percent in patients who do not receive the drug.

Neural prostheses

Neural prostheses are used to compensate for lost function resulting from SCI. These sophisticated electrical and mechanical devices connect with the nervous system to supplement or replace lost motor and sensory functions. Neural prostheses contain many intricate components, such as implanted stimulators, electrodes, leads and connectors, sensors, and programming systems. There are many technical considerations in selecting each component. The electronic components must be as small as possible. Biocompatibility between electrodes and body tissue is also required to prevent the patient from being harmed by contact with the device. One device, a neural prosthesis that allows rudimentary hand control, was approved by the United States Food and Drug Administration (FDA). Patients control the device using shoulder muscles. With training, most patients can open and close their hand in two different grasping movements and lock the grasp in place by moving their shoulder in different ways.

Rehabilitation

Rehabilitation techniques can greatly improve patients' health and quality of life by helping them learn to use their remaining abilities. They start by setting functional goals. Functional goals are a realistic expectation of activities that a person with SCI eventually should be able to do with a particular level of injury. These goals are set during rehabilitation with the medical team. They help the patient with SCI learn new ways to manage his/her daily activities and stay healthy. Developing independence is especially important to kids, particularly teenagers. Many hospitals have SCI units geared to help patients develop their independence, and SCI treatment centers are operational in several states with special programs for children. The SCI units include kitchens and laundry facilities and other equipment so that patients can learn independent living skills, such as cooking meals or ironing clothes. A spinal cord injury can also affect the nerves and muscles and can cause bowel and bladder problems and skin problems. Children are prepared for these changes during rehabilitation and are taught the self-care skills needed to deal with these problems. Parents of spinal cord injured children also need to learn how to take care of their spinal-cord injured child. Having a spinal cord injury does not mean that children have to stop participating in games and enjoyable activities. Most SCI units have recreational therapists on staff to show kids how to **play** wheelchair basketball, volleyball, and tennis, as well as specially adapted games.

Alternative treatment

People with spinal cord injuries caused by traumatic events have in the past been considered hopeless cases destined to a life of paralysis. But in the last decades of the twentieth century there were dramatic advances in spinal cord regeneration research. For example, Swiss scientist Martin Schwab actually managed to heal spinal cords in rats and restored their ability to walk. At the Swedish Karolinska Institute, scientists succeeded in constructing a bridge of slender nerve filaments to connect a once-severed spinal cord in rats that subsequently were able to flex their legs. These developments and others offer paralyzed people some hope. In the early 2000s envisioned treatments include an immune therapy procedure that has been tested in Israel with human subjects and possibilities for mechanical neural prostheses.

Acupuncture is a more conservative form of alternative treatment with documented evidence for the reduction of SCI-related **muscle spasms**, increased level of sensation, improved bladder and bowel function, improvement in lower limb paralysis, with younger patients reported to have better outcomes.

Nutritional concerns

Because of the changes that occur in the body after SCI, parents need to understand the role that nutrition can play in the overall health of a child following a spinal cord injury.

Special health concerns resulting from SCI are as follows:

- Bowel management. Individuals with SCI may have neurogenic bowel, with the result that the messages from the brain that control the downward muscular movements of the bowel are either absent or not working properly, making it difficult for stool to move through the intestines. SCI diets accordingly include high fiber and plenty of fluids to regulate bowel movements.
- Heart problems. SCI presents a greater risk for cardiovascular and heart problems, hence the necessity to limit salt and cholesterol intake.
- Pressure ulcers. Pressure ulcers are always a concern to individuals with SCI and a diet high in protein, **vitamins**, and **minerals** is recommended to promote skin healing.
- Kidney or bladder stones. Individuals with SCI may be prone to developing calcium stones. Certain beverages can cause crystals to form in the urine and excessive consumption of dairy products is accordingly avoided with water highly recommended as the best drink.

- Urinary tract infection. The loss of normal bladder function after SCI places an individual at risk for urinary tract infection. A high fluid intake every day has been shown to reduce the problem of infections.
- Weight control. After SCI, the metabolic rate is usually lower. Metabolic rate is how fast a body burns ingested calories. A lower muscle mass and a decrease in activities cause a lower metabolic rate, meaning that fewer calories are needed each day to maintain a desirable weight. After rehabilitation, the ideal body weight of a person with SCI is lower than for a nondisabled individual. Dieticians normally decrease the amount of calories by 5 percent for those with paraplegia and 10 to 15 percent for those with tetraplegia (quadriplegia).

Prognosis

The prognosis of SCI depends on the location and extent of injury. Once the initial injury heals, functional improvements may continue for at least six months. Any disability that remains after that point is likely to be permanent. Injuries of the neck above C4 with significant involvement of the diaphragm have worse outcomes. Although SCI often results in permanent disability, rehabilitation can maximize the level of function and help patients adapt and lead independent, productive lives.

According to the American Association of Neurological Surgeons, mortality from SCI is influenced by several factors, the most important being the severity of associated injuries. Because of the force that is required to fracture the spine, it is not uncommon for the patient to suffer significant damage to the chest and/or abdomen. Many of these associated injuries are fatal. For isolated SCIs, the mortality after one year is roughly 5 to 7 percent. If a patient survives the first 24 hours after injury, the probability of survival for ten years is approximately 75 to 80 percent. Likewise, the ten-year survival rate for patients who survived the first year after injury is 87 percent.

Prevention

The following guidelines have been shown to help prevent SCI:

- use of safe driving practices
- avoidance of situations that may become violent
- keeping firearms locked away
- taking precautions to prevent falls around the home (walkways free from obstacles, non-slip materials in bathtubs, etc)
- use of proper **safety** equipment for sports

The American Academy of Orthopedic Surgeons (AAOS) also recommends that playgrounds be made safe to prevent spinal cord injuries. It offers the following checklist to help parents assess the safety of their child's playground:

- Are any pieces of playground equipment missing supports, anchors, or footings?
- Are any supports, anchors, or footings damaged or loose?
- Has the wood started to splinter or rot?
- Are surface materials missing or damaged?
- Are there any missing, loose, or damaged nuts and bolts on the equipment?
- Are any seats broken?
- Are swing hangers and chains broken or worn?
- Are hooks, rings, or links misshapen or deformed?
- Are there any broken, missing, or loose steps?
- Are any ladder rungs missing, broken, or loose?
- Are tree roots visible or rocks sticking up that could cause a child to trip and fall?

If the answer to any of these questions is "Yes," this playground is not safe for a child. The AAOS recommends that the playground be reported to local park or school officials or to contact a local orthopedic surgeon to enquire as how to build a safe, accessible playground for the area.

Parental concerns

In most cases, SCI requires that the home be modified to be fully accessible to the injured child. Bathrooms need to be fitted with a shower chair, grab bars, a shower wand, a tub lift, or a shower bench. Grab bars should be installed on three sides of the shower, and non-skid strips should be applied to the bottom of the shower or tub. Bedrooms should be located for convenient access to the bathroom and adequate space should be provided around the bed for wheelchair access with convenient storage near the bed for braces, prostheses, and clothing. Light switches should be lowered for easy access and ramps should be built to facilitate displacements.

See also Computed tomography; Magnetic resonance imaging.

Resources

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KEY TERMS

Axon—A long, threadlike projection that is part of a neuron (nerve cell).

Central nervous system—Part of the nervous system consisting of the brain, cranial nerves, and spinal cord. The brain is the center of higher processes, such as thought and emotion and is responsible for the coordination and control of bodily activities and the interpretation of information from the senses. The cranial nerves and spinal cord link the brain to the peripheral nervous system, that is the nerves present in the rest of body.

Computed tomography (CT)—An imaging technique in which cross-sectional x rays of the body are compiled to create a three-dimensional image of the body's internal structures; also called computed axial tomography.

Dendrite—A threadlike extension of the cytoplasm of a neuron that conducts electrical impulses toward the cell body of the neuron. Usually it spreads out into many branches..

Gray matter—Areas of the brain and spinal cord that are comprised mostly of unmyelinated nerves.

Magnetic resonance imaging (MRI)—An imaging technique that uses a large circular magnet and radio waves to generate signals from atoms in the body. These signals are used to construct detailed images of internal body structures and organs, including the brain.

Methylpredisolone—A steroid drug. Methylpredisolone administered within eight hours of acute spinal cord trauma is the first drug shown to improve recovery from spinal cord injury.

Myelin—A fatty sheath surrounding nerves throughout the body that helps them conduct impulses more quickly.

Myelogram—An x-ray image of the spinal cord, spinal canal, and nerve roots taken with the aid of a contrast dye.

Spina bifida—A birth defect (a congenital malformation) in which part of the vertebrae fail to develop completely so that a portion of the spinal cord, which is normally protected within the vertebral column, is exposed. People with spina bifida can suffer from bladder and bowel incontinence, cognitive (learning) problems, and limited mobility.

Vertebrae—Singular, vertebra. The individual bones of the spinal column that are stacked on top of each other. There is a hole in the center of each bone, through which the spinal cord passes.

White matter—A substance, composed primarily of myelin fibers, found in the brain and nervous system that protects nerves and allows messages to be sent to and from the brain and various parts of the body. Also called white substance.

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International Spinal Cord Regeneration Center. PO Box 451, Bonita, California 91902. Web site: <www.electriciti.com/~spinal>.

National Association for Home Care (NAHC). 228 7th Street SE, Washington, DC 20003. Web site: <www.nahc.org>.

National Institute of Neurological Disorders and Stroke (NINDS). PO Box 5801, Bethesda, MD 20824. Web site: <www.ninds.nih.gov>.

National Spinal Cord Injury Association (NSCIA). 6701 Democracy Blvd, Suite 300–9, Bethesda, MD 20817. Web site: <www.spinalcord.org>.

Spinal Cord Society. 19051 County Highway 1, Fergus Falls, MN 56537–7609. Web site: <http://users.aol.com/scsweb>.

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Spinal muscular atrophy

Definition

Spinal muscular atrophy is a term that describes a number of different conditions, all of which have in common the gradual deterioration of the voluntary muscles.

Description

Several different conditions fall under the name spinal muscular atrophy (SMA). These include SMA type I, also called Werdnig-Hoffmann; SMA type II; SMA type III, also called Kugelberg-Welander disease; Kennedy syndrome, or progressive spinobulbar muscular atrophy; and congenital SMA with arthrogryposis.

Demographics

The autosomal recessive forms of spinal muscular atrophy are the most common inherited cause of infant death. Each type of spinal muscular atrophy has an incidence of about 10 to 15 cases in every 100,000 live births.

Causes and symptoms

All types of spinal muscular atrophy are genetic diseases. Most of the syndromes are autosomal recessive, meaning that they have no predilection for either sex. Parents of children with SMA usually carry the gene for the disease but have no symptoms themselves. A child who receives two genes (one from each parent) will express the symptoms of the disease.

Although the entire sequence of abnormalities that causes spinal muscular atrophy was not delineated as of 2004, there is thought to be an absence or deficiency of a specific protein necessary for the proper functioning of the nerve cells responsible for movement (motor neurons).

SMA type I (Werdnig-Hoffmann disease)

SMA type I is usually noted prior to birth, due to a decrease in the baby's movements in utero, or early in life. Babies with this type of SMA have decreased muscle and trunk tone, resulting in floppiness of the limbs and weak arm and leg movements. They have difficulty with swallowing and, therefore, with feeding, and they have breathing problems. These children are unable to learn to sit or to stand. The disease is usually fatal prior to the age of two.

SMA type II

Symptoms of SMA type II are usually noted in a child between three and 15 months of age. Symptoms include breathing problems; weak and floppy limbs; involuntary jerking and twitching of muscles in the arms, legs, and tongue; abnormal reflexes. Children with SMA type II may eventually be able to sit, but they are unable to learn to stand or to walk.

SMA type III (Kugelberg-Welander disease)

Children with SMA type III begin to experience symptoms between the ages of two and 17 years. Problems develop that hamper the child's ability to walk, run, climb stairs, and rise from a chair. Twitches and tremors may develop in the child's fingers.

Kennedy syndrome (progressive spinobulbar muscular atrophy)

This form of spinal muscular atrophy only affects men; it is an X-linked recessive disorder, meaning that the defective gene is passed from mother to son.

Individuals with Kennedy syndrome begin to develop symptoms between the ages of 15 and 60 years. Characteristic symptoms include increasing weakness of the tongue and facial muscles, problems with swallowing, impaired speech, and increased size of the male breast (gynecomastia). The severity of the symptoms of Kennedy syndrome progress gradually.

Congenital SMA with arthrogryposis

This is one of the rarest forms of spinal muscular atrophy. It is present at birth, and children exhibit severe contractures of the joints, resulting in limb deformity; spinal curvature; deformities of the chest wall; difficulties breathing; abnormally small jaw; and upper eyelid droop (ptosis).

Diagnosis

Diagnosis is by a combination of clinical observation; blood tests that reveal an increased level of creatine kinase (which appears in the blood when muscle tissue is being broken down); distinctive abnormalities on muscle biopsy; characteristic electromyographic and nerve conduction abnormalities; and genetic testing.

Treatment

There are no cures for any of the forms of spinal muscular atrophy. The treatments involve addressing the symptoms and attempting to improve quality of life. Medical treatment may be necessary for recurrent **pneumonia** and other respiratory infections. Surgery may be necessary for spinal curvature and severe contractures. Physical therapy, occupational therapy, and other types of rehabilitation programs may help individuals achieve the highest level of functioning possible.

Prognosis

The prognosis for spinal muscular atrophy is variable. Life expectancy is dependent on the degree of respiratory impairment present. Because of the slow progression of symptoms, individuals with Types III or Kennedy syndrome may have normal life spans.

Prevention

There is no way to prevent spinal muscular atrophy. However, genetic counseling is crucial so that parents can make informed decisions about having children. In general, when a **family** has already had a child with SMA, each subsequent pregnancy has a 25 percent chance of producing another child with SMA. Prenatal testing is available. Parents must then decide whether to use the information to help them prepare for the arrival of a baby with SMA or to terminate the pregnancy.

Parental concerns

Caring for a child with SMA can be very challenging and emotionally draining. Support groups, respite care, and help to support other siblings in the family can be important adjunct measures.

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NIH Neurological Institute., PO Box 5801, Bethesda, MD 20824. Web site: <www.ninds.nih.gov>.

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Spinal tap *see* **Cerebrospinal fluid (CSF) analysis**

Spinocerebellar ataxia *see* **Friedrich's ataxia**

Spirometry *see* **Pulmonary function test**

Sports

Definition

Sports are group games and individual activities involving physical activity and skills.

Description

Sports help children develop physical skills, get **exercise**, make friends, have fun, learn to **play** as a member of a team, learn to play fair, and improve **self-esteem**.

Participation in sports is a great way of staying active and offers wonderful rewards for mental health. Being involved in sports has been proven to help children learn valuable skills for dealing with life's ups and downs. They teach youth how to interact with others and work as a team. This skill facilitates working with others in other ways such as on a class project or a school play. Sports also help students become more independent and feel better about themselves. The result is positive self-esteem and self-confidence, which are extremely important for determining later happiness and success.

Sports also offer an enjoyable, exciting environment in which to learn how to handle both failure and success. Everyone wins and loses some of the time in both sports and other endeavors. Winning feels great and empowering but can also cause a young person to feel pressure and **anxiety** in the next attempt to win. Losing usually produces feelings of sadness, depression, and disappointment. Learning how to cope with these different feelings fosters good mental health.

Another aspect of sports that contributes to a healthy mind is goal-setting. Young people who have goals are more likely to be self-motivated and are usually able to accomplish more because they know what they need to do in order to get ahead. Without goals, adolescents tend to lack direction and focus. In sports, goal setting is essential for improving individually and working as a team. This is also true in other pursuits. For example, if a student wants to get better grades, reaching specific goals, such as studying for a certain period of time each night, is the most likely way to achieve them.

SPORTSMANSHIP American sports culture has increasingly become a business. The highly stressful and competitive attitude prevalent at colleges and in professional sports affects the world of children's sports and athletics, creating an unhealthy environment. The attitudes and behavior taught to children in sports carry over into adulthood. Parents should take an active role in helping their child develop good sportsmanship, according to a 2002 health advisory issued by the journal *Clinical Reference Systems*.

To help adolescents get the most out of sports, parents need to be actively involved. Quoting from the American Academy of Child & Adolescent Psychiatry Web site, parental involvement includes the following steps:

- providing emotional support and positive feedback
- attending all or some games and talking about them afterward
- having realistic expectations for your child
- learning the sport and supporting your child's involvement
- helping your child talk with you about experiences with the coach and other team members
- helping your child handle disappointments and losing
- modeling respectful spectator behavior

EXTREME SPORTS Extreme sports in the early 2000s are becoming increasingly popular among young people. They offer the thrill of facing difficult challenges and overcoming obstacles. Extreme sports get the heart racing and put the body and mind to the test in the face of danger. However, with the many physical and mental benefits of extreme sports comes the risk of injuries. It is essential to work with a trained instructor and use the necessary **safety** equipment when doing any kind of extreme sport.

Extreme sports are not for everyone. However, those looking for bigger challenges in their quest for physical fitness have many options, including rock and ice climbing, surfing, whitewater rafting, wakeboarding, water-skiing, mountain-bike racing, bicycle stunt-riding, skydiving, skateboarding, and extreme snowboarding. There are many camps around the country that teach extreme sports to kids and teenagers. Anyone can find the nearest extreme sports camp or more general information by typing "extreme sports" on any Internet search engine. There are thousands of Web sites devoted to these activities.

Infancy

An infant is capable of participating in only a limited amount of athletic activity. Still, many parents worry about their child's motor skill development and wonder how they can help develop these skills. The American Academy of Pediatrics (AAP) advises parents that normal play with adults is more than enough physical stimulus to encourage normal development of motor skills. In years of research, no one has produced any evidence that increased stimulation of infants increases development of motor skills in later years.

Swimming is perhaps the only sport infants are really able to participate in. While infants instinctively hold their breath when immersed in water, pediatricians warn that they also swallow water, which can produce hazardous side effects. The AAP advises that infants should not participate in swimming activities until they are at least four months old.

Toddlerhood

Toddlers are naturally curious and exploratory, leading them to develop independence skills such as walking and talking. These should be encouraged by adults, as should frequent interaction with other children their own age. Athletic activity at this age should be free form and spontaneous, with adult interference or direction held to a minimum. The AAP suggests that adult intervention, such as teaching a child to throw and catch a baseball, has little effect on later motor skills development, and they warn that the repetition of such practicing often stifles the natural urge to play creatively. It has also been shown that until children reach ages of five to seven, their vision is not sufficiently developed to follow objects that are moving quickly through their line of sight, such as thrown balls.

Preschool

Children are not little adults when it comes to sports and physical activities. As reported in Heidi Splete's article on age-appropriate sports skills, Sally Harris, a pediatrician at the Palo Alto Medical Clinic in Palo Alto, California, asserts that early childhood sports should focus on skill development rather than competitiveness. Activities should allow children to learn by trial and error with minimal instruction. Competition is mostly a distraction for preschool-age children. Appropriate athletic activities for children of this age are dance, beginning gymnastics (primarily tumbling), and swimming. Free-form play with peers is probably most important, both for its socializing effect and for the creative expression it offers.

Sports activity in early childhood should have three basic components, according to Harris. They are acquisition of basic motor skills, social development by the child's interaction with coaches and teammates, and **cognitive development** in understanding and following instructions and executing strategy and tactics.

School age

By the age of five or six, children begin rapidly developing motor skills. Also, posture and balance become automatic, and reaction times become faster. However, learning complex rules is often difficult and trying to teach a child a sport requiring a great deal of instruction, such as baseball, football, or soccer, may only cause frustration and a lack of interest. A child's inability in these areas can also cause a sense of failure and provoke a life-long aversion to organized sports. One good way to get a child interested in sports during these years is to engage in physical activity the whole **family** can participate in, such as taking long walks or bicycle rides. Most pediatricians suggest that complex team sports that require coaching or memorization should be postponed until a child reaches the age of nine or ten. Between the ages of six and nine years, beginning soccer and baseball are appropriate sports, especially if the focus is on getting children interested in sports or physical activity.

By the time a child reaches **adolescence**, his or her interest in sports is most likely at its peak. Children of this age often collect sports memorabilia, wear clothes resembling the uniforms of their favorite players, and spend larger amounts of time watching, participating in, and talking about sports. At ages 10 through 12, children can improve traditional athletic skills and master complex motor skills. They are able to play sports involving strategies and teamwork, but growth spurts can bring physical and emotional changes that parents and coaches should be aware of, according to Harris.

In the last several decades of the twentieth century, there was a dramatic decrease in the number of school districts that require physical education classes for students. As a result, the U.S. Department of Health and Human Services set an objective to increase the number of children six years of age and older who exercise on a daily basis at light to moderate levels for at least 30 minutes.

A 2002 survey of student participation in extracurricular sports activities at middle schools showed a typical program was offered on average 3.6 hours per week. It also revealed that 26.7 percent of boys and 22.9 percent of girls participated in the activities. The most commonly offered activities at middle schools surveyed were basketball (31.7%), track and field (10.3%), soccer (9.4%), tennis (6.7%), and football (5.4%).

Since the middle schools offered a small number of sports activity programs, the survey recommends middle schools add a variety of noncompetitive activities, such as dance, aerobics, martial arts, jogging, walking, and **yoga**. Providing programs that appeal to a wider range of students at all grade levels of middle and high school would likely increase participation in extracurricular sports and physical activity programs.

The social benefits of athletics are especially important for young girls. In fact, it has been argued that girls are more in need of the benefits of athletics than boys. Adolescent girls tend to have lower self-esteem than boys, and many suffer from the false belief that their bodies are useful only to the extent that they are attractive to boys. Statistics compiled by the Women's Sports Foundation also demonstrate that young female athletes receive substantial benefits from participation in sports.

They found that girls who participated in school athletics are 92 percent less likely to use drugs, including tobacco and alcohol; and 80 percent less likely to get pregnant. Additionally, they are three times more likely to graduate from college.

Common problems

The most common problem in adolescent sports is sports-related injuries. An estimated 30 million children in the United States play in organized sports but about 35 percent drop out each year, usually due to physical injury or emotional stress. Each year, hospital emergency rooms see more than 2.6 million sports-related injuries in young people, according to an article in the April 8, 2002 issue of *U. S. News & World Report*.

Among children ages 5 to 14 years, the top **sports injuries** annually are: bicycling, 336,250; basketball, 193,400; football, 185,740; baseball and softball, 117,250; and soccer, 85,430. The number of other sports injuries include skateboarding, 49,930; hockey, 25,400; and gymnastics, 26,950.

Among young people ages 15 to 24 years, the top sports injuries are: basketball, 277,00; football, 171,290; bicycling, 95,720; baseball and softball, 88,340; and soccer, 68,790, according to the article. Other sports injuries included general exercising, 38,560; snowboarding, 29,700; hockey, 28,070; and skateboarding, 27,470.

Parental concerns

The National Athletic Training Association encourages parents to ask questions of coaches when their children become involved in sports. These questions include the following:

- What is the level of the coach's education? Does it include training in **cardiopulmonary resuscitation** (CPR) and first aid?
- What does the coach do when an injury happens? What is the protocol for returning to play following an injury?
- Is there an on-site athletic healthcare provider or consulting team physician? Does the coach knows about any health conditions of the child and have phone numbers where parents can be reached in an emergency?
- Are there emergency medications available for children with **asthma** or allergies?
- What are the inclement weather guidelines, especially for lightning storms and extreme heat?
- Is the athletic equipment safe, properly fitted, and in good condition?
- Are there any supervised preseason and in-season conditioning programs?

When to call the doctor

If a child receives a soft tissue injury, such as a strain or sprain, or a bone injury, the best immediate treatment is ice, compression, elevation of the injury, and rest. Get professional treatment if any injury is severe, such as a fracture, profuse bleeding, dislocated joint, prolonged swelling, or prolonged or severe **pain**. Playing rigorous sports in the heat requires close monitoring of both body and weather conditions. Heat injuries are always dangerous and can be fatal. Children perspire less than adults and require a higher core body temperature to trigger sweating. Heat-related illnesses include **dehydration**, heat exhaustion (**nausea**, **dizziness**, weakness, **headache**, pale and moist skin, heavy perspiration, normal or low body temperature, weak pulse, dilated pupils, disorientation, and fainting spells), and heat stroke (headache, dizziness, confusion, and hot dry skin, possibly leading to blood vessel collapse, coma, and death). Professional medical help should be sought for heat stroke, heat exhaustion, and any other heat-related illnesses that do not quickly clear up.

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Children participating in a basketball game. *(© Tom & Dee Ann McCarthy/Corbis.)*

KEY TERMS

Cardiopulmonary resuscitation (CPR)—An emergency procedure designed to stimulate breathing and blood flow through a combination of chest compressions and rescue breathing. It is used to restore circulation and prevent brain death to a person who has collapsed, is unconscious, is not breathing, and has no pulse.

Cognitive ability—Relating to the process of acquiring knowledge by using reasoning, intuition, or perception.

Dehydration—An excessive loss of water from the body. It may follow vomiting, prolonged diarrhea, or excessive sweating.

Heat exhaustion—A condition of physical weakness or collapse often accompanied by nausea, muscle cramps, and dizziness, that is caused by exposure to intense heat.

Heat stroke—A serious condition that results from exposure to extreme heat. The body loses its ability to cool itself. Severe headache, high fever, and hot, dry skin may result. In severe cases, a person with heat stroke may collapse or go into a coma.

Motor skills—Controlled movements of muscle groups. Fine motor skills involve tasks that require dexterity of small muscles, such as buttoning a shirt. Tasks such as walking or throwing a ball involve the use of gross motor skills.

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Sports injuries

Definition

Sports injuries result from acute trauma or repetitive stress associated with athletic activities. Sports injuries

can affect bones or soft tissue such as ligaments, muscles, and tendons.

Description

Children are more likely to suffer sports injuries than adults since a child's vulnerability is heightened by immature reflexes, an inability to recognize and evaluate risks, and underdeveloped coordination.

In 2002, about 20.3 million Americans suffered a sports injury. The highest rate is among children age 5 to 14 years (59.3 per 1000 people). As many as 20 percent of children who **play** sports get hurt, and about 25 percent of their injuries are classified as serious. Boys age 12 through 17 are the highest risk group. More than 775,000 boys and girls under age 14 are treated in hospital emergency rooms each year for sports-related injuries. Between one half and two thirds of childhood sports injuries occur during practice or in the course of unorganized athletic activity.

Types of sports injuries

About 95 percent of sports injuries are minor soft tissue traumas. The most common sports injury is a bruise (contusion). It is caused when blood collects at the site of an injury and discolors the skin.

Sprains account for one third of all sports injuries. A sprain is a partial or complete tear of a ligament, a strong band of tissue that connects bones to one another and stabilizes joints.

A strain is a partial or complete tear of a muscle (tissue composed of cells that enable the body to move) or a tendon (strong connective tissue that links muscles to bones).

Inflammation of a tendon (tendinitis) and inflammation of one of the fluid-filled sacs that allow tendons to move easily over bones (bursitis) usually result from minor stresses that repeatedly aggravate the same part of the body. These conditions often occur at the same time.

SKELETAL AND BRAIN INJURIES **Fractures** account for 5 to 6 percent of all sports injuries. The bones of the arms and legs are most apt to be broken. Sports activities rarely involve fractures of the spine or skull. The bones of the legs and feet are most susceptible to stress fractures, which occur when muscle strains or contractions make bones bend. Stress fractures are especially common in ballet dancers, long-distance runners, and in people whose bones are thin.

Shin splints are characterized by soreness and slight swelling of the front, inside, and back of the lower leg and by sharp **pain** that develops while exercising and gradually intensifies. Shin splints are caused by overuse or by stress fractures that result from the repeated foot pounding associated with activities such as aerobics, long-distance running, basketball, and volleyball.

A compartment syndrome is a potentially debilitating condition in which the muscles of the lower leg grow too large to be contained within membranes that enclose them. This condition is characterized by **numbness and tingling**. Untreated compartment syndrome can result in long-term loss of function.

Brain injury is the primary cause of fatal sports-related injuries. A **concussion** can result from even minor blows to the head. A concussion can cause loss of consciousness and may affect balance, comprehension, coordination, hearing, memory, and vision.

TREATMENT Treatment for minor soft tissue injuries generally consists of compressing the injured area with an elastic bandage, elevation, ice, and rest.

Anti-inflammatory medications, taken by mouth or injected into the swelling, may be used to treat bursitis. Anti-inflammatory medications and exercises to correct muscle imbalances are often used to treat tendinitis. If the athlete keeps stressing inflamed tendons, they may rupture, and casting or surgery is sometimes necessary to correct this condition. Orthopedic surgery may be required to repair serious **sprains and strains**.

Controlling inflammation as well as restoring normal use and mobility are the goals of treatment for overuse injuries. Athletes who have been injured are usually advised to limit their activities until their injuries are healed. The physician may suggest special exercises or behavior modifications for athletes who have had several injuries. Athletes who have been severely injured may be advised to stop playing completely.

Preschool

Appropriate athletic activities for children of this age are dance, beginning gymnastics (primarily tumbling), and swimming. The most common injuries are sprains and strains of soft tissue such as muscles and tendons.

School age

No matter what the form of specific training or sport activity, stretching and flexibility drills should be included in any pre-participation or warm-up program, even in the very young. Many studies have documented a very low incidence of injury in the total spectrum of

youth sporting endeavors, according to the American Orthopaedic Society for Sports Medicine (AOSSM).

The occurrence of injury in the pre-puberty athlete has been documented as being much lower than in the post-puberty athlete, and lower in post-puberty than in the young adult. This is probably due to the fact that the younger athlete has a lower ratio of kinetic energy to body mass, which means the more immature the physical body, the lower the speed and power.

Since the magnitude of injury is almost always directly related to energy expended in a traumatic event, the younger athlete is less likely to get injured than his older counterpart. The athletic injuries that do occur are usually minor contusions and sprains. Fractures, dislocations, and major ligament injuries can happen but are more common in older age groups. Scientific studies have failed to document a significant increase in injuries to the growth areas of bones in young athletes. Only in extreme cases, such as young gymnasts in intense training for long periods of time, are some athletes at risk for growth plate injuries.

CONTACT VERSUS NON-CONTACT SPORTS The most notable examples of contact sports practiced in the United States are football, ice hockey, wrestling, and basketball. In each of these sports the athlete's body is used to physically control the opponent and, thus, to influence the play of the game. Using the body in this manner creates the opportunity for injury.

The majority of injuries in these contact sports are **bruises** and scrapes. The more significant injuries such as fractures, dislocations, or major ligament damage occur in the post-pubescent athlete. Parents should be responsive to complaints of pain and discomfort from athletes in all age groups and be aware that any athlete who is not playing up to skill level may be suffering from a significant injury.

In non-contact sports, major fractures, dislocations, or soft-tissue injuries are usually associated with accidental rather than intended collisions. Minor sprains, muscle pulls, blisters, and overuse syndrome are commonly seen injuries in non-contact sports, according to the AOSSM.

The overuse syndrome is usually related to sports requiring repetitive, high-stress motion such as tennis, swimming, track, golf, and baseball. Injury occurs as a result of constant repetition of a particular movement. Stress fractures, shin splints, and tendonitis are examples of overuse injuries.

The treatment in each case entails early recognition of the problem, followed by abstinence from competition or at least a decrease or change in training until the affected area is totally symptom free. Training intensity and duration can then increase again. Return to the previous level of training should be gradual and well planned. If the symptoms of overuse persist beyond a few days of rest or if they recur, a physician should evaluate the athlete.

Common problems

Common causes of sports injuries include athletic equipment that malfunctions or is used incorrectly, falls by athletes, forceful high-speed collisions between players, and wear and tear on areas of the body that are continually subjected to stress. Symptoms include instability or obvious dislocation of a joint, pain, swelling, and weakness.

Parental concerns

Every child who plans to participate in organized athletic activity should have an annual pre-season sports physical. This special examination is performed by a pediatrician or **family** physician who carefully evaluates the site of any previous injury, possibly recommends special stretching and strengthening exercises to help growing athletes create and preserve proper muscle and joint interaction, and pays special attention to the cardiovascular and skeletal systems.

Telling the physician which sport the athlete plays helps the physician determine which parts of the body are subjected to the most stress. The physician then is able to suggest to the athlete steps to take to minimize the chance of getting hurt.

Other injury-reducing game plans include:

- being in shape
- knowing and obeying the rules that regulate the activity
- not playing when tired, ill, or in pain
- not using steroids, which can improve athletic performance but cause life-threatening problems
- taking good care of athletic equipment and using it properly
- wearing appropriate protective equipment

When to call the doctor

A physician, pediatrician, sports medicine physician, or orthopedic surgeon should evaluate symptoms that persist, intensify, or reduce the athlete's ability to play without pain. Prompt diagnosis often can prevent

Teenage girl with a knee injury from playing soccer. *(© Tom Stewart/Corbis)*

minor injuries from becoming major problems or causing long-term damage.

A doctor should examine anyone who has the following symptoms:

- people who are prevented from playing by severe pain associated with acute injury
- people whose ability to play has declined due to chronic or long-term consequences of an injury
- people whose injury has caused visible deformities in an arm or leg.

The physician will perform a physical examination, ask how the injury occurred, and what symptoms the patient has experienced. X rays and other imaging studies of bones and soft tissues may be ordered. Anyone who has suffered a blow to the head should be examined immediately, and at five-minute intervals until normal comprehension has returned. The initial examination measures the athlete's awareness, concentration, and short-term memory. Subsequent evaluations of concussion assess **dizziness**, **headache**, **nausea**, and visual disturbances. In most cases, a physician should be consulted for athletes with head injuries.

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Anti-inflammatory—A class of drugs, including nonsteroidal anti-inflammatory drugs (NSAIDs) and corticosteroids, used to relieve swelling, pain, and other symptoms of inflammation.

Bursitis—Inflammation of a bursa, a fluid-filled cavity or sac. In the body, bursae are located at places where friction might otherwise develop.

Cardiovascular—Relating to the heart and blood vessels.

Compartment syndrome—A condition in which the blood supply to a muscle is cut off because the muscle swells but is constricted by the connective tissue around it.

Concussion—An injury to the brain, often resulting from a blow to the head, that can cause temporary disorientation, memory loss, or unconsciousness.

Kinetic energy—The energy that the body has because of its motion.

Repetitive stress injury—An injury resulting from a repeated movement such as typing or throwing a ball.

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Ken R. Wells

Sprains and strains

Definition

A sprain is damage to or tearing of ligaments or a joint capsule. A strain refers to damage to or tearing of a muscle.

Description

When excessive force is applied to a joint, the ligaments that hold the bones together may be torn or damaged. This action results in a sprain, and its seriousness depends on how badly the ligaments are torn. Any joint can be sprained, but the most frequently injured joints are the ankle, knee, and finger.

Strains are tears in the muscle. Sometimes called pulled muscles, they usually occur because of overexertion or improper lifting techniques. Straining the muscles of the back is common.

Demographics

Sprains and strains are common. Anyone can have them. Children under age eight are less likely to have sprains than are older people. Children's ligaments are tighter, and their bones are more apt to break before a ligament tears. People who are active in **sports** suffer more strains and sprains than less active people. However, being overweight and generally inactive also increases the chance of developing a strain or sprain. Repeated sprains in the same joint make the joint less stable and more prone to future sprains.

Causes and symptoms

Any unfamiliar activity that stresses a muscle or joint may cause a strain or sprain. Heavy lifting, falls, and playing a sport without warming up or conditioning are common causes. There are three grades of sprains.

Grade I sprains are mild injuries in which there is no tearing of the ligament and no joint function is lost, although there may be tenderness and slight swelling. Grade II sprains are caused by a partial tear in the ligament. These sprains are characterized by obvious swelling, extensive bruising, **pain**, difficulty bearing weight, and reduced function of the joint. Grade III, or third degree, sprains are caused by complete tearing of the ligament where there is severe pain, loss of joint function, widespread swelling and bruising, and the inability to bear weight. These symptoms are similar to those of bone **fractures**.

Strains can range from mild muscle stiffness to great soreness. Strains result from overuse of muscles, improper use of the muscles, or as the result of injury in another part of the body when the body compensates for pain by altering the way it moves.

When to call the doctor

Parents should call the doctor if their child experiences intense pain and swelling that does not improve within 24 to 48 hours; if their child cannot bear weight on the joint; if the child cannot use the muscle at all; or if there is a popping sensation in the joint when it is moved.

Diagnosis

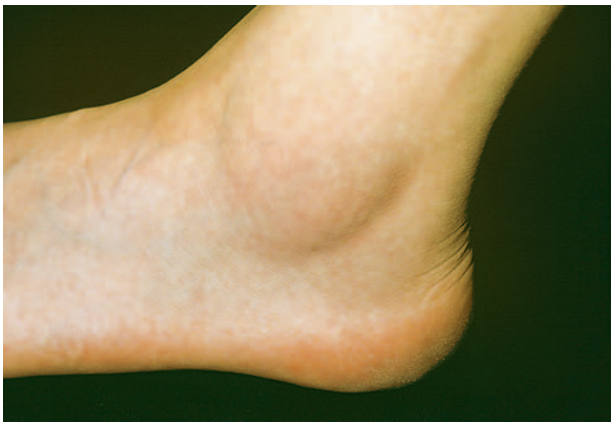
Grade I sprains and mild strains are usually self-diagnosed. Grade II and III sprains are often seen by a physician, who x rays the area to differentiate between a sprain and a fracture. An MRI may be done to look for ruptured ligaments in a joint.

Treatment

Grade I sprains and mild strains can be treated at home. Basic first aid for sprains consists of RICE: Rest; Ice for 48 hours; Compression (wrapping in an elastic bandage); and Elevation of the sprain above the level of the heart. Over-the-counter pain medication such as **acetaminophen** (Tylenol) or ibuprofen (Motrin) can be taken for pain.

In addition to RICE, people with grade II and grade III sprains in the ankle or knee often need to use crutches until the sprains have healed enough to bear weight. Sometimes, physical therapy or home exercises are needed to restore the strength and flexibility of the joint.

Grade III sprains are usually immobilized in a cast for several weeks to see if the sprain heals. Pain medication is prescribed. Surgery may be necessary to relieve pain and restore function. Athletic people under age 40 are the most likely candidates for surgery, especially with grade III knee sprains. For complete healing, physical therapy usually follows surgery.



Swelling is a symptom of a sprained ankle. *(© Dr. P. Marazzi/ Photo Researchers, Inc.)*

Alternative treatment

Alternative practitioners endorse RICE and conventional treatments. In addition, nutritional therapists recommend vitamin C and bioflavonoids to supplement a diet high in whole grains, fresh fruits, and vegetables. Anti-inflammatories such as bromelain (a proteolytic enzyme from pineapples) and tumeric (*Curcuma longa*) may also be helpful. The homeopathic remedy arnica (*Arnica montana*) may be used initially for a few days, followed by ruta (*Ruta graveolens*) for joint-related injuries or Rhus toxicodendron for muscle-related injuries. If surgery is needed, alternative practitioners can recommend pre- and post-surgical therapies that enhance healing.

Prognosis

Moderate sprains heal within two to four weeks, but it can take months to recover from severe ligament tears. Until the early 2000s, tearing the ligaments of the knee meant the end to an athlete's career. Subsequent improved surgical and rehabilitative techniques offer the possibility of complete recovery. However, once a joint has been sprained, it never is as strong as it was before.

Prevention

Sprains and strains can be prevented by warming-up before exercising, using proper lifting techniques, wearing properly fitting shoes, and taping or bracing the joint.

KEY TERMS

Ligament—A type of tough, fibrous tissue that connects bones or cartilage and provides support and strength to joints.

Parental concerns

Parents should be aware that repeated spraining of a joint weakens it. It may be necessary for the child to do exercises to strengthen the joint after a serious sprain. Parents should allow plenty of time for strains and sprains to heal before allowing their child to return to strenuous athletics.

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Tish Davidson, A.M.

Sprue *see* **Celiac disease**

SSRIs *see* **Antidepressants**

Stanford-Binet intelligence scales

Definition

The Stanford-Binet **intelligence** scale is a standardized test that assesses intelligence and cognitive abilities in children, beginning at age two, and in adults.

Purpose

The Stanford-Binet intelligence scale is used as a tool in school placement, in determining the presence of a learning disability or a **developmental delay**, and in tracking intellectual development. In addition, it is sometimes included in neuropsychological testing to assess the brain function of individuals with neurological impairments.

Description

The Stanford-Binet intelligence scale is a direct descendent of the Binet-Simon scale, the first intelligence scale created in 1905 by psychologist Alfred Binet (1857–1911) and Theophilus Simon. Lewis Terman (1877–1956) published the Stanford-Binet scale initially in 1916. As of 2004, the scale had been revised five times—in 1937, 1960 (with a scoring change of this version in 1973), 1986, and 2003.

Beginning with the fourth revision (1986), the test underwent design changes to include a larger, more diverse, representative sample in order to minimize the gender and racial inequities that had been criticized in earlier versions of the test. Originally designed for children only, with the fifth edition (2003) the Stanford-Binet can be used on anyone older than two years of age.

The Stanford-Binet scale tests intelligence across six areas: general intelligence, knowledge, fluid reasoning, quantitative reasoning, visual-spatial processing, and working memory. These areas are covered by ten subtests that include activities measuring both verbal and non-verbal intelligence. Activities include verbal absurdities, picture absurdities, verbal analogies, form patterns, procedural knowledge, sentence and word memory, position and direction, early reasoning, and quantitative reasoning.

All test subjects take two initial routing tests: a vocabulary test and a matrices test (which assesses non-verbal reasoning). The results of these tests, along with the subject's age, determines the number and level of subtests to be administered.

Total testing time is around 45 to 60 minutes, depending on the child's age and the number of subtests given. Raw scores are based on the number of items answered and are converted into a standard age score corresponding to age group, similar to an IQ measure.

Precautions

Intelligence testing requires a clinically trained examiner. The Stanford-Binet intelligence scale should

Teenage girl taking an intelligence test. *(© Lew Merrim/ Science Source/Photo Researchers, Inc.)*

KEY TERMS

Norms—A fixed or ideal standard; a normative or mean score for a particular age group.

Representative sample—A random sample of people that adequately represents the test-taking population in age, gender, race, and socioeconomic standing.

Standard deviation—A measure of the distribution of scores around the average (mean). In a normal distribution, two standard deviations above and below the mean includes about 95% of all samples.

Standardization—The process of determining established norms and procedures for a test to act as a standard reference point for future test results.

be administered, scored, and interpreted by a trained professional, preferably a psychologist.

Children with physical disabilities may require certain accommodations when taking the test, such as extra time for tasks, rest breaks, or instructions received in an alternate format (e.g., signing for a deaf child). The examiner should be made aware of a child's potential limitations before the day of the test so that appropriate accommodations are available.

Normal results

Scoring for the Stanford-Binet generates a verbal IQ score (VIQ), a non-verbal IQ score (NIQ), and a full-scale IQ (FSIQ). It is a standardized test, meaning that norms are established during the design phase of the test by administering the test to a large, representative sample of the test population (in the case of the fifth edition, data from the 2000 U.S. census were used). The test has a mean, or average, standard score of 100 and a standard deviation of 15 for composite scores (subtests have a mean of 10 and a standard deviation of 3). The standard deviation indicates how far above or below the norm the subject's score is. For example, an eight-year-old is assessed with the Stanford-Binet scale and achieves a standard age score of 115. The mean score of 100 is the average level at which all eight-year-olds in the representative sample performed. This child's score would be one standard deviation above that norm.

While standard age scores provide a reference point for evaluation, they represent an average of a variety of skill areas. A trained psychologist evaluates and interprets an individual's performance on the scale's subtests to discover strengths and weaknesses and offer recommendations based upon these findings.

Parental concerns

Test **anxiety** can have a negative impact on a child's performance, so parents should attempt to take the stress off their child by making sure they understand that it is the effort and attention they give the test, not the final score, that matters. Parents can also ensure that their children are well-rested on the testing day and have a nutritious meal beforehand.

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Paula Ford-Martin

Staphylococcal infections

Definition

Staphylococcal (staph) infections are communicable diseases caused by certain bacteria and generally characterized by the formation of abscesses. They are the leading cause of primary infections originating in hospitals in the United States.

Description

Classified since the early twentieth century as among the deadliest of all disease-causing organisms, staphylococcal bacteria exist on the skin or inside the nostrils of 20 to 30 percent of healthy people. It is sometimes found in breast tissue, the mouth, and the genital, urinary, and upper respiratory tracts.

Staph bacteria are usually harmless; however, when an injury or a break in the skin enables the organisms to invade the body and overcome the body's natural defenses, consequences can range from minor discomfort to death.

Demographics

Infection is most apt to occur in newborns; children whose immune systems have been undermined by radiation treatments, **chemotherapy**, or medication; those with surgical incisions or skin disorders; and among people with serious illnesses such as **cancer**, diabetes, and lung disease. Children are also more at risk for staph infections if they have HIV/AIDS or other diseases that compromise immune function, have a catheter or implanted prosthetics, are hospitalized, have open **wounds**, or live in close quarters with a large group of others. It is not clear exactly how many staph infections occur each year, but it is the most common infection that begins in the hospital.

Causes and symptoms

Staph infections produce pus-filled pockets (abscesses) located just beneath the surface of the skin or deep within the body. A localized staph infection is confined to a ring of dead and dying white blood cells and bacteria. The skin above it feels warm to the touch. Most of these abscesses eventually burst, and pus that leaks onto the skin can cause new infections.

A small fraction of localized staph infections enter the bloodstream and spread through the body. In children, these systemic (affecting the whole body) or disseminated infections frequently affect the ends of the long bones of the arms or legs, causing a bone infection called osteomyelitis. Other abscesses that can develop from staph infection include those of the brain, heart, kidneys, liver, lungs, or spleen.

Staphylococcus aureus

Named for the golden color of the bacteria grown under laboratory conditions, *Staphylococcus aureus* is a hardy organism that can survive in extreme temperatures or other inhospitable circumstances. About 70 to 90 percent of the population carry this type of staph in their nostrils at some time in their lives. Although present on the skin of only 5 to 20 percent of healthy people, as many as 40 percent of individuals carry it elsewhere, such as in the throat, vagina, or rectum, for varying periods of time, from hours to years, without developing symptoms or becoming ill.

S. aureus flourishes in hospitals, where it infects healthcare personnel and infects patients who have had surgery, have open wounds, have acute **dermatitis**, insulin-dependent diabetes, or dialysis-dependent kidney disease, or who receive frequent allergy-desensitization injections. Staph bacteria can also contaminate bedclothes, catheters, and other objects.

S. aureus causes a variety of infections. Boils and inflammation of the skin surrounding a hair shaft (folliculitis) are the most common. Toxic shock (TSS) and scalded skin syndrome (SSS) are among the most serious.

TOXIC SHOCK **Toxic shock syndrome** is a life-threatening infection characterized by severe **headache**, **sore throat**, **fever** as high as 105°F (40.5°C), and a sunburn-like rash that spreads from the face to the rest of the body. Symptoms appear suddenly. They also include **dehydration** and watery **diarrhea**.

Inadequate blood flow to peripheral parts of the body (shock) and loss of consciousness occur within the first 48 hours. Between the third and seventh day of illness, skin peels from the palms of the hands, soles of the feet, and other parts of the body. Kidney, liver, and muscle damage often occur.

SCALDED SKIN SYNDROME Rare in adults and most common in newborns and other children under the age of five, scalded skin syndrome originates with a localized skin infection. A mild fever and/or an increase in the number of infection-fighting white blood cells may occur.

A bright red rash spreads from the face to other parts of the body and eventually forms scales. Large, soft blisters develop at the site of infection and elsewhere. When they burst, they expose inflamed skin that looks as if it had been burned.

MISCELLANEOUS INFECTIONS *S. aureus* can also cause the following:

- arthritis
- bacteria in the bloodstream (bacteremia)
- pockets of infection and pus under the skin (carbuncles)
- tissue inflammation that spreads below the skin, causing **pain** and swelling (cellulitis)
- inflammation of the valves and walls of the heart (endocarditis)
- inflammation of tissue that encloses and protects the spinal cord and brain (meningitis)
- inflammation of bone and bone marrow (osteomyelitis)
- pneumonia

Other strains of staphylococci

S. EPIDERMIDIS Capable of clinging to tubing (such as that used for intravenous feeding), prosthetic devices, and other non-living surfaces, *S. epidermidis* is the organism that most often contaminates devices that provide direct access to the bloodstream.

The primary cause of bacterial infection in hospital patients, this strain of staph is most likely to infect cancer patients, whose immune systems have been compromised and high-risk newborns receiving intravenous supplements.

S. epidermidis also accounts for two of every five cases of prosthetic valve endocarditis. Prosthetic valve endocarditis is inflammation that occurs as a complication of the implantation of an artificial valve in the heart. Although contamination usually occurs during surgery, symptoms of infection may not become evident until a year after the operation. More than half of the patients who develop prosthetic valve endocarditis die.

Causes and symptoms

Staph bacteria can spread through the air, but infection is almost always the result of direct contact with open sores or body fluids contaminated by these organisms. Staph bacteria often enter the body through inflamed hair follicles or oil glands. Or they penetrate skin damaged by **burns**, cuts and scrapes, infection, insect **bites**, or wounds.

Multiplying beneath the skin, bacteria infect and destroy tissue in the area where they entered the body. Staph infection of the blood (staphylococcal bacteremia) develops when bacteria from a local infection infiltrate the lymph glands and bloodstream. These infections, which can usually be traced to contaminated catheters or intravenous devices, cause persistent high fever. They may cause shock. They also can cause death within a short time.

When to call the doctor

The following are common symptoms of staph infection:

- pain or swelling around a cut or an area of skin that has been scraped
- boils or other skin abscesses
- blistering, peeling, or scaling of the skin (This symptom is most common in infants and young children.)
- enlarged lymph nodes in the neck, armpits, or groin

A **family** physician should be notified whenever the following symptoms are present:

- Lymph nodes in the neck, armpits, or groin become swollen or tender.
- An area of skin that has been cut or scraped becomes painful or swollen, feels hot, or produces pus. These symptoms may mean the infection has spread to the bloodstream.

- A boil or carbuncle appears on any part of the face or spine. Staph infections affecting these areas can spread to the brain or spinal cord.
- A boil becomes very sore. Usually a sign that infection has spread, this condition may be accompanied by fever, chills, and red streaks radiating from the site of the original infection.
- Boils develop repeatedly. This type of recurrent infection could be a symptom of diabetes.

Diagnosis

Blood tests that show unusually high concentrations of white blood cells can suggest staph infection, but diagnosis is based on laboratory analysis of material removed from pus-filled sores and on analysis of normally uninfected body fluids such as blood and urine. Also, x-rays can enable doctors to locate internal abscesses and estimate the severity of infection. Needle biopsy (removing tissue with a needle, then examining it under a microscope) may be used to assess if any bones are infected.

Treatment

Superficial staph infections can generally be cured by keeping the area clean, using soaps that leave a germ-killing film on the skin, and applying warm, moist compresses to the affected area for 20 to 30 minutes three or four times a day.

Severe or recurrent infections may require a seven to 10 day course of treatment with penicillin or other oral **antibiotics**. The location of the infection and the identity of the causal bacteria determine which of several effective medications should be prescribed.

In case of a more serious infection, antibiotics may be administered intravenously for as long as six weeks. Intravenous antibiotics are also used to treat staph infections around the eyes or on other parts of the face.

Surgery may be required to drain or remove abscesses that form on internal organs or on shunts or other devices implanted inside the body.

Alternative treatment

Alternative therapies for staph infection are meant to strengthen the immune system and prevent recurrences. Among the therapies believed to be helpful for the person with a staph infection are **yoga** (to stimulate the immune system and promote relaxation), acupuncture (to draw heat away from the infection), and herbal remedies. Herbs that may help the body overcome, or withstand, staph infection include the following:

- Garlic (*Allium sativum*). This herb is believed to have antibacterial properties. Herbalists recommend consuming three garlic cloves or three garlic oil capsules a day, starting when symptoms of infection first appear.
- Cleavers (*Galium aparine*). This anti-inflammatory herb is believed to support the lymphatic system. It may be taken internally to help heal staph abscesses and reduce swelling of the lymph nodes. A cleavers compress can also be applied directly to a skin infection.
- Goldenseal (*Hydrastis canadensis*). Another herb believed to fight infection and reduce inflammation, goldenseal may be taken internally when symptoms of infection first appear. Skin infections can be treated by making a paste of water and powdered goldenseal root and applying it directly to the affected area. The preparation should be covered with a clean bandage and left in place overnight.
- Echinacea (*Echinacea* spp.). Taken internally, this herb is believed to have antibiotic properties and is also thought to strengthen the immune system.
- Thyme (*Thymus vulgaris*), lavender (*Lavandula officinalis*), or bergamot (*Citrus bergamot*) oils. These oils are believed to have antibacterial properties and may help to prevent the scarring that may result from skin infections. A few drops of these oils are added to water and then a compress soaked in the water is applied to the affected area.
- Tea tree oil (*Melaleuca* spp.). Another infection-fighting herb, this oil can be applied directly to a boil or other skin infection.

Prognosis

Most healthy people who develop staph infections recover fully within a short time. Others develop repeated infections. Some become seriously ill, requiring long-term therapy or emergency care. A small percentage die.

Doctors and researchers are becoming increasingly concerned about staph infections that are resistant to antibiotics. A bacterium that is considered resistant is one that can no longer be treated effectively using the antibiotics that are commonly prescribed for that type of infection. Resistant staph infections can usually be treated effectively with other antibiotics. Children who are most at risk for resistant staph infections are those who have been in the hospital or have serious underlying medical conditions. According to the Centers for Disease Control, although it is not clear how many cases of resistant staph infections occur each year, they are thought to be very rare. They recommend

KEY TERMS

Abscess—A localized collection of pus in the skin or other body tissue caused by infection.

Endocarditis—Inflammation of the inner membrane lining heart and/or of the heart valves caused by infection.

treating all infections promptly and only prescribing antibiotics when there is an underlying bacterial cause for the disease (antibiotics are not effective against viruses) to help reduce the occurrence of bacteria becoming resistant to antibiotics.

Prevention

Healthcare providers and patients should always wash their hands thoroughly with warm water and soap after treating a staph infection or touching an open wound or the pus it produces. Pus that oozes onto the skin from the site of an infection should be removed immediately. This affected area should then be cleansed with antiseptic or with antibacterial soap.

To prevent infection from spreading from one part of the body to another, it is important to shower rather than bathe during the healing process. Because staph infection is easily transmitted from one member of a household to others, towels, washcloths, and bed linens used by someone with a staph infection should not be used by anyone else. They should be changed daily until symptoms disappear, and laundered separately in hot water with bleach. Children should frequently be reminded not to share brushes, combs, or hair accessories.

Parental concerns

Staph infections are most likely to occur after a child has had surgery or a wound of some kind. A good way to help prevent staph infections of wounds is to keep the wound clean and dry. Children who have staph infections, especially skin infections, should be kept away from others whom they are likely to infect, and their bedding, clothes, and other things that may have touched the wound should be cleaned with hot soapy water and bleach.

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Tish Davidson, A.M.
Maureen Haggerty

STDs *see* **Sexually transmitted diseases**

Stealing

Definition

Stealing is taking another person's property without permission.

Description

Stealing is taking someone's property without permission. Very young children do not understand the concept of personal property. When they see something they want, they simply take it. Young children generally take things for immediate use only, whereas older children will take them "for keeps." Since they have no sense of personal property, young children should not be accused of stealing when they take another person's things without permission. However, the concept of stealing should be explained right from the start, even before the child can understand. If a parent, teacher, or other adult simply tells the child, "Don't take Sally's crayon," the child will believe only that taking Sally's crayon is wrong, while taking a crayon from Juan, or a cookie from Sally, is okay. A child must be told repeatedly that taking other people's things is wrong in order to develop an understanding of the broader concept of stealing.

Most children have a basic sense of "mine" and "not mine" by the age of two and can therefore begin to learn respect for other people's possessions. However, a true understanding of the harmful nature of stealing does not begin to develop until about age five to seven. At this age, children are deterred from stealing mostly by their **fear** of parental disapproval. Internal motivations of con-

science and guilt do not develop until the middle childhood years. Once the recognition of property boundaries develops, stealing becomes an intentional act that must be addressed more deliberately.

Children steal for a number of reasons. Young children, or older children who have not developed sufficient self-control, may steal to achieve instant gratification when an object cannot be obtained immediately by honest means. Older children may steal to gain a sense of power, to acquire status with peers who resist authority, to get attention, to take revenge on someone who has hurt them, to alleviate boredom, or to vent unresolved feelings of anger or fear. Children who steal are often expressing displaced feelings of **anxiety**, rage, or alienation resulting from a disruption in their life, such as a parent's **divorce** or remarriage.

People who feel excluded or disconnected from society have fewer qualms about stealing, because they have less sense of respect, trust, or responsibility in relation to the community. They may even purposely steal in retaliation for the **pain** they feel society has inflicted on them. Studies have shown a direct correlation between stealing and alienation. Community-building programs in U.S. high schools have greatly reduced the incidence of theft by developing a sense of unity among the students and faculty. When a child feels integrated into a community, he or she is more likely to support all members of that community. Stealing becomes less tempting in a mutually supportive environment.

A child who is caught stealing for the first time should be treated compassionately; the focus should be on the reason(s) for the act rather than on the act itself. Parents, teachers, or other adult caregivers need to discern if the child lacks self-control, is angry (and with whom), needs attention, is bored, feels pressured by peers to cross boundaries, feels alienated from the community, has poor **self-esteem**, or needs to develop more positive moral values. A habitual stealer is expressing a serious internal problem that needs close attention. Children at risk of becoming habitual stealers often times have the following characteristics: low self-esteem; strong desires and weak self-control (impulsiveness); a lack of sensitivity to others; are angry, bored, or feel disconnected; spend a great deal of time alone; have recently experienced a significant disruption in their lives. Stealing is a behavior problem, not a character problem. The behavior can be corrected if the underlying difficulty is resolved.

Preschool

Children under the age of five generally are not sufficiently able to understand the concept of property to realize that they are stealing. Even though they might not understand, parents of children this age should make the child give back whatever was stolen and should explain why stealing is bad and how it hurts other people. The child should not be labeled bad, but the lesson should be made clear that stealing is wrong.

Elementary school

Children in elementary school generally are developed enough to understand that stealing is wrong and why it is wrong. When elementary school children steal, it is generally because they have seen something that they want, and they lack well-developed self-control. Children in this age group who are caught stealing should be made to take the item back or should be made to find ways to make enough money to pay for what they have stolen. Usually if a parent or other adult forces the child to apologize to the person from whom they stole, the embarrassment is enough to deter repeated episodes of stealing.

Middle and high school

Older children steal for different reasons than younger children. They want to feel powerful or want something expensive to try to keep up with their peers, or they may be distressed about a situation at home. Or they may want to fit in with a group. One fourth of all people caught shoplifting are between the ages of 13 and 17. In most cases children outgrow this behavior, but it still needs to be dealt with in a serious manner. Children who steal are not necessarily delinquents; however, children over the age of 15 who steal may have serious underlying troubles that need to be dealt with by a mental health professional.

Common problems

Though children who steal do so for a number of different reasons, stealing should always be treated seriously. If there is an underlying cause, such as unhappiness at home, then resolving the underlying problem usually resolves the stealing behavior, although the stealing itself should never be ignored.

Parental concerns

Just because a child has stolen does not mean he or she is going to grow up to live a life of crime. Children who steal are often helpful around the house, get good grades, and are otherwise good kids. Stealing, or a suspicion of stealing, needs to be dealt with in a serious manner, but once the matter has been dealt with, it should not

be brought up again. In this way the child has a chance to start over with a clean slate.

When to call the doctor

If stealing is accompanied by other problems, such as difficulty interacting with peers or poor grades, it may be a sign of a serious underlying problem. If a child steals after the age of 15 or has gotten caught stealing more than once or there is a suspicion of underlying emotional or drug problems that might be causing the stealing, a mental health professional should be consulted.

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Steinert's disease *see* **Mytonic dystrophy**

Stepfamilies

Definition

A stepfamily is formed by the marriage or long-term cohabitation of two individuals, when one or both have at least one child from a previous relationship living part-time or full-time in the household. The individual who is not the biological parent of the child or children is referred to as the stepparent. Stepfamilies are also called blended families.

Purpose

Stepfamilies merge unrelated parents and children into a **family** unit that, with time and emotional work, can function as effectively as a traditional nuclear family. For children previously living in a single-parent family, a stepfamily can provide a more structured family environment with positive influences from two parental figures. For parents, a stepfamily can provide social support for new couples and new, emotionally rewarding relationships with biological and stepchildren.

Description

A stepfamily is a family unit in which one or both adult partners have children from a previous relationship. Stepfamilies can be formed after a **divorce** or death of a parent in a nuclear family or when a single parent chooses a long-term partner. Although in the past, marriage was usually required to define a stepfamily, marriage is not always a prerequisite for parents and children living together in the same household. Many adult partners choose to live together (cohabitation) on a long-term basis rather than marry. Children can be full-time or part-time members of a stepfamily, depending on the custody arrangement between the biological parents. Children may also be part of two stepfamilies if both parents remarry. The following terms are used to define members of a stepfamily:

- stepparent: a non-biological parent
- stepchild: a non-biological child brought into the family by marriage or cohabitation with the biological parent
- stepsiblings (stepbrother, stepsister): siblings who are not related biologically, whose parents are married to each other or cohabiting long-term
- custodial parent: the biological parent awarded primary custody by a court during divorce proceedings
- non-custodial parent: the biological parent awarded part-time custody or visitation rights by a court during divorce proceedings
- half-siblings: children who share biologically one parent
- stepgrandparents: non-biological grandparents
- mutual child: a biological child of the remarried or cohabiting couple

There are key differences between the dynamics in a stepfamily and the dynamics of a first-time nuclear family:

- Stepfamilies ultimately result from a loss, death of a parent/spouse, divorce, end of a long-term relationship, changes in lifestyle (e.g., moving, loss of job), and, therefore, involve grief on the part of both parents and children. This grief may remain unresolved and affect stepfamily relationships.
- Children in stepfamilies are members of two households and, as a result, may experience confusion, **discipline** issues, loss of stability, and conflicting feelings of loyalty.
- The role of the stepparent and status in the family is often unclear with regard to authority, level of involvement with the stepchild, and discipline. In addition, no legal relationship exists between stepparents and stepchildren.
- Stepparents must assume parental roles before there is an emotional bond with the stepchild and are often required to make instant adjustments to a parental role. In contrast, biological parents bond with their child as the child grows.
- Stepfamilies must cope with outside influences and ongoing change due to issues with the other biological parent and family members.

According to statistics from the United States Census Bureau and the Stepfamily Foundation, one in three Americans is involved in a stepfamily situation, and 1,300 new stepfamilies form each day. In addition, 50 percent of children under age 13 as of 2004 lived with one biological parent and the parent's partner. As of 2004, it is estimated that there are more stepfamilies than traditional nuclear families in the United States. The number of stepfamilies is underestimated because the U.S. Census Bureau did not as of 2004 recognize that a child can be a member of two stepfamilies; only the household where the child lives the majority of the time is counted. Because in most divorces, primary custody is awarded to the biological mother, most stepfamilies involve stepfathers who become the full-time stepparent. In rare cases, a biological father is awarded primary custody, and a stepmother can become a full-time stepparent.

Precautions

Stepfamilies are increasingly referred to as blended families, by the media and others. Stepfamily researchers, family therapists, and the Stepfamily Association of America (SAA) view this term as inaccurate because it infers that members of a stepfamily blend into an entirely new family unit, losing their individuality and attachment to other outside family members. The term stepfamily is preferred because the derivation of the prefix "step-" originates from the Old English word "steop-" which means "bereave." The term stepchild used to refer to orphans who lost their parents, and stepfather/stepmother used to refer to individuals who became parents to an orphan. Because other family types (biological, single-parent, foster, adoptive) are defined by the parent-child relationship, the SAA believes that the term stepfamily more accurately reflects that relationship and is consistent with other family definitions. Viewing the stepfamily as a blended family can lead to unrealistic expectations, confused and conflicted children, difficult adjustment, and in many cases, failure of the marriage and family.

Preparation

Divorce, remarriage, and the formation of a stepfamily are traumatic events for children. Transition can be eased by including children in discussions and preparations for the stepfamily's future. For example, for couples getting remarried, children can be included in the actual wedding ceremony (not just as ringbearers and flower girls) and given tokens, like a piece of jewelry or special gift (like the wedding rings that their parents exchange), that symbolize the joining of the new family.

Individual therapy for children whose parents are going through a divorce and remarriage can be helpful. Group **family therapy** with all members of the stepfamily can help identify issues that may undermine successful family functioning. Because grandparents can influence stepfamily dynamics, educating stepgrandparents about stepfamily issues can also help. Roles of the noncustodial parent and stepparent must be clearly defined to avoid unnecessary conflicts. Reading information on stepfamilies and joining a stepfamily support group can help ensure future success. With cooperation and understanding among stepfamily members, a stepfamily can function successfully and even heal emotional scars of past divorce.

Risks

A National Institutes of Health (NIH) study of stepfamilies found that a stepfamily has a unique natural life cycle, takes several years to develop into a family unit, and is at greatest risk for failure during its first two years. According to U.S. Census Bureau statistics, the average marriage in the United States only lasts seven years, and one of every two marriages ends in divorce. Stepfamilies are at greater risk for failure and broken marriage due to the increased stresses of stepfamily life. These stresses include the unclear role and authority of the stepparent,

financial responsibility for stepchildren, conflict between custodial and noncustodial parents, and emotional tensions.

A study by British and Canadian researchers found that children in stepfamilies and **single-parent families** had more behavioral and emotional problems compared with children in intact biological families and that stresses within the family were more influential than family type in contributing to children's psychological problems. Adolescents are especially vulnerable to psychological and emotional problems resulting from a combination of **puberty** and family stresses. Medical professionals, such as pediatricians, psychologists, and therapists, can provide resources and referrals for adolescents requiring treatment and/or therapy for depression, oppositional defiance disorder, and unresolved feelings of anger, resentment, and loss.

Parental concerns

While stepmothers face some of the same issues that stepfathers face, both part-time and full-time stepmothers have a more difficult role in the stepfamily and are often expected to be more involved with their stepchild due to socialization pressures (being a mother), societal expectations, and expectations from their husband. Joining a stepmother support group can be helpful in working out frustrations and problems in the stepmother role.

Children in stepfamilies are subject to multiple parental influences and may become confused and conflicted about how they fit into each family and which parent is responsible for discipline. All parents—biological and stepparents—should strive to work out such issues for the benefit of their children. Minimizing conflicts between all parents can help children adjust to stepfamily life.

For various reasons, society does not always view stepparents as having the same responsibilities as biological parents. Employers, other family members, friends, and neighbors may have difficulty understanding and relating to stepfamily issues. One workplace psychologist estimates that businesses in the United States lose more than $10 billion annually due to problems related to stepfamily issues, working parents, and other marital stresses. Although many employers do offer employee assistance programs with substance abuse counseling, child care, and family/marriage counseling, divorced parents, working stepparents, and working live-in partners rarely seek counseling.

Parents and stepparents should be concerned during the first two years after the stepfamily is formed, since this has been identified as a crucial time period for stepfamily success. To help strengthen the stepfamily, parents can establish new and enjoyable family traditions, recognize that children need to stay in touch with noncustodial parents, and focus on being open with family communication. Organizations such as the Stepfamily Association of America offer resources and ideas for building stepfamily bonds, such as celebrating National Stepfamily Day every September and engaging in pleasurable family activities, like movie and pizza night.

KEY TERMS

Cohabitation—Sexual partners living together outside of marriage.

Nuclear family—The basic family unit, consisting of a father, a mother, and their biological children.

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Jennifer E. Sisk, M.A.

Steroids, anabolic *see* **Anabolic steroids**

Stimulant drugs

Definition

Stimulant drugs are drugs that excite the central nervous system.

Description

There are several drugs used as stimulants. Although in large part they share the same properties, their use is determined by how well they are absorbed from the gastrointestinal tract. These drugs are related to the body's normal stimulant hormones epinephrine and norepinephrine.

- Injectable stimulants are used to stimulate the heart or breathing. Epinephrine (adrenalin) is the most common.
- Topical stimulants are used as **decongestants**, since they cause blood vessels to contract. They are also used to stop superficial bleeding by contracting the capillaries and for relief of **conjunctivitis**. They may be applied to the skin, inhaled, or applied in the form of drops as nose drops or eye drops.
- Oral stimulants, including the two drugs in this class (**methylphenidate** [Ritalin] and amphetamine) are used to treat extreme daytime sleepiness also known as **narcolepsy** and for their calming effect in attention-deficit hyperactivity disorder (ADHD).
- Caffeine, a stimulant found in foods and drinks, is used to promote wakefulness and alertness.

The orally active stimulants were formerly used as an aid to dieting but were of little value for this purpose. They may still be used in the most extreme cases of **obesity** but are no longer routinely prescribed for this purpose. Some were widely used as decongestants for colds and **allergies**. They are subject to abuse, and amphetamines and methylphenidate are controlled substances in the United States.

Pemoline (Cylert) is also a member of this class but is rarely used because of its potential for causing severe liver problems. This drug should be reserved for treatment of children whose ADHD cannot be controlled with either first or second line drug therapy and whose condition is so severe that the potential benefits justify the risk.

Stimulant drugs, in addition to their proper medicinal use, are subject to abuse. The drugs commonly abused are methylphenidate, amphetamine, and methamphetamine. A related drug, 3,4-methylenedioxymethamphetamine (better known as ecstasy or MDMA), is also widely abused. Unlike methylphenidate and amphetamine, MDMA has no legitimate therapeutic use.

Cocaine is chemically different from the traditional stimulants but provides similar effects. It is used medicinally as a local anesthetic but is not available for self-administration. Cocaine has become a major drug abuse problem.

General use

The most common use of methylphenidate and amphetamine in children is for control of **attention-deficit/hyperactivity disorder**. This is a condition marked by general restlessness, excessive activity, and inability to concentrate on a topic. Children who have this problem are unable to concentrate on schoolwork and fall behind their classmates. They are frequently disruptive. For this condition, the stimulants have a reverse activity and have a calming, rather than a stimulating effect.

Precautions

Stimulant drugs are subject to abuse and development of tolerance. This does not appear to be a problem, however, when the drugs are appropriately used for a proper diagnosis of ADHD.

When used to treat young children, there is some evidence that stimulant drugs reduce the rate of growth. This may be made up for by a growth spurt when the drugs are discontinued.

Stimulant drugs increase blood pressure.

Side effects

The side effects for stimulant drugs are different when they are used as stimulants and when they are used for their calming effect in ADHD. The effects listed below are those seen when amphetamines and/or methylphenidate are used to treat attention-deficit hyperactivity disorder:

- reduction in rate of growth
- exacerbation of related problems such as Tourette's disorder
- appetite suppression

There are many additional side effects seen when amphetamines or methylphenidate are used or abused for their stimulant properties, but these effects are not normally seen when the stimulants are used for a proper diagnosis of ADHD and dosed appropriately.

Some of the adverse effects that may result from stimulant abuse are increased wakefulness, increased physical activity, decreased appetite, increased respiration, high **fever**, euphoria, irritability, insomnia, confusion, tremors, convulsions, **anxiety**, paranoia, and aggressiveness. The high fever and convulsions may be fatal. Long term abuse of stimulants may result in permanent brain damage that causes involuntary, Parkinson-like movements.

Methamphetamine causes increased heart rate and blood pressure and can cause irreversible damage to blood vessels in the brain, producing strokes. Other effects of methamphetamine include respiratory problems, irregular heartbeat, and extreme anorexia. Its use can result in cardiovascular collapse and death.

The reports of growth suppression associated with amphetamines and methylphenidate are not definitive but appear to be valid. This growth suppression is balanced by a growth spurt when the drugs are discontinued. For this reason, stimulants should only be administered during school hours and discontinued during summer and holiday periods.

Interactions

Stimulant drugs have no interactions with drugs that are normally prescribed for children.

Parental concerns

When used to treat ADHD, methylphenidate and amphetamines do not have the adverse effects associated with these drugs when they are abused.

ADHD is a difficult diagnosis and may be confused with normal childhood energy. A diagnosis should be made, and drug therapy initiated, only by a qualified professional experienced in this condition.

Because of the potential for abuse, methylphenidate and amphetamines must be kept out of reach of children, particularly visitors and older siblings of a child being treated for ADHD.

Because of the risk of growth suppression, stimulant drugs should only be administered during school periods. They should not be used to calm an active child for the convenience of parents or babysitters.

Children who fail a trial of one stimulant may respond to another drug in the same class. A child who does poorly on methylphenidate may respond to amphetamines and vice versa.

Approximately 15 to 30 percent of children with ADHD have underlying Tourette's disorder, a condition marked by vocal and motor **tics**. Starting treatment with methylphenidate or amphetamines may unmask the condition, and the tics will become apparent. This is not an effect of the drug, but rather a consideration of the underlying problem.

There is some dispute over the lowest age at which stimulant therapy may be appropriately started, but it seems agreed that these drugs should not be used to treat children under the age of three years.

Sometimes, drugs which are properly prescribed for ADHD are diverted and used by other children as recreational drugs. If a child who has been well stabilized on stimulants for ADHD begins to get worse, consider the possibility that the drugs are being sold to others, rather than being used therapeutically. Stimulant drugs should be administered by a parent, guardian, school nurse, or other responsible person. This will both guard against diversion and assure that children are not forgetting to take their medication.

The effects of cocaine are generally similar to those of amphetamines.

Signs of possible stimulant abuse, regarding amphetamines and cocaine, include dilated pupils, frequent lip licking and dry mouth, excessive activity, and lack of **sleep**. The drug abuser becomes talkative, but the discussion lacks continuity or coherence, and the subject changes frequently.

See also Caffeine.

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KEY TERMS

Attention deficit hyperactivity disorder (ADHD)—A condition in which a person (usually a child) has an unusually high activity level and a short attention span. People with the disorder may act impulsively and may have learning and behavioral problems.

Tic—A brief and intermittent involuntary movement or sound.

Tourette syndrome—A neurological disorder characterized by multiple involuntary movements and uncontrollable vocalizations called tics that come and go over years, usually beginning in childhood and becoming chronic. Sometimes the tics include inappropriate or obscene language (coprolalia).

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Samuel Uretsky, PharmD

Stings *see* **Bites and stings**

Stomach flu *see* **Gastroenteritis**

Stomatitis

Definition

Stomatitis is an inflammation of the mucous lining of the mouth, which may involve the cheeks, gums, tongue, lips, and roof or floor of the mouth. The word "stomatitis" literally means inflammation of the mouth.

Description

Stomatitis is an inflammation of the lining of any of the soft-tissue structures of the mouth. It is usually a painful condition, associated with redness, swelling, and occasional bleeding from the affected area. The inflammation may be caused by conditions in the mouth itself, such as poor hygiene, from **burns** from hot food or drinks, or by conditions that affect the entire body, such as medications, allergic reactions, or infections. Children may develop stomatitis at any point in their development, from infancy to **adolescence**. The two most common types seen in children are herpes stomatitis, which is caused by the **herpes simplex** virus, and aphthous stomatitis, more often referred to as **canker sores**.

Transmission

Depending upon its cause, stomatitis may or may not be contagious. Herpes stomatitis is considered contagious. Children may be exposed through kissing, sharing food, or playing in close contact with others who have an active herpes infection, such as a **cold sore**. Aphthous stomatitis is not contagious.

Demographics

Though stomatitis may occur at any time during a child's growth, different types affect children at different times. Herpes stomatitis can occur anywhere between six months and five years of age but is most common in children one to two years old who have not been exposed to the herpes virus before. Aphthous stomatitis begins in childhood or adolescence, with peak onset in those aged ten to 19 years. Aphthous ulcers may be more common in females than males. Children of higher socioeconomic status may be more affected than those who are from lower socioeconomic groups.

Causes and symptoms

Causes

A number of factors can cause stomatitis. Cheek biting, braces, or jagged teeth may persistently irritate the oral structures. Chronic mouth breathing due to plugged nasal airways may cause dryness of the mouth tissues, which in turn leads to irritation. The cause of herpes stomatitis is the herpes virus type 1 (not to be confused with genital herpes, which is caused by the herpes virus type 2 and is a sexually transmitted disease). The cause

of aphthous stomatitis is unknown, although several factors are suspected. There may be an inherited tendency to develop canker sores and there may also be an immune system link. In addition, they may be triggered by emotional stress; nutritional deficiencies of iron, **folic acid**, or vitamin B12; menstrual periods; **food allergies**; or viral infections. They may occur with no identifiable cause.

Symptoms

Stomatitis is characterized by **pain** or discomfort in the mouth and the presence of open sores or ulcers in the mouth. Herpes stomatitis may cause the following symptoms:

- **fever**, sometimes as high as 101–104°F (38.3–40°C), which may precede the appearance of blisters and ulcers by one or two days
- irritability and restlessness
- blisters in the mouth, often on the tongue or cheeks or roof of the mouth, which then pop and form ulcers (These ulcers are usually small [about one to five millimeters in diameter], grayish white in the middle, and red around the edges.)
- swollen gums, which may be irritated and bleed
- pain in the mouth
- drooling
- difficulty swallowing
- foul-smelling breath

Aphthous stomatitis may cause the following symptoms:

- burning or tingling sensation in the mouth prior to the onset of other symptoms
- skin lesions on the mucous membranes of the mouth, which begin as a red spot or bump, then develop into an open ulcer, which is usually small (one to two millimeters to one centimeter in diameter) (The ulcers can be single or break out in clusters. The ulcers are painful, and the center appears white or yellow with a fibrous texture. The border of the sore may be bright red.)

When to call the doctor

Parents should call the doctor if any of the following occur:

- inability to drink or swallow
- high temperature
- fussiness and inability to settle down
- symptoms not improved after three days

If the child appears dehydrated, parents should seek immediate medical attention. Signs include dry lips, the absence of tears when crying, a sinking soft spot on an infant's head, and no urination in eight hours or very dark urine. Parents should also seek care if the child is very weak, tired, or difficult to waken.

Diagnosis

Stomatitis is diagnosed by the doctor based primarily upon the appearance of the mouth sores. Both herpes and aphthous stomatitis have lesions that are unique in appearance. Although laboratory studies are seldom performed, the physician may order further blood tests or cultures of the lesions in order to confirm the diagnosis and rule out other causes.

Treatment

The treatment of stomatitis is based upon the problem causing it. For all types, local cleansing and good **oral hygiene** is fundamental. Sharp-edged foods such as peanuts, tacos, and potato chips should be avoided. A soft-bristled toothbrush should be used, and the teeth and gums should be brushed carefully. If toothbrushing is too painful, the child should rinse out his mouth with plain water after each meal. Local factors, such as sharp teeth or braces, can be addressed by a dentist or orthodontist.

Herpes stomatitis treatment

In herpes stomatitis, the most important part of treatment is for parents to keep their child drinking as normally as possible. Bland fluids such as apple juice, liquid flavored gelatin, or lukewarm broth are easiest to drink. Sucking on a Popsicle or sherbet may be soothing. Citrus juices and spicy or salty foods should be avoided. In the event of severe disease, the doctor may use intravenous fluids to prevent **dehydration**. **Acetaminophen** may be used for temperatures over 101°F (38.3°C) and to address pain. Medicines that numb the mouth, like viscous lidocaine or topical anesthetics only last for a brief time and, by numbing the mouth, may cause your child to further injure damaged tissues without knowing it. **Antibiotics** are of no help in treating herpes stomatitis. However, if the case is particularly severe, the doctor may prescribe an antiviral medication such as acyclovir which, if given at the beginning of the outbreak, may help clear things up faster.

Aphthous stomatitis treatment

Medical treatment is usually not necessary for aphthous stomatitis, unless the ulcers are severe (larger

than one centimeter or lasting longer than two weeks). In this case medical evaluation and treatment may be indicated, and topical or oral tetracycline may be given. However, tetracycline is usually not prescribed for children until after all of their permanent teeth have erupted, as it can permanently discolor teeth that are still forming. Avoid hot or spicy foods to minimize discomfort. Mild mouth washes such as salt water or over-the-counter mouthwashes may help. Over-the-counter topical medications applied to the ulcerated area may reduce discomfort and sooth the area. To prevent bacterial infections from developing, parents should encourage their child to brush and floss teeth regularly.

Alternative treatment

Placing a spent tea bag on a canker sore may provide comfort. Sodium lauryl sulfate (SLS), a component of some toothpastes, is a potential cause of canker sores. In one study, most recurrent canker sores were eliminated just by avoiding SLS-containing toothpaste for three months.

Nutritional concerns

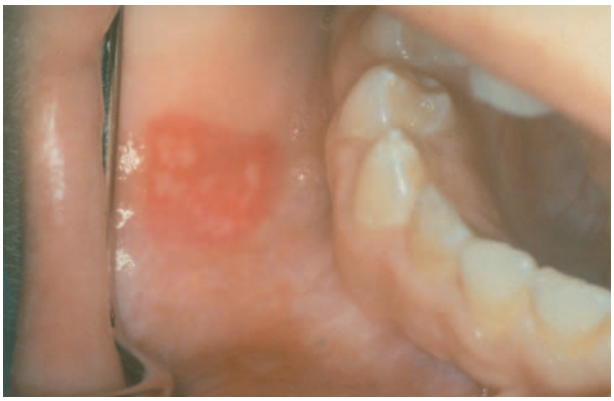
Some physicians may recommend a variety of dietary measures to treat stomatitis. These may include eating cottage cheese, buttermilk, and yogurt, as well as foods high in B **vitamins**. Some doctors may recommend supplementation with folic acid, iron, or vitamin B12.

Prognosis

The prognosis for the resolution of stomatitis is based upon the cause of the problem. Many mouth ulcers are benign and resolve without specific treatment. In the case of herpes stomatitis, complete recovery is expected within ten days without any medical intervention. Oral acyclovir may speed up recovery. Most children are minimally inconvenienced by aphthous stomatitis, because attacks are usually infrequent and only last a few days.

Prevention

Stomatitis caused by irritants can be prevented by good oral hygiene, regular dental checkups, and good dietary habits. Because so many adults and children carry the herpes virus, and because they can pass it on even if they have no symptoms, there is no practical way to prevent herpes stomatitis. Parents can, however, discourage their child from kissing, sharing food, or playing in close contact with people who have an active herpes infection.



This patient is afflicted with stomatitis, a common inflammatory disease of the mouth. *(Photograph by Edward H. Gill, Custom Medical Stock Photo Inc.)*

Canker sores may be minimized by teaching children to avoid trauma, even minor trauma, to the mouth, such as hard toothbrushes and rough foods. If the doctor has determined that the child has a nutritional deficiency, parents can insure that the child is taking the appropriate supplements and eating the recommended foods. Avoiding stressful situations may also be beneficial.

Parental concerns

Most cases of stomatitis in children are benign and resolve within a relatively short period of time. Children with herpes stomatitis may return to school or **day care** when their fever is gone and the mouth sores are healed. Since aphthous stomatitis is not contagious, there is no need to curtail a child's activities unless they have developed signs of complications, such as infection.

See also Canker sores.

Resources

PERIODICALS

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ORGANIZATIONS

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KEY TERMS

Aphthous stomatitis—A specific type of stomatitis presenting with shallow, painful ulcers. Also known as canker sores.

Herpes stomatitis—A form of stomatitis caused by the herpes 1 virus, usually seen in young children.

Stomatitis—Inflammation of the mucous lining of any of the structures of the mouth, including the cheeks, gums, tongue, lips, and roof or floor of the mouth.

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Strabismus

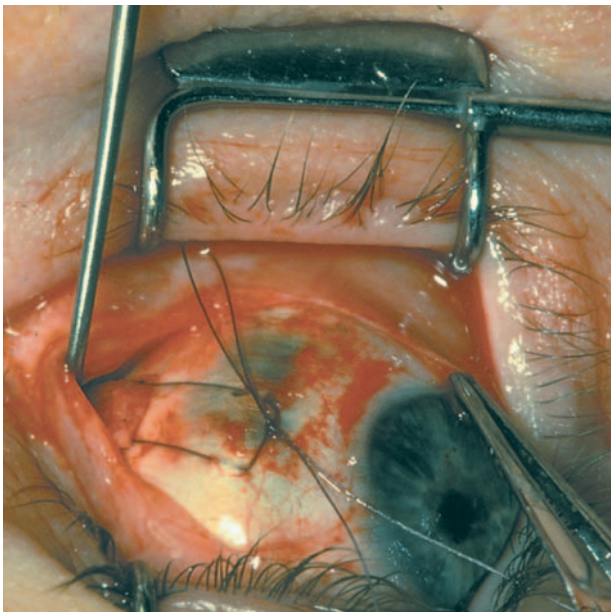
Definition

Strabismus is a condition in which the eyes do not align in the same direction. It is also called crossed eyes or squint.

Description

With normal vision, both eyes focus on the same spot and send the brain the same message. This binocular fixation (both eyes looking directly at the same object) is necessary to see three-dimensionally and to aid in depth perception. When an eye is misaligned, the brain receives two different images. Young children learn to ignore distorted messages from a misaligned eye, but adults with strabismus often develop double vision (diplopia). A baby's eyes should be straight and parallel by three or four months of age. A child who develops strabismus after the age of eight or nine years is said to have adult-onset strabismus.

Esotropia is the most common type of strabismus. It occurs when the eyes turn inward. Infantile esotropia develops in children under the age of six months.



A close-up of ophthalmic surgery being performed to correct strabismus. *(Photograph by Michael English, M.D. Custom Medical Stock Photo, Inc.)*

Accommodative esotropia develops in children under age three who cross their eyes when focusing on objects nearby. This usually occurs in children who are moderately to highly farsighted (hyperopic). Congenital esotropia is a very rare form of strabismus that occurs with certain birth defects.

Another common form of strabismus is exotropia, sometimes called walleye, where the eyes turn outward. It may only be noticeable when a child looks at distant objects, daydreams, or is tired or sick. Other strabismus conditions include hypertrophia, where the eyes turn upward, and hypotropia, where the eyes turn downward.

With strabismus, in some cases the eye turn occurs always in the same eye; however, sometimes the turn alternates from one eye to the other. Most children with strabismus have comitant strabismus, which means that no matter where they look, the degree of deviation does not change. In incomitant strabismus, the amount of misalignment depends upon which direction the eyes are pointed.

False strabismus (pseudostrabismus) occurs when a child appears to have a turned eye; however, this appearance may actually be due to other factors:

- extra skin that covers the inner corner of the eye
- a broad, flat nose

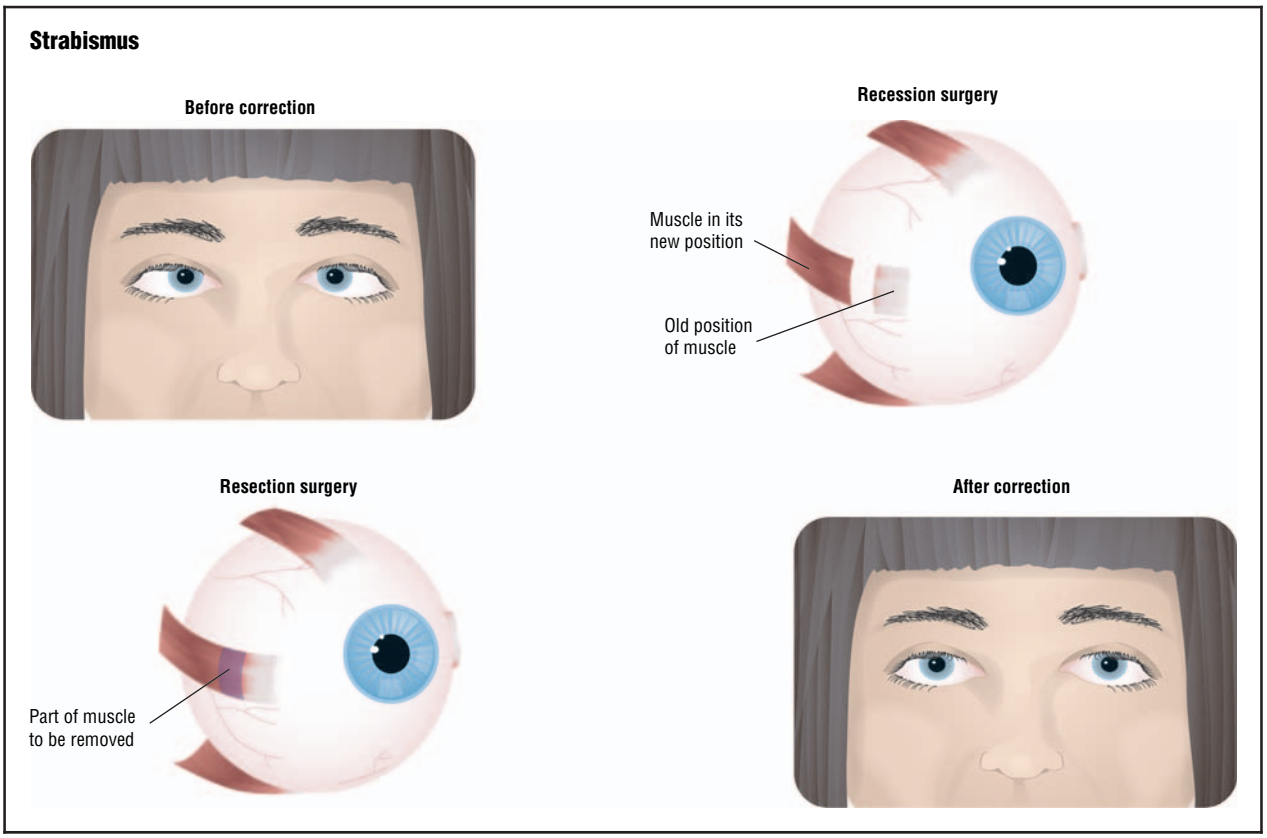


Illustration of patient with strabismus, before and after corrective surgery. During the surgery, the eye muscle may be lengthened (recession surgery) or shortened (resection surgery). *(Illustration by GGS Information Services.)*

• eyes set unusually close together or far apart

False strabismus usually disappears as the child's face grows. An eye doctor (ophthalmologist) needs to determine whether the eye turn is true strabismus or pseudostrabismus.

Demographics

Strabismus affects 5 percent of people in the United States or more than 12 million individuals, most of them children. Infantile esotropia affects about 1 percent of full term, healthy babies and a higher percentage of babies born prematurely or born with other facial defects. Congenital esotropia is rare but does not occur at a higher rate in premature babies. Strabismus occurs equally in boys and girls and shows no variation in racial or ethnic distribution. Most strabismus develops in young children, although a few diseases may cause it to develop in adults.

Causes and symptoms

Strabismus can be caused most often by a defect in the part of the brain that controls eye movement. It is caused less frequently by a defect in the muscles that control eye movement. It is especially common in children who have the following:

• brain tumors
• cerebral palsy
• Down syndrome
• hydrocephalus
• other disorders that affect the brain

Diseases that cause partial or total blindness can cause strabismus. So can extreme farsightedness, cataracts, eye injury, or having much better vision in one eye than the other.

The most obvious symptom of strabismus is an eye that is not always straight. The deviation can vary from day to day or during the day. People who have strabismus often squint in bright sunlight or tilt their heads to focus their eyes.

When to call the doctor

Parents should call their doctor whenever they notice their child's eyes appear misaligned, even if the child is very young. A baby whose eyes have not straightened by the age of four months should be examined to rule out serious disease. Strabismus is not a condition that a child will outgrow without medical intervention. Pediatricians can refer parents to an ophthalmologist (eye specialist) skilled in evaluating the vision of very young children.

Diagnosis

Every baby's eyes should be examined by the age of nine months. A pediatrician, **family** doctor, ophthalmologist, or optometrist licensed to use diagnostic drugs uses drops that dilate the pupils and temporarily paralyze eye-focusing muscles to evaluate visual status and ocular health. Early diagnosis is important. Some eye turns may result from a tumor. Untreated strabismus can damage vision and possibly result in lazy eye (**amblyopia**).

Treatment

Preserving or restoring vision and improving appearance may involve one or more of the following:

- glasses to aid in focusing and straighten the eye(s)
- patching to force infants and young children to use and straighten the weaker eye
- eye drops or ointments as a substitute for patching or glasses or to make glasses more effective
- surgery to tighten, relax, or reposition eye muscles
- medication injected into an overactive eye muscle to allow the opposite muscle to straighten the eye
- vision training (also called eye exercises)

Prognosis

Early consistent treatment usually improves vision and appearance. The most satisfactory results are achieved if the condition is corrected as early as possible and before the age of seven.

Prevention

Strabismus cannot be prevented, but it can be corrected with early intervention.

Parental concerns

Parents are often concerned that eye turn is indicative of other vision problems. Sometimes strabismus does accompany other vision defects, so a complete eye examination by a pediatric ophthalmologist is advisable. Delay only increases the difficulty in correcting strabismus, so parents should not wait to see if their child outgrows the condition.

KEY TERMS

Amblyopia—Decreased visual acuity, usually in one eye, in the absence of any structural abnormality in the eye.

Ophthalmologist—A physician who specializes in the anatomy and physiology of the eyes and in the diagnosis and treatment of eye diseases and disorders.

Resources

BOOKS

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ORGANIZATIONS

American Academy of Ophthalmology. PO Box 7424, 655 Beach Street, San Francisco, CA 94120–7424. Web site: <www.aao.org>.

American Academy of Optometry. 6110 Executive Boulevard, Suite 506, Rockville, MD 20852. Web site: <www.aaopt.org>.

American Academy of Pediatric Ophthalmology and Strabismus (AAPOS). PO Box 193832 San Francisco, CA 94119. Web site: <www.med-aapos.bu.edu>.

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Stranger anxiety

Definition

Stranger anxiety is **fear** or wariness of people with whom a child is not familiar.

Description

An infant learns to recognize her parents within the first few months of birth by sight, sound, and smell. Until about six months of age, the baby will usually seem interested in other adults as well, engaging in games such as peek-a-boo. After six months, many babies undergo a period of fear and unhappiness around anyone except their parents. The child may burst into tears if an unknown person makes eye contact or shriek if left even momentarily in the care of an unfamiliar person.

This stranger anxiety is a normal part of a child's **cognitive development**. It usually begins at around eight or nine months and generally lasts into the child's second year. Normal **separation anxiety** develops during this same period. Both of these responses arise because the baby has reached a stage of mental development where she can differentiate her caretakers from other people, and she has a strong preference for familiar faces. Rather than indicating emotional difficulties, the emergence of a fear of strangers in the second half of the first year is an indicator of mental development.

Infants may react immediately and vigorously to strangers, especially if approached suddenly or picked up by someone unfamiliar. The child may be particularly upset around people who look different to her, for example, people with glasses or men with beards. The setting and way in which the stranger approaches the child can influence how the child may respond. If the stranger approaches slowly when the caregiver is nearby, smiling and speaking softly, offering a toy, the infant will sometimes show interest rather than distress. However, the degree of distress shown by an infant to a stranger varies greatly from baby to baby, a finding that many believe to be rooted in the **temperament** of the infant. A genetic basis for the development of stranger anxiety has also been shown by twin research. Identical **twins** show more similar onset of stranger distress than fraternal twins.

As infants acquire more experience in dealing with unfamiliar persons at **family** outings or in **day care**, their anxiety about strangers diminishes. Young children show a wide variety of responses depending on the situation, their past experiences, and their natural level of sociability.

Common problems

Stranger anxiety can be upsetting to friends and relatives, who may feel rebuffed by a suddenly shy child. The baby may reject a caregiver she was previously comfortable with or grow hysterical when relatives visit. It can also be a frustrating time for the child's parents, since the baby may reject the parent who is not the principal caregiver. Parents should respect the child's fear as much as possible and allow her to approach people as she is able. If the child does not want to be hugged by or sit with a relative, it is unwise to force her. Eventually children outgrow their fear and become more tolerant of strangers.

Parental concerns

All parents are concerned about teaching their children to be wary when approached by unfamiliar adults. However, parents need to find a balance between concern and encouragement of their child's natural curiosity and friendliness, while at the same time teaching them that they should always rely on parental guidance and approval in dealing with strangers.

When to call the doctor

While stranger distress and separation anxiety are normal for infants and toddlers, should a parent become concerned if they persist into the toddler or **preschool** years? The answer to this question depends in part on the nature of the child's response, its intensity, and persistence over time. For example, it is commonplace for preschoolers to show some distress on meeting new people and separating from their parents during the first week or two of daycare or in a new setting. Typically this settling in period does not last too long. If older children persists in showing excessive distress and anxiety on meeting new people, to the point where it interferes with their social development, parents should discuss this pattern with their pediatrician, who may make a referral to a child psychologist for further evaluation.

See also Separation anxiety; School phobia/school refusal.

Resources

ORGANIZATIONS

American Academy of Pediatrics. 141 Northwest Point Boulevard, Elk Grove Village, IL 60007–1098. Web site: <www.aap.org>.

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Peter LaFrenier

Strawberry marks *see* **Birthmarks**
Strength training *see* **Exercise**
Strep culture *see* **Throat culture**

Strep throat

Definition

Streptococcal **sore throat**, or strep throat, as it is more commonly called, is a bacterial infection of the mucous membranes lining the throat or pharynx.

Description

Strep throat is caused by a type of bacteria called group A streptococci. The tonsils may also become infected (**tonsillitis**). Left untreated, strep throat may develop into **rheumatic fever** or other serious conditions.

Demographics

Strep throat accounts for between 5 and 10 percent of all sore throats. Although anyone can get strep throat, it is most common in school-age children. People who smoke, who are fatigued, run down, or who live in damp, crowded conditions are also more likely to become infected. Children under age two and adults who are not around children are less likely to get the disease.

Strep throat occurs most frequently between November to April. The disease passes directly from person to person by coughing, sneezing, and close contact. On rare occasions, the disease is passed through food, when a food handler infected with strep throat accidentally contaminates food by coughing or sneezing. Statistically, if someone in the household is infected, one out of every four other household members may get strep throat within two to seven days.

Causes and symptoms

A person with strep throat suddenly develops a painful sore throat one to five days after being exposed to the streptococcus bacteria. The **pain** is indistinguishable from sore throats caused by other diseases.

The infected person usually feels tired and has a **fever**, sometimes accompanied by chills, **headache**, muscle aches, swollen lymph glands, and **nausea**. Young children may complain of abdominal pain. The tonsils look swollen and are bright red, with white or yellow patches of pus on them. Sometimes the roof of the mouth is red or has small red spots. Often a person with strep throat has bad breath.

Despite these common symptoms, strep throat can be deceptive. It is possible to have the disease and not show any of these symptoms. Many young children complain only of a headache and stomachache, without the characteristic sore throat.

Occasionally, within a few days of developing the sore throat, an individual may develop a fine, rough, sunburn-like rash over the face and upper body and have a fever of 101–104°F (38.3–40°C). The tongue becomes bright red, with a flecked, strawberry-like appearance. When a rash develops, this form of strep throat is called **scarlet fever**. The rash is a reaction to toxins released by the streptococcus bacteria. Scarlet fever is no more dangerous than strep throat and is treated the same way. The rash disappears in about five days. One to three weeks later, patches of skin may peel off, as might occur with a **sunburn**, especially on the fingers and toes.

Untreated strep throat can cause rheumatic fever. This is a serious illness, although it occurs rarely. One outbreak appeared in the United States in the mid-1980s. Rheumatic fever occurs most often in children between the ages of five and 15 and may have a genetic component, since it seems to run in families. Although the strep throat that causes rheumatic fever is contagious, rheumatic fever itself is not.

Rheumatic fever begins one to six weeks after an untreated streptococcal infection. The joints, especially the wrists, elbows, knees, and ankles become red, sore, and swollen. The infected person develops a high fever and possibly a rapid heartbeat when lying down, paleness, shortness of breath, and fluid retention. A red rash over the trunk may come and go for weeks or months. An acute attack of rheumatic fever lasts about three months.

Rheumatic fever can cause permanent damage to the heart and heart valves. It can be prevented by promptly treating **streptococcal infections** with **antibiotics**. It

does not occur if all the streptococcus bacteria are killed within the first ten to 12 days after infection.

In the 1990s, outbreaks of a virulent strain of group A streptococcus were reported to cause a toxic-shock-like illness and a severe invasive infection called necrotizing fasciitis, which destroys skin and muscle tissue. Although these diseases are caused by group A streptococci, they rarely begin with strep throat. Usually the streptococcus bacteria enters the body through a skin wound. These complications are rare. However, since the death rate in necrotizing fasciitis is 30 to 50 percent, it is wise to seek prompt treatment for any streptococcal infection.

Diagnosis

Diagnosis of a strep throat by a doctor begins with a physical examination of the throat and chest. The doctor will also look for signs of other illness, such as a sinus infection or **bronchitis**, and seek information about whether the patient has been around other people with strep throat. If it appears that the patient may have strep throat, the doctor will do laboratory tests.

There are two types of tests to determine if a person has strep throat. A rapid strep test can only determine the presence of streptococcal bacteria but will not tell if the sore throat is caused by another kind of bacteria. To perform a rapid strep test or a **throat culture**, a nurse will use a sterile swab to reach down into the throat and obtain a sample of material from the sore area. The procedure takes only a few seconds but may cause gagging. The results are available in about 20 minutes. The advantage of this test is the speed with which a diagnosis can be made.

The rapid strep test has a false negative rate of about 20 percent. In other words, in about 20 percent of cases where no strep is detected by the rapid strep test, the patient actually does have strep throat. Because of this margin of error, when a rapid strep test is negative, the doctor often does a throat culture.

For a throat culture a sample of swabbed material is cultured, or grown, in the laboratory on a medium that allows technicians to determine what kind of bacteria are present. Results take 24 to 48 hours. The test is very accurate and will show the presence of other kinds of bacteria besides streptococci. It is important not to take any leftover antibiotics before visiting the doctor and having a throat culture. Even small amounts of antibiotics can suppress the bacteria and mask its presence in the throat culture.

In the event that rheumatic fever is suspected, the doctor does a blood test. Results of this test, called an antistreptolysin-O test, tell the doctor whether the person has recently been infected with strep bacteria. This information helps the doctor distinguish between rheumatic fever and rheumatoid arthritis.

Treatment

Strep throat is treated with antibiotics. Penicillin is the preferred medication. Oral penicillin must be taken for 10 days. Patients need to take the entire amount of antibiotic prescribed and not discontinue taking the medication when they feel better. Stopping the antibiotic early can lead to a return of the strep infection. Occasionally, a single injection of long-acting penicillin (Bicillin) is given instead of ten days of oral treatment.

About 10 percent of the time, penicillin is not effective against the strep bacteria. When this happens a doctor may prescribe other antibiotics such as amoxicillin (Amoxil, Pentamox, Sumox, Trimox), clindamycin (Cleocin), or a cephalosporin (Keflex, Durocef, Ceclor). Erythromycin (Eryzole, Pediazole, Ilosone), another inexpensive antibiotic, is given to people who are allergic to penicillin. Scarlet fever is treated with the same antibiotics as strep throat.

Without treatment, the symptoms of strep throat begin subsiding in four or five days. However, because of the possibility of getting rheumatic fever, it is important to treat strep throat promptly with antibiotics. If rheumatic fever does occur, it is also treated with antibiotics. Anti-inflammatory drugs, such as steroids, are used to treat joint swelling. Diuretics are used to reduce water retention. Once the rheumatic fever becomes inactive, children may continue on low doses of antibiotics to prevent a reoccurrence. Necrotizing fasciitis is treated with intravenous antibiotics.

Prognosis

Patients with strep throat begin feeling better about 24 hours after starting antibiotics. Symptoms rarely last longer than five days.

People remain contagious until after they have been taking antibiotics for 24 hours. Children should not return to school or childcare until they are no longer contagious. Food handlers should not work for the first 24 hours after antibiotic treatment, because strep infections are occasionally passed through contaminated food. People who are not treated with antibiotics can continue to spread strep bacteria for several months.

About 10 percent of strep throat cases do not respond to penicillin. People who have even a mild sore throat after a 10-day treatment with antibiotic should return to their doctor. An explanation for this problem may be that the person is just a carrier of strep and that something else is causing the sore throat.

Taking antibiotics within the first week of a strep infection will prevent rheumatic fever and other complications. If rheumatic fever does occur, the outcomes vary considerably. Some cases may be cured. In others there may be permanent damage to the heart and heart valves. In rare cases, rheumatic fever can be fatal.

Necrotizing fasciitis has a death rate of 30 to 50 percent. Patients who survive often suffer a great deal of tissue and muscle loss. Fortunately, this complication of a streptococcus infection is very rare.

Prevention

There is no way to prevent getting a strep throat. However, the risk of getting one or passing one on to another person can be minimized by the following precautions:

- washing hands well and frequently, especially after nose blowing or sneezing and before food handling
- disposing of used tissues properly
- avoiding close contact with someone who has a strep throat
- not sharing food and eating utensils with anyone
- not smoking

Parental concerns

Children who have strep throat should be kept out of daycare, school, activities, and other public places until they have been taking their antibiotic for a full 24 hours. This will help decrease the likelihood of passing on the infection to others.

Parents who are caring for a child with strep will want to take the following steps:

- Give the child **acetaminophen** or ibuprofen for pain. Aspirin should not be given to children because of its association with **Reye's syndrome**, a serious disease.
- Encourage the child to gargle with warm double strength tea or warm salt water, made by adding one teaspoon of salt to eight ounces of water, to relieve sore throat pain.
- Make sure that the child drinks plenty of fluids but avoids acidic juices like orange juice because they irritate the throat.

KEY TERMS

Lactobacillus acidophilus—Commonly known as acidophilus, a bacteria found in yogurt that changes the balance of the bacteria in the intestine in a beneficial way.

- Offer the child soft, nutritious foods like noodle soup and avoid spicy foods.
- Help the child avoid exposure to people who are smoking.
- Encourage the child to rest until the fever is gone, then allow him or her to gradually resume activities.
- Use a room humidifier, as it may make sore throat sufferers more comfortable.
- Be aware that antiseptic lozenges and sprays may aggravate the sore throat rather than improve it.

Resources

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Streptococcal infections

Definition

Streptococcal (strep) infections are communicable diseases that develop when bacteria of the family Streptococcus invade parts of the body and contaminate blood or tissue.

Description

Most people have some form of strep bacteria in their body at some point. A person who hosts bacteria

without showing signs of infection is considered a carrier.

Types of infection

Primary strep infections invade healthy tissue and most often affect the throat. Secondary strep infections invade tissue already weakened by injury or illness. They frequently affect the bones, ears, eyes, joints, or intestines. Both primary and secondary strep infections can travel from affected tissues to lymph glands, enter the bloodstream, and spread throughout the body. Numerous strains of streptococcal bacteria have been identified. Types A, B, C, D, and G are most likely to make people sick.

GROUP A Group A strep (GAS) is the form of streptococcal bacteria most apt to be associated with serious illness. Two of the most severe invasive GAS infections are necrotizing fasciitis or flesh-eating bacteria (destruction of muscle tissue and fat) and **toxic shock syndrome** (a rapidly progressive disorder that causes shock and damages internal organs). GAS is also the type of strep responsible for **strep throat**. Strep throat is common and not usually serious. If untreated, however, strep throat can develop into **rheumatic fever** which can permanently damage the heart and other organs.

GROUP B Group B strep (GBS) most often affects pregnant women, infants, the elderly, and chronically ill adults. Streptococcal infection occurs when bacteria contaminate cuts or open sores or otherwise penetrate the body's natural defenses. The bacteria can be passed from pregnant women to their newborns during **childbirth**.

GROUP C Group C strep (GCS) is a common source of infection in animals. It rarely causes human illness.

GROUP D Group D strep (GDS) is a common cause of wound infections in hospital patients. GDS is also associated with the following:

- abnormal growth of tissue in the gastrointestinal tract
- urinary tract infection (UTI)
- womb infections in women who have just given birth

GROUP G Normally present on the skin, in the mouth and throat, and in the intestines and genital tract, Group G strep (GGS) is most likely to lead to infection in alcoholics and in people who have **cancer**, **diabetes mellitus**, rheumatoid arthritis, and other conditions that suppress immune-system activity.

GGS can cause a variety of infections, including the following:

- bacteria in the bloodstream (bacteremia)
- inflammation of the connective tissue structure surrounding a joint (bursitis)
- endocarditis, a condition that affects the lining of the heart chambers and the heart valves
- **meningitis**
- inflammation of bone and bone marrow (osteomyelitis)
- inflammation of the lining of the abdomen (peritonitis)

Causes and symptoms

GAS

GAS is transmitted by direct contact with saliva, nasal discharge, or open **wounds** of someone who has the infection. Chronic illness, kidney disease treated by dialysis, and steroid use increase vulnerability to infection. About one of five people with GAS infection develops a sore, inflamed throat and pus on the tonsils (strep throat). The majority of those infected by GAS either have no symptoms or develop enlarged lymph nodes, **fever**, **headache**, **nausea**, **vomiting**, weakness, and a rapid heartbeat.

Flesh-eating bacteria is characterized by fever, extreme **pain**, swelling, and redness at a site where skin is broken. Symptoms of toxic shock include abdominal pain, confusion, **dizziness**, and widespread red skin rash.

GBS

A pregnant woman who has GBS infection can develop infections of the bladder, blood, and urinary tract, and deliver a baby who is infected or stillborn. The risk of transmitting GBS infection during birth is highest in a woman whose labor begins before the thirty-seventh week of pregnancy or lasts more than 18 hours or who has the following conditions:

- has a GBS urinary-tract infection
- has already given birth to a baby infected with GBS
- develops a fever during labor

More than 13 percent of babies who develop GBS infection during birth or within the first few months of life develop neurological disorders. An equal number of them die.

Miscellaneous symptoms

Other symptoms associated with strep infections include the following:

- anemia
- elevated white blood cell counts

- inflammation of the epiglottis (epiglottitis)
- heart murmur
- high blood pressure
- infection of the heart muscle
- kidney inflammation (nephritis)
- swelling of the face and ankles

Demographics

Between 10,000 and 15,000 invasive GAS infections occur in the United States every year. In 1999, there were 300 cases of toxic shock associated with GAS infection and 600 cases of necrotizing fasciitis. There are millions of cases of strep throat every year, and similar numbers of cases of relatively mild skin infections. Strep throat is most common among school-age children and people who live in group settings (for example, dorms, boarding schools, the military).

Since first emerging in the 1970s, GBS has been the primary cause of life-threatening illness and death in newborns. GBS exists in the reproductive tract of 20 to 25 percent of all pregnant women. Although no more than 2 percent of these women develop invasive infection, if untreated 40 to 73 percent transmit bacteria to their babies during delivery. About 12,000 of the 3.5 million babies born in the United States each year develop GBS disease in infancy. About 75 percent of them develop early-onset infection. Sometimes evident within a few hours of birth and always apparent within the first week of life, this condition causes inflammation of the membranes covering the brain and spinal cord (meningitis), **pneumonia**, blood infection (sepsis), and other problems.

Late-onset GBS develops between the ages of seven days and three months. It often causes meningitis. About half of all cases of this rare condition can be traced to mothers who are GBS carriers. The cause of the others is unknown. GBS has also been linked to a history of breast cancer. Approximately 5 percent of babies who develop GBS die. However, those who survive often have debilitating problems after the disease. Infections caused by the other types of strep are rare.

When to call the doctor

If the child has a fever and **sore throat**, a wound that seems to be infected, a rash, is acting very sick, or has any other symptoms of strep infection, the doctor should be consulted.

Diagnosis

Strep bacteria can be obtained by swabbing the back of the throat, the vagina, the rectum, or the infected area with a piece of sterile cotton. A blood sample can also be taken. Microscopic examination of the smear can identify which type of bacteria has been collected. A rapid strep test may be done to test for step throat infection. This kind of test gives results within the hour. A sample may also be sent to a lab for traditional culturing, which takes from one to two days, because this form of testing is more accurate than the rapid strep test.

Treatment

Penicillin and other **antibiotics** are used to treat GAS and other types of strep infection. It usually takes less than 24 hours for antibiotics to eliminate an infected person's ability to transmit the infection, but antibiotics always need to be taken for the full course prescribed by the doctor to prevent reinfection or other complications.

Guidelines developed by the American Academy of Obstetrics and Gynecology (AAOG), the American Academy of Pediatrics (AAP), and the Centers for Disease Control and Prevention (CDC) recommend administering intravenous antibiotics during labor to a woman at high risk of passing GBS infection on to her child and offering the medication to any pregnant woman who wants it.

Initiating antibiotic therapy at least four hours before birth allows medication to become concentrated enough to protect the baby during passage through the birth canal. Babies infected with GBS during or shortly after birth need to be treated right away, but they may still die. Those who survive often require lengthy hospital stays and develop vision or hearing loss and other permanent disabilities.

Alternative treatment

Conventional medicine is very successful in treating strep infections. However, several alternative therapies, including homeopathy and botanical medicine, may help relieve symptoms or support the person with a strep infection. For example, several herbs, including garlic (*Allium sativum*), echinacea (*Echinacea* spp.), and goldenseal (*Hydrastis canadensis*), are believed to strengthen the immune system, thus helping the body fight a current infection, as well as helping prevent future infections.

Prognosis

GAS is responsible for more than 2,000 deaths a year. About one in five people infected with flesh-eating

bacteria die. So do three out of every five people who develop streptococcal toxic shock syndrome. Strep throat, however, is almost never fatal, although left untreated it can result in diseases such as rheumatic fever that can affect the heart.

Early-onset GBS kills 15 percent of the infants it affects. Late-onset disease claims the lives of 10 percent of babies who develop it. GBS infections are fatal in about 20 percent of the men and non-pregnant women who develop them. About 10 to 15 percent of non-GAS strep infections are fatal. Antibiotic therapy, begun when symptoms first appear, may increase a patient's chance of survival.

Prevention

Washing the hands frequently, especially before eating and after using the bathroom, and keeping wounds clean can help prevent strep infection. Exposure to infected people should be avoided, and a family physician should be notified if the child develops an extremely sore throat or pain, redness, swelling, or drainage at the site of a wound or break in the skin.

Until vaccines to prevent strep infection become available, 12 monthly doses of oral or injected antibiotics may prevent some types of recurrent infection if necessary. Pregnant women should be screened for GBS during the last few weeks of pregnancy. About one fourth of pregnant women are thought to carry GBS in their vaginal or rectal tracts. If GBS is found to be present, antibiotics can be administered intravenously during labor. This greatly reduces the chance of GBS being transmitted from mother to baby when the baby is in the birth canal. The chances are believed to be reduced from one in 200 that the baby will develop GBS infection to one in 4000.

Parental concerns

Strep infections can develop into life-threatening or debilitating problems if not treated promptly. Ensuring the child takes the full course of antibiotics prescribed by the doctor even if the symptoms have gone away can prevent life-threatening complications such as rheumatic fever. Pregnant women should be screened for GBS during the last weeks of pregnancy to help ensure that GBS does not infect their newborns.

Resources

BOOKS

Laskey, Elizabeth. *Strep Throat*. Chicago: Heinemann Library, 2003.

KEY TERMS

Bacteremia—Bacterial infection of the blood.

Bursitis—Inflammation of a bursa, a fluid-filled cavity or sac. In the body, bursae are located at places where friction might otherwise develop.

Osteomyelitis—An infection of the bone and bone marrow, usually caused by bacteria.

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Stridor

Definition

Stridor is a term used to describe noisy breathing in general and to refer specifically to a high-pitched crowing sound associated with **croup**, respiratory infection, and airway obstruction.

Description

Stridor is a symptom, not a disease. It occurs when air is forced through breathing passages narrowed by the following:

- illness
- infection
- the presence of **foreign objects**
- congenital throat abnormalities

The sound is usually loud enough to be heard at a distance, although sometimes only during deep breathing and can occur on inhaling, exhaling, or both. It can be a symptom of a life-threatening respiratory emergency.

Demographics

Stridor is most common in children. Croup, an inflammation of the trachea (windpipe) and larynx (voice box), is the most common cause of stridor in children under age two. Young children also frequently develop acute stridor by inhaling a foreign object, often food such as hot dogs, popcorn, or hard candy. Stridor as a complication of bacterial infections is also common in children under age eight.

Congenital stridor is caused by abnormalities in the airways that cause them to partially collapse when the child breathes. It is present at birth and usually becomes obvious within the first six weeks of life.

Causes and symptoms

During childhood, stridor is usually caused by infection of the cartilage flap (epiglottis) that covers the opening of the trachea to prevent material from entering the lungs and **choking** a person during swallowing. It can also be caused by foreign objects, such as a food or a coin, that a child has tried to swallow.

Laryngomalacia is the most common cause of congenital stridor, accounting for 75 percent of stridor in newborns. It seems to be caused by a collapse of tissue around the larynx and usually occurs in newborns that have no other health problems. It produces a rapid, low-pitched form of stridor that may be heard when a baby inhales. This condition develops soon after birth and usually does not require medical attention. It normally disappears as the child matures and almost always by the time the child is 18 months old.

Causes of stridor in adolescents and adults include the following:

- abscess or swelling of the upper airway
- paralysis or malfunction of the vocal cords
- tumor
- enlargement of the thyroid gland goiter
- swelling of the voice box (laryngeal edema)
- narrowing of the windpipe (tracheal stenosis)

When to call the doctor

Acute stridor, especially when caused by inhaling a foreign object, can be a life-threatening emergency. Emergency medical care should be sought immediately if the individual is showing any signs of difficulty breathing or is turning blue, is unconscious, or is thought to have inhaled a foreign object. In other cases, a doctor should be consulted on a non-emergency basis whenever stridor develops in a newborn or when stridor accompanies other signs of illness such as a **fever**.

Diagnosis

When stridor is present in a newborn, pediatricians and neonatologists also look for evidence of heart defects or neurological disorders that may cause paralysis of the vocal cords. Paralysis of the vocal cords can be life threatening. If examinations do not reveal other reasons for the baby's noisy breathing, the air passages are assumed to be the cause of the problem.

Listening to an older child or adult breathe usually enables pediatricians, **family** physicians, and pulmonary specialists to estimate where an airway obstruction is located. The timing and location of the noisy breathing, whether the sound is intermittent, occurs during eating, is better or worse when lying or standing, as well as the presence or absence of fever or other signs of infection and similar information help in determining the cause of stridor. It is sometimes difficult in children for doctors to differentiate between stridor and wheezing caused by **asthma**. However, a history of the breathing problem and careful examination can usually help them make the distinction.

The extent of the obstruction can be calculated by assessing several features in the patient:

- complexion
- chest movements
- breathing rate
- level of consciousness

X rays and direct examination of the voice box (larynx) and breathing passages using a laryngoscope or bronchoscope indicate the exact location of the obstruction or inflammation. **Computed tomography** (CT) scans and **magnetic resonance imaging** (MRI) scans also may be useful, especially if surgery is needed.

Flow-volume loops and pulse oximetry are diagnostic tools used to measure how much air flows through the breathing passages and how much oxygen is available. **Pulmonary function tests** may also be performed.

Treatment

Treatment of stridor depends on the underlying cause of the breathing difficulty. Life-threatening emergencies may require the insertion of a breathing tube through the mouth and nose (tracheal intubation) or the insertion of a breathing tube directly into the windpipe (tracheotomy) and surgery to remove a foreign object.

KEY TERMS

Congenital—Present at birth.

Laryngomalacia—A birth defect that causes the tissues around the larynx to partially collapse and narrow the air passageway, causing noisy breathing.

Laryngoscope—An endoscope that is used to examine the interior of the larynx.

Bacterial infections are treated with **antibiotics**. Congenital stridor is usually left untreated and resolves on its own.

Prognosis

The outcome of stridor depends on its cause. Death by suffocation may occur when a foreign object blocks the airway. Otherwise the outcome for most cases of stridor is good to excellent, depending on the cause.

Prevention

Adults must keep small, easily swallowed objects such as coins, beads, and hard, round candies away from young children so that they do not try to swallow them. Taking precautions against colds and bronchial infections (washing hands, not sharing dishes, avoiding sick people) can cut down on stridor from infective causes. Congenital stridor is not preventable.

Parental concerns

Congenital stridor in a newborn can sound frightening to parents, but it is rarely a cause for concern or medical intervention.

See also Croup; Foreign objects; Vocal cord dysfunction.

Resources

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Stroke

Definition

A stroke, also called a cerebral infarction, is a life-threatening condition marked by a sudden disruption in the blood supply to the brain.

Description

A disruption in the blood supply to the brain starves the brain of oxygen-rich blood and causes the nerve cells in that area to become damaged and die within minutes. The body parts controlled by those damaged brain cells lose their ability to function.

Depending on the area of the brain that is affected, a stroke can alter many aspects of a child's functioning such as speech, movement, behavior and learning. A stroke also may cause weakness or paralysis on one side of the body. The loss of function may be mild or severe, temporary or permanent.

If medical treatment begins within hours after symptoms are recognized, brain damage can be limited and the risk of permanent medical effects can be decreased.

Types of stroke

An ischemic stroke—the most common form of stroke in children under age 15—is caused by a blocked or narrowed artery. In children, blockages may be caused by a blood clot, injury to the artery, or rarely in children, atherosclerosis (build-up of fatty deposits on the blood vessel walls). A cerebral thrombosis is a blood clot that develops at the clogged part of the blood vessel. A cerebral embolism is a blood clot that travels to the clogged

blood vessel from another location in the circulatory system.

A hemorrhagic stroke—the more common form of stroke in infants and children under age two—occurs when a weakened blood vessel leaks or bursts, causing bleeding in the brain tissue or near the surface of the brain.

Two types of weakened blood vessels usually cause hemorrhagic stroke, including:

- aneurysm: ballooning of a weakened area of a blood vessel
- arteriovenous malformations: cluster of abnormal blood vessels

A transient ischemic attack (TIA), also called a "mini stroke," is characterized by a short-term blood vessel obstruction or clot that tends to resolve itself quickly, usually within 10–20 minutes, or up to 24 hours. A TIA usually does not require intervention. However, a TIA is a strong indicator of an ischemic stroke and should be evaluated in the same way as a stroke to prevent a more serious attack.

In children, strokes can be categorized as:

- prenatal stroke: occurring before birth
- neonatal or perinatal stroke: occurring in infants less than 30 days old
- pediatric or childhood stroke: occurring in children aged 15 and under

Demographics

Childhood stroke is relatively rare, occurring in about two to three of every 100,000 children aged one to 14 per year. In comparison, stroke occurs in about 100 of every 100,000 adults per year. The rate of ischemic stroke and hemorrhagic stroke is similar among children aged one to 14.

Stroke occurs more frequently in children under age two, and peaks in the perinatal period. In the National Hospital Discharge Survey from 1980-1998, the rate of stroke for infants less than 30 days old (per 100,000 live births per year) was 26.4, with rates of 6.7 for hemorrhagic stroke and 17.8 for ischemic stroke.

More fatal strokes occur in African-American children than white children, mirroring the racial differences of stroke in adults. Compared to the stroke risk of white children, African-American children have an increased relative risk of 2.12, Hispanics a decreased relative risk of 0.76 and Asians have a similar risk. Boys have a 1.28-fold higher risk of stroke than girls and have a higher case-fatality rate for ischemic stroke than girls. The increased risk among African Americans is not explained by the presence of sickle cell disease, nor is the excess risk among boys explained by trauma.

Research conducted by the National Institute of Neurological Diseases and Stroke (NINDS) indicates a "stroke belt," or geographical area where fatal strokes are more predominant. This stroke belt includes Alabama, Arkansas, Georgia, Indiana, Kentucky, Louisiana, Mississippi, North Carolina, South Carolina, Tennessee, and Virginia. Researchers examined death certificates over a 19-year period and found a 21 percent higher risk of death from stroke in people under age 20 in the stroke belt states had compared with the same age group in other states. During the same period, people over age 25 in the stroke belt region had a 20 percent higher risk of death from stroke. Because the overall rate of stroke in children is low, researchers warn parents in these states not to be too alarmed. However, the findings indicate further investigation is needed.

Causes and symptoms

Causes

The cause of childhood stroke is unknown in one-third of cases, and an underlying medical condition or multiple conditions appear to contribute to over half of the cases. The most common causes of stroke are congenital (present at birth) and acquired heart diseases, and **sickle cell anemia**.

About 10–15 percent of children with sickle cell disease suffer a stroke, usually ischemic stroke. Sickle cell disease is a blood disorder in which the blood cells cannot carry oxygen to the brain because the blood vessels to the brain are either narrowed or closed.

One rare cause of stroke is an extreme case of the **chickenpox** virus, which causes a narrowing of blood vessels in the head for some children.

RISK FACTORS Although **obesity**, **high cholesterol**, high blood pressure, atherosclerosis, and **smoking** are common stroke risk factors in adults, they rarely contribute to stroke risk in children. Risk factors for childhood stroke include a **family** history of stroke, cardiovascular disease or diabetes, as well as the presence of the conditions listed below.

Some of the more common congenital heart diseases that increase the risk of childhood stroke include:

- aortic and mitral valve stenosis
- atrial septal defect
- patent ductus arteriosus (PDA)

- patent foramen ovale
- inherited blood clotting disorders
- ventricular septal defect
- hypercoagulable states

Some of the acquired heart conditions that increase the risk of childhood stroke include:

- bacterial meningitis
- endocarditis
- arrhythmia and atrial fibrillation
- artificial heart valve
- myocarditis
- cardiomyopathy
- rheumatic heart disease
- embolism
- anoxia
- antiphospholipid antibody syndrome
- encephalitis
- blood vessel disease
- certain blood disorders, such as hemophilia
- inborn errors of metabolism
- illicit drug use
- teenage pregnancy
- teen use of **oral contraceptives** (birth control pills)

Possible traumas that increase the risk of childhood stroke include birth injury or trauma, **child abuse,** or other injury or trauma.

Because of the wide range of secondary conditions that contribute to stroke, it is difficult for researchers to assess the relative contribution of each risk factor to the problem of cerebrovascular disease as a whole, according to the Child Neurology Society Ad Hoc Committee on Stroke in Children. In addition, this variability also hinders clinical research.

Symptoms

In infants and very young children, stroke symptoms are sudden and include:

- seizures
- coma
- paralysis on one side of the body
- **nausea** or vomiting

In older children, stroke symptoms are sudden and include:

- numbness or weakness of the face, arm, or leg, especially on one side of the body
- confusion or difficulty speaking or understanding speech
- vision difficulties, often in one eye
- hearing problems, often in one ear
- difficulty walking, **dizziness** or loss of balance or coordination
- severe **headache**
- difficulty swallowing
- nausea or vomiting
- painful or stiff neck

Other stroke signs and symptoms include:

- sudden severe headache with unknown cause
- sudden nausea or vomiting
- warm, flushed, clammy skin
- slow, full pulse
- appearance of unequal pupils
- facial "droop" on one side
- salivary drool
- urinary incontinence

If the child seems to recover quickly from these stroke symptoms, a TIA may have occurred. All neurological symptoms should serve as a stroke "warning sign" and could indicate a pending, more serious attack. The child should receive prompt evaluation so necessary preventive therapies can be initiated.

WHEN TO CALL THE DOCTOR If a child has any of the symptoms listed above, the parent or caregiver should immediately dial 9-1-1 to seek emergency care. It is important not to wait to see if symptoms subside; a stroke is a medical emergency. Until the paramedics arrive, the parent or caregiver should follow these first aid guidelines:

- Make sure the child is in a comfortable posture, lying on his or her side, so the airway does not become obstructed by drool or mucus.
- Talk reassuringly to the child, even if he or she is unconscious.
- Do not leave the child alone—constantly observe the child.
- Cover the child with a blanket or remove clothing as needed to maintain the child's normal body temperature.
- Do not give the child any medication, including aspirin; medication will be given later as needed.

Diagnosis

In most children, the diagnosis of stroke is delayed by more than 24 hours from the onset of symptoms. This delay is thought to occur because there is a lack of general awareness by physicians and families of cerebrovascular disorders in children. However, early recognition and treatment of a stroke could improve management, reduce the risk of brain damage and permanent disability, help prevent a recurrence, and initiate a proper treatment and rehabilitation program to maximize functional recovery.

The diagnosis of pediatric stroke generally occurs in the emergency room and includes:

- personal and family medical history
- review of current medications
- evaluation of other health problems
- physical examination
- brief neurological exam
- diagnostic tests

The medical history helps the physician evaluate the presence of other conditions or disorders that might have caused the stroke. The child's family medical history is evaluated to determine if there is a history of cardiovascular or neurological diseases that might increase the risk of blood clots.

The brief neurological exam includes a review of the patient's mental status, motor and sensory system, deep tendon reflexes, coordination, and walking pattern (gait). The cranial nerve function also will be evaluated and includes a review of the patient's visual function and eye movement, strength of facial muscles, the gag reflex, tongue and lip movements, ability to smell and taste, hearing, and sensation and movement of the face, head, and neck.

Questions about the child's condition may include:

- What symptoms occurred?
- When were the symptoms first noticed?
- How long did the symptoms last?
- What functions were affected?

During the physical exam, the child's pulse, blood pressure, and height and weight are checked and recorded.

Diagnostic tests include:

- Blood tests: Test used to detect the presence of any chemical abnormalities, infection, or blood clotting that may have caused the stroke.
- Magnetic resonance imaging (MRI) scan: An imaging technique that provides a detailed picture of the brain without the use of x rays. MRI uses a large magnet, radio waves and a computer to produce these images.
- Computed tomography (CT) scan: An imaging technique that shows the blood vessels in the brain. A CT scan is used to identify the area of the brain affected and to detect signs of swelling.
- Chest x ray: X rays are used to detect an enlarged heart, vascular abnormalities, or lung problems.
- Angiogram: An invasive imaging technique used to examine the blood vessels in the brain. An angiogram is only performed if the CT or MRI scans do not show conclusive results.
- Echocardiogram (echo): A graphic outline of the heart's movement, valves and chambers, used to determine if the stroke was caused by a blood clot traveling from the heart to the brain. Echo is often combined with Doppler ultrasound and color Doppler. During the echo, an ultrasound transducer (hand-held wand placed on the skin of the chest) emits high-frequency sound waves to produce pictures of the heart's valves and chambers.

MRI is more sensitive than CT scanning for the diagnosis of an ischemic stroke within 24 hours. However, the two tests are comparable when used to evaluate the effects of a hemorrhagic stroke.

In rare cases or when carotid artery disease is suspected, additional tests may include a carotid ultrasound or cerebral or carotid angiogram. Other tests to diagnose stroke may include a transcranial Doppler ultrasound and neurosonogram. In a transcranial Doppler ultrasound, sound waves are used to measure blood flow in the vessels of the brain. In a neurosonogram, ultra high frequency sound waves are used to analyze blood flow and possible blockages in the blood vessels in or leading to the brain.

If a pediatric stroke is diagnosed, additional tests may be performed to assess the overall function

- Electroencephalogram (EEG): Electrodes (small, sticky metal patches attached to the scalp) are connected by wires (leads) to an electroencephalograph machine to chart the brain's continuous electrical activity.
- Evoked potentials study: Wires attached to the scalp, neck, and limbs are connected to a computer to measure the electrical activity in certain areas of the brain and spinal cord when specific sensory nerve pathways are stimulated. The brain's electrical response to visual, auditory, and sensual stimulation is recorded.

Treatment

Initial treatment depends on the type of stroke. For an ischemic stroke, initial emergent treatment focuses on restoring blood flow to the brain. For a hemorrhagic stroke, the goal of initial treatment is to control the bleeding. Children with a hemorrhagic stroke may be transferred to a center with neurosurgical facilities so the proper treatment, such as decompression or **hydrocephalus** drainage, can be provided by skilled specialists.

Emergency-room treatment may include: oxygen to ensure the brain is getting the maximum amount, control of body temperature, **assessment** and treatment of breathing difficulties, intravenous fluids to prevent or treat **dehydration**, and medications to control blood pressure and prevent blood clotting. Blood transfusions may be used to treat children with sickle cell disease.

Treatment team

Treatment should be provided by a pediatric neurologist and a multi-disciplinary team of specialists that may include a physical therapist, occupational therapist, speech therapist, social worker, and other specialists as needed to meet the child's individual needs.

Medications

Adult stroke patients who receive treatment within three hours after the onset of stroke symptoms may receive a "clot-busting" medication called t-PA. However, the diagnosis of stroke is rarely made within three hours, so the use of this drug in children is uncommon.

Anticoagulant medications, including heparin or warfarin and low-dose aspirin, may be used to reduce the risk of blood clot formation. Although experience with these medications in children suggests they are safe, their use in children remains controversial because of the risk of Reye's syndrome. Sometimes the potential benefits of these medications outweigh the small risk of side effects. Researchers agree that further studies are needed to determine the proper dosage and effectiveness of aspirin and other anticoagulant medications for treating stroke in children.

The most important medication guidelines are: 1) Ensure your child takes all medications exactly as prescribed; 2) Never discontinue any medication without first talking to the child's doctor, even if the medication does not seem to be working or is causing unwanted side effects; and 3) Follow-up with the child's health care provider as recommended to monitor the effects of the medication. Frequent blood tests are required for people taking anticoagulants to evaluate the dosage and effects of the medication.

Other stroke medications that are still being tested in clinical trials include:

- Citicoline as a treatment for ischemic stroke. Studies have shown both acute and long-term neuroprotective properties of citicoline in animal models of stroke and in several human clinical trials.
- Epoetin, a synthetic version of human erythropoietin, as a treatment for ischemic stroke. Epoetin aids the body in producing red blood cells and is currently used to treat anemia associated with kidney disease or caused by some drugs.
- Early administration of magnesium to serve as a potential neuroprotective agent. Studies have shown neuroprotectant properties of magnesium sulfate in animal models of stroke, and improved outcomes following magnesium sulfate treatment in humans have been observed following small pilot studies.

Rehabilitation

After the child's condition has stabilized, rehabilitation is initiated. Rehabilitation includes physical, occupational, and speech therapy. Therapy is usually initiated as soon as possible after a stroke and is often the most intense in the early stages of recovery. Clinicians should work with the child and the parents or caregivers to develop an individual treatment plan. Specific treatment goals will vary from one child to the next but will focus on restoring maximum function and independence, helping the child return to normal activities, and improving the child's quality of life. The child's progress after rehabilitation will depend upon which area of the brain was affected, the cause of stroke, the extent of injury, and the presence of other medical conditions.

Physical therapy includes stretching exercises, muscle group strengthening exercises, and range of motion exercises to preserve flexibility and range of motion. Exercises should be practiced daily, as recommended by the physical therapist. A physical therapist can instruct the patient on proper posture guidelines to maintain proper alignment of the hips and back. Balancing rest and **exercise** is also important.

Occupational therapy may include splints, casts, or braces on the affected arm or leg to enable proper limb positioning, prevent joint stiffness, and maintain flexibility and range of motion. An occupational therapist can recommend assistive equipment and devices to help the child with activities of daily living, such as bathing, dressing, and eating. If a walker or wheelchair are needed, an occupational therapist can provide specific instructions.

Physical and occupational therapists can provide guidelines on how to adapt the child's home and school environments to ensure **safety** and comfort.

Speech therapy will focus on the child's specific needs which may include any or all aspects of language use, such as speaking, reading, writing, and understanding the spoken word. Speech and language problems (aphasia) usually occur when a stroke affects the right side of the body.

Behavioral problems and learning disabilities, such as difficulties with attention or concentration, may become apparent when the child goes to school, so specific treatments and educational assistance may be needed to address these problems. A formal assessment can help parents identify potential behavioral and learning problems.

Surgery

The need for surgical treatment for pediatric stroke will depend on a number of factors, including the type of stroke, extent of damage from stroke, the child's age, and potential benefits and risks. Sometimes urgent surgery is necessary soon after the child is admitted to the emergency room to remove a blood clot and restore oxygen flow to the brain tissue.

Treatment options for hemorrhagic stroke may include surgery, sterotactic radiotherapy, or interventional neuroradiology to treat the underlying aneurysm or arteriovenous malformation.

There are several surgical procedures to repair an aneurysm that may have caused a hemorrhagic stroke. A clip may be placed across the neck of the aneurysm (like a clip at the end of a balloon) to stop the bleeding. A newer approach is to thread a long, thin tube through the artery that leads to the aneurysm. Then a tiny coil is fed through the tube into the aneurysm "balloon" to fill the space and seal off the bleeding.

An interventional procedure called carotid angioplasty may be performed to treat a blockage or blockages in the carotid arteries. During the procedure, a tiny balloon at the end of a long, thin tube (called a catheter) is pushed through the artery to the blockage. When the balloon is inflated, it opens the artery. In addition, a mesh tube (called a stent) may be placed inside the artery to help hold it open.

Carotid endarterectomy is a surgical procedure performed to remove a blockage from the carotid artery. During the operation, the surgeon scrapes away plaque from the wall of the artery so blood can flow freely through the artery to the brain.

Intracranial bypass surgery is a surgical procedure performed to restore blood flow around a blocked blood vessel in the brain. During the surgery, a healthy blood vessel, on the outside of the scalp, is re-routed to the part of the brain that is not getting enough blood flow. This new blood vessel bypasses the blocked vessel and provides an additional blood supply to areas of the brain that were deprived of blood. When blood flow is restored, the brain works normally, and the symptoms disappear. This procedure is not as common as the other surgical treatments listed above to treat pediatric stroke but it may be used to treat recurrent TIAs.

Alternative treatment

Alternative and complementary therapies include approaches that are considered to be outside the mainstream of traditional health care.

Techniques that induce relaxation and reduce stress, such as **yoga**, Tai Chi, meditation, guided imagery, and relaxation training, may be helpful in controlling blood pressure. Acupuncture and biofeedback training also may help induce relaxation. Before learning or practicing any particular technique, it is important for the parent/caregiver and child to learn about the therapy, its safety and effectiveness, potential side effects, and the expertise and qualifications of the practitioner. Although some practices are beneficial, others may be harmful to certain patients.

Alternative treatments should not be used as a substitute for medical therapies prescribed by a doctor. Parents should discuss these techniques and treatments with the child's doctor to determine the remedies that may be beneficial for the child.

Nutritional concerns

Dietary guidelines are individualized, based on the child's age, diagnosis, overall health, and level of functioning. Specific nutritional problems, such as swallowing or feeding difficulties, may be a concern in some patients and should be managed by a team of specialists including a speech therapist. Early identification, treatment, and correction of specific feeding problems will improve the health and nutritional status of the child.

A child's self-feeding skills can impact his or her health outcome. One study indicated that 90 percent of children with good to fair motor and feeding skills reached adulthood. In contrast, a lack of self-feeding skills was associated with a six-fold increase in mortality (rate of death).

Maintaining a healthy weight is important to prevent the development of chronic diseases such as diabetes, high blood pressure (**hypertension**), and heart disease.

Tube feedings may be required in some patients with **failure to thrive**, aspiration **pneumonia**, difficulty swallowing, or an inability to ingest adequate calories orally to maintain nutritional status or promote growth.

A well-balanced and carefully planned diet will help maintain general good health for children who have suffered a stroke. In general, children should follow the same low-fat, high fiber diet that is recommended for the general population.

In children older than age two, the following low-fat dietary guidelines are recommended:

- Total fat intake should comprise 30 percent or less of total calories consumed per day.
- Calories consumed as saturated fat should equal no more than 8-10 percent of total calories consumed per day.
- Total cholesterol intake should be less than 300 mg/dl per day.

If the child has high blood pressure, the DASH diet is recommended. The "Dietary Approaches to Stop Hypertension (DASH)" study, sponsored by the National Institutes of Health (NIH), showed that elevated blood pressures were reduced by an eating plan that emphasized fruits, vegetables, and low-fat dairy foods and was low in saturated fat, total fat, and cholesterol. The DASH diet includes whole grains, poultry, fish, and nuts. Fats, red meats, sodium, sweets, and sugar-sweetened beverages are limited. Sodium should also be reduced to no more than 1,500 milligrams per day.

Prognosis

Cerebrovascular disorders are among the top 10 causes of death in children, with rates highest in the first year of life. From 1979 to 1998 in the United States, childhood mortality from stroke declined sharply, by 58 percent, with reductions in all major subtypes: ischemic stroke decreased by 19 percent, subarachnoid hemorrhage by 79 percent, and intracerebral hemorrhage by 54 percent.

Some children survive a pediatric stroke with no life-long consequences. In other children, long-term complications of stroke may develop right away or within months to years after a stroke. According to a 2000 study published in the *Journal of Child Neurology*, the outcome of childhood stroke was a moderate or severe deficit in 42 percent of cases. Adverse outcomes after childhood stroke—including death in 10 percent, recurrence in 20 percent, and neurological deficits in two-thirds of survivors—can be reduced with available stroke treatments.

When a stroke affects a child whose brain is still developing, it is thought that the developing brain may be able to compensate for the functions that were lost as a result of a stroke.

Recovery from stroke is different with each child. Overall, the degree of permanent disability after a stroke is less in children than in adults. Speech and language problems usually improve rapidly in the first year after a stroke. Children may only have minor delays in the development of coordinated movement or in cognitive functioning. Almost all children recover the ability to walk independently after a stroke, unless there is another condition that causes disability. Recovery of function in the affected arm and hand is usually the most significant movement problem after a stroke. Most children who suffer from a stroke can expect to lead independent lives as adults.

Prevention

Despite current treatment, one out of 10 children with ischemic stroke will have a recurrence within five years. Although there is a high risk of repeat strokes in patients with sickle cell anemia, the risk can be reduced with regular blood transfusions. If no cause of the stroke was identified, the risk of a recurrence is low. If a cause was identified, the underlying condition should be treated, and anticoagulant or low-dose aspirin therapy may be initiated, depending on the child's diagnosis.

There is no screening for stroke, but screening exists for many of its risk factors. To prevent stroke, risk factors should be treated and managed by the child's primary care doctor or specialist. The doctor can advise if specific preventive treatment is needed.

Management of high cholesterol—especially high LDL (low-density lipoprotein) levels—high blood pressure and diabetes can help reduce the risk of a stroke.

Nutritional concerns

An adequate intake of **folic acid** (vitamin B9) has been linked to the prevention of stroke and heart disease by lowering homocysteine, an amino acid related to the early development of cardiovascular disease when high levels are present in the blood. Dietary sources of folic acid include: vegetables, especially green vegetables; potatoes; cereal and cereal products; fruits; and organ meats (liver or kidney). It is best to eat fresh fruits and vegetables whenever possible to get the most **vitamins**.

Recommended daily intake in micrograms (mcg) for folic acid supplements (oral tablets) include: 25–100 mcg in newborns to age three; 75–400 mcg in children aged four to six; 100-400 mcg in children aged seven to 10; and 150–400 mcg in children aged 11 and above.

Vitamin K is an important nutrient needed to regulate normal blood clotting. A diet deficient in vitamin K can cause prolonged blood-clotting time and easy bleeding and bruising. Vitamin K is found in: alfalfa, asparagus, broccoli, Brussels sprouts, cabbage cheddar cheese, green tea, green leafy lettuce, liver, seaweed, spinach, and turnip greens. Recommended daily intake for vitamin K supplements (for patients not on anticoagulant therapy) include: 10 mcg in newborns to age three; 20 mcg in children aged four to six; 30 mcg in children aged seven to 14; 65 mcg in boys and 55 mcg in girls aged 15–18; 70–80 mcg for males over age 18 and 60–65 mcg for females over age 18. If the patient is taking anticoagulant medications, vitamin K supplements are not recommended, and foods high in vitamin K are limited, since they counteract the action of the medication.

Vitamin E and beta carotene supplements were once thought to help decrease the risk of stroke and prevent the development of heart disease, but newer studies disprove their effectiveness. Researchers at The Cleveland Clinic Heart Center performed a meta-analysis of seven large randomized trials of vitamin E (given alone or in combination with other antioxidants) and eight of beta carotene. All trials included 1,000 or more patients and follow-up ranged from 1.4 to 12 years. The doses of vitamin E given in these trials ranged from 50–800 international units (IU) and 15–50 milligrams (mg) for beta carotene. The meta-analysis reviewed the effect of these antioxidants on death from cardiovascular disease or from any other cause ("all-cause mortality").

Their findings, published in the June, 2003 issue of *The Lancet* journal, do not support the continued use of vitamin E supplementation nor the inclusion of vitamin E in further studies. Regardless of the dosage given or the patient population, Vitamin E did not provide any benefit in lowering mortality compared to control treatments, and it did not significantly decrease the risk of cardiovascular death or stroke (cerebrovascular accident). In addition, they recommend that vitamin supplements containing beta carotene be "actively discouraged" because of the small but statistically significant increased risk of death. Researchers discourage further study of beta carotene because of the mortality risk.

Even though studies have demonstrated that vitamin E and beta carotene supplements do not reduce stroke risk, foods rich in antioxidants are still encouraged because they also contain beneficial nutrients such as flavonoids and lycopenes that are not usually included in standard oral vitamin supplements. A diet rich in antioxidant-containing foods, such as fruits, vegetables and whole grains, is linked to a reduced risk of cardiovascular disease.

Dietary supplements should not be used as a substitute for medical therapies prescribed by a doctor. Parents should discuss these **nutrition** supplements with the child's doctor to determine the remedies that may be beneficial for the child.

Parental concerns

It is common for a child to feel sad or depressed after a stroke. These emotions may be the result of not knowing what to expect or not being able to do simple tasks without becoming overly tired. Temporary feelings of sadness are normal, and should gradually go away within a few weeks, as the child starts a rehabilitation program and returns to some of his or her normal routines and activities.

When a depressed mood is severe and accompanied by other symptoms that persist every day for two or more weeks, the parent should ask for a referral to a mental health professional who can help the child cope and recover. There are many treatments for depression. A healthy lifestyle including regular exercise, proper **sleep**, a well-balanced diet, as well as relaxation and stress management techniques can help manage depression. Major depressive disorder may be treated with **antidepressants**, psychotherapy (supportive counseling or "talk therapy"), or a combination of both.

Regular follow-up visits with the child's health care provider will help identify and manage risk factors and other medical conditions. If the child has a known medical condition that increases the risk of stroke, it is important for parents and caregivers to learn the warning signs and symptoms of stroke in children and infants. If the child experiences any unexpected neurological problem, the parent should have the child evaluated by a physician. Lastly, it is important for parents to carefully follow the child's treatment plan, including following the medication schedule exactly as prescribed.

Resources

BOOKS

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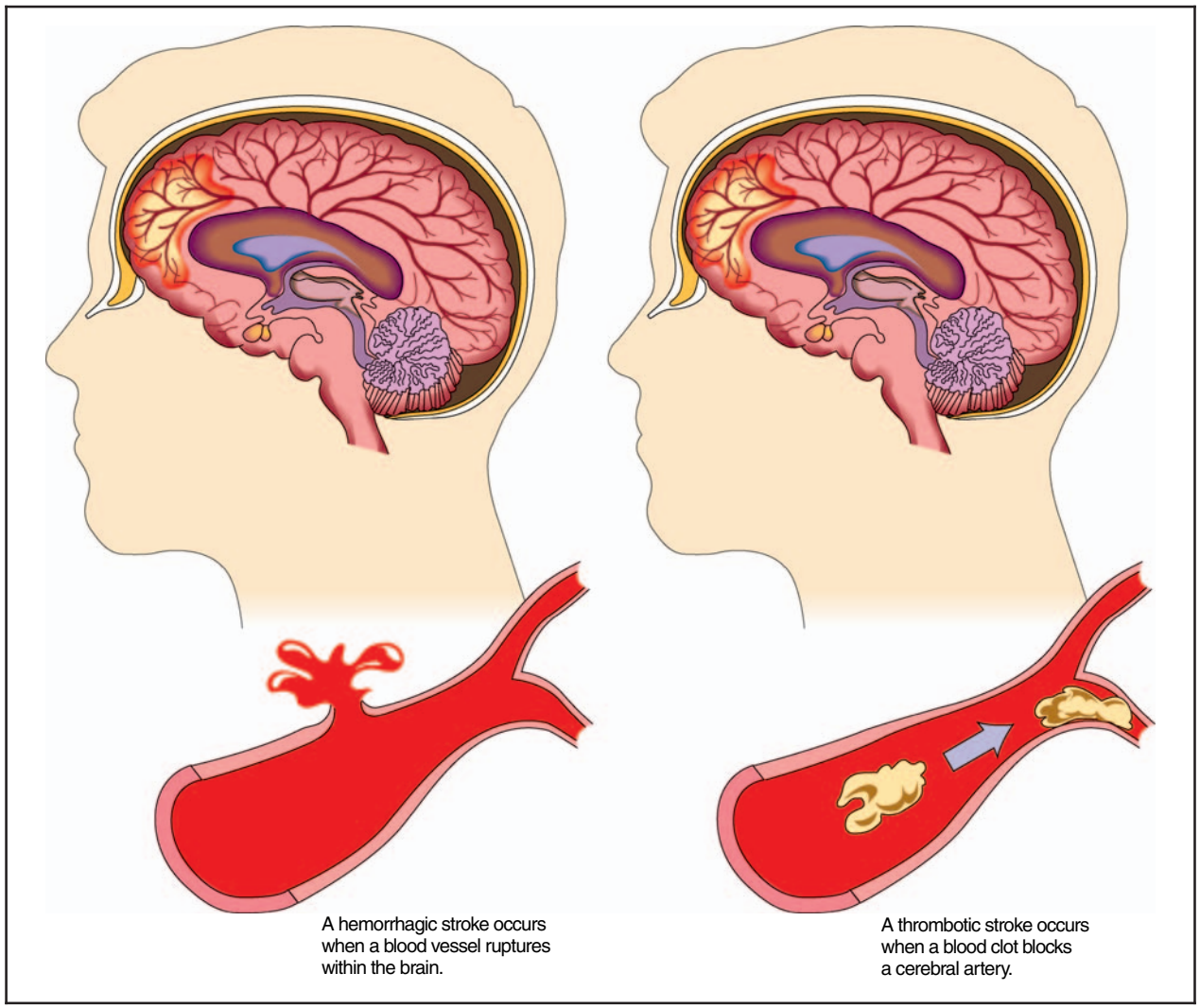

A hemorrhagic stroke (left) compared to a thrombotic stroke (right). *(Illustration by Hans & Cassidy.)*

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Kirkham, FJ. "Stroke in Childhood." Archives of Disease in Childhood. 81 (July, 1999): 85-89.

Nicolaides, P. and R.E. Appleton. "Stroke in Children." Developmental Medicine and Child Neurology. 38:2 (February, 1996): 172-180.

ORGANIZATIONS

American Stroke Foundation. 11902 Lowell, Overland Park, KS 66213. (913) 649-1776. <http://www.americanstroke.org>.

KEY TERMS

Activities of daily living (ADL)—The activities performed during the course of a normal day, for example, eating, bathing, dressing, toileting, etc.

Aneurysm—A weakened area in the wall of a blood vessel which causes an outpouching or bulge. Aneurysms may be fatal if these weak areas burst, resulting in uncontrollable bleeding.

Anoxia—Lack of oxygen.

Antibody—A special protein made by the body's immune system as a defense against foreign material (bacteria, viruses, etc.) that enters the body. It is uniquely designed to attack and neutralize the specific antigen that triggered the immune response.

Antiphospholipid antibody syndrome—An immune disorder that occurs when the body recognizes phospholipids (part of a cell's membrane) as foreign and produces abnormal antibodies against them. This syndrome is associated with abnormal blood clotting, low blood platelet counts, and migraine headaches.

Aorta—The main artery located above the heart that pumps oxygenated blood out into the body. The aorta is the largest artery in the body.

Aortic valve—The valve between the heart's left ventricle and ascending aorta that prevents regurgitation of blood back into the left ventricle.

Aortic valve stenosis—Narrowing of the aortic valve.

Aphasia—The loss of the ability to speak, or to understand written or spoken language. A person who cannot speak or understand language is said to be aphasic.

Arteriosclerosis—A chronic condition characterized by thickening, loss of leasticity, and hardening of the arteries and the build-up of plaque on the arterial walls. Arteriosclerosis can slow or impair blood circulation. It includes atherosclerosis, but the two terms are often used synonymously.

Artery—A blood vessel that carries blood away from the heart to the cells, tissues, and organs of the body.

Atrial—Referring to the upper chambers of the heart.

Atrial fibrillation—A type of heart arrhythmia in which the upper chamber of the heart quivers instead of pumping in an organized way. In this condition, the upper chambers (atria) of the heart do not completely empty when the heart beats, which can allow blood clots to form.

Atrial septal defect—An opening between the right and left atria (upper chambers) of the heart.

Cardiologist—A physician who specializes in diagnosing and treating heart diseases.

Central nervous system—Part of the nervous system consisting of the brain, cranial nerves, and spinal cord. The brain is the center of higher processes, such as thought and emotion and is responsible for the coordination and control of bodily activities and the interpretation of information from the senses. The cranial nerves and spinal cord link the brain to the peripheral nervous system, that is the nerves present in the rest of body.

Cerebrospinal fluid—The clear, normally colorless fluid that fills the brain cavities (ventricles), the subarachnoid space around the brain, and the spinal cord and acts as a shock absorber.

Decompression—A decrease in pressure from the surrounding water that occurs with decreasing diving depth.

Dysphagia—Difficulty in swallowing.

Echocardiogram—A record of the internal structures of the heart obtained from beams of ultrasonic waves directed through the wall of the chest.

Electrocardiagram (ECG, EKG)—A record of the electrical activity of the heart, with each wave being labeled as P, Q, R, S, and T waves. It is often used in the diagnosis of cases of abnormal cardiac rhythm and myocardial damage.

Embolus—Plural, emboli. An embolus is something that blocks the blood flow in a blood vessel. It may be a gas bubble, a blood clot, a fat globule, a mass of bacteria, or other foreign body that forms somewhere else and travels through the circulatory system until it gets stuck.

Encephalitis—Inflammation of the brain, usually caused by a virus. The inflammation may interfere with normal brain function and may cause seizures, sleepiness, confusion, personality changes, weakness in one or more parts of the body, and even coma.

Graft—A transplanted organ or other tissue.

Heart attack—Damage that occurs to the heart when one of the coronary arteries becomes narrowed or blocked.

Hemiparesis—Weakness on one side of the body.

KEY TERMS *(contd.)*

Hemiplegia—Paralysis of one side of the body.

Hydrocephalus—An abnormal accumulation of cerebrospinal fluid within the brain. This accumulation can be harmful by pressing on brain structures, and damaging them.

Hypercoagulable states—Also called thromboembolic state or thrombophilia. A condition characterized by excess blood clotting.

Hypertension—Abnormally high arterial blood pressure, which if left untreated can lead to heart disease and stroke.

Intracerebral hemorrhage—A cause of some strokes in which vessels within the brain begin bleeding.

Ischemia—A decrease in the blood supply to an area of the body caused by obstruction or constriction of blood vessels.

Mitral valve stenosis—Narrowing of the mitral valve.

Neurologist—A doctor who specializes in disorders of the nervous system, including the brain, spinal cord, and nerves.

Neurosurgeon—Physician who performs surgery on the nervous system.

Occupational therapist—A healthcare provider who specializes in adapting the physical environment to meet a patient's needs. An occupational therapist also assists patients and caregivers with activities of daily living and provide instructions on wheelchair use or other adaptive equipment.

Patent ductus arteriosus—A congenital defect in which the temporary blood vessel connecting the left pulmonary artery to the aorta in the fetus doesn't close after birth.

Patent foramen ovale (PFO)—A congenital heart defect characterized by an open flap that remains between the two upper chambers of the heart (the left and right atria). This opening can allow a blood clot from one part of the body to travel through the flap and up to the brain, causing a stroke.

Physiatrist—A physician who specializes in physical medicine and rehabilitation.

Physical therapist—A healthcare provider who teaches patients how to perform therapeutic exercises to maintain maximum mobility and range of motion.

Reye's syndrome—A serious, life-threatening illness in children, usually developing after a bout of flu or chickenpox, and often associated with the use of aspirin. Symptoms include uncontrollable vomiting, often with lethargy, memory loss, disorientation, or delirium. Swelling of the brain may cause seizures, coma, and in severe cases, death.

Stent—A slender hollow catheter or rod placed within a vessel or duct to provide support or to keep it open.

Subarachnoid hemorrhage—A collection of blood in the subarachnoid space, the space between the arachnoid and pia mater membranes that surround the brain. This space is normally filled with cerebrospinal fluid. A subarachnoid hemorrhage can lead to stroke, seizures, permanent brain damage, and other complications.

Unilateral neglect—Also called one-sided neglect. A side effect of stroke in which the stroke survivor ignores or forgets the weaker side of the body caused by the stroke.

Vein—A blood vessel that returns blood to the heart from the body. All the veins from the body converge into two major veins that lead to the right atrium of the heart. These veins are the superior vena cava and the inferior vena cava. The pulmonary vein carries the blood from the right ventricle of the heart into the lungs.

Ventricle septal defect—A hole in the wall (septum) between the lower chambers of the heart.

Ventricles—The lower pumping chambers of the heart. The ventricles push blood to the lungs and the rest of the body.

Ventricles of the brain—The spaces within the brain where cerebrospinal fluid is made.

American Stroke Association, A Division of American Heart Association, 7272 Greenville Ave., Dallas, TX 75231. (888) 4-STROKE (787653). E-mail: strokeassociation@heart.org. <http://www.strokeassociation.org>.

Children's Hemiplegia and Stroke Association. 4101 W. Green Oaks, Ste. 305, PMB 149, Arlington, TX 76016. (817) 492-4325. E-mail: info@chasa.org. <http://www.chasa.org>.

National Heart, Lung and Blood Institute. National Institutes of Health, Building 1, 1 Center Dr., Bethesda, MD 20892. E-mail: NHLBIinfo@rover.nhlbi. <http://www.nhlbi.nih.gov>.

National Institute on Disability and Rehabilitation Research, Office of Special Education and Rehabilitative Services, U.S. Department of Education, 400 Maryland Ave. S.W., Washington, DC 20202-7100. (202) 245-7640. <http://www.ed.gov/about/offices/list/osers/nidrr/>.

National Institute of Neurological Disorders and Stroke (NINDS), National Institutes of Health. P.O. Box 5801, Bethesda, MD 20824. (800) 352-9424 or (301) 496-5751. <http://www.ninds.nih.gov/about_ninds/>.

National Rehabilitation Information Center (NARIC).4200 Forbes Blvd., Ste. 202, Lanham, MD 20700. (800) 346-2742 or (301) 459-5900. <http://www.naric.com>.

National Stroke Association. 9707 E. Easter Ln., Englewood, CO 80112-3747. (800) STROKES (787-6537) or (303) 649-9299. <http://www.stroke.org>.

Stroke Clubs International. 805 12th St. Galveston, TX 77550. (409) 762-1022 attn. Ellis Williamson. E-mail: strokeclub@aol.com.

WEB SITES

The Brain Attack Coalition. <www.stroke-site.org>

The Brain Matters, American Academy of Neurology Foundation. <www.thebrainmatters.org>

Different Strokes—A Charity for Younger Stroke Survivors. <www.differentstrokes.co.uk/>

HeartCenterOnline. <www.heartcenteronline.com>

HemiHelp—Information and Support for Children and Young People with Hemiplegia. <www.hemihelp.org.uk>

Pediatric Stroke Network. <www.pediatricstrokenetwork.com>

Angela M. Costello

Sturge-Weber syndrome

Definition

Sturge-Weber syndrome (SWS) is a rare, congenital (present at birth), noninherited disorder characterized by the vascular malformation (birthmark) called a port wine stain, usually seen on an infant's face. Sturge-Weber also is characterized by neurological abnormalities including seizures, weakness on one side of the body, **developmental delay**, and glaucoma (increased pressure within the eye). Other terms for SWS are: encephalotrigeminal angiomatosis, encephalofacial angiomatosis, or Sturge-Weber-Dimitri syndrome.

Sturge-Weber syndrome is named for the British physicians William A. Sturge (1850–1919), who first described the condition, and Frederick Parkes Weber (1863–1962) who demonstrated its intracranial calcifications.

Description

SWS is a rare congenital disorder whose most apparent indication is a port wine stain on the face that is associated with neurological abnormalities. The port wine stain is a benign tumor just under the surface of the skin, made up of overabundant blood vessels (angiomas). Port wine stain may affect either or both sides of the face and can vary in size. Other neurological abnormalities may be present, including angioma on the brain's surface.

Demographics

The incidence of SWS is estimated at one per 50,000 live births in the United States. No regional or gender differences have been noted. An estimated 13 percent of individuals with SWS will not have the port wine stain. In addition, some children with port wine stain may not have Sturge-Weber syndrome.

Causes and symptoms

The exact cause and incidence of Sturge-Weber syndrome was as of 2004 not understood. It is not thought to be genetic.

Frequency of symptoms

A child born with SWS has a higher likelihood of the following clinical signs of the disorder:

- port wine stain: 8–15 percent
- bilateral (both sides) brain involvement: 15 percent
- seizures: 72–93 percent
- hemipareis (weakness on one side of body): 25–56 percent
- hemianopsia (loss of half of the field of vision): 44 percent
- headaches: 44–62 percent
- developmental delay/mental retardation: 50–75 percent
- glaucoma (increased pressure within the eye): 30–71 percent
- choroidal hemangioma (nonmalignant blood vessel tumors in the eye): 40 percent

The following manifestations of SWS may be present:

- Port wine stain: The port wine stain is caused by excess capillaries (tiny blood vessels) just below the skin's surface. It may vary in color, shape, and location on the

face. Sometimes the port wine stain covers other parts of the body as well as the face.

- Seizures: Angiomas on the surface of the brain cause seizures in nearly all children with SWS. As the child grows, the affected part of the brain can atrophy (waste away). Deposits of calcium also may occur. This can cause seizures to become more frequent and to last longer.
- Hemipareis (weakness on one side of body): In SWS, this results from frequent seizures.
- Hemianopsia (loss of half of the field of vision): Angiomas can affect the optic nerve, causing blindness in half of the eye.
- Headaches: About one-third of children aged ten years and younger with SWS suffer from migraines.
- Developmental delay/mental retardation: Seizures are responsible for learning difficulties in two out of three children with SWS.
- Glaucoma (increased pressure within the eye): Glaucoma is present in 70 percent of children with SWS whose upper eyelids have port wine stain. Fluid produced within the eye (aqueous humor) cannot exit normally. This leads to increased pressure within the eye and eventual damage to the optic nerve.
- Choroidal hemangioma (nonmalignant blood vessel tumors in the eye): Noncancerous tumors can grow within the eye on the choroid blood vessel, the vessel that nourishes the eye. If the tumor is in the central area of vision, visual function can be affected.

When to call the doctor

An infant born with a port wine stain will be immediately evaluated by healthcare staff. In some cases, infants with SWS will not have a port wine stain present at birth. In these cases, suspicion of SWS may not arise until a child has a seizure or other neurological problem.

Diagnosis

Clinical diagnosis of SWS begins with the observation of port wine stain in an infant. The port wine stain may not be obvious in children of color. Not all children with port wine stain will have SWS, however; and some children with SWS will not have port wine stain. In the absence of port wine stain, other neurological abnormalities will help determine the diagnosis. Seizures may be the first symptoms of SWS in a child, usually by the first year. The seizures are usually frequent and may be prolonged. If glaucoma is involved, there may be no symptoms in older children. Infants may avoid bright light as a result of enlarged corneas.

If neurological involvement is suspected, the following tests may be used to help make a diagnosis:

- X ray of the skull to show calcifications (calcium deposits)
- CT scan of the skull to show calcifications, abnormal veins, and brain atrophy
- MRI to show angiomas (benign tumors made up of blood vessels)
- single-photon emission **computed tomography** to measure blood flow in the brain
- EEG to evaluate seizures

Treatment

Treatment for SWS depends on the disorder involved.

- Port wine stain: Laser treatment is used to lighten or remove port wine stain. Pulsed-dye laser therapy successfully treats port wine stain without significant scarring. Treatment should start as soon as possible. Multiple treatments will be necessary.
- Seizures: Drug therapy may be used to control seizures. However, the seizures often are resistant to treatment. In some cases, early surgical removal of the part of the brain with the abnormal blood vessels may be considered.
- Vision problems: Drug therapy may be used to treat glaucoma. Photodynamic therapy also is used to treat choroidal hemangiomas that affect the eye.
- Headaches: Medications may be taken to treat migraines. Children age two and under should not take aspirin due to the risk of Reye syndrome.
- Developmental delay and learning problems: A wide range of treatment options is available to children with developmental delay and learning problems associated with SWS.
- Hemipareis (weakness on one side of body): Hemipareis can be treated with physical and occupational therapy.

Prognosis

SWS is not a fatal disease. The prognosis for SWS depends on the specific neurological abnormalities present. Some abnormalities associated with SWS may worsen with age. Successful treatment of seizures improves the outlook for children with SWS.

Prevention

There was as of 2004 no known way to prevent SWS. Nothing a parent has done or did not do causes the disorder.

Parental concerns

The seizures that are often present with SWS can place children in potentially dangerous situations.

See also Seizure disorder.

Resources

PERIODICALS

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Thomas-Sohl, K. A., et al. "Sturge-Weber syndrome: a review." *Pediatric Neurology* 30 (May 2004): 303–10.

ORGANIZATIONS

American Academy of Dermatology. 930 E. Woodfield Rd., Schaumburg, IL 60168. Web site: <www.aad.org/>

Children's Hemiplegia and Stroke Association. Suite 305, PMB 149 4101 W. Green Oaks. Arlington, TX 76016. Web site: <www.hemikids.org/hemiplegia.htm>.

FACES: The National Craniofacial Association. PO Box 11082, Chattanooga, TN 37401. Web site: <www.faces-cranio.org/>.

National Association for Rare Disorders. 55 Kenosia Avenue, PO Box 1968, Danbury, CT 06813–1968. Web site: <www.rarediseases.org/info/contact.html>.

Sturge-Weber Foundation. PO Box 418, Mount Freedom, NJ 07970. Web site: <www.sturge-weber.com/>.

WEB SITES

"NINDS Sturge-Weber Syndrome Information Page." *National Institute of Neurological Disorders and Stroke*, 2001. Available online at <www.ninds.nih.gov/health_and_medical/disorders/sturge_doc.htm> (accessed November 30, 2004).

Sturge-Weber Foundation. Available online at <www.sturge-weber.com> (accessed November 30, 2004).

"Sturge-Weber Syndrome." *eMedicine*, 2001. Available online at <www.emedicine.com/neuro/topic356.htm#section~workup> (accessed November 30, 2004).

KEY TERMS

Angioma—A tumor (such as a hemangioma or lymphangioma) that mainly consists of blood vessels or lymphatic vessels.

Anomaly—Something that is different from what is normal or expected. Also an unusual or irregular structure.

Capillaries—The tiniest blood vessels with the smallest diameter. These vessels receive blood from the arterioles and deliver blood to the venules. In the lungs, capillaries are located next to the alveoli so that they can pick up oxygen from inhaled air.

Choroidal hemangioma—A nonmalignant blood vessel tumor in the eye.

Hemianopsia—Loss of half of the field of vision.

Hemiparesis—Weakness on one side of the body.

"Sturge-Weber Syndrome." *Epilepsy Action*. Available online at <www.epilepsy.org.uk/info/sturge.html> (accessed November 30, 2004).

Christine Kuehn Kelly

Stuttering

Definition

Stuttering is a speech problem characterized by repetitions; pauses; or drawn-out syllables, words, and phrases. Stutterers are different than people experiencing normal fluency problems because a stutterer's disfluency is more severe and consistent than that of people who do not stutter.

Description

Normal **language development** in a child usually includes a period of disfluency. Children might repeat syllables or words once or twice. Sometimes, children experiencing normal disfluencies hesitate during speech or use fillers, including "um," with frequency. These developmental problems usually happen between one and five years of age. Often, parents are concerned about the disfluency they hear in their children.

A child with mild stuttering, however, will repeat sounds more than twice. Parents and teachers often notice the child's facial muscles become tense and he or she might struggle to speak. The child's voice pitch might rise with repetitions, and some children experience occasional periods when airflow or voice stops for seconds at a time. Children with more severe stuttering stutter through more than 10 percent of their speech. This child exhibits considerable tension and tries to avoid stuttering by using different words. In these children, complete blocks of speech are more common than repetitions or prolongations, during which children lengthen syllables or words.

Stuttering does not affect **intelligence**. Teens often experience more noticeable problems with stuttering as they enter the dating scene and increase their social interactions. Stuttering can severely affect one's life. Often, adults who are concerned about stuttering choose their careers based on the disability.

The degree of stuttering is often inconsistent. Stutterers can be fluent in some situations. Many find that they stop stuttering when singing or doing other activities involving speech. Some have good and bad days when it comes to stuttering. On good days, a stutterer might be able to talk fluently using words that usually cause him to repeat, pause or prolong sounds, syllables, parts of words, entire words, or phrases.

Demographics

More than 3 million Americans stutter and four times more males are affected than females. Stuttering usually begins in childhood when the child is developing language skills, and it rarely develops in adulthood with only 1 percent of the population affected by the disorder. Approximately 25 percent of all children experience speech disfluencies during development that concern their parents because of their severity.

Causes and symptoms

There is no known cause of stuttering. Some believe that it has a physical cause and that it might be related to a breakdown in the neurological system. Stuttering starts early in life and often is inherited. Brain scan research has revealed that there might be abnormalities in the brains of stutterers, while they are stuttering. Myths about why stuttering occurs abound. Some cultures believe that stuttering is caused by emotional problems, tickling an infant too much, or because a mother ate improperly during breastfeeding. None has been proven to be true. It is believed that some drugs might induce stuttering-like conditions. These include **antidepressants**, **antihistamines**, tranquilizers, and selective serotonin reuptake inhibitors.

When to call the doctor

The child's doctor should be contacted if parents have concerns about the speech patterns of their child. The doctor may refer parents to a speech-language specialist for evaluation if needed.

Diagnosis

Speech and language therapists diagnose stuttering by asking stutterers to read out loud, pronounce specific words, and talk. Some also order hearing tests. The tests will determine whether a person needs speech therapy.

Treatment

As of 2004, researchers did not understand what causes stuttering. However, progress has been made regarding what contributes to the development of the disability; therefore, in some cases it can be prevented in childhood with the help of therapy early on. Therapy can help people of all ages suffering from the speech disability. While not an overnight cure, therapy can offer positive results and more fluent speech patterns. The goals of therapy are for the stutterer to reduce stuttering frequency, decrease the tension and struggle of stuttering, become educated about stuttering, and learn effective communications skills, such as making eye contact, to further enhance speech. The therapy focuses on helping stutterers to discover easier and different ways of producing sounds and expressing thoughts. The success of therapy depends largely on the stutterer's willingness to work at getting better.

The duration of stuttering therapy needed varies among stutterers. Sometimes, stutterers find intermittent therapy useful throughout their lives.

Parents, teachers and others can help ease stuttering. These include: talking slowly, but normally, clearly, and in a relaxed manner to a stutterer; answering questions after a pause to encourage a relaxed transaction; trying not to make stuttering worse by getting annoyed by a person's stuttering; giving stutterers reassurance about their stuttering; and encourage the stutterer to talk about his or her stuttering.

Electronic fluency aids help some stutterers when used as an adjunct to therapy. Medications, such as antipsychotics and neuroleptics, have been used to treat stuttering with limited success.

Some people use relaxation techniques to help their stuttering.

Prognosis

As of the early 2000s no answers had been found to explain the causes of stuttering; still, much has been learned about what contributes to stuttering's development and how to prevent it in children. People who stutter can get better through therapy. Winston Churchill, Marilyn Monroe, Carly Simon, James Earl Jones, and King George VI were childhood stutterers who went on to live successful professional lives.

Prevention

The location of some genes appears to predispose people to stuttering. While genetic factors do not explain all stuttering, genetics may help to uncover the disability's causes. Speech therapy, especially that performed at a young age, can stop the progression of stuttering.

Parental concerns

Many children experience brief episodes of stuttering. In many cases, these are transitory and disappear without treatment. Parents should be aware that some stuttering is quite normal when a child feels under pressure to talk. Thus, parents should wait to allow the child to communicate at his or her own speed, and not pressure the child to talk or make fun of the stutter.

KEY TERMS

Antipsychotic drug—A class of drugs used to control psychotic symptoms in patients with psychotic disorders such as schizophrenia and delusional disorder. Antipsychotics include risperidone (Risperdal), haloperidol (Haldol), and chlorpromazine (Thorazine).

Disfluency—An interruption in speech flow.

Neuroleptics—Antipsychotic drugs that affect psychomotor activity.

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Styes and chalazia

Definition

Styes and chalazia are infections and inflammations of the tiny oil glands on the eyelids. A sty, or external hordeolum, is a common childhood infection of an oil gland on the surface of the upper or lower eyelids at the base of the eyelash. An internal hordeolum is an infection deeper inside the eyelid.

A chalazion is an inflammation or blockage of the deep oil glands within the eyelid that develops into a small hard mass.

Description

Styes and internal hordeola begin as a red, pimple-like bump on the eyelid. The eye may water and itch, and the eyelid may be swollen and painful. Styes come to a head in about three days when they open and drain. Healing is rapid. Internal hordeola are larger, last longer, and are more painful.

Chalazia develop within the Meibomian glands, or oil glands of the eyelid. There are approximately one hundred of these glands located underneath each row of eyelashes. Obstruction and infection, which often are the result of bacteria, cause the gland and the area around it to swell. Chalazia are slow growing, usually over two to three weeks, and can last several months.

A chalazion first appears as a firm lump under the skin but is usually not painful. If the oil gland is blocked and inflammation spreads beyond the eyelid, the condition can interfere with vision. Sometimes, a chalazion develops after a sty has healed.

Growths on the eyelid that are not red and painful are usually cysts and should be evaluated by a doctor. Sometimes, they are removed. In most cases, they are not.

Transmission

Staphylococcus aureus bacteria are thought to be responsible for most styes. Rubbing the eyes, especially when the sty is oozing pus, can spread the infection along the eyelid and cause other styes. There is also a chance that if transmitted to the face or other parts of the body the bacteria in the sty can cause **impetigo**, a contagious skin infection.

Demographics

Styes are more common in children than adults. Once a child has one sty, there is an increased chance of the child having another later on. They also seem to recur in children with lowered immune resistance such as children with diabetes. **Acne** also seems to trigger styes in some adolescents.

Chalazia occur more often in adults than children, and in men more than women. As is the case with styes, having acne seems to predispose some adolescents to having chalazia.

Causes and symptoms

Styes and internal hordeola in children are usually caused by *Staphylococcus aureus* bacterial infections that are transmitted from a child's eyes and nose. In most cases, the bacteria enter the eye through unwashed hands or contaminated contact lens. Bacteria may live on the eyelids or eyelash hair follicles themselves and begin to grow when the oil gland of a hair follicle becomes blocked.

Symptoms

Styes appear as red bumps on the eyelid and may cause **itching** or tearing. Sometimes children report feeling as if something is in the eye. Both styes and internal hordeola are usually painful. The eyelid may look red and be swollen. Vision is sometimes blurred, and the eyes may be sensitive to light.

Though chalazia may appear as large unsightly lumps deep within the eyelid, they are usually not painful. On rare occasions, if a chalazion becomes quite big, it can press on the cornea. If the chalazion is on the upper eye lid, it can produce various vision problems including astigmatism, a distortion of the lens that causes fuzzy vision.

When to call the doctor

It is important to call the doctor if the child has a **fever**, **pain** in the eye, swelling or redness over the entire eyelid, or a painful sty persists for one to two weeks. It may be necessary to call a doctor if the child experiences no improvement after three days of home care. In addition, if the child experiences vision problems, a doctor should be consulted.

Conjunctivitis, which appears as redness in the white of the eye, is a serious condition that must have a doctor's attention. In most cases, **antibiotics** or antibiotic ointments are prescribed.

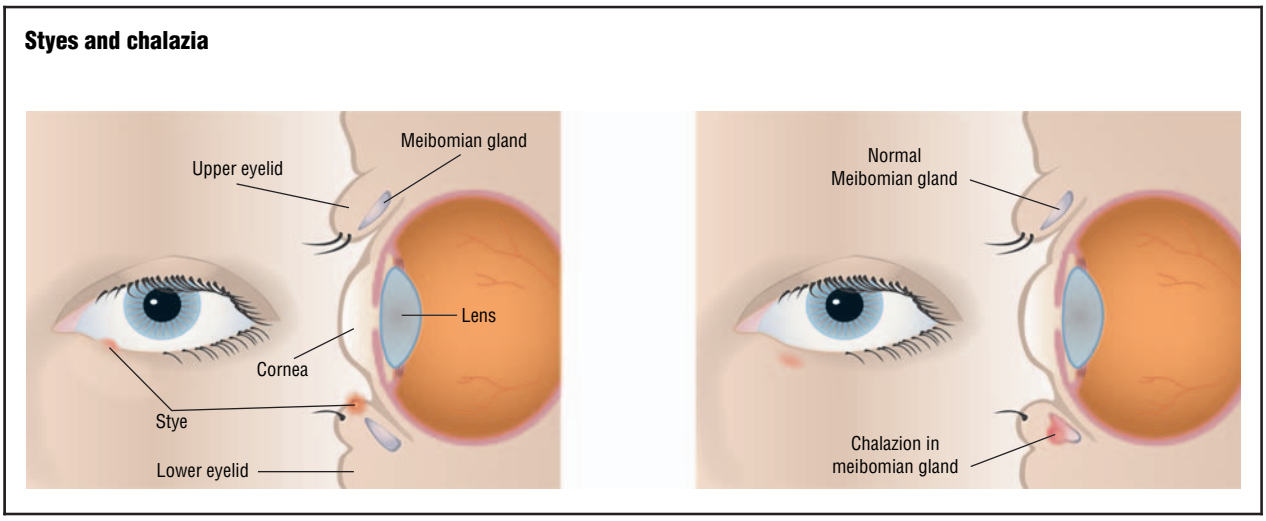

Illustration of a sty (left) and a chalazion. The sty appears on the margin of the eyelid, while the chalazion occurs deeper within the Meibomian gland of the eyelid. *(Illustration by GGS Information Services.)*

Stubborn chalazia that do not go away within six weeks or continue to enlarge require medical attention. They may need to be removed surgically, using local anesthesia.

Diagnosis

The doctor determines whether a child has a sty by visually examining the appearance of the eyelid. If the bump is hard and is located deep within the eyelid, it probably is a chalazion. For a patient whose sty has not healed with home treatment, the doctor may test the fluid in the eye to determine the type of bacteria present and prescribe treatment accordingly.

Treatment

The usual treatment for styes begins at home with the application of a warm washcloth soaked in fairly warm water. Heat is important to increase the blood supply to the eye, helping the immune system to fight off the infection. It also opens the blocked oil gland and helps remove pus. The water should be hot but not too hot. If a finger cannot remain in the water comfortably, it is too hot. Washcloths should not be heated in a microwave oven. Salt or rosewater may be added to the warm water.

The warm washcloth is applied to the eyelid of the closed eye for 10 to 15 minutes three to four times a day. Relief is usually felt within 24 hours.

The child and the parent should avoid popping the sty as one would a pimple. This can irritate the eyelid further and spread the bacteria. By allowing the style to break on its own and leak out pus, the sty can drain. Daily washing and the use of warm compresses help eliminate the bacteria released by the burst sty.

Some doctors recommend using over-the-counter eye washes, such as Bausch and Lomb Eye Wash or Collyrium Eye Wash, or medicated pads made especially for the eyes to clean around the eyelid. Others suggest washing the eyelid and the eyelash area with a mild soap wash, made from equal parts of baby shampoo and water. The soap is applied by dipping a cotton ball or clean cloth into the soap wash and washing the eye area gently. This action decreases the risk of infection, especially if the child has recurring styes. This procedure also helps prevent further styes.

Sometimes, antibiotic ointment, sulphonimide, or drops are used. Ointment is applied by putting a thin layer over the sty, usually at bedtime. To insert eye drops into the eye of an older child or adolescent, pull the lower lid down to create a pouch and then put the drops into the pouch. For a younger child, the child can lie down and then close his or her eyes. The eye drops are put into the corner of the affected eye closest to the nose. Then, the child should open his or her eyes so that the eye drops can roll into the eye. Ointment or eye drops usually are prescribed after a sty has been lanced.

The doctor may also decide to surgically drain the sty by lancing it. Usually, not all of the pus is removed if there is acute inflammation. Doing that can deform the eyelid. Sometimes, if a single eyelash is involved, it may be removed to promote healing and drainage.

If the infection does not respond to treatment or spreads to other eye areas or to the lymph nodes in front of the ear, the doctor may prescribe oral antibiotics, such as erythromycin, dicloxacillin, or cephalexin.

Internal hordeola are usually just monitored by the doctor. They may need to be opened and drained or may require antibiotics.

Chalazia are given the same treatment as styes.

Until the sty or chalazion is healed, the child should not wear **contact lenses** and the adolescent should not wear eye makeup.

Alternative treatment

Homeopathic practitioners prescribe oral homeopathic medications that reduce the bacterial growth within the sty and chalazion and thereby heal the inflammation. These medications also eliminate the itching and scaling often experienced along the eyelash line. In addition, homeopathic oral medications are aimed at boosting the immune system. Homeopathic practice usually does not use topical ointments or drops but does recommend using an antibacterial eyewash made from five drops of tincture of goldenseal in a cup of warm water. This is swabbed on the eyelid with a cotton ball. (Since goldenseal can stain, it should be used carefully.)

Prognosis

Most styes heal with minimal treatment in about a week.

Recurring styes may be an indication of a chronic eye infection called *Staphylococcus blepharitis*. In this condition, the eyelids are crusty, reddened, and swollen. The eyes may itch and burn. The base of the eyelashes may have dry scales that flake like dandruff. This condition can cause loss of eyelashes. Both *Staphylococcus blepharitis* and recurring styes can cause scarring of the eyelid.

Prevention

Cleaning the eye with a warm washcloth helps prevent the pores from clogging and a sty from forming. Using a mild, soap wash made with baby shampoo and water will also clean the area and help prevent styes from recurring.

Maintaining good hygiene is essential to preventing styes. Not touching or rubbing the eyes with the hands, especially if they are unwashed, can also prevent styes. Providing separate washcloths and towels for each child will keep bacteria from spreading. In addition, teenagers should not share makeup, especially mascara, eyeshadow, and eyeliner. Eye makeup should be replaced at least every six months because bacteria can grow in these cosmetics and cause infection.

Protecting the eyes from dust and air pollution by using safety glasses when doing dusky work outdoors, such as raking leaves, can also help prevent eye infections. These protections are also good when children are working with craft materials or other materials that may produce airborne particles.

Children's contact lens care should be monitored to maintain cleanliness. Bacteria can contaminate contact lenses or be transmitted by lenses when they are removed or inserted into the eyes. This is a common vector for bacterial transmission into the eye.

Stress has been a contributing factor to the formation of styes, especially in children with immune system disorders. Therefore, stress management techniques could help prevent styes from forming.

Parental concerns

Parents should make sure children wash their hands often and use fresh linens. Contact lenses need to be kept clean and eyes need to be protected when someone is doing dusty chores or art projects. If a child has diabetes

KEY TERMS

Autoimmune disorder—One of a group of disorders, like rheumatoid arthritis and systemic lupus erythematosus, in which the immune system is overactive and has lost the ability to distinguish between self and non-self. The body's immune cells turn on the body, attacking various tissues and organs.

Chalazion—A condition in which clogging of the Meibomiam gland causes a cyst inside the eyelid.

Conjunctivitis—Inflammation of the conjunctiva, the mucous membrane covering the white part of the eye (sclera) and lining the inside of the eyelids also called pinkeye.

Hordeolum—The medical term for sty, an infection or small abscess formation in a hair follicle of the eyelids.

Sty—An external hordeolum caused by an infection of an oil gland on the eyelid.

or an autoimmune disorder, good eye hygiene is as critical as monitoring wound healing and foot care.

See also Conjunctivitis; Eye glasses and contact lenses; Eye and vision development.

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Janie Franz

Subdural hematoma

Definition

A subdural hematoma is a collection of blood in the space between the outer and middle layers of the covering of the brain. It is most often caused by torn, bleeding veins as a result of a head trauma.

Description

The covering of the brain (meninges) has three main layers. The outside is a tough, fibrous covering called the dura mater. The middle layer is the arachnoid mater, and the layer closest to the brain tissue is the pia mater. Subdural hemotamas occur when blood collects in the space between the dura mater and the arachnoid mater. Subdural hematomas usually occur because veins on the inside of the dura that connect the brain cortex and the venous sinuses (bridging veins) are ruptured as the result of a blow to the head. Symptoms can occur within minutes to hours.

Subdural hematomas in children and adolescents are usually abrupt onset or acute and are brought about by accident or injury. Another type of subdural hematoma called a chronic subdural hematoma can occur in people over age 60. However, what follows applies to acute subdural hematomas in children only.

Subdural hemotamas range from fatal or life threatening to small with only minor effects, depending on the quantity of blood released and the amount of injury to other brain tissues. With small subdural hematomas, the blood may slowly be reabsorbed over several weeks without much damage. Larger hematomas, however, can gradually get bigger even though the bleeding has stopped. This enlargement increases pressure inside the skull and can compress the brain, possibly resulting in permanent brain damage or death if the blood is not drained away and the pressure relieved through surgical intervention.

Demographics

In the United States, head injuries are the leading cause of accidental death and permanent disability in people under age 45. Not all these head injuries involve subdural hematoma, but it is the most common type of bleeding in the brain to result from trauma.

Infants are more prone to subdural hematoma than toddlers and older children, because the brain of infants has more room than the brain of older children to move around in the skull when shaken or hit. The neck muscles of infants are also less developed and unable to hold the head steady when shaken.

Children with blood clotting disorders are at an especially high risk of developing bleeding in the brain.

Causes and symptoms

In infants and children, subdural hematoma is often seen in physical **child abuse**. Its presence is one of the defining parameters (along with retinal hemorrhage) of **shaken baby syndrome**. Infants rarely fall until they start learning to walk, so falls account for only a small number of subdural hematomas in infants. However, many subdural hematomas in toddlers result from accidental falls, as they learn to walk and climb. In older children, a fall in which they hit their head is a common cause of subdural hematoma. All age groups are susceptible to developing subdural hematomas from vehicle accidents. In young children, even if the head does not contact a solid surface, the shaking, whiplash movement from some vehicle crashes causes blood vessels to burst in the brain.

Symptoms of subdural hematoma tend to fluctuate and include the following:

- **headache**
- episodes of confusion and drowsiness
- one-sided weakness or paralysis

- lethargy
- enlarged or asymmetric pupils
- convulsions
- increased intracranial pressure
- loss of consciousness after **head injury**
- coma

When to call the doctor

Individuals who show any immediate symptoms of subdural hematoma should be taken to the emergency room. Infants and children should be checked by a doctor if they have had a hard fall or accident in which they have hit their head or if child abuse or shaken baby syndrome is suspected.

Diagnosis

Diagnosis is made based on history, external signs and symptoms of head injury (although external injuries may not always be present), and confirmed through magnetic resonance imaging (MRI). X rays may be done so the doctor can look for skull fracture.

Treatment

Small hematomas that do not cause symptoms may not need to be treated. Otherwise, the hematoma should be surgically removed. Liquid blood can be drained from burr holes drilled into the skull. The surgeon may have to open a section of skull (craniotomy) to remove a large clot and/or to tie off the bleeding vein.

Corticosteroids and diuretics may be given to help control brain swelling, depending on the age of the child and the extent of the injury. After surgery, anticonvulsant drugs such as phenytoin may help control or prevent seizures, which can begin as late as two years after the head injury.

Prognosis

The outcome of subdural hematoma depends on how promptly treatment is received and how much damage the brain has received. Head injuries have a high mortality rate. The mortality rate for all patients with acute subdural hematoma is about 60 percent. Even when recovery occurs, permanent disability can occur. Headache, amnesia, attention problems, **anxiety**, and personality changes may continue for some time after surgery.

KEY TERMS

Corticosteroids—A group of hormones produced naturally by the adrenal gland or manufactured synthetically. They are often used to treat inflammation. Examples include cortisone and prednisone.

Diuretics—A group of drugs that helps remove excess water from the body by increasing the amount lost by urination.

Prevention

Preventing blunt head trauma from falls, child abuse, and assaults is the most effective way of preventing subdural hematoma.

Parental concerns

Research in the early 2000s suggests that some of the effects of brain injury do not show up in children until several years after the injury. These include the development of social and academic skills. Parents should be alert to this possibility.

See also Child abuse.

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Carol A. Turkington

Substance abuse and dependence

Definition

Substance abuse is a pattern of behavior that displays many adverse results from continual use of a substance. Substance dependence is a group of behavioral and physiological symptoms that indicate the continual, compulsive use of a substance in self-administered doses despite the problems related to the use of the substance.

Description

The characteristics of abuse are a failure to carry out obligations at home or work, continual use under circumstances that present a hazard (such as driving a car), and legal problems such as arrests. Use of the drug is persistent despite personal problems caused by the effects of the substance on the self or others. In substance dependence, as the patient's tolerance for the drug increases, increased amounts of a substance are needed to achieve the desired effect or level of intoxication. Withdrawal is a physiological and psychological change that occurs when the body's concentration of the substance declines in a person who has been a heavy user.

Substance abuse and dependence cuts across all lines of race, culture, educational, and socioeconomic status, leaving no group untouched by its devastating effects. An estimated 13 million Americans abuse or are dependent on an illegal substance. Substance abuse is an enormous public health problem, with far-ranging effects throughout society. In addition to the toll substance abuse can take on one's physical health, substance abuse is considered to be an important factor in a wide variety of social problems, affecting rates of crime, domestic violence, **sexually transmitted diseases** (including HIV/AIDS), unemployment, homelessness, teen pregnancy, and failure in school. An estimated 20 percent of the total yearly cost of health care in the United States is spent on the effects of drug and alcohol abuse.

A wide range of substances can be abused. The most common classes include the following:

- opioids, including such prescription **pain** killers as morphine and demerol, as well as illegal substances such as heroin
- benzodiazapines, including prescription drugs used for treating **anxiety**, such as valium
- sedatives or "downers," including prescription barbiturate drugs commonly referred to as tranquilizers
- stimulants or "speed," including prescription amphetamine drugs used as weight loss drugs and in the treatment of attention deficit disorder
- cannabinoid drugs obtained from the hemp plant, including marijuana and hashish
- cocaine-based drugs, including cocaine and "crack"
- hallucinogenic or psychedelic drugs, including lysergic acid diethylamide (LSD) or "acid," phencyclidine (PCP) or "angel dust," 3-4 methylenedioxymethamphetamine (MDMA) or "ecstasy," and other PCP-type drugs
- inhalants, including gaseous drugs used in the medical practice of anesthesia, as well as such common substances as paint thinner, gasoline, and glue
- alcoholic drinks
- cigarettes, cigars, and other tobacco products

Those substances of abuse that are actually prescription medications may have been obtained on the street by fraudulent means or may have been a legal, medically indicated prescription that a person begins to use without regard to the directions of his or her physician.

A number of important terms must be defined in order to have a complete discussion of substance abuse. Drug tolerance refers to a person's body being accustomed to the symptoms produced by a specific quantity of a substance. When a person first begins taking a substance, he or she will note various mental or physical reactions brought on by the drug (some of which are the very changes in consciousness that the individual is seeking through substance use). Over time with repeated use, the same dosage of the substance produces fewer of the desired feelings. In order to continue to feel the desired effect of the substance, progressively higher drug doses must be taken.

Demographics

The National Survey on Drug Use and Health (NSDUH) is conducted annually by the Substance Abuse and Mental Health Services Administration (SAMHSA) of the U.S. Department of Health and Human Services. In 2003, the study found the rate of substance dependence or abuse was 8.9 percent for youths aged 12 to 17 and 21 percent for persons aged 18 to 25. Among persons with substance dependence or abuse, illicit drugs accounted for 58.1 percent of youths and 37.2 percent of persons aged 18 to 25. In 2003, males were almost twice as likely to be classified with substance dependence or abuse as females (12.2% versus 6.2%). Among youths aged 12 to 17, however, the rate of substance dependence or abuse among females (9.1%) was similar to the rate among males (8.7%). The rate of substance dependence or abuse was highest among Native Americans and Alaska Natives (17.2%). The next highest rates were among Native Hawaiians and other Pacific Islanders (12.9%) and persons reporting mixed ethnicity (11.3%). Asian Americans had the lowest rate (6.3%). The rates among Hispanics (9.8%) and whites (9.2%) were higher than the rate among blacks (8.1%).

Rates of drug use showed substantial variation by age. For example, in 2003, 3.8 percent of youths aged 12 to 13 reported current illicit drug use compared with 10.9 percent of youths aged 14 to 15 and 19.2 percent of youths aged 16 to 17. As in other years, illicit drug use in 2003 tended to increase with age among young persons, peaking among 18 to 20-year-olds (23.3%) and declining steadily after that point with increasing age. The prevalence of current alcohol use among adolescents in 2003 increased with increasing age, from 2.9 percent at age 12 to a peak of about 70 percent for persons 21 to 22 years old. The highest prevalence of both binge and heavy drinking was for young adults aged 18 to 25, with the peak rate of both measures occurring at age 21. The rate of binge drinking was 41.6 percent for young adults aged 18 to 25 and 47.8 percent at age 21. Heavy alcohol use was reported by 15.1 percent of persons aged 18 to 25 and 18.7 percent of persons aged 21. Among youths aged 12 to 17, an estimated 17.7 percent used alcohol in the month prior to the survey interview. Of all youths, 10.6 percent were binge drinkers, and 2.6 percent were heavy drinkers, similar to the 2002 numbers.

In 2003 rates of illicit drug use varied significantly among the major racial-ethnic groups. The rate of illicit drug use was highest among Native Americans and Alaska Natives (12.1%), persons reporting two or more races (12%), and Native Hawaiians and other Pacific Islanders (11.1%). Rates were 8.7 percent for African Americans, 8.3 percent for Caucasians, and 8 percent for Hispanics. Asian Americans had the lowest rate of illicit drug use at 3.8 percent. These rates were unchanged from 2002. Native Americans and Alaska Natives were more likely than any other racial-ethnic group to report the use of tobacco products in 2003. Among persons aged 12 or older, 41.8 percent of Native Americans and Alaska Natives reported using at least one tobacco product in the past month. The lowest current tobacco use rate among racial-ethnic groups in 2003 was observed for Asian Americans (13.8%), a decrease from the 2002 rate (18.6%).

Young adults aged 18 to 25 had the highest rate of current use of cigarettes (40.2%), similar to the rate in 2002. Past month cigarette use rates among youths in 2002 and 2003 were 13 percent and 12.2 percent, respectively, not a statistically significant change. However, there were significant declines in past year (from 20.3% to 19%) and lifetime (from 33.3% to 31%) cigarette use among youths aged 12mto 17 between 2002 and 2003. Among persons aged twelve or older, a higher proportion of males than females smoked cigarettes in the past month in 2003 (28.1% versus 23%). Among youths aged 12 to 17, however, girls (12.5%) were as likely as boys (11.9%) to smoke in the past month. There was no change in cigarette use among boys aged 12 to 17 between 2002 and 2003. However, among girls, cigarette use decreased from 13.6 percent in 2002 to 12.5 percent in 2003.

Causes and symptoms

There is not thought to be a single cause of substance abuse, though scientists are as of 2004 increasingly convinced that certain people possess a genetic predisposition which can affect the development of addictive behaviors. One theory holds that a particular nerve pathway in the brain (dubbed the "mesolimbic reward pathway") holds certain chemical characteristics which can increase the likelihood that substance use will ultimately lead to substance **addiction**. Certainly, however, other social factors are involved, including **family** problems and **peer pressure**. Primary **mood disorders** (bipolar), **personality disorders**, and the role of learned behavior can influence the likelihood that a person will become substance dependent.

The symptoms of substance abuse may be related to its social effects as well as its physical effects. The social effects of substance abuse may include dropping out of school or losing a series of jobs, engaging in fighting and violence in relationships, and legal problems (ranging from driving under the influence to the commission of crimes designed to obtain the money needed to support an expensive drug habit).

When to call the doctor

The earlier one seeks help for their child or teen's substance abuse or dependence problems, the better. Regarding the matter of determining if a teen is experimenting or moving more deeply into the drug culture, parents must be careful observers, particularly of the little details that make up a teen's life. Dramatic change in appearance, friends, or physical health may be signs of trouble. If parents believe their child may be drinking or using drugs, they should seek help through a substance abuse recovery program, family physician, or mental health professional.

Diagnosis

The most difficult aspect of diagnosis involves overcoming the patient's denial. Denial is a psychological trait that prevents a person from acknowledging the reality a situation. Denial may cause a person to be completely unaware of the seriousness of the substance use or may cause the person to greatly underestimate the degree of the problem and its effects on his or her life. A physical examination may reveal signs of substance abuse in the form of needle marks, tracks, trauma to the inside of the nostrils from snorting drugs, unusually large or small pupils. With the person's permission, substance use can also be detected by examining in a laboratory an individual's blood, urine, or hair. This drug testing is limited by sensitivity, specificity, and the time elapsed since the person last used the drug.

Treatment

Treatment has several goals, which include helping a person deal with the uncomfortable and possibly life-threatening symptoms associated with withdrawal from an addictive substance (called detoxification), helping a person deal with the social effects which substance abuse has had on his or her life, and efforts to prevent relapse (resumed use of the substance). Individual or group psychotherapy is sometimes helpful.

Detoxification may take from several days to many weeks. Detoxification can be accomplished suddenly, by complete and immediate cessation of all substance use or by slowly decreasing (tapering) the dose that a person is taking, to minimize the side effects of withdrawal. Some substances absolutely must be tapered, because "cold turkey" methods of detoxification are potentially life threatening. Alternatively, a variety of medications may be used to combat the unpleasant and threatening physical symptoms of withdrawal. A substance (such as methadone in the case of heroine addiction) may be substituted for the original substance of abuse, with gradual tapering of this substituted drug. In practice, many patients may be maintained on methadone and lead a reasonably normal life style. Because of the rebound effects of wildly fluctuating blood pressure, body temperature, heart and breathing rates, as well as the potential for bizarre behavior and hallucinations, a person undergoing withdrawal must be carefully monitored.

Alternative treatment

Alternative treatments thought to improve a person's ability to stop substance use include acupuncture and hypnotherapy. Ridding the body of toxins is believed to be aided by hydrotherapy (bathing regularly in water containing baking soda, sea salt or Epsom salts). Hydrotherapy can include a constitutional effect where the body's vital force is stimulated and all organ systems are revitalized. Elimination of toxins is aided as well as by such herbs as milk thistle, burdock, and licorice. Anxiety brought on by substance withdrawal is thought to be lessened by using other herbs, for example valerian, vervain, skullcap, and kava.

Prognosis

After a person has successfully withdrawn from substance use, the even more difficult task of recovery begins. Recovery refers to the life-long efforts of a person to avoid returning to substance use. The craving can be so strong, even years and years after initial withdrawal has been accomplished, that a previously addicted person may be virtually forever in danger of slipping back into substance use. Triggers for such a relapse include any number of life stresses (problems on the job or in the marriage, loss of a relationship, death of a loved one, financial stresses), in addition to seemingly mundane exposure to a place or an acquaintance associated with previous substance use. While some people remain in counseling indefinitely as a way of maintaining contact with a professional who can help monitor behavior, others find that various support groups or twelve-step programs such as Narcotics Anonymous and Alcoholics Anonymous are the most helpful ways of monitoring the recovery process and avoiding relapse.

Prevention

Prevention is best aimed at teenagers, who are at very high risk for substance experimentation. Education regarding the risks and consequences of substance use, as well as teaching methods of resisting peer pressure, are important components of a prevention program. Furthermore, it is important to identify children at higher risk for substance abuse (including victims of physical or

sexual abuse, children of parents who have a history of substance abuse, especially alcohol, and children with school failure or attention deficit disorder). These children may require a more intensive prevention program.

Parental concerns

Parents and guardians need to be aware of the power they have to influence the development of their kids throughout the teenage years. **Adolescence** brings a new and dramatic stage to family life. The changes that are required are not just the teen's to make; parents need to change their relationship with their teenager. It is best if parents are proactive about the challenges of this life cycle stage, particularly those that pertain to the possibility of experimenting with and using alcohol and drugs. Parents should not be afraid to talk directly to their kids about drug use, especially if they have had problems with drugs or alcohol themselves. Parents should give clear, no-use messages about **smoking**, drugs, and alcohol. It is important for kids and teens to understand that the rules and expectations set by parents are based on parental love and concern for their well-being. Parents should also be actively involved and demonstrate interest in their teen's friends and social activities. Spending quality time with teens and setting good examples are essential. Even if problems such as substance abuse already exist in the teen's life, parents and families can have a positive influence on their teen's behavior.

KEY TERMS

Addiction—The state of being both physically and psychologically dependent on a substance.

Dependence—A state in which a person requires a steady concentration of a particular substance to avoid experiencing withdrawal symptoms.

Detoxification—The process of physically eliminating drugs and/or alcohol from the system of a substance-dependent individual.

High—The altered state of consciousness that a person seeks when abusing a substance.

Street drug—A substance purchased from a drug dealer. It may be a legal substance, sold illicitly (without a prescription, and not for medical use), or it may be a substance which is illegal to possess.

Tolerance—A condition in which an addict needs higher doses of a substance to achieve the same effect previously achieved with a lower dose.

Withdrawal—The characteristic withdrawal syndrome for alcohol includes feelings of irritability or anxiety, elevated blood pressure and pulse, tremors, and clammy skin.

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Sucrose intolerance *see* **Carbohydrate intolerance**

Sudden infant death syndrome

Definition

Sudden infant death syndrome (SIDS) is the sudden, unexpected death of a seemingly normal, healthy infant under one year of age that remains unexplained after a thorough postmortem investigation, including an autopsy and a review of the case history.

Description

SIDS is a defined medical disorder that is listed in the *International Classification of Diseases, 9th Revision* (ICD-9). The first published research about sudden infant death appeared in the mid-nineteenth century. Since then, researchers and healthcare providers have struggled to define the syndrome and determine its causes. The key characteristics of SIDS include:

- infant less than one year of age
- infant seemingly healthy (no preceding symptoms)
- complete investigation fails to find a cause of death
- no associated **child abuse** or illness

Demographics

In the United States, SIDS was the third leading cause of postneonatal deaths (those occurring between the ages of 28 days and one year) in 2001. According to the National Center for Health Statistics, 2,234 infants in the United States died of SIDS in 2001, or 8.1 percent of total infant deaths. (In the late 1990s, many sources placed the annual total number of deaths as high as 6,000 due to possible under-reporting.) Ninety percent of SIDS deaths occur during the first six months of life, mostly between the ages of two and four months. SIDS also occurs about 1.5 times more frequently in boys than girls. The rate of SIDS in African-American infants is twice as high as that of Caucasians, a fact often attributed to the lower quality of prenatal care received by many African-American mothers.

Causes and symptoms

Studies have identified many risk factors for SIDS, but the actual cause of the disorder remains a mystery. Although investigators are still not sure whether the immediate cause of SIDS deaths is due to respiratory failure or cardiac arrest, patterns of infant **sleep**, breathing, and arousal are a major focus of research in the early 2000s. It is known that young infants often stop breathing for short periods of time, then gasp and start again. Some researchers and physicians believe that SIDS involves a flaw in the mechanism that is responsible for resumption of breathing.

Aside from its occurrence during sleep, the other most striking feature of SIDS is its narrow age distribution, which has prompted researchers to examine the developmental changes that take place between the ages of two and four months, especially between the ages of two and four months, when most SIDS deaths occur. A growing number of experts believe that rather than a single cause, there are a number of different conditions that can cause or contribute to SIDS. This picture is complicated still further by the interaction of possible physical abnormalities with a number of environmental and developmental factors known to increase the risk of SIDS. Premature infants and low birth weight babies in general are known to be at increased risk of developing SIDS, as are infants born to teenage mothers, poor mothers, and mothers who for any reason have had inadequate prenatal care. Other risk factors include maternal **smoking** during pregnancy, exposure to smoking in the home after birth, formula feeding rather than breastfeeding, and prior death of a sibling from SIDS (although this is thought to be due to shared environmental risk factors rather than genetic predisposition). Many SIDS deaths occur in babies who have recently had colds (a possible reason that SIDS is most prevalent in winter, the time when upper respiratory infections are most frequent).

As of 2004, the most significant risk factor discovered for SIDS was placing babies to sleep in a prone position (on their stomachs). Studies have reported that anywhere from 28 percent to 52 percent of infants who die of SIDS are found lying face down. Another finding reinforcing the connection between SIDS and prone-sleeping is the fact that SIDS rates are higher in Western cultures, where women have traditionally placed children on their stomachs, than in Eastern ones, where infants usually sleep on their backs. The cause-effect

relationship between prone-sleeping and SIDS is not fully understood. However, it is known that when infants sleep on their backs they are more prone to arousal, and SIDS is often thought to involve a failure to rouse from sleep. In addition, prone-sleeping raises a baby's temperature, which is another risk factor for the disorder.

When to call the doctor

Parents or caregivers should immediately call for emergency care if a child is found not breathing or without a pulse or is unable to be aroused from sleep.

Diagnosis

In most cases, three techniques are used in an attempt to determine the cause of an infant's death. These are:

- Death scene investigation. A thorough examination of the scene of death, including recording baby's position, collecting items from the surrounding area, and interviewing **family** members and/or caregivers, can sometimes point to an external cause of death.
- Autopsy. The autopsy, usually performed by a medical examiner or coroner, focuses on finding any identifiable cause of death. While parents may reject the idea of an autopsy because they feel it violates their infant's remains, it is often the only tool that can definitively rule out other potential causes of death.
- Review of family history. Healthcare providers or police interview parents and/or caregivers in order to determine the child's medical and family history, in an attempt to rule out possible illness, child abuse, or other cause of death.

Treatment

Because SIDS affects seemingly healthy infants, and death is the first symptom of the disorder, it is not possible to treat an infant who is truly affected by SIDS. If life support is implemented and the child is resuscitated, emergency care will be provided in an attempt to stabilize the child. Healthcare personnel perform a complete medical exam and record the child's medical history to exclude other potential causes.

Prognosis

By definition the prognosis for babies affected by SIDS is invariably death. In some rare cases, emergency care providers are able to resuscitate an infant who is seemingly lifeless; the prognosis remains poor in these cases.

Prevention

In the 1990s a number of countries initiated campaigns aimed at getting parents to put their infants to sleep on their backs or sides. In the United States, the American Academy of Pediatrics (AAP) in 1992 issued an official recommendation that infants be put to bed on their backs (supine position) or on their sides (lateral position). In 1994 the Public Health Service launched its "Back to Sleep" campaign, targeting parents, other care givers, and healthcare personnel with brochures advocating supine or lateral infant sleeping and also including information about other risk factors for SIDS. By the mid-1990s it was apparent that this and similar campaigns worldwide had had a significant—in many cases dramatic—impact in reducing the number of deaths from SIDS. In a number of countries the incidence of SIDS dropped by 50 percent or more. SIDS deaths in Great Britain were reduced by 91 percent between 1989 and 1992; in Denmark they declined by 72 percent between 1991 and 1993; and they were reduced by 45 percent in New Zealand between 1989 and 1992.

In the United States, the AAP recommendations reduced the incidence of front-sleeping in infants from over 70 percent in 1992 to 24 percent in 1996. A decline in SIDS rates, already observed in the 1980s, tripled its previous pace between 1990 and 1994, with SIDS deaths falling 10 to 15 percent between 1992 and 1994. Links between SIDS and other aspects of an infant's sleep environment have also emerged. The best known is the finding that soft, padded sleep surfaces can endanger infants by obstructing breathing or creating air pockets that trap their expelled carbon dioxide, which they can then inhale.

Some research also suggests that co-sleeping (having an infant sleep with the mother in her bed) can help regulate an infant's sleep pattern in ways that reduce the risk of SIDS. (Like supine infant sleeping, co-sleeping is also prevalent among Asian populations, which have a low incidence of SIDS.) Infants who share their mothers' beds become accustomed to frequent minor arousals when the mother shifts position, and their own sleep tends to be lighter and more even than that of infants who sleep alone in their cribs and are more prone to the heavier, but sporadic, breathing that stops and then starts up again with a gasp. Experts speculate that this lighter sleep not only makes it less likely for an infant to stop breathing but also that such an infant, with the "practice" gained from more frequent arousals every night, can be aroused more easily when any respiratory distress does occur. In addition, infants who co-sleep with their mothers are naturally more likely to sleep on their backs or sides, which also reduces the risk of SIDS.

In December 1996 the AAP issued the following updated recommendations regarding infant sleep:

- Infants should be put to sleep in a nonprone position. The supine position (on their backs) is safest, but sleeping on their sides can also significantly reduce the risk of SIDS. When infants sleep on their sides, the bottom arm should be extended to prevent them from rolling over on to their stomachs.
- Soft sleeping surfaces should be avoided, and a sleeping infant should not be placed on soft objects such as pillows or quilts.
- It may be better for parents, with the guidance of their pediatrician, to depart from these recommendations in the case of infants with certain health problems, such as gastroesophageal reflux (GER).
- Infants should spend some time lying on their stomachs when they are awake and supervised by an adult.

Other precautions parents can take include obtaining adequate prenatal care; avoiding exposing infants to cigarette smoke, either pre- or postnatally; breastfeeding instead of formula feeding; and not allowing an infant to become overheated while sleeping. Another measure taken by some parents is the use of a portable battery-operated monitor that sounds an alarm in response to significant deviations in infants' respiration or heart rates while they are asleep. Monitoring is based on the belief that if parents can quickly reach an infant who has stopped breathing, they can either get him breathing again themselves or call for emergency assistance. There has been no substantiated link between monitoring and the decrease in SIDS, and infants have, in fact, died while being monitored. Nevertheless, monitors provide peace of mind for many parents, especially those who have already lost an infant to SIDS or whose baby has special risk factors for the disorder. Medical opinion is generally in favor of monitoring only for newborns who have had episodes of apnea (cessation of breathing) or for any infant who has had a precipitous, life-threatening interruption of breathing or cardiovascular function.

Parental concerns

Losing a child—a traumatic experience for any parent—is especially difficult for those who lose a child to SIDS because the death is so sudden and its cause can often not be determined. Parents of a child who dies of SIDS do not gain a medical explanation of their infant's death. Although such an understanding does not lessen their loss, it can serve an important function in the healing process, one that SIDS parents do not have. In addition to the emotions that normally accompany grief, such as denial, anger, and guilt, SIDS parents may experience certain other reactions unique to their situation. They may become fearful that another unexpected disaster will strike them or members of their families. After the death of a child from SIDS, parents often become overprotective of the infant's older siblings and of any children born subsequently. Some **fear** having another child, due to misgivings that the tragedy they have experienced may repeat itself. Parents of children who die of SIDS often make major changes in their lives during the period following the death, such as relocating or changing jobs, as a way to avoid confronting painful memories or as a way to protect themselves against the SIDS death of another baby by changing the circumstances of their lives as much as possible.

SIDS deaths place a great strain on marriages. Parents' individual ways of coping with their grief may prevent them from giving each other the support they need, creating an emotional distance between them. Nevertheless, the **divorce** rate among SIDS parents appears to be no higher than that for the general population, and in one survey half the respondents reported that their marriages had ultimately been strengthened by the experience.

A SIDS death also has a significant effect on the infant's siblings. Young children often experience developmental regressions in **toilet training** or other areas. Some fear going to sleep, which they associate with the death of their baby brother or sister. As with any death in the family, children need to be reassured that they are not guilty in any way. Many pose difficult questions to their parents, wanting to know why the baby died or where he has gone, or even whether they are going to die, too. Children may also come to feel jealous of the attention paid to the infant who has died or resentful of the disruption the death has caused in their family's life. Most parents report that their way of caring for their remaining children changes after the family experiences a SIDS death. Having young children (or infants born later on) sleep with them at night makes some parents feel more confident of preventing a second tragedy from occurring. In addition to overprotecting their children and worrying about their health, SIDS parents may also spoil them and find it hard to say no to their requests. On the positive side, many parents simply value their remaining children more, spend more time with them, and become closer to them. In a minority of cases, however, the reverse happens, and parents feel emotionally distant from their surviving children. In addition, fear of being hurt sometimes makes it difficult for some parents to bond with babies born later.

Many parents of infants who die of SIDS are helped by participating in local support groups, where they can share their feelings and experiences with others who

KEY TERMS

Apnea—The temporary absence of breathing. Sleep apnea consists of repeated episodes of temporary suspension of breathing during sleep.

Co-sleeping—Having an infant sleep with the mother in her bed.

Gastroesophogeal reflux—Backward flow of stomach contents into the esophagus.

have undergone the same experience. Counseling can also be beneficial, especially with a mental health professional experienced in dealing with parental grief.

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National SIDS/Infant Death Resource Center. 2070 Chain Bridge Rd., Suite 450, Vienna, VA 22182. Web site: <www.sidscenter.org>.

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Sugar diabetes *see* **Diabetes mellitus**

Sugar intolerance *see* **Carbohydrate intolerance**

Suicide and suicidal behavior

Definition

Suicide is the act of ending one's own life. Suicidal behavior are thoughts or tendencies that put a person at risk for committing suicide.

Description

Suicide, attempted suicide, and thoughts of committing suicide are, as of the early 2000s, growing problems among adolescents in the United States and much of the world. It is the third leading cause of death among 15 to 19 year olds in the United States and the sixth leading cause of death among 10 to 14 year olds. About 2 percent of adolescent girls and 1 percent of adolescent boys attempt suicide each year in the United States. Another 5 to 10 percent of children and teens each year come up with a plan to commit suicide.

Psychologists have identified the teenage years as one of the most difficult phases of human life. Although they are often seen as a time in which to enjoy life, hang out with friends, and perform other activities that adults would not usually do, the teenage period can be difficult. Many changes in the human mind take place during **puberty**. Apart from facing the onset of sexual maturity, teenagers must also make key decisions about their future, develop their identities, change schools, and meet new friends. They may have to cope with a wide range of personal and social challenges. Many young people have difficulty dealing with stress these experiences may elicit.

The most common reasons for suicide or suicidal behavior among children and adolescents involve personal conflict or loss, most frequently with parents or romantic attachments. **Family** discord, physical or sexual abuse, and an upcoming legal or disciplinary crisis are also commonly associated with completed and attempted suicide. Adolescents who complete suicide show relatively high suicidal intent, and many are intoxicated at the time of death. The most serious suicide attempters leave suicide notes, show evidence of planning, and use an irreversible method, such as a gunshot to the head. Most adolescent suicide attempts, though, are of relatively low intent and lethality, and only a small number of these individuals actually want to die. Usually, suicide attempters want to escape psychological **pain** or unbearable circumstances, gain attention, influence others, or communicate strong feelings, such as anger or love.

Suicidal behavior is rare in children prior to puberty, probably because of their relative inability to plan and

execute a suicide attempt. Psychiatric risk factors, such as depression and substance abuse, become more frequent in **adolescence**, contributing to the increase in the frequency of suicidal behavior in older children. Some view the transition from primary to middle school as particularly stressful, especially for girls. Also, parental monitoring and supervision decrease with increasing age, so that adolescents may be more likely to experience emotional difficulties without their parents' knowledge.

Repeated suicide attempts are common, but rates vary. Follow-up studies ranging from one to 12 years found a re-attempt rate among adolescents of 5 to 6 percent per year, with the greatest risk within the first three months after the initial attempt. Factors associated with a higher re-attempt rate included chronic and severe psychiatric disorders, such as depression and substance abuse; hostility and aggression; non-compliance with treatment; poor levels of social skills; family discord, neglect, or abuse; and parental psychiatric disorders.

Highest risk

Four out of five teenagers who successfully commit suicide are male, but the average female teenager is prone to attempt suicide four more times during her teen years than the average male. White teenage males are more likely to commit suicide than other ethnic groups, but as of the early 2000s teenage suicide among blacks is also increasing. Teenagers who have unsuccessfully tried to commit suicide in the past are more likely to attempt suicide in the future. The odds increase after each failed attempt. There are two groups of teens that are at a particularly high risk for committing suicide: Native Americans, and gay, lesbian, bisexual, and transgendered teens.

In Native American, including Native Alaskan, youth ages 15 to 24 years, suicide is the second leading cause of death, according to a 2001 survey by the Bureau of Indian Affairs. The survey also showed that 16 percent of Native American youth attempted suicide in the preceding year. Among Native American high school students, suicide attempts were most associated with poor school performance, poor physical health, a history of family or friends who committed or attempted suicide, family problems, and physical and sexual abuse.

Gay and bisexual male teens, which represent about 10 percent of the male teen population, are six to seven times more at risk for attempting suicide than their heterosexual peers. Several surveys show gay and lesbian youth account for 30 percent of all suicides among teens, according to the U.S. Department of Health and Human Services. Yet most studies of teen suicide have not been concerned with identifying sexual orientation.

A 1997 study by the Massachusetts Department of Education found that 46 percent of high school students who identified themselves as gay, lesbian, or bisexual, had attempted suicide in the past year compared to 8.8 percent of their heterosexual peers. Of the gay, lesbian, and bisexual teens, 23.5 percent required medical care as a result of their suicide attempt compared to 3.3 percent of heterosexual students who attempted suicide.

Common problems

The following are common risk factors for teenage suicide:

- Psychological problems: Depression, previous attempts at suicide, and having received psychiatric care in the past.
- Personal failure: Unmet high standards set by the teen or parents, including failing grades in school or poor performance in sports.
- Recent loss: Death of a close friend or family member, **divorce**, **abandonment** by a parent, pregnancy, and the breakup with a boyfriend or girlfriend.
- Substance abuse: Abuse of alcohol and other drugs as forms of self-medication for overwhelming depression. A combination of depression, substance abuse, and lowered impulse control can lead to suicide or attempted suicide. Substance abuse in other family members can also lead to suicide.
- Household guns: Easy access to a gun. Children of law enforcement officers have a much higher suicide rate because of the accessibility of guns in their houses. The most common method of suicide among teens is gunshot.
- Violence: Violence against the teen either at home or outside the home, including physical, emotional, or sexual abuse, or bullying. Violence at home or against the youth teaches teens that the way to resolve conflict is through violence, and suicide is the ultimate act of self-violence.
- Communication problems: The inability to discuss anger or other uncomfortable feelings with family members or friends. These feelings can include loneliness, rejection, and awareness of one's gay or bisexual sexual orientation.

Parental concerns

Parents who are concerned that their child is or may be suicidal should seek help immediately, such as from a psychiatrist, psychologist, or counselor. Therapists and counselors can listen to the child talk about his or her

problems and may be able to suggest ways to cope which the teen will find useful.

There are a number of ways parents can help children and teens deal with loneliness, depression, and suicidal feelings. First, they can let the child do the talking, and listen carefully. They can let the child know they take his or her feelings and thoughts seriously. They can try to identify the root of the problem. Second, they can ask direct questions, such as "Are you thinking of committing suicide?" or "Are you thinking of ending your life?" Third, they can stay with the child. Parents should not leave their child alone if the child says he or she wants to commit suicide. By staying with the child, the parent may be protecting the child's life.

When to call the doctor

Many doctors recommend that teenagers be taken to a hospital immediately after they express the desire to commit suicide. At the least, immediate psychological help should be sought. There are many methods, both medical and psychological, of helping teenagers who consider committing suicide. Most teenagers who think of suicide believe their problems are too hard to solve or too embarrassing to talk about, so it is important for their helpers to show they are trustworthy and able to listen. Seeing a psychologist is widely recommended as well. A psychologist may be able to improve a teenager's vision of life by listening to the young person and conveying optimism regarding the future.

Doctors recommend that helpers not ask the teenager's reason for thinking of suicide; rather, helpers should listen and wait for the teenager to trust enough to talk openly about the problem. Helpers should, however, be understanding of the teenager's situation. Doctors also recommend that helpers not mention "reasons for living," as doing so might generate more depressing thoughts in the teenager.

There are many telephone hot lines available, on national, state, and local levels, to help teenagers who are considering suicide. Two national, 24-hour, toll-free suicide hotlines are: 800–784–2433 and 800–999–9999. Gay, lesbian, bisexual, or transgendered teens thinking of suicide can get help at 800–850–8078.

KEY TERMS

Puberty—The point in development when the ability to reproduce begins. The gonads begin to function and secondary sexual characteristics begin to appear.

Transgendered—Any person who feels their assigned gender does not completely or adequately reflect their internal gender, such as a biological male who perceives himself to be female.

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Suicide Awareness Voices of Education. 7317 Cahill Road, Suite 207, Minneapolis, MN 55424. Web site: <www.save.org>.

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Sulfonamides

Definition

Sulfonamides, sometimes called sulfa drugs, are medicines that prevent the growth of bacteria in the body. The sulfonamides have largely been replaced by the **antibiotics** which generally are safer and more effective.

Description

Sulfonamides are used to treat many kinds of infections caused by bacteria and certain other microorganisms. Physicians may prescribe these drugs to treat urinary tract infections, ear infections, frequent or long lasting **bronchitis**, bacterial **meningitis**, certain eye infections, *Pneumocystis carinii* **pneumonia**, traveler's **diarrhea**, and a number of other kinds of infections. These drugs will not work for colds, flu, and other infections caused by viruses.

Description

Although there were many sulfonamides, relatively few are in use as of 2004:

- Sulfisoxazole (Gantrisin) is used to treat urinary tract infections. In combination with erythromycin, sulfisoxazole may be used to treat ear infections in children.
- Trimethoprim/sulfamethoxazole (Bactrim, Septra) is a combination of two sulfonamides used together. The combination is more effective than giving either drug alone in a larger dose. The combination is commonly used to treat urinary tract infections and other infections that cannot be treated with antibiotics.
- Sulfadiazine may be used to protect people with **rheumatic fever** from infections. It is used to treat **toxoplasmosis**. An ointment containing silver sulfadiazine is widely used for treatment of burns.
- Sulfasalazine (Azulfadine) is used to treat infections of the colon and intestine.

General use

The most common use for sulfonamides in adults is treatment of urinary tract infections. In children, sulfonamides have more limited use. Sulfisoxazole may be used for prophylaxis of ear infections and prevention of meningococcal infections. Sulfasakazube is used to treat children over the age of two years with ulcerative colitis.

Precautions

Sulfonamides should never be used in infants under the age of two months. They should also be used with extreme care in patients with liver problems, kidney problems, and some types of anemia.

Side effects

Although such side effects are rare, some people have severe and life-threatening reactions to sulfonamides. These include sudden, severe liver damage; serious blood problems; breakdown of the outer layer of the skin; and a condition called Stevens-Johnson syndrome, in which people get blisters around the mouth, eyes, or anus. People should call a physician immediately if any of the following signs of a dangerous reaction occur:

- skin rash or reddish or purplish spots on the skin
- other skin problems, such as blistering or peeling
- fever
- sore throat
- cough
- shortness of breath
- joint pain
- pale skin
- yellow skin or eyes

This medicine may cause **dizziness**. Sulfonamides may also cause blood problems that can interfere with healing and lead to additional infections. This medicine may increase sensitivity to sunlight. Even brief exposure to sun can cause severe **sunburn** or rash. While being treated with this medicine, people should avoid being in direct sunlight. Very rarely, systemic sulfonamides may even cause kidney stones.

The most serious adverse effects of sulfonamides cannot be predicted. Some steps can minimize some of the less severe adverse effects. Because sulfonamides are

not very soluble, they should always be taken with a full glass of water. Moreover, sulfonamides increase sensitivity to sunlight and increase the risk of sunburn. People taking sulfonamides by mouth should avoid direct sunlight and stay covered up. They should not rely on **sunscreens**. This risk does not apply, however, to people using sulfonamide eye or ear drops. Oral sulfonamides should always be taken in evenly spaced doses to maintain a steady blood level throughout the day.

Interactions

Sulfonamides may interact with a large number of other medicines. When interaction occurs, the effects of one or both of the drugs may change or the risk of side effects may be greater. People who take sulfonamides should let their physician know all other medicines they are taking. Among the drugs that may interact with sulfonamides are:

- acetaminophen (Tylenol)
- medicine for overactive thyroid
- other medicines used to treat infections
- birth control pills
- medicines for diabetes, such as glyburide (Micronase)
- anticoagulants, such as warfarin (Coumadin)
- amantadine (Symmetrel)
- water pills (diuretics) such as hydrochlorothiazide (HCTZ, HydroDIURIL)
- the anticancer drug methotrexate (Rheumatrex)
- antiseizure medicines such as valproic acid (Depakote, Depakene)

The list above does not include every drug that may interact with sulfonamides but is limited to drugs that might be used in treatment of children and adolescents. Parents should be sure to check with a physician or pharmacist before combining sulfonamides with any other prescription or nonprescription (over-the-counter) medicine.

Parental concerns

Parents giving their children eye or ear drops should be sure they know the proper way to administer these drops. Parents should review the technique with a physician or nurse to be sure the medication is given properly. If children are taking sulfonamides by mouth, parents should be sure that the children are drinking a full glass of water with each dose. Because some of the adverse effects of sulfonamides may be very serious, parents should report any suspicious symptoms to their physician promptly.

See also Cystitis; Penicillins; Tetracyclines.

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Sunburn

Definition

Sunburn is an inflammation of the skin caused by overexposure to ultraviolet radiation from the sun.

Description

Sunburn is caused by exposure to the ultraviolet (UV) rays of the sun. There are two types of ultraviolet rays, UVA and UVB. UVB radiation causes most sunburn (about 85%). However, most UVB rays are absorbed by **sunscreens**, but only about half the UVA rays are absorbed.

Although sunburn itself is not a serious health problem in the short term, skin **cancer** from sun overexposure is in the early 2000s a growing problem in the United States. Both UVA and UVB radiation play a role in the development of a form of skin cancer called malignant melanoma. According to the American Cancer Society, melanoma accounts for only 4 percent of all skin cancer, but 79 percent of skin cancer deaths, or about 7,900 deaths annually in the United States. In addition, more than 1 million Americans develop nonmelanoma skin cancer each year, although deaths from this form of cancer are much more rare (about 1,000 per year).

Skin contains a protective pigment called melanin. The darker the skin tone, the more melanin is present. Fair-skinned people are most susceptible to sunburn, because their skin produces only small amounts of the melanin. However, even the darkest-skinned people can get sunburn and skin cancer.

Infants are most susceptible to sunburn and should be kept out of the sun at all times. Children are more susceptible than adults, and because of their outdoor activities get three times more sun exposure on average than adults. It is estimated that one-half to three-quarters of an individual's total number of lifetime sunburns occur in childhood and **adolescence**.

Long-term effects of repeated sun overexposure and burning can cause premature aging and wrinkling of the skin. Overexposure can increase the risk of skin cancer, especially a serious burn in childhood. Individuals at highest risk for developing melanoma are those who have intermittent severe (blistering) sunburns in youth or adolescence.

Occasionally an allergic response to a drug will cause a skin reaction resembling sunburn in the absence of sun exposure.

KEY TERMS

Antibiotics—Drugs that are designed to kill or inhibit the growth of the bacteria that cause infections.

Prophylaxis—Protection against or prevention of a disease. Antibiotic prophylaxis is the use of antibiotics to prevent a possible infection.

Stevens-Johnson syndrome—A severe inflammatory skin eruption that occurs as a result of an allergic reaction or respiratory infection.

Toxoplasmosis—A parasitic infection caused by the intracellular protozoan *Toxoplasmosis gondii*. Humans are most commonly infected by swallowing the oocyte form of the parasite in soil (or kitty litter) contaminated by feces from an infected cat; or by swallowing the cyst form of the parasite in raw or undercooked meat.

Demographics

Infants and children are more likely to get sunburned than adults. Individuals who live in areas where the climate is mostly sunny year round (Arizona, southern California) are at higher risk both for sunburn and skin cancer. Those living at high altitudes are also at higher risk. The chance of being sunburned increases about 4 percent or every 1,000 feet (300 meters) rise in altitude. Fair-skinned, pale, freckled individuals are more likely to get sunburned than individuals with darker skin. Sunburn is extremely common. One poll found that in the summer of 1997, 13 percent of children had developed a sunburn in the preceding week.

Causes and symptoms

The ultraviolet rays in sunlight destroy cells in the outer layer of the skin, damaging tiny blood vessels underneath. When the skin is burned, the blood vessels dilate and leak fluid. Cells stop making certain proteins because their DNA is damaged by the ultraviolet rays. Repeated DNA damage can lead to cancer.

When UV rays burn the skin, immune system defenses that identify the burned skin as foreign are triggered. At the same time, the UV rays transform a substance on the skin that interferes with this immune response. While this keeps the immune system from attacking a person's own skin, it also means that any malignant (cancerous) cells in the skin will be able to grow freely.

Sunburn causes skin to turn red and blister. Symptoms appear from one to 24 hours after sun exposure and peak several days later, after which dead skin cells peel off. In severe cases, the burn may occur with sunstroke (**vomiting**, **fever**, and collapse). Severe cases of sunburn may require **hospitalization**.

When to call the doctor

The doctor should be called any time there are symptoms of heatstroke, **dehydration**, blurred vision (possible sun damage to the eyes), chills, fever, vomiting, or blistering associated with sun exposure.

Diagnosis

Sunburn is easily diagnosed by visual inspection of the skin. No laboratory tests are needed.

Treatment

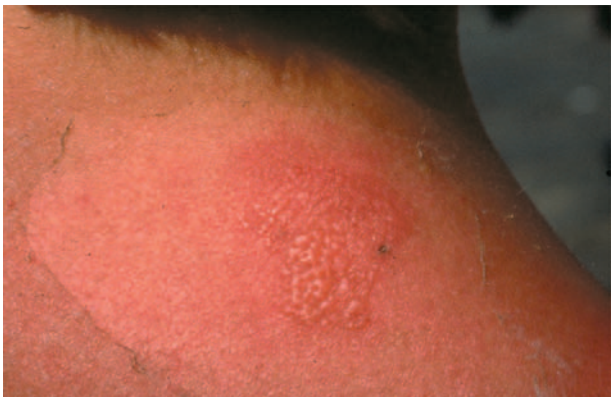
In most cases, treatment involves making the sunburned person more comfortable. The individual should get out of the sun and protect tender skin against more sun exposure for at least one week. **Pain** can be treated with **acetaminophen** (Tylenol) or **nonsteroidal anti-inflammatory drugs** (NSAIDs) such as ibuprofen. Individuals with moderate sunburn over a large area should drink extra water to avoid dehydration. In addition, discomfort may be reduced by using the following:

- calamine lotion
- sunburn cream or spray
- cool tap water compress
- colloidal oatmeal baths
- moisturizer creams to reduce skin peeling

People who are severely sunburned should see a doctor who may prescribe corticosteroid cream to speed healing. Extreme sunburns that blister may require treatment in a hospital burn unit and intravenous fluids to prevent dehydration. Individuals who develop sunburn as the result of a drug reaction should see a doctor promptly.

Alternative treatment

Over-the-counter preparations containing aloe (*Aloe barbadensis*) are an effective treatment for sunburn, easing pain and inflammation while also relieving dryness of the skin. A variety of topical herbal remedies applied as lotions, poultices, or compresses may also help relieve the effects of sunburn. Calendula (*Calendula officinalis*) is one of the most frequently recommended to reduce inflammation.



Patient with a second-degree sunburn on the back of the neck. *(Custom Medical Stock Photo Inc.)*

Prognosis

Short-term prognosis is excellent. Moderately burned skin should heal within a week. While the skin will heal after sunburn, the risk of skin cancer increases with exposure and subsequent **burns**. Even one bad burn in childhood carries an increased risk of skin cancer.

Prevention

Infants under the age of six months should be kept strictly out of the sun. Sunscreens have not been approved for use by infants. Everyone age six months and older should use a water-resistant sunscreen having a sun protective factor (SPF) of at least 15, with an SPF of 30 or more strongly recommended for children. Sunscreen should be applied 15–30 minutes before going outside, as it takes that long to bond effectively with the skin and become effective. Sunscreen should be reapplied every two hours (more often after swimming).

In addition, people should take the following steps:

- Limit sun exposure to 15 minutes the first day, even if the weather is hazy, slowly increasing exposure daily.
- Reapply waterproof sunscreen after swimming for more than 80 minutes, after toweling off, or after perspiring heavily, or every two hours if not swimming.
- Avoid the sun between 10 A.M. and 3 P.M. when the sun is strongest and most direct.
- Wear a hat or cap to protect the face.

KEY TERMS

Malignant melanoma—The most serious of the three types of skin cancer, malignant melanoma arises from the melanocytes, the skin cells that produce the pigment melanin.

Sunscreen—A product that blocks the damaging rays of the sun. Good sunscreens contain either para-aminobenzoic acid (PABA) or benzophenone, or both. Sunscreen protection factors range from two to 45.

Sunstroke—Heatstroke caused by direct exposure to the sun in which body temperature increases to dangerously high levels.

- Use sunscreen when participating in snow activities such as skiing where sunlight is reflected off the snow.
- Wear an opaque shirt on water, because reflected rays are intensified.

Parental concerns

Parents, concern about their child's sun exposure is usually influenced by their own experience with tanning and sunburn. Until the early 2000s, a tan was considered healthy rather than an increased cancer risk. Many adolescents still desire a tanned look but should be discouraged from as much sun exposure as possible. Those who insist on tanning should be encouraged to tan gradually and avoid burns.

See also Heat disorders.

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Sunscreens

Definition

Sunscreens are products applied to the skin to protect against the harmful effects of the sun's ultraviolet (UV) rays.

Description

Many brands of sunscreens are available, containing a variety of ingredients. The active ingredients work by absorbing, reflecting, or scattering some or all of the sun's rays. Most sunscreen products contain combinations of ingredients. Sunscreen products are sold as lotions, creams, gels, oils, sprays, sticks, and lip balms, and can be bought without a physician's prescription.

The U.S. Food and Drug Administration requires sunscreen products to carry a sun protection factor (SPF) rating on their labels. This number tells how well the sunscreen protects against burning. The higher the number, the longer a person can stay in the sun without burning.

There are three types of ultraviolet light, based on their wavelength: UVA, UVB, and UVC. UVC has the shortest wavelength and is blocked by the earth's ozone layer. Concerns about the depletion of the ozone layer focus on the serious health effects that increased exposure to UVC light would have.

UVB light is the next shortest wavelength and is called the tanning light since it is light in this range that promotes creation of the skin pigment melanin that creates a tan. UVB light only penetrates the outermost layer of the skin, but it promotes basal and squamous cell carcinoma and may worsen the effects of UVA.

Ultraviolet A is long-wave radiation generated by the sun that penetrates more deeply than UVB, causes wrinkling and leathering of the skin and damages con-

nective tissue. UVA is the light that causes melanoma, the most serious skin **cancer**.

Several types of chemicals are used as sunscreens. They vary by the degree of protection they can provide and the types of ultraviolet light they can block:

- Cinnamates, such as octyl methoxcinnamate, give low levels of protection, and are only effective against UVB light.
- Para-amino benzoic acid (PABA) compounds, including PABA, padimate O (octyl dimethyl PABA),and glyceryl PABA, are effective only against UVB light.
- Salicylates, octylsalicylate, and homosalate offer moderate levels of protection against both UVA and UVB light, but the range of light waves against which they protect is relatively narrow.
- Benzophenones, including oxybenzone and dioxybenzone, protect against a broader range of ultraviolet light than the salicylates and are more useful for broad spectrum protection.
- Physical sunscreens are really sun blockers and include titanium dioxide, red petrolatum, and zinc oxide. Preparations containing these blockers are thick ointments and are usually reserved for skin areas at high risk of burn, such as the nose.

Other compounds, such as Parsol 1789 (avobenzone), Eusolex 8020, and menthyl anthranilate appear to be valuable broad spectrum agents. In one study, the combination of 3 percent butyl methoxydibenzoylmethane and 7 percent padimate O was the most effective of all sunscreens tested.

In addition to the chemical used as a sunscreen, the vehicle can be important in determining how well a product works. Unfortunately, thick, greasy ointments seem to work better than vanishing creams, lotions, or liquids.

General use

Users should carefully read the instructions that come with the sunscreen. Some of these products need to be applied as long as one or two hours before sun exposure. Others should be applied 30 minutes before exposure and frequently during exposure.

Users should apply sunscreen liberally to all exposed parts of the skin, including the hands, feet, nose, ears, neck, scalp (if the hair is thin or very short), and eyelids. However, they should avoid getting sunscreen in the eyes, as it can cause irritation. Use a lip balm containing sunscreen to protect the lips. Reapply sunscreen liberally every one or two hours—more frequently when perspiring heavily. People should reapply sunscreen after they go in the water.

Precautions

Sunscreen alone will not provide full protection from the sun. When possible, people should wear a hat, long pants, a long-sleeved shirt, and sunglasses. They should try to stay out of the sun between 10 A.M. and 2 P.M. (11 A.M. to 3 P.M. daylight saving time), when the sun's rays are strongest. The sun can damage the skin even on cloudy days, so people should get in the habit of using a sunscreen every day. They need to be especially careful at high elevations and in areas with surfaces that reflect the sun's rays, such as off sand, water, concrete, and snow.

Sunlamps, tanning beds, and tanning booths were once thought to be safer than the sun, because they give off mainly UVA rays. However, UVA rays are now known to cause serious skin damage and may increase the risk of melanoma. Health experts advise people not to use these tanning devices.

People with fair skin, blond, red, or light brown hair, and light colored eyes are at greatest risk for developing skin cancer. So are people with many large skin **moles**. These people should avoid exposure to the sun as much as possible. However, even dark skinned people, including African Americans and Hispanic Americans, may suffer skin damage from the sun and should be careful about exposure.

Side effects

The most common side effects are drying or tightening of the skin. This problem does not need medical attention unless it does not improve. Other side effects are rare, but possible. If any of the following symptoms occur, people should check with a physician as soon as possible:

- acne
- burning, **itching**, or stinging of the skin
- redness or swelling of the skin
- rash, with or without blisters that ooze and become crusted
- pain in hairy parts of body
- pus in hair follicles

The side effects of sunscreens cannot be prevented but can be minimized by testing a sunscreen on a small area of the body before all-over applications.

Young girl applies sunscreen to her face to protect herself from sun damage. *(© Lowell Georgia/Corbis.)*

KEY TERMS

Hair follicle—The root of a hair (that portion of a hair below the skin surface) together with its epithelial and connective tissue coverings.

Melanoma—A tumor, usually of the skin.

Ozone—A form of oxygen with three atoms in its molecule (O_3), produced by an electric spark or ultraviolet light passing through air or oxygen. A layer of ozone about 15 mi (24 km) above Earth's surface helps protect living things from the damaging effects of the sun's ultraviolet rays. Ozone is used therapeutically as a disinfectant and oxidative agent.

Pus—A thick, yellowish or greenish fluid composed of the remains of dead white blood cells, pathogens, and decomposed cellular debris. It is most often associated with bacterial infection.

Ultraviolet (UV) radiation—A portion of the light spectrum with a wavelength just below that of visible light. UV radiation is damaging to DNA and can destroy microorganisms. It may be responsible for sunburns, skins cancers, and cataracts in humans. Two bands of the UV spectrum, UVA and UVB, are used to treat psoriasis and other skin diseases.

Interactions

Anyone who is using a prescription or nonprescription (over-the-counter) drug that is applied to the skin should check with a physician before using a sunscreen.

Parental concerns

Sunscreens should not be used on children under six months of age because of the risk of side effects. Instead, children this young should be kept out of the sun. Children over six months of age should be protected with clothing and sunscreens of at least SPF 15, preferably lotions. Sunscreens containing alcohol should not be used on children because they may irritate the skin.

Before using a new sunscreen, particularly a newer formulation, it should be tested on a small area of skin. These products have some risk of causing **rashes** and other side effects.

Sunscreens should always be applied before a trip to the beach or into some other setting with intense sun exposure. Parents who start to apply sunscreen to their children upon arrival at these settings will exceed their own sun exposure limits before they begin to apply sunscreen to themselves.

Parents should consider using two to three different sunscreens at one time, to get the best results with the fewest problems. Liquids may be best for the scalp, since they can penetrate the hair. Lotions may be most appropriate for most of the body. Ointments may be the best choice for the nose and other parts of the face.

Users should always check expiration dates and not use a sunscreen past its expiration. Reapply sunscreens as directed. Children may benefit from a waterproof sunscreen. There have been claims that these sunscreens may cause eye damage, but this appears to be a hoax. There is no basis for this allegation in the medical literature.

Although sunscreen is useful, it is no replacement for subprotective clothing. While a good sunscreen has an SPF of 15 or above, denim fabric has an SPF of 1700. In contrast, a white T-shirt only has an SPF of 15, and, when wet, has an effective SPF of only 10. Sunglasses are also useful for eye protection.

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Sunstroke *see* **Heat disorders**

Sweat test

Definition

A sweat test, sometimes called a sweat chloride test, is a procedure used to measure the amount of sodium and/or chloride (salt) excreted by a person's sweat glands.

Purpose

The sweat test is used to diagnosis **cystic fibrosis** (CF). CF is an incurable, inherited disorder that affects glands in the lungs, intestines, bile duct, and pancreas, as well as the sweat glands. The sweat test is administered as soon as CF is suspected, either because of **family** history or symptoms, such as frequent colds, recurrent lung infections, recurrent **diarrhea**, difficulty absorbing food, and slower-than-normal growth.

Because prompt diagnosis and treatment can often ease the severity of CF, sweat tests may be administered as early as the first week of life. This timing is recommended only when a family history of CF exists or the newborn exhibits symptoms specific to the disorder. However, newborns may not make enough sweat to accurately perform the test; hence, it may be repeated when they are older. Diagnosis of CF is made based on two or more sweat tests with abnormal chloride readings. Although sweat tests are highly accurate, diagnosis may be confirmed with genetic testing.

To have CF, a child must inherit a gene for the disorder from both parents. Because siblings of CF patients have a 25 percent chance of having the disorder, they should also be tested. However, the sweat test can determine only if the child has the disorder. It cannot determine whether a child is a carrier of a single CF gene that can be passed on to the next generation.

Description

Individuals with CF produce a higher than normal level of sodium chloride (salt) in their sweat. This measurement does not normally change with diet, medication, or environmental factors, making it a good diagnostic tool. The sweat test uses a process called iontophoresis. With iontophoresis, a very small, painless electric current is used to help draw sweat to the surface of the skin, where it can be collected and analyzed. The amount of electric current is tiny, and the test is safe and painless for all ages.

For infants a sweat test is done on the right thigh; for children and adults, the right forearm is used. After the area is washed and dried, two metal electrodes are attached and fastened with straps. Two gauze pads, one soaked in salt water or bicarbonate and the other in pilocarpine, a drug that stimulates sweating, are placed under the electrodes. A tiny electric current is applied to the skin for five to ten minutes to carry the pilocarpine into the skin. This stimulates the sweat glands to begin working. The procedure is painless; the child feels only a slight tingling or tickling.

After about ten minutes, the electrodes are removed. The skin is washed with distilled water and dried again. A dry piece of filter paper is taped to the area where the pilocarpine was applied. The paper is then covered with wax or a sheet of plastic, so that evaporation does not occur. The filter paper is called a sweat patch. After 30 to 45 minutes, the plastic is removed and the paper is placed in a sealed bottle. The entire process takes between 60 and 90 minutes.

It is important that the test be performed in a certified lab because the reliability of the test is operator-dependent. In the certified lab, the sweat patch is weighed and analyzed for sodium and/or chloride content. In children, normal sodium levels are less than 70 milliequivalents per milliliter (mEq/L). A sodium level greater than 90 mEq/L is indicative of CF. Normal

chloride reading is less than 40 mEq/L. A chloride reading of greater than 60 mEq/L is indicative of CF. Readings falling between these numbers are borderline and require additional testing. Results are usually available within one to two working days.

Precautions

To ensure accuracy, sweat tests should be analyzed by a laboratory certified by the Cystic Fibrosis Foundation. Some other conditions such as malfunction of the adrenal gland or kidney failure can produce abnormal chloride readings. However, these conditions have distinct symptoms that differ substantially from CF. A sweat test is never used to diagnose these conditions.

Preparation

Before the sweat test, children should not **exercise** heavily or become overheated. There are no dietary restrictions; children may eat normally before the test. The results are not affected by medication; children may take their medication on the day of the test.

Aftercare

The test area may be red and sweaty for several hours after the test.

Risks

Although there is virtually no risk of electrical shock from a sweat test, it should never be conducted on the left side of the body, nor should it be given in the chest area, because there is a very small chance that the electric current could affect the heart. The current should come from a battery-powered unit rather than from a direct current.

Parental concerns

Parents can expect to stay with their child during the test.

See also Cystic fibrosis.

KEY TERMS

Iontophoresis— Application of a small electric current to the skin.

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Sweating, excessive *see* **Hyperhidrosis**
Swimmer's ear *see* **Otitis externa**
Swollen glands *see* **Lyme disease**
Syndactyly *see* **Polydactyly and syndactyly**
Syphilis, nonvenereal *see* **Bejel**
Syringomyelia *see* **Chiari malformation**

T

Talipes *see* **Clubfoot**

Tantrums

Definition

A tantrum is an episode of extreme anger and frustration characterized by crying, screaming, and violent body motions, including throwing things, falling to the floor, and banging one's head, hands, and feet against the floor.

Description

Tantrums, also called temper tantrums, can occur by the age of 15 months, but are most frequent between the ages of two and four. All children have them at some point, and active, strong-willed youngsters may have as many as one or two a week. Generally, tantrums are an expression of frustration. Children may be frustrated by their inability to perform an activity they are attempting, such as buttoning a coat. Tantrums may also be an expression of frustration at the lack of control children have over their lives, such as at bedtime when children want to continue playing instead of going to bed. Occasionally a tantrum may also be an attempt to gain attention from a parent or other caregiver, or it may be an attempt to manipulate the situation in some way.

Aside from taking any measures needed to prevent danger to children, parents should try to ignore the tantrum and let it run its course. If the upset has occurred over something the child wants and has been denied, it is tempting to give in to the child's wishes, but doing so can be harmful because it teaches children that they can get what they want by having a tantrum. Frequently, tantrums occur in a public place, which is especially unsettling for parents. Children become over stimulated or tire more easily in busy public spaces such as supermarkets and malls and may use the tantrum as an attempt to regain parental attention that is focused elsewhere. In spite of their embarrassment, parents should treat a public tantrum in essentially the same way they treat one at home. Whenever possible, they should remove the child to the car or some other private space to avoid inconveniencing others and attracting any more unwelcome attention, after which they should ignore the tantrum and let it run its course.

While a parent cannot stop tantrums once they are in progress, it is sometimes possible to prevent them by being alert to certain danger signs, especially fatigue, hunger, and irritability. In these cases, they can change plans to give the child a needed rest, food, or change of scene. For example, a child who is getting cranky at a party or other event at which the parent is present can be taken home early. The archetypal shopping tantrum over the candy bar at the checkout counter or the elaborate toy can sometimes be countered by proposing an alternative treat or purchase instead of the flat denial that sends the child into a tantrum. Emotional upsets that occur when children are left with a babysitter or at daycare are usually a sign of **separation anxiety** and can be alleviated by preparing children in advance for the separation and giving them the opportunity to become familiar with the babysitter or daycare setting ahead of time. Keeping walking trips short can prevent tantrums over a child's demand to be carried.

Toddlerhood

Children between the ages of two and four are the most likely to have tantrums and to have them the most often. They have not acquired the verbal skills necessary to adequately express their emotions or even, in many situations, to make themselves understood. In addition, they can only use words to demand what they want, not to negotiate for it. They love to explore, but often they do not understand which places or objects are off limits and are scolded as a result. Although they are developing rapidly, they still lack the motor skills to do many things they would like to do. They want to be independent but still require continuous supervision and assistance, and their preferences are often unrecognized, ignored, or

refused by their caregivers. There is also a great deal of ambivalence and indecision associated with this stage of life, meaning that there is internal conflict as well as tension between the toddler and his or her environment. The tantrum occurs because the small child, who is still learning to cope with her feelings, is simply unable to contain strong emotions of anger, frustration, or disappointment. In some cases, children are actively discouraged from showing these feelings, which creates even more tension.

School-age children

School-age children tend to have tantrums less often, but many children still have them occasionally. At this age, frustration with inability to do homework may often be the cause of tantrums. Parents should let their child calm down and then offer to help them and give encouragement. It can be helpful to remind the child that the task that causes frustration will become easier to perform with practice. If the child has tantrums at school, a doctor's advice should be sought because it could be a symptom of other another problem such as a learning disability.

Common problems

Having tantrums is a normal part of growing up; however, they are not socially acceptable behavior. Consequently, the most common problems with tantrums are problems for the parents. The tantrums often take place in public, which can be embarrassing and make them harder to deal with calmly. If the child actually hurts himself or others or has very frequent tantrums, it may be a sign of behavior problems, and the child should be assessed by a pediatrician.

Parental concerns

Most children do not actually hurt themselves or others during tantrums, although it may seem like they are going to. Holding the breath cannot actually hurt a child; the child will breathe involuntarily before harm occurs. A child's tantrums can, however, challenge parents' ability to remain calm. Tantrums may occur in busy places such as restaurants and grocery stores, and the child is more likely to be tired. It can also be very distressing for parents to see the child so upset and out of control. Parents who are concerned about their ability to calmly deal with the child's temper tantrums may talk to the child's pediatrician about ways to cope more effectively with this natural part of the child's development.

When to call the doctor

The child's doctor should be called if the child hurts herself, the parent, other people, or objects during a tantrum. If the child has more than five tantrums a day, or the tantrums reoccur in school, the doctor should also be consulted.

See also Separation anxiety.

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Tish Davidson, A.M.

Tattoos *see* **Piercing and tattoos**

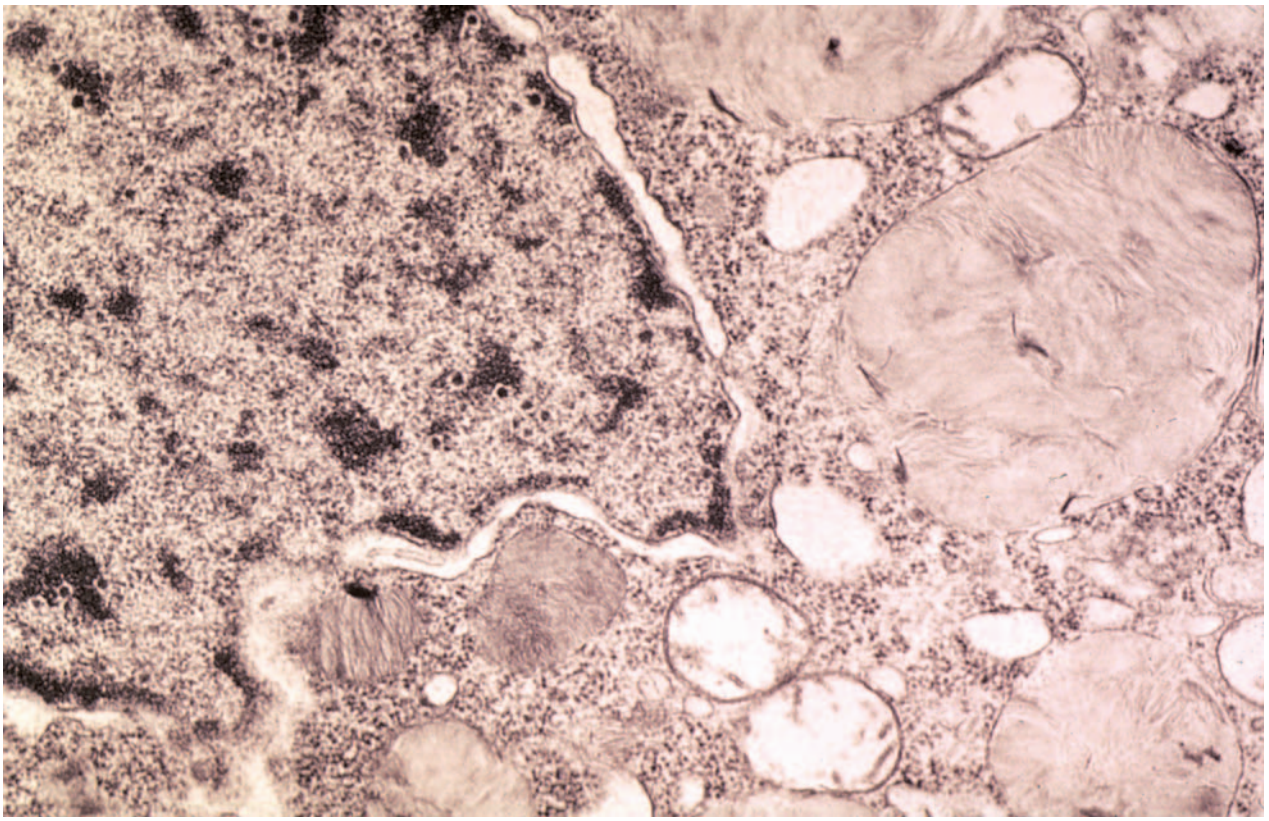
Tay-Sachs disease

Definition

Tay-Sachs disease is a genetic disorder caused by a missing enzyme that results in the accumulation of a fatty substance in the nervous system. This disease causes disability and death.

Description

Gangliosides are fatty substances necessary for the proper development of the brain and nerve cells (nervous system). Under normal conditions, gangliosides are continuously broken down, so that an appropriate balance is maintained. In Tay-Sachs disease, the enzyme necessary



Section of brain tissue from patient with Tay-Sachs disease. *(© 1992 IMS Creative. Custom Medical Stock Photo, Inc.)*

for removing excess gangliosides is missing. This situation allows gangliosides to accumulate throughout the brain and is responsible for the disability associated with the disease.

Demographics

Tay-Sachs disease is particularly common among Jewish people of Eastern European and Russian (Ashkenazi) origin. About one out of every 2,500 to 3,600 babies born to Ashkenazi Jewish couples have the disease. In the general population about one out of every 320,000 babies born has Tay-Sachs disease. Approximately one in 30 Ashkenazi Jews is a carrier of the gene that causes the disease. Tay-Sachs is also more common among certain French-Canadian, Pennsylvania Dutch, and Cajun families.

Causes and symptoms

Tay-Sachs is caused by a defective gene. Genes are located on chromosomes and serve to direct specific developments and processes within the body. The genetic defect in Tay-Sachs disease results in the lack of an enzyme called hexosaminidase A. Without this enzyme, gangliosides cannot be broken down. They build up within the brain, interfering with nerve functioning. Because Tay-Sachs is a recessive disorder, only people who receive two defective genes (one from the mother and one from the father) will actually have the disease. People who have only one defective gene and one normal gene are called carriers. They carry the defective gene and thus the possibility of passing the gene and/or the disease onto their offspring.

When a carrier and a non-carrier have children, none of their children will actually have Tay-Sachs. The statistical probability is that 50 percent of their children will be carriers themselves. When two carriers have children, their children have a 25 percent chance of having normal genes, a 50 percent chance of being carriers of the defective gene, and a 25 percent chance of having two defective genes. Only the individual with two defective genes actually has the disease.

Classic Tay-Sachs disease strikes infants around the age of six months. Up until this age, the baby appears to develop normally. When Tay-Sachs begins to show itself, the baby stops interacting with other people and

KEY TERMS

Ganglioside—A fatty (lipid) substance found within the brain and nerve cells.

develops a staring gaze. Normal levels of noise startle the baby to an abnormal degree. By about one year of age, the baby has very weak, floppy muscles and may be completely blind. The head is quite large. Children with Tay-Sachs also have other symptoms, such as loss of peripheral (side) vision, inability to breathe and swallow, and paralysis as the disorder progresses. Seizures become a problem between ages one and two, and the baby usually dies by about age four.

A few variations from this classical progression of Tay-Sachs disease are possible:

- Juvenile hexosaminidase A deficiency: Symptoms appear between ages two and five; the disease progresses more slowly, with death by about 15 years.
- Chronic hexosaminidase A deficiency: Symptoms may begin around age five or may not occur until between 20 and 30 years of age. The disease is milder. Speech becomes slurred. The individual may have difficulty walking due to weakness, **muscle cramps**, and decreased coordination of movements. Some individuals develop mental illness. Many have changes in intellect, hearing, or vision.

When to call the doctor

If the child has any noticeable problems that might be associated with Tay-Sachs disease or appears to stop developing normally after a period of normal development, the doctor should be consulted.

Diagnosis

Examination of the eyes of a child with Tay-Sachs disease reveals a characteristic cherry-red spot at the back of the eye in an area called the retina. Tests to determine the presence and quantity of hexosaminidase A can be performed on the blood, specially treated skin cells, or white blood cells. A carrier has about half the normal level of hexosaminidase A present, while an individual with the disease has no hexosaminidase A at all.

Treatment

Providing good, supportive care and treating the symptoms as they arise is the only way to treat Tay-Sachs; there is no way to treat the disease itself.

Prognosis

The prognosis for a child with classic Tay-Sachs disease is death. Because the chronic form of Tay-Sachs was discovered near the end of the 2000s, prognosis for this type of the disease was, as of 2004, not completely known.

Prevention

There is no known way to prevent Tay-Sachs disease. It is, however, possible to identify carriers of the disease and provide them with genetic counseling and appropriate information concerning the chance of their offspring having Tay-Sachs disease. When the levels of hexosaminidase A are half the normal level, a person is a carrier of the defective gene. Blood tests of carriers reveal reduction of hexosaminidase A.

When a woman is already pregnant, tests can be performed on either the cells of the fetus (**amniocentesis**) or the placenta (chorionic villus sampling) to determine whether the baby will have Tay-Sachs disease.

Parental concerns

If parents are thinking of having a child and believe they might be carriers of Tay-Sachs, they should be screened so that they can assess their options. Children born with infantile Tay-Sachs, even with the best available care, usually die before the age of five. Children born with juvenile Tay-Sachs usually die before the age of 15.

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ORGANIZATIONS

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National Tay-Sachs and Allied Diseases Association. 2001 Beacon Street, Suite 204, Brighton, MA 02135. Web site: <www.ntsad.org>.

Tish Davidson, A.M.

Teething *see* **Dental development**

Television habits

Definition

Television habits consist of patterns of behavior determined by the amount of time and importance individuals give to watching television broadcasts and recorded videos and DVDs.

Description

Ever since the 1ate 1940s when television first became available, social scientists have been interested in its effect on behavior. Originally seen as entertainment for adults and older children, television in the twenty-first century is watched by all age groups, including infants. More than 98 percent of homes in the United States have at least one television set. Many have more. One study found that 32 percent of children ages two to seven had television sets in their bedroom. This number increased to 65 percent for children ages eight to 18.

Although television can be an educational tool for children, exposing them to information and situations that they cannot experience first hand, social scientists and the American Academy of Pediatrics (AAP) have raised specific concerns about the effect of television watching on child development. Areas of concern include:

- inability of young children to distinguish between television fantasy and reality
- exposure to television violence, especially where violence is not shown to have any serious consequences
- exposure to age-inappropriate sexual situations
- effect of advertising on children
- glamorization of unrealistic body images
- promotion of alcohol use and the glamorization of cigarette and cigar smoking
- increased **obesity** when passive television watching replaces active **play**
- time taken away from school work and traditional hobby activities
- the short, segmented, hyperactive nature of children's television programming that may decrease attention span and contribute to attention deficit disorder (ADD) in children
- verifiable health risks of excessive television watching to children as concluded by the AAP

Factors that increase the likelihood of heavy television viewing by children include low socioeconomic status, living in a single parent household, and being born to a teenage mother. Viewing time is also increased by parental beliefs that television viewing does not hurt children and improves their vocabulary and imagination. Heavy parental television viewing, multiple television sets at home, television in the child's bedroom, and using television to distract young children all increase the likelihood that children will become heavy television and video watchers.

Infancy

Before the 1990s, few television programs were designed for children under the age of three. However, the success of programs such as "Teletubbies" aimed at children under age two, opened a new market to programmers. As of 2004, studies showed that by the age of 11 months, infants are watching a combined average of 75 minutes of television programming and videos daily. Between the ages of 12 and 23 months, this amount increases to almost two hours daily.

In the first two years of life the brain develops rapidly, and children learn new physical, mental, and social skills. Infants need interaction with caregivers who respond to them and interact with them. Watching television is a passive activity that does not meet these needs. The AAP recommends that television and video viewing for children under age two be discouraged and severely limited. They suggest that reading, singing, playing, and interacting with adults fosters proper, age-appropriate brain development that can be inhibited by too much television watching. They recommend that televisions be removed from childcare areas, because they are a distraction to both caregivers and children.

Toddlerhood

By the age of three, the average American child is watching more than two hours of television and videos daily. Toddlerhood is a time when motor skills develop and children begin actively moving and exploring their environment. Pediatricians are concerned that television time takes away from activities needed for physical and mental development. They recommend children of this age watch only a few selected programs or videos, after which the television should be turned off.

Preschool

Many educational programs such as "Sesame Street" are aimed at preschoolers. Properly selected and watched in moderation, these programs can increase reading readiness and number awareness and promote positive social behavior such as sharing and taking turns.

However, preschoolers are highly influenced by advertisements on commercial television. Until the age of about eight, children lack the ability to differentiate between fantasy and reality both in programming and in advertising. In addition, a three-year study of television violence found that nearly 66 percent of television programs contain violence and that children's television programs contain the most violence of all programming. **Preschool** children are often frightened by what they see on television, including the news.

School age

American school-age children watch on average 4.5 hours of television a day. Repeated studies have shown that children who watch a lot of television perform more poorly in school. In addition, there is a definite link between television watching and childhood obesity. The link is strongest among African-American and Latino children. This link is thought to result from several causes. First, television viewing is a passive activity. Children who are watching are not actively playing or exercising. Second, there is a strong tendency to eat snacks while watching television, compounding the problem. Third, advertising is often aimed at promoting high calorie, less healthful foods. The eating habits of children who watch a lot of television are influenced by this advertising.

School-age children are also influenced by the content of the programming they watch. Many studies have linked real-life violence to the repeated viewing of violence on television. Some experts theorize that children become immune to violence after seeing it repeatedly on television. For example, the average child will have seen over 8,000 television murders before finishing grade school. Other experts believe that because the hero uses violence to triumph as much as 80 percent of the time on television, children develop the idea that force is an acceptable way to solve problems. Finally, the aftermath of violence and the harm it does are rarely shown in a realistic way on television. Although television is not the only cause of social violence, most social scientists believe it is a significant contributing factor. Likewise, television's portrayal of sexual situations and the lack of on-screen consequences of sexual activity, the promotion of unrealistic body image, stereotypical gender roles, and under-representation of minorities have been found in multiple studies to promote unhealthy and unrealistic views among children and adolescents.

Common problems

The most common problems surrounding television viewing involve children being frightened by what they see, substituting television watching for health interactions with friends and **family**, and viewing material that is age-inappropriate.

Parental concerns

The most common parental concern surrounding television habits is controlling the amount and quality of television a child watches. Often parents who are heavy television viewers themselves see nothing wrong with turning on the television in the morning and letting it stay on all day. They may also be unwilling to give up watching their favorite programs, even when these programs are age-inappropriate for their children. This problem can be solved by taping programs for later viewing after children are asleep.

The new television rating system and v-chip technology required by law in all televisions with a 13-inch screen or larger manufactured after 2000 is intended to help parents control what their children watch by allowing them to lock out certain programs. However, studies have shown that very few parents understand or use the television rating system in determining what their children may see and that very few actually use the v-chip technology to lock out specific programs.

The American Academy of Pediatrics suggests the following steps to parents for controlling the television viewing habits of their children.

- Watch only specific programs; do not turn the television on and let it run.
- Remove televisions from children's bedrooms.
- Limit television viewing to two hours or less daily.
- Do not watch television during meals.
- Watch television with children and discuss what is shown.
- Substitute reading time or play time for television time.
- Teach children the difference between advertisements and program content.

When to call the doctor

Parents may want to consider seeking the advice of a pediatrician and/or child and adolescent psychiatrist if

Healthy television habits

Limit television viewing to a maximum of two hours per day.

Plan programs to watch by looking in the TV guide ahead of time.

Keep videotapes or DVDs of children's programming on hand as a backup when there is nothing appropriate for children on television.

Keep television sets out of children's rooms.

Keep the television off during meals.

Watch shows with your child, and discuss them afterward.

Criticize the negative behaviors that some TV characters exhibit, such as violence or smoking.

Explain that commercials are designed to make the viewer want something, and that not all advertised products are good or needed.

SOURCE: American Academy of Pediatrics and National Institute on Media and the Family, 2002.

(Table by GGS Information Services.)

their child appears addicted to television and videos. **Addiction** may be suggested if watching television takes priority over schoolwork and family interactions and replaces normal social activities with friends. Parents should also be concerned if they feel their child is developing unrealistic expectations about body image from television or is excessively attracted to and talks about imitating violent, gory events on television or in the movies.

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ORGANIZATIONS

American Academy of Pediatrics. 141 Northwest Point Boulevard, Elk Grove Village, IL 60007–1098. Web site: <www.aap.org>.

Tish Davidson, A.M.

Temper tantrums *see* **Tantrums**

Temperament

Definition

Individual differences in human motivation and emotion that appear early in life, usually thought to be biological in origin. Temperament is sometimes considered the biological or physiological component of personality, which refers to the sum total of the physical, emotional, mental, spiritual, and social dimensions of an individual.

Description

History

Ancient Greek and Roman physicians invoked nature, claiming that the proportions of the various humors or fluids in the bodies influenced personality. They thought that there were four basic temperaments—sanguine (cheerful), choleric (irritable), melancholic (gloomy), and phlegmatic (apathetic)—which were determined by the predominance of blood, yellow bile, black bile, and phlegm respectively in the person's physical constitution. The ancient theory survives in the form of such expressions as "being in a bad (or good) humor."

The theory of four bodily humors did not survive the rise of scientific medicine in the seventeenth century as an explanation for differences in human temperament, but it has not been replaced by any single universally accepted theory of personality either. During most of the twentieth century, political ideology, discoveries about the learning or conditioning capabilities of infants, and the emergence of psychoanalytic theory, which emphasized the importance of early experience, all combined to discredit biological explanations for human motivation and emotion. Nurture and socialization became the favored explanations of differences in temperament.

There was, however, a resurgence of interest in the contribution of temperament to children's development after the 1950s. Temperament came to be summarized as the biological dimension of personality. It was seen as a predisposition that allows two individuals to experience the same objective event very differently within the range of normal behavior and development.

Specific approaches to temperament

THE NEW YORK LONGITUDINAL STUDY Suspecting that inherent individual differences among their young patients contributed to their developmental paths, two child psychiatrists, Alexander Thomas and Stella Chess, designed a study that challenged the nature-nurture dichotomy. Beginning in 1956 and ultimately publishing their research in Temperament and Development in 1977, Thomas and Chess collected longitudinal data from over 100 children, following them from infancy through early adulthood. Using extensive clinical interviews to gather information about children's behavior as well as parents' values and expectations, they examined what they termed the goodness of fit between the individual child and his or her environment.

Thomas and Chess found that children could be rated on each of nine dimensions even in infancy:

- Activity level: The child's general level of energy and movement—whether he or she is quiet, always "on the go," or somewhere in-between.
- Rhythmicity: The child's regular biological patterns of appetite and sleep—whether the child gets hungry or tired at predictable times.
- Approach/withdrawal: The child's usual response to new people or situations—whether the child is eager for new experiences or shy and hesitant.
- Adaptability: The child's ability and pace in adjusting to changes in schedules or transitions from one activity to another.
- Threshold of responsiveness: The child's level of sensitivity to such physical stimuli as sounds, smells, and lights. For example, some children are easily startled by sudden noises while others are less sensitive to them. Some children are pickier about food than others.
- Intensity: The child's responses to people or events. Some children react strongly and loudly to even minor events while others are less demonstrative or openly emotional.
- Quality of mood: The child's overall worldview, whether positive or negative. Some children tend to focus on the negative aspects of a situation while others are more positive or hopeful. Some children tend to approach life in a serious or analytical fashion while others respond to their immediate impressions of situations.
- Distractibility: The child's ability to pay attention to tasks or instructions even when the child is not particularly interested in them. Some children have shorter attention spans than others.
- Persistence: The child's ability to continue with an activity in the face of obstacles or problems. Some children are more easily discouraged by difficulties than others.

Thomas and Chess combined the patterns of children's ratings on each of these nine dimensions to distinguish three major temperamental types:

- Easy children: About 40 percent of the NYLS sample displayed a temperamental profile marked by regularity, ease of approach to new stimuli, adaptability to change, mild to moderate mood intensity, and a generally positive mood. This profile characterizes what Thomas and Chess call the easy child.
- Difficult children: About 10 percent of children showed a very different profile and were called difficult children. They had irregular patterns of eating and sleeping, withdrew from new stimuli, did not adapt easily to change, and reacted intensely to changes. Their overall mood was often negative.
- Slow-to-adapt children: Children who were slow to warm up comprised the third temperamental group, about 15 percent of Thomas and Chess's sample. These children tended to withdraw from new stimuli and had difficulty adapting to change, but their reactions were of mild intensity and gradually became either neutral or positive with repeated exposures to the new event or person.

Some researchers prefer the terms flexible, active or feisty, and cautious instead of the somewhat judgmental terms of easy, difficult, and slow-to-adapt, respectively.

Clearly, these three temperamental types that Thomas and Chess identified did not include all of the variations seen in children across the entire sample. About one third of the children showed mixed profiles. Nonetheless, these temperamental classifications became highly influential in child development research. Perhaps the greatest contribution of the NYLS, however, was Thomas and Chess's emphasis on "goodness of fit"; that is, they maintained that the child's temperament by itself was not the most important consideration in his or her growth and development, but the extent to which that temperament agreed with the values, expectations, and

style of the child's environment, whether **family**, child-care setting, school, or culture. For example, a quiet and serious child fits in well with a family of scholars or intellectuals, whereas an intense, active, and easily distracted child may not be accepted as readily in the same family context. In terms of culture, some ethnic groups place a high value on self-control and relating well to others, while other groups emphasize assertiveness and independence. A child who has a high energy level and reacts intensely to persons and events will have a better fit with the second group than with the first. The notion of goodness of fit also helps to explain why some children in a given family seem to get along better with their parents than their siblings do. Even though temperament is thought to be rooted in biology, different children in the same family may have very different temperaments.

TRAIT APPROACHES Some approaches to the study of temperament emphasize traits; that is, they assume that temperamental qualities can be rated as persisting within individuals across time in a variety of situations. In 1984, as published in their book, *Temperament: Early Developing Personality Traits*, Arnold Buss and others considered temperaments to be heritable and stable personality profiles—profiles that are genetically influenced and relatively unchanging over time. These researchers used maternal questionnaires to gather information on children's emotionality, activity, and sociability, traits they regarded as the fundamental dimensions of temperament. Interestingly, Buss and Plomin suggested that children who are rated as extreme on these dimensions may be qualitatively different from those whose scores lie closer to the middle.

Basic emotions were at the core of H. Hill Goldsmith and Joseph Campos's conception of temperament. In 1983, in an essay included in *Socio-Emotional Development,* they described temperament in terms of individual differences in experiencing and expressing such primary emotions as anger, **fear**, and pleasure. Goldsmith and Campos, however, emphasized the speed and intensity of children's responses to stimuli as well as the specific emotions involved. Their evaluations were based on three measurements: threshold (the amount of stimulation the child requires before responding); latency to respond (how rapidly the child reacts to the stimulus); and intensity of response.

In 2004, Mary Rothbart emphasized reactivity and self-regulation as core processes in organizing temperamental profiles. These processes, she believed, can be seen in six significant infant behaviors: smiling; distress when confronted by limitations; fear; activity level; soothability, and duration of orienting (how long the baby plays with or pays attention to a single object). Her Infant Behavior Questionnaire (IBQ), which was developed in the early 1980s, remained, as of 2004, one of the most widely used methods of assessing temperament in infants between the ages of three months and 12 months. In the first version of the IBQ, published in 1981, parents were asked to rate the frequency of these temperament-related behaviors in their child over a two-week period. The revised version of the IBQ, known as the IBQ-R, was developed by Rothbart and her colleague Masha Gartstein in the early 2000s. The IBQ-R expanded the original six measures of temperament to 14. The new measurements include the following:

- Approach: The infant's excitement and looking forward to a pleasurable experience or activity.
- Vocal reactivity: The baby's level of vocal responses to stimuli in its daily routine.
- Perceptual sensitivity: The infant's ability to detect low-intensity stimuli in its environment.
- Sadness.
- High-intensity pleasure: The infant's reactions to pleasurable stimuli or activities of high intensity, such as loud music or bright lights.
- Low-intensity pleasure.
- Cuddliness: The infant's physical and emotional responses to being held or cuddled by a parent or caregiver.
- Rate of recovery from distress: How long it takes the infant to return to a normal level of emotion after an exciting or upsetting experience and how readily the child falls asleep.

In contrast to Goldsmith and Campos, Rothbart emphasized cognitive processes in children as the key to understanding temperament rather than emotions by themselves. For Rothbart and her colleagues, the infant's ability to focus its attention is the basis of its later ability to regulate its reactions to people and events. In Rothbart's view, what she calls the attentional system allows the child to regulate his or her outward behavior as well as internal reactions to stimuli. As children mature, they develop the ability to turn their attention to alternative strategies when they are frustrated and to make plans in order to achieve their goals. Different patterns of self-regulation in turn help to explain differences in temperament.

Goldsmith and Rothbart collaborated to develop an **assessment** tool to gauge temperamental dimension based on systematic observations of behaviors elicited under standard laboratory conditions (for example, how a child reacts to a mechanical spider). The development of an observational protocol or test for assessing

temperamental characteristics offers an advantage over reliance on questionnaires. When parents describe their children's behavior, they are influenced by their feelings about the child as well as their observations. In addition, the parents' reports include many sources of information such that reports of the child's behavior cannot be easily separated from the parents' biases, values, or expectations.

TYPE APPROACHES Another major approach to the study of temperament distinguishes among types of people characterized by different patterns of behavior. In the 1990s, in *Galen's Prophecy*, Jerome Kagan and his colleagues studied two types of children whom they defined as inhibited and uninhibited (or exuberant) respectively. Kagan's group studied the development of these two types of children through **adolescence** as well as the infant profiles that predicted the children's behavior at later ages. At early ages, inhibited children cling to their mothers and may cry and hesitate when confronted with unfamiliar persons or events. These children appear to be timid and shy and represent about 20 percent of volunteer Caucasian samples. Uninhibited or exuberant children, on the other hand, approach new events and persons without hesitation or trepidation. They appear fearless and sociable and represent about 40 percent of volunteer samples. Kagan's observations of these children over time indicated that these characteristic profiles tended to continue, although the display of temperamental tendencies varied in accordance with the child's developmental level. An older inhibited child or teenager, for example, may not cling to his or her mother or cry when coming to an unfamiliar laboratory but may hesitate to talk to the examiner and may smile infrequently.

Interestingly, Kagan found that the behavioral profiles of these children were accompanied by physiologic profiles that suggested different levels of reactivity in the children's central nervous systems, particularly in regard to fear and stress reactions. Inhibited, compared to uninhibited, children tended to have higher and more stable heart rates, higher levels of stress-related hormones like cortisol and norephinephrine, larger changes in blood pressure in response to stressors, and measurable tension in their voices when speaking under mildly stressful conditions. These differences seemed to support the contention that temperamental categories have a biological dimension.

Although young infants are not sufficiently mature to demonstrate timidity in response to new experiences, the reactivity of the structures in the human nervous system that are thought to underlie inhibited and uninhibited temperaments may appear at early ages. When infants are exposed to variations in the sights and sounds in their environment, some become aroused and demonstrate this arousal by moving their arms and legs and fretting or crying. Other infants remain calm, relatively motionless, and do not cry. Those who are highly reactive to stimulation tend to become inhibited in their reactions to novelty and uncertainty at later ages. Those whose reactivity level is low in infancy tend to grow into children who remain relaxed in novel situations so that they appear outgoing and uninhibited.

MALLEABILITY OF TEMPERAMENT Malleability refers to the extent to which temperament can be influenced or reshaped by later life events. The reader should note that the continuity of temperamental profiles from infancy through later ages is a group phenomenon; that is, individual children may change and become more or less inhibited while the groups of children remain distinct on average. Neither temperament nor biology is destiny. Temperament and environment both influence development, although relatively few researchers have studied the interaction of these two influences as of the early 2000s.

Research in early 2000s about temperament

In the early 2000s, research on temperament in children and adolescents is making use of new brain imaging technology to expand understanding of the biological processes that influence emotional self-regulation and task-related activities. This technology is known as functional **magnetic resonance imaging** (fMRI). Functional magnetic resonance imaging is based on the fact that activity in a specific part of the brain is accompanied by an increased flow of blood to that region. As the blood flow increases, the amount of deoxyhemoglobin, a form of hemoglobin that has lost its oxygen content, decreases in the affected area of the brain. Since the amount of deoxyhemoglobin in the blood affects the magnetic resonance image signal, it can be used as the source of the signal for fMRI. This discovery means that fMRI studies can be conducted without injecting radioactive materials into a subject's blood. In addition, it means that usable MRI images can be obtained in a very short period of time (1.5–2 minutes on average) rather than the longer periods of testing required when radioactive materials are used.

FMRI has many beneficial applications, ranging from more accurate planning for brain surgery to more effective **pain management**. In terms of the study of temperament, fMRI allows researchers to study such complex brain activities as problem-solving as well as visual and auditory (hearing) perception. In 2003, the National Institute of Mental Health (NIMH) began a

study that uses fMRI technology on 60 children and adolescents between the ages of nine and 16. The study is designed to test the hypothesis that differences in temperament related to differences in brain functioning put some children at an increased risk of certain psychiatric disorders later in life. The type of child that Kagan's research group identified as inhibited, for example, appears to have the same pattern of disturbed nerve cell activity that has been identified in adults diagnosed with mood or **anxiety** disorders. Specifically, inhibited children seem to have a higher level of activity in a part of the brain called the amygdala, which regulates emotion, and a lower than average level of activity in the prefrontal cortex, which governs a person's ability to express emotions. Exuberant children, on the other hand, are thought to have a relatively high level of activity in the prefrontal cortex in response to certain stimuli.

In addition to its usefulness in studying the parts of the brain that are activated by sensory perception, thinking, and emotional responses to various stimuli, fMRI may also be helpful in distinguishing between problem behaviors in children that are rooted in temperament and behaviors that indicate a psychological problem. As of the early 2000s, research in the area of temperament has not been closely coordinated with research in childhood psychiatric disorders; as a result, both the causes and treatments of these disorders were, as of 2004, not well understood. Child psychiatrists have already observed that avoidant personality disorder (APD) and generalized anxiety disorder (GAD) are closely linked to the inhibited type of temperament as described in Kagan's work. To give another example, such temperamental traits as irritability and strong negative reactivity are thought to contribute to the development of **oppositional defiant disorder** in some children. Lastly, attention deficit/hyperactivity disorder is thought to be heavily influenced by genetic factors affecting the child's temperament, including the production and metabolism of certain neurotransmitters in the brain that affect the child's ability to focus his or her attention.

Common problems

The following are some of the problems that may arise in connection with differences in children's temperaments:

- Parents tend to regard certain characteristics as negative rather than as potentially positive. For example, a child's slowness to adapt may be seen as a drawback rather than as a protection against the dangers of impetuosity or being overly influenced by peer pressure.
- Behavioral problems are related to a poor fit between parent and child. Pediatricians often see families in which a vicious circle of negative interactions develops. The most common example is an angry reaction to a difficult child's aggressiveness or restlessness that takes the form of scolding or spanking. The child reacts to the parents' negative actions by increased aggressiveness, temper **tantrums**, or stubbornness. Another common pattern is the shy or inhibited child who becomes even more withdrawn when parents react to the **shyness** by lecturing or shaming the child.
- Favoritism becomes a factor when some parents find it much easier to relate to a child with a flexible temperament or one whose temperament matches their own than to a child who does not fit in as well. They may ignore the child they find less agreeable or punish him or her unfairly.

Parental concerns

Common parental concerns about evaluations of their children's temperament include the following:

- Fears about labeling or stigmatization: Some parents are concerned about the reactions of teachers or other adults if their child is identified as "difficult." This fear is one reason why some researchers prefer to describe children in this category as "active" or "feisty" rather than to use the negative term difficult.
- Concerns about fairness: Parents whose children have different temperaments are sometimes concerned that treating the children differently will be perceived as unfair or unjust.
- Concerns about the parent-child bond: Some parents worry about their ability to relate to a child with a difficult temperament or one whose temperament is different from their own. They may feel guilty about their negative emotional reactions toward such a child and doubt their ability to be good parents.

When to call the doctor

As has already been mentioned, it is not always easy for parents to distinguish between a child with a "difficult" temperament whose behaviors are still within the normal range and a child with a psychiatric disorder. Some guidelines that have been given by pediatricians include the following:

- The specific problem behavior(s) cannot be attributed to the child's developmental stage (such as "the terrible twos").
- The child's problematic behaviors occur frequently.
- The child has several problematic behaviors.
- The child's behaviors are interfering with his or her social and intellectual development.

KEY TERMS

Amygdala—An almond-shaped brain structure in the limbic system that is activated in stressful situations to trigger the emotion of fear. It is thought that the emotional overreactions in Alzheimer's patients are related to the destruction of neurons in the amygdala.

Cortisol—A steroid hormone secreted by the adrenal cortex that is important for maintenance of body fluids, electrolytes, and blood sugar levels. Also called hydrocortisone.

Goodness of fit—A term first used by Thomas and Chess to describe the importance of children's interactions with their environment as well as their basic temperament in understanding their later growth and development.

Inhibited—A type of child defined by Jerome Kagan and his colleagues as having a low level of responsiveness to strangers, a reluctance to initiate activities, and requiring a long time to relax in new situations. Children with inhibited temperaments appear to be more susceptible to anxiety disorders, depression, and certain personality disorders in their later years.

Malleability—A term that refers to the adaptability of human temperament; the extent to which it can be reshaped.

Neurotransmitter—A chemical messenger that transmits an impulse from one nerve cell to the next.

Norepinephrine—A hormone secreted by certain nerve endings of the sympathetic nervous system, and by the medulla (center) of the adrenal glands. Its primary function is to help maintain a constant blood pressure by stimulating certain blood vessels to constrict when the blood pressure falls below normal.

Personality—The organized pattern of behaviors and attitudes that makes a human being distinctive. Personality is formed by the ongoing interaction of temperament, character, and environment.

Protocol—A plan for carrying out a scientific study or a patient s course of treatment.

Reactivity—The level or intensity of a person's physical or emotional excitability.

Temperament—A person's natural disposition or inborn combination of mental and emotional traits.

Threshold—The minimum level of stimulation necessary to produce a response.

Trait—A distinguishing feature of an individual.

Type—A category used to define personality, usually based on a theory of some kind. Inhibited and uninhibited are examples of personality types.

See also Attention-deficit/hyperactivity disorder (ADHD); Magnetic resonance imaging; Personality development.

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Doreen Arcus, PhD

Testicular torsion

Definition

Testicular torsion is the twisting of a testis (testicle) such that the spermatic cord becomes twisted, cutting off blood flow to the testis.

Description

The testes are suspended in the scrotum by a single bundle of tissues called the spermatic cord. Normally this bundle of tissue holds the testes in place. Each testis receives blood through the spermatic cord. When the testicle is not held firmly in place it can twist, creating a kink in the spermatic cord. When this happens blood supply to the testis is cut off. The resulting situation is an emergency, because the testis will die within hours if the blood supply is not restored.

Demographics

There is approximately one case of testicular torsion in every 4,000 men under age 25 in the United States. There are two times in life when torsion is most common, although it can occur at any age. Testicular torsion is most common in the first year of life and during **adolescence**. Torsion is more common in adolescents that it is in newborns.

Causes and symptoms

Testicular torsion is caused by the rotating of the testicle is such a way that the blood flow to it is cut off. Symptoms of testicular torsion are sudden severe **pain** in the scrotum, swelling and/or discoloration of the scrotum, **nausea**, and **vomiting**. Approximately 40 percent of patients with testicular torsion reported having a similar pain sometime before, but at that time the pain resolved without treatment.

When to call the doctor

Testicular torsion is an emergency, and the child should be taken to the doctor or emergency room immediately if he shows the signs or symptoms of testicular torsion. The chance that the testicle will be saved is directly linked to how long the testicle is without blood flow. If the torsion occurs for less than six hours, there is a high chance the testicle can be saved. If the torsion occurs for more than 24 hours, it is very unlikely that the testicle can be saved.

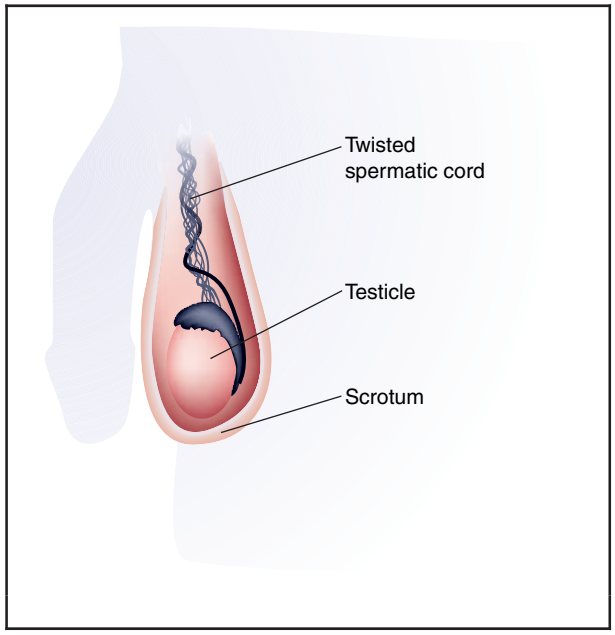
Diagnosis

The doctor usually first performs a visual examination of the scrotum. The affected testis may appear to be slightly higher than the unaffected one. The scrotum may be swollen or discolored. If the doctor is unsure, diagnostic tests may also be performed.

One such diagnostic test is a nuclear scan of the scrotum. In this procedure, a tiny amount of radioactive fluid is injected into the blood and detected as it flows through the scrotum and testicles. Torsion is indicated if the radioactive fluid does not flow through the sore testis. Ultrasound scan accompanied by a contrast agent can also be used to diagnose testicular torsion. Other diagnostic tests may be performed to help the doctor determine if torsion has occurred.

Treatment

Surgery performed within the first six hours has an 80 to 100 percent chance of saving the affected testis. This likelihood goes down the longer blood flow to the testis has been cut off. After 24 hours, it is very unlikely that the testicle can be saved. During the procedure, the surgeon untwists the cord and secures the testis in place



A rare condition, testicular torsion occurs when the spermatic cord is twisted and cuts off the blood supply to the testicle. *(Illustration by Argosy, Inc.)*

so that it cannot rotate again. This securing is called orchiopexy. The other testicle is also secured during the surgery to prevent future testicular torsion, because children who have had one episode of testicular torsion are likely to experience it again. If the testicle has not been untwisted in time and is dead, the surgeon will remove it.

While waiting for surgery, the doctor may try to restore blood flow to the testis by hand. This can help to save the testis if surgery is not possible right away. Surgery is still necessary, however, even if blood flow to the testis has been restored, because it is very likely that torsion will occur again.

Prognosis

If the torsion is relieved within six hours, it is very likely that the testis will recover normal blood flow and function. If the torsion continues for more than 24 hours, it is unlikely that the testis can be saved. One testis is all that is necessary for normal growth and maturation, as well as normal fertility later in life.

Prevention

The only way to prevent torsion is to surgically anchor the testes so that they cannot move. This is frequently done after an occurrence of torsion, both to the torsed testis and the unaffected testis.

KEY TERMS

Orchiopexy—A surgical procedure that places an undescended testicle in the scrotum and/or attaches a testicle to the scrotum.

Scrotum—The external pouch containing the male reproductive glands (testes) and part of the spermatic cord.

Spermatic cord—The tissue that suspends the testis inside the scrotum.

Parental concerns

Testicular torsion is usually very painful. If the torsion is not corrected quickly, the testis usually loses function. Only one healthy and functioning testis is required for normal growth and development and for normal fertility later in life.

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Tests for pregnant women *see* **Antenatal testing**

Tetanus

Definition

Tetanus, also called lockjaw, is a serious disease of the nervous system that can cause uncontrolled **muscle**

spasms and death. It is caused by toxins (poisons) produced by the bacterium *Clostridium tetani*.

Description

Tetanus occurs when the body is infected with spores of the bacterium *C. tetani*. This bacterium is found worldwide in soil and animal manure. The spores can remain alive in the soil for years and are resistant to heating and chemical destruction. They are more common in hot, damp environments than in cold or dry ones.

Once spores enter the body through a break in the skin, they begin producing bacteria. These bacteria multiply in areas where there is little oxygen present and produce a toxin that affects the nervous system. The toxin spreads along the nerves of the body, causing the nerves to fire (react). This results in muscle spasms and convulsions.

Transmission

The bacteria that cause tetanus enter the body through a scrape, cut, or wound, in about 70 percent of cases. The most susceptible **wounds** are those that are caused by blunt trauma such as crushing or by **bites**. The bacteria can also enter at the site of a burn, bedsore, or **frostbite**, or be introduced into the body during surgery. In developing countries, newborns often contract tetanus from contaminated instruments used to tie off the umbilical cord after birth. Often the site where the bacteria enter is insignificant, does not become swollen or red, and does not require medical attention. Any time between two and 50 days later (most commonly between days seven and 21 days), the individual begins to show the signs of tetanus.

The severity of the disease is related to several factors:

- The sooner symptoms appear, the more severe the disease.
- If the point of entry was in the head or face, symptoms are more severe.
- The very young and the very old suffer more severe symptoms and higher death rates.

Demographics

With almost universal **vaccination** starting in the 1940s, tetanus has become rare in the United States. Fewer than 50 cases have been reported annually since 1995. Worldwide, the disease is common, especially in newborns in developing parts of Asia, Africa, and South America where immunization is not universally available. The disease can affect individuals of any race, age, or gender.

Causes and symptoms

Since the incubation period can range from several days to many weeks, individuals often do not associate their initial symptoms with wound infection. The first sign of tetanus is a tightening of the jaw muscles that gives the disease its common name, lockjaw. This symptom is followed by waves of back spasms. The spasms then extend to the arms producing clenched fists and to the legs. Any stimulus, such as noise or light, can set off a round of convulsions. Other symptoms include drooling, increase in blood pressure (**hypertension**), irregular heart beat, inability to open the mouth, high **fever**, kidney failure, and respiratory failure.

When to call the doctor

Tetanus is a medical emergency, and individuals should be taken to the emergency room as soon as symptoms are noticed. About 75 percent of individuals with tetanus are first seen by a dentist or oral surgeon for **pain** and stiffness in the jaw and mouth region.

Diagnosis

Diagnosis of tetanus is based on presenting symptoms rather than laboratory tests. Less than one-third of the time can the bacteria that causes the disease be cultured from a wound.

Treatment

Treatment begins immediately in the emergency room or intensive care unit of a hospital. There are five aspects of treatment. Initially the patient is placed in a dark, quiet room and given a sedative, usually a drug in the benzodiazepine family, through direct injection into a vein (IV) in an effort to reduce muscle spasms. A tube may be inserted in to the trachea (tracheotomy) in order to keep the airways open.

The second aspect of treatment is to clean and disinfect any wounds and remove any dead flesh.

The third aspect of treatment involves killing the bacteria producing the toxin using antimicrobial drugs given as an injection. The drug of choice is metronidazole (Flagyl), with penicillin the second choice.

Fourth, the toxin already circulating in the blood must be neutralized so that it causes no further damage

to the nervous system. This is done with injections of human tetanus immunoglobulin (TIG).

Finally, complications of the disease are managed. This may involve IV fluid replacement, use of a respirator, or kidney dialysis. Contracting tetanus does not provide immunity against future infections, so tetanus immunizations are also given.

Prognosis

Individuals who develop symptoms within a few days of infection have close to a 100 percent mortality rate. The mortality rate for infections originating in the head and in newborns is also very high. The sooner an individual is treated, the more likely he or she is to survive. Overall, the death rate in the United States is 10 percent. Worldwide it is 45 percent. According the United States Centers for Disease Control, the average hospital stay is 16 days. Recovery for those who survive is normally complete after about four weeks.

Prevention

Tetanus is completely preventable by immunization. The recommendation in the United States, as of 2004, is to immunize children against tetanus on the following schedule:

- initial vaccination at two months of age
- repeat at four months of age
- repeat at six months of age
- repeat at 12 to 15 months of age
- repeat at four to six years of age
- booster dose given every 10 years there after, normally at ages 15, 25, 35, etc.

Receiving the complete schedule of multiple vaccinations is necessary to ensure full protection. For children, vaccination against tetanus is normally included in a vaccine called DTaP that protects against **diphtheria**, tetanus, and **whooping cough** (acellular pertussis). Many school districts require proof of vaccination before a child may enroll.

Other prevention measures involve prompt cleaning and protection of wounds and hygiene measure such as washing well after exposure to soil containing animal manure. Sterile conditions during surgery also help prevent infection.

KEY TERMS

Intravenous—Into a vein; a needle is inserted into a vein in the back of the hand, inside the elbow, or some other location on the body. Fluids, nutrients, and drugs can be injected. Commonly called IV.

Toxin—A poisonous substance usually produced by a microorganism or plant.

Trachea—The windpipe. A tube composed of cartilage and membrane that extends from below the voice box into the chest where it splits into two branches, the bronchi, that lead to each lung.

Tracheotomy—An surgical procedure in which the surgeon cuts directly through the patient's neck into the windpipe below a blockage in order to keep the airway open.

Umbilical cord—The blood vessels that allow the developing baby to receive nutrition and oxygen from its mother; the blood vessels also eliminate the baby's waste products. One end of the umbilical cord is attached to the placenta and the other end is attached to the baby's belly button (umbilicus).

Nutritional concerns

Food is not given by mouth to individuals who are having muscle spasms for **fear** they will breathe the food into their lungs. During this time, they are fed intravenously.

Parental concerns

Some parents hesitate to vaccinate their children for religious reasons or because they fear side effects of the vaccination. The bacteria that cause tetanus are so common and the disease is so serious that protection against acquiring tetanus outweighs any risks associated with vaccination.

See also Vaccination.

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Tetracyclines

Definition

Tetracyclines are a group of **antibiotics** that are useful in treatment of many bacterial infections.

Description

Tetracyclines are called broad-spectrum antibiotics because they can be used to treat a wide variety of infections. Physicians may prescribe these drugs to treat eye infections, **pneumonia**, gonorrhea, **Rocky Mountain spotted fever**, urinary tract infections, certain bacteria that could be used in biological weapons, and other infections caused by bacteria. The medicine is also used to treat **acne**. The tetracyclines will not work for colds, flu, and other infections caused by viruses. Tetracyclines are generally a low-cost alternative among antibiotics.

There are five drugs in the tetracycline class:

- demeclocycline
- doxycycline
- minocycline
- oxytetracycline
- tetracycline

General use

All tetracyclines are used for treatment of infections in patients over the age of eight years. They may be used in several forms, including capsules, injections, ointments, eye and ear drops.

Tetracyclines are bacteriostatic. They do not kill bacteria; they prevent bacteria from growing, so that the body's natural defenses are better able to deal with an infection. For this reason, tetracyclines are not used in patients with impaired immune systems.

Although all tetracyclines are similar, and can do most of the same work, there are some differences. Doxycycline requires only one dose a day and can be used even when the patient has kidney problems. Demeclocycline and minocycline penetrate the skin better than other tetracyclines and may be preferred for treatment of acne. Demeclocycline is effective for the syndrome of inappropriate anti-diuretic hormone (SIDAH), although it is not officially approved for this purpose.

In addition to their role in treating infections, tetracyclines have a wide range of other uses. These include protection against some types of malaria and treatment of some of the infections that might be used in bioterrorism. Some tetracycline derivatives have been useful in **cancer** therapy. Tetracyclines have been useful in prevention of gum diseases of the mouth.

Precautions

Tetracyclines should normally not be used in children under the age of eight because some tetracyclines can be absorbed into the bones and teeth and give the teeth a mottled appearance. Some experts believe that tetracyclines should be avoided in children younger than ten.

Side effects

Not all tetracyclines have the same side effects, but the following list includes some of the most common problems:

- dizziness and lightheadedness
- **diarrhea**
- stomach upset
- nausea
- vomiting
- photosensitivity
- fungus infections
- tooth discoloration
- mouth irritation
- skin discoloration

On rare occasions tetracyclines may cause more severe adverse effects, including kidney damage and drug-induced lupus.

Patients taking tetracyclines should avoid prolonged sun exposure. Standard **sunscreens** are not adequate to

protect against severe **sunburn** in patients taking tetracyclines.

Interactions

Tetracyclines should not be used at the same time the patient is receiving a live vaccine. The antibiotics may prevent the vaccine from growing, and this may keep the vaccine from producing immunity.

Moreover, tetracyclines may reduce the effectiveness of **oral contraceptives**.

Many antibiotics share tetracyclines' interaction with neuromuscular blocking agents. Tetracyclines should not be used at the same time as neuromuscular blocking agents since the antibiotics can increase the strength of the neuromuscular blocker, which can make breathing difficult. While this interaction is severe, it is rare, since the neuromuscular blocking agents are usually used only in surgery.

Tetracyclines should not be taken at the same time as foods containing calcium or foods containing iron, magnesium, or aluminum. The metals bind to the tetracycline, and the combination has reduced effect on bacteria.

The common interaction between tetracyclines and **minerals** can be avoided by taking tetracycline on an empty stomach, one hour before or two hours after meals, with water.

Parental concerns

Although it is recommended that tetracyclines not be given to children under the age of eight, the drug is sometimes required in severe infections. Tetracyclines may be required for children who have developed infections either in hospitals or while traveling overseas.

Parents should carefully check the expiration date of tetracycline and not use the drug past the expiration date. Expired tetracycline has been known to cause a severe kidney problem called Fanconi syndrome. Expired tetracycline should be disposed of, not saved.

Because tetracyclines can cause photosensitization, patients taking these drugs should use sunscreen and avoid direct sunlight.

Because of their interaction with metals, tetracyclines should always be taken on an empty stomach with only water. Patients should particularly avoid calcium-containing dairy products and antacids as well as multivitamin-mineral supplements.

KEY TERMS

Antibiotics—Drugs that are designed to kill or inhibit the growth of the bacteria that cause infections.

Bacteriostatic—An agent that prevents the growth of bacteria.

Fanconi's syndrome—A group of disorders involving kidney tubule malfunction and glucose, phosphate, and bicarbonate in the urine. Two forms of this syndrome have been identified: an inherited form and an acquired form caused by vitamin D deficiency or exposure to heavy metals.

Photosensitization—Development of oversensitivity to sunlight.

Tetracyclines inhibit the growth of many bacteria and other microorganisms which can lead to overgrowth of other microorganisms. Possible symptoms are discoloration of the tongue and diarrhea. Parents should report these problems to the prescriber immediately.

Parents should alert all health-care professionals about all drugs their children are taking. Both tetracycline and oral contraceptives are used to treat acne in teenage girls, but these drugs should not be used together.

See also Penicillins.

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Samuel Uretsky, PharmD

Tetralogy of Fallot

Definition

Tetralogy of Fallot is a common syndrome of congenital heart defects.

Description

The heart is two pumps in one. The ventricle on the left side pumps blood full of oxygen through the body; the ventricle on the right side pumps the same blood through the pulmonary artery to the lungs to take up oxygen. The left ventricle operates at pressures about four times as high as the right ventricle. Blood is supposed to flow through one side, then the other.

Tetralogy of Fallot is a condition that is characterized by several congenital heart defects occurring at once. They include: ventricular septal defect (abnormal passageway between the right and left ventricles), displaced aorta, narrowed pulmonary valve, thickened right ventricle wall.

Each defect acts in combination with the others to create a malfunction of the heart. The problem starts very early in the uterus with a narrowed pulmonary valve and a hole between the ventricles. This is not particularly a problem for a fetus because hardly any blood flows through the lungs until birth. It is only after birth that the defects pose a problem. The blood that is supposed to start flowing through the lungs cannot easily get there because of the narrowed valve; however, the hole between the ventricles remains open. Because of the opening between ventricles, much of the blood that comes back to the heart needing oxygen is sent out without being properly oxygenated. In addition, the right heart has to pump at the same pressure as the left side. Several changes follow. First, the baby turns blue (cyanotic) because of the deoxygenated blood that bypasses the lungs. Deoxygenated blood is darker and appears blue through the skin. Second, the right side of the heart (ventricle) hypertrophies (gets more muscular) from the extra exercise demanded of it. Next, the low oxygen causes the blood to get thicker and clot more easily. Clots in the veins can now pass through the hole in the heart and directly enter the aorta, where they can do much more damage than in the lungs such as causing infarcts in the brain. In addition, these anomalies make the lining of the heart more susceptible to infection (endocarditis), which can damage valves and lead to blood poisoning (septicemia).

Demographics

Researchers estimate that tetralogy of Fallot occurs in approximately one in every 2000 births. In the United States, almost 10 percent of **congenital heart disease** is tetralogy of Fallot. Boys are slightly more likely to have this malformation than girls.

Causes and symptoms

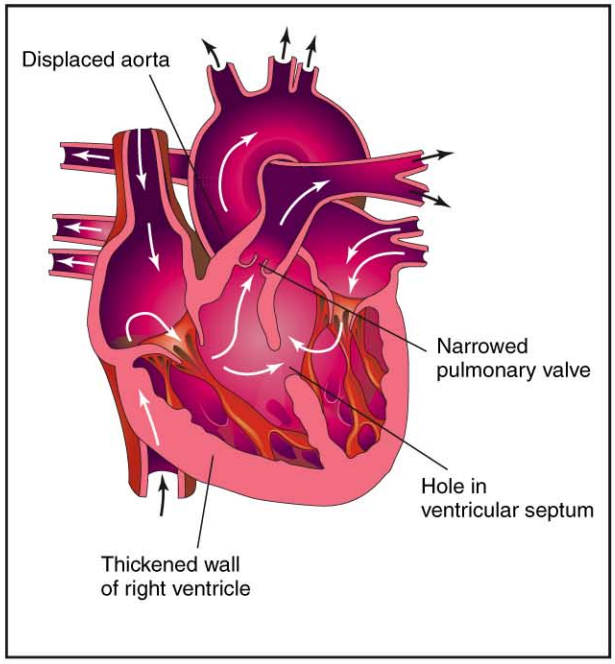
Tetralogy of Fallot is a congenital defect with unknown causes. Babies with tetralogy of Fallot are blue at birth or cyanotic. Sometimes the blue color appears only when they cry. They also have detectable **heart murmurs**. Infants with mild forms can have surgery postponed until they are older. Infants with more severe symptoms often have attacks of worsened cyanosis. During attacks, they turn very blue, have shortness of breath, and can faint. These symptoms usually occur during heightened activity, such as crying.

Diagnosis

A complete evaluation of the circulation is required, testing the blood for its oxygen content. Three diagnostic tests are performed: an echocardiogram, a chest x ray, and an electrocardiogram.

Treatment

Correction of the defects is done through open heart surgery. Surgery must be carefully timed with attention to the progression of the disease process, the size of the infant,



Tetralogy of Fallot is a common syndrome of congenital heart defects. This condition, present *in utero,* is caused by the narrowing of the pulmonary artery and a hole between the ventricles. When the baby is born and begins to breathe on its own, the baby turns cyanotic, or blue, due to the deoxygenated blood that bypasses the lungs because the narrowed pathway and the hole between the ventricles has remained open. *(Illustration by Electronic Illustrators Group.)*

KEY TERMS

Aorta—The main artery located above the heart that pumps oxygenated blood out into the body. The aorta is the largest artery in the body.

Cyanotic—Marked by a bluish tinge to the skin that occurs when the blood oxygen level drops too low. It is one of the types of congenital heart disease.

Deoxygenated blood—Blood that does not contain oxygen.

Endocarditis—Inflammation of the inner membrane lining heart and/or of the heart valves caused by infection.

Infarct—An area of dead tissue caused by inadequate blood supply.

Paroxysmal hypercyanoic attacks—Sudden episodes of cyanosis resulting from the circulation of deoxygenated blood to the body.

Septicemia—A systemic infection due to the presence of bacteria and their toxins in the bloodstream. Septicemia is sometimes called blood poisoning.

Systemic circulation—Refers to the general blood circulation of the body, not including the lungs.

Ventricles—The lower pumping chambers of the heart. The ventricles push blood to the lungs and the rest of the body.

and the size of the various defects. There are temporary surgical procedures that can prolong the time before corrective surgery, while the baby grows larger and stronger.

During surgery, the pulmonary valve is widened, the ventricular septal defect is closed, and any interim correction is removed.

Prognosis

Surgical correction has a high rate of success, returning the child to near-normal health.

Prevention

There is no known prevention for tetralogy of Fallot.

Parental concerns

Tetralogy of Fallot is a complex congenital malformation; however, open heart surgery is highly effective in correcting it. Most children have an excellent outcome and a normal healthy life. For most children, activity level, appetite, and growth eventually return to normal. Open heart surgery to repair tetralogy of Fallot is usually performed in children between the ages of six months and two years. Children with severe tetralogy of Fallot will begin the process of surgical correction in infancy. For children in whom the condition is milder, corrective surgery may be postponed until the child is older and has grown. While waiting for corrective surgery, children may experience episodes called paroxysmal hypercyanotic attacks, in which the child may cry intensely or become restless, turn blue (especially around the lips, fingernails, and toenails), and sometimes faint. These attacks can be quite serious and may require emergency medical care. For infants, holding the baby on the parent's shoulder with the infant's knees tucked underneath him may help reduce the symptoms. Older children may crouch in a squatting position.

After the child's heart surgery, parents should follow all instructions given by the healthcare team. Most children will continue to be seen by a team of doctors including the pediatrician, cardiologist, and pediatric cardiac surgeon.

When to call the doctor

Following open heart surgery, parents should call the doctor if any of the following occurs:

- fever of 101.5°F (38.6°C) or higher
- swelling or puffiness around the child's eyes, arms, or legs
- redness or swelling, cloudy yellow drainage, or an opening at the incision site
- rapid breathing
- increased fatigue or tiredness
- dry **cough** that was not present before surgery
- decreased appetite or refusal to eat
- increased pain

Resources

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Thalassemia

Definition

Thalassemia describes a group of inherited disorders characterized by reduced or absent amounts of hemoglobin. Hemoglobin is the protein in red blood cells that carries oxygen throughout the body. There are two basic groups of thalassemia disorders: alpha thalassemias and beta thalassemias. These conditions cause varying degrees of anemia, which can range from insignificant to fatal.

Description

Thalassemia is a genetic disorder. It cannot be acquired from contact with other people or from the environment. In all types of thalassemia, the quantity of hemoglobin produced is reduced or absent. This circumstance affects the ability of the blood to carry oxygen to all parts of the body. Although both alpha and beta thalassemias affect hemoglobin, these diseases affect the body in distinctly different ways. Hemoglobin is made up of three components: alpha globin, beta globin, and heme. Thalassemias are classified according to the globin that is deficient.

Alpha thalassemia

Individuals inherit from each parent a gene controlling alpha globin production. Two spots (called loci) on these genes control alpha globin production. Alpha thalassemias result from changes (mutations) in these genes. There are two main types of alpha thalassemia disease: hemoglobin H disease and alpha thalassemia major. The two diseases are quite different from beta thalassemia, as well as from one another.

Individuals with hemoglobin H disease have inherited one completely defective gene and one gene that has one rather than two functional loci. This circumstance substantially reduces the amount of alpha globin that the body produces. As a result, individuals with hemoglobin H disease can experience events of hemolytic anemia—anemia caused by the rapid breakdown of the red blood cells. These events are thought to be triggered by various environmental causes, such as infection and/or exposure to certain chemicals. Hemoglobin H disease is milder than alpha thalassemia and usually milder than beta thalassemia.

Individuals with alpha thalassemia major have inherited two completely defective genes, one from each parent. Alpha thalassemia major, sometimes called hemoglobin Barts or hydrops fetalis, is a fatal disease that results in severe anemia that begins even before birth. Most affected babies do not survive to be born or die shortly after birth.

Beta thalassemia

Beta thalassemia, also called Cooley's anemia, is the most well known type of thalassemia. It is caused by a change in the gene for the beta globin component of hemoglobin. Beta thalassemia causes variable anemia that can range from moderate to severe, depending in part on the exact genetic change underlying the disease.

Beta thalassemia major causes severe anemia that usually occurs within three to six months after birth. If

left untreated, severe anemia can result in stunted growth and development, as well as other characteristic physical complications that can lead to a dramatically decreased life expectancy. In developed countries, screening in the newborn period usually identifies beta thalassemia before symptoms have developed. Children who are identified early can be started on ongoing blood transfusion therapy as needed.

Beta thalassemia minor describes a disease where only one gene of the pair that control beta hemoglobin production is defective. There are few or mild events. However, the individual can pass the defective gene on to his or her offspring

Beta thalassemia intermedia is a clinical term that describes the disease in individuals who have moderate anemia that only requires blood transfusions intermittently.

Demographics

The thalassemias are among the most common genetic diseases worldwide. Both alpha and beta thalassemia have been described in individuals of almost every ancestry, but the conditions are more common among certain ethnic groups. Unaffected carriers of all types of thalassemia traits do not experience health problems.

Determining the prevalence for alpha thalassemia is difficult due to limitations in diagnostic testing. In the United States, up to 30 percent of African Americans are thought to be carriers for alpha thalassemia traits, meaning that they show no symptoms of the disorder but can pass the trait to their offspring. Despite this estimate, the number of babies born with hemoglobin H disease or alpha thalassemia major is very low. The highest frequency of alpha thalassemia diseases occurs in individuals of Southeast Asian and Chinese descent. Individuals of Greek, Middle Eastern, and North African descent also carry genes for the disease more frequently than individuals of Northern European descent. One study of 500 pregnant women in northern Thailand estimated a frequency of one in 500 pregnancies affected by alpha thalassemia major, for example. Prevalence of alpha thalassemia disease is significantly lower in the United States owing primarily to immigration patterns. However, at least one state, California, has observed growing hemoglobin H disease rates that are high enough to justify universal newborn screening for the condition.

Beta thalassemia trait is seen most commonly in people with the following ancestry: Mediterranean (including North African, and particularly Italian and Greek), Middle Eastern, Indian, African, Chinese, and Southeast Asian (including Vietnamese, Laotian, Thai, Singaporean, Filipino, Cambodian, Malaysian, Burmese, and Indonesian). It is difficult to obtain accurate prevalence figures for various types of thalassemia within different populations.

Two studies reflect prevalence figures that can be helpful counseling families and determining who to screen for beta thalassemia. Between the years of 1990 and 1996, the State of California screened over 3.1 million newborns for beta thalassemia. Approximately one in 114,000 infants had beta thalassemia major, with prevalence rates being highest among Asian Indians (about one in 4,000), Southeast Asians (about one in 10,000), and Middle Easterners (about one in 7,000). The pattern observed in California is expected to be different in other areas of the United States and the world. For example, Italians are underrepresented in this population when compared to the population of the East Coast of the United States.

Causes and symptoms

Humans normally make several types of hemoglobin. An individual's stage in development determines whether he or she makes primarily embryonic, fetal, or adult hemoglobins. All types of hemoglobin are made of three components: heme, alpha globin, and beta globin. All types of thalassemia are caused by changes in either the alpha- or beta-globin gene. These changes cause little or no globin to be produced. All types of thalassemias are recessively inherited, meaning that a genetic change must be inherited from both the mother and the father to produce the disease in the child. The severity of the disease is influenced by the exact thalassemia mutations inherited, as well as other genetic and environmental factors. There are rare exceptions, notably with beta thalassemia, where globin gene mutations exhibit a dominant pattern of inheritance in which only one gene needs to be altered in order to see disease expression.

Alpha thalassemia

Most individuals have four normal copies of the alpha globin gene, two copies on each chromosome 16. These genes make the alpha globin component of normal adult hemoglobin, which is called hemoglobin A. Alpha globin is also a component of fetal hemoglobin. Since there are four genes (instead of the usual two) to consider when looking at alpha globin gene inheritance, there are several alpha globin types that are possible.

Absence of one functioning alpha globin gene leads to a condition known as silent alpha thalassemia trait. This condition causes no health problems and can be

detected only by special genetic testing. Alpha thalassemia trait occurs when two alpha globin genes are missing or not functioning. There are no associated health problems, although the trait status may be detected by more routine blood screening.

Hemoglobin H disease results from the deletion of three of the four alpha globin genes. Hemoglobin H symptoms can also be a part of a unique condition called alpha thalassemia **mental retardation** syndrome. This syndrome can be caused by a deletion of a significant amount of chromosome 16, affecting the alpha globin genes. This situation is usually not inherited, but rather occurs sporadically in the affected individual. Affected individuals have mild hemoglobin H disease, mild-to-moderate mental retardation, and characteristic facial features, as well as various other developmental processes that mimic hemoglobin H disease.

Alpha thalassemia major results from the deletion of all four alpha globin genes, such that there are no functioning alpha globin genes. In this situation, there is a 25 percent chance for alpha thalassemia major in each of such a couple's children.

Beta thalassemia

Most individuals have two normal copies of the beta globin gene, which is located on chromosome 11 and makes the beta globin component of normal adult hemoglobin. There are approximately one hundred genetic mutations that have been described that cause beta thalassemia, designated as either beta0 or beta+ mutations. No beta globin is produced with a beta0 mutation, and only a small fraction of the normal amount of beta globin is produced with a beta+ mutation.

When an individual has one normal beta globin gene and one with a beta thalassemia mutation, he or she is said to carry the beta thalassemia trait. Carrying the trait is generally thought not to cause health problems, although some women with beta thalassemia trait may have an increased tendency toward anemia during pregnancy.

When both parents carry the beta thalassemia trait, there is a 25 percent chance that each of their children will inherit beta thalassemia disease by inheriting two beta thalassemia mutations, one from each parent. The clinical severity of the beta thalassemia disease depends largely on whether the mutations inherited are beta0 thalassemia or beta+ thalassemia mutations. Two beta0 mutations generally lead to beta thalassemia major, and two beta+ thalassemia mutations generally lead to beta thalassemia intermedia, a milder form of the disease. Inheritance of one beta0 and one beta+ thalassemia mutation tends to be less predictable.

Symptoms

Hemoglobin H disease

Hemoglobin H disease is a relatively mild form of thalassemia that may go unrecognized. It is not generally considered a condition that will reduce one's life expectancy. Education is an important part of managing the health of an individual with hemoglobin H disease. It is important to be able to recognize the signs of severe anemia that require medical attention. It is also important to be aware of the medications, chemicals, and other exposures to avoid due to the theoretical risk they pose of precipitating a severe anemia event. When severe anemia occurs, it is treated with blood transfusion therapy. For many individuals with hemoglobin H disease, this is rarely required. For those with a more severe form of the disease, the need for transfusions may be intermittent or ongoing, perhaps on a monthly basis, and require desferoxamine treatment. This treatment removes excess iron from the body. Individuals with this more severe form of the disease may also have an increased chance of requiring removal of an enlarged and/or overactive spleen.

Alpha thalassemia major

Because alpha globin is a necessary component of hemoglobin, absence of all functioning alpha globin genes leads to serious medical consequences that begin even before birth. Affected fetuses develop severe anemia as early as the first trimester of pregnancy. The placenta, heart, liver, spleen, and adrenal glands may all become enlarged. Fluid can begin collecting throughout the body as early as the start of the second trimester, causing damage to developing tissues and organs. Growth retardation is also common. Affected fetuses usually miscarry or die shortly after birth. In addition, women carrying affected fetuses are at increased risk of developing complications of pregnancy and delivery. Up to 80 percent of such women develop toxemia, a disturbance of metabolism that can potentially lead to convulsions and coma. Other maternal complications include premature delivery and increased rates of delivery by **cesarean section**, as well as hemorrhage after delivery.

Beta thalassemia major is characterized by severe anemia that can begin several months after birth. In the United States and other developed countries beta thalassemia is identified and treated early and effectively. Therefore, the following discussion of symptoms applies primarily to affected individuals in the past and in some underdeveloped countries as of the early 2000s. If

untreated, beta thalassemia major can lead to severe lethargy, paleness, and growth and **developmental delay**. The body attempts to compensate by producing more blood, which is made inside the bones in the marrow. However, this effort is ineffective without the needed genetic instructions to make enough functioning hemoglobin. Instead, obvious bone expansion and changes occur that cause characteristic facial and other changes in appearance, as well as increased risk of **fractures**. Severe anemia taxes other organs in the body such as the heart, spleen, and liver, which must work harder than usual. This stress can lead to heart failure, as well as enlargement and other problems of the liver and spleen. When untreated, beta thalassemia major generally results in childhood death, usually due to heart failure. In developed countries, diagnosis is usually made early, often before symptoms have begun. This factor allows for treatment with blood transfusion therapy, which can prevent most of the complications of the severe anemia caused by beta thalassemia major.

Individuals with beta thalassemia intermedia have a more moderate anemia that may only require treatment with transfusion intermittently, such as when infections stress the body. As a person with beta thalassemia intermedia gets older, however, the need for blood transfusions may increase to the point that they are required on a regular basis. When this occurs the disease becomes more similar to beta thalassemia major. Other genetic and environmental factors can influence the course of the disease as well. For example, co-inheritance of one or two alpha thalassemia mutations can tend to improve some of the symptoms of beta thalassemia disease, which results in part from an imbalance in the amount of alpha- and beta-globin present in the red blood cells.

When to call the doctor

Signs of thalassemia diseases are often noted by the doctor during newborn screening. Parents should contact their doctors if they suspect any developmental delays, especially if the parents belong to one of the ethnic groups at higher risk for the disease.

Diagnosis

Diagnosis of thalassemia can occur under various circumstances and at various ages. Several states offer thalassemia screening as part of the usual battery of blood tests done on newborns. This arrangement allows for early identification and treatment. Thalassemia can be identified before birth using prenatal diagnosis. Chorionic villus sampling (CVS) can be done as early as 10 weeks of pregnancy. It involves removing a sample of the placenta and testing the cells. CVS carries a risk of causing a miscarriage that is between 0.5 percent and 1 percent. **Amniocentesis** is generally done between 15 and 22 weeks of pregnancy but can sometimes be offered earlier. Two to three tablespoons of the fluid surrounding the baby are removed. This fluid contains fetal cells that can be tested. The risk of miscarriage associated with amniocentesis ranges from 0.33 to 0.5 percent.

Pregnant women and couples may choose prenatal testing in order to prepare for the birth of a baby that may have thalassemia. Alternately, knowing the diagnosis during pregnancy allows for the option of pregnancy termination. Preimplantation genetic diagnosis (PGD) is a relatively new technique that involves in-vitro fertilization followed by genetic testing of one cell from each developing embryo. Only the embryos unaffected by the disease are transferred back into the uterus.

Thalassemia may be suspected if an individual shows signs that are suggestive of the disease. In all cases, however, laboratory tests are essential to confirm the exact diagnosis and to allow for the provision of accurate genetic counseling about recurrence risks and testing options for parents and affected individuals. Screening is likewise recommended to determine trait status for individuals of high-risk ethnic groups.

The following tests are used to screen for thalassemia disease and/or trait:

- complete blood count
- hemoglobin electrophoresis
- free erythrocyte-protoporphyrin (or ferritin or other studies of serum iron levels)

A complete blood count will identify low levels of hemoglobin, small red blood cells, and other red blood cell abnormalities that are characteristic of a thalassemia diagnosis. Since thalassemia trait can sometimes be difficult to distinguish from iron deficiency, tests to evaluate iron levels are important.

Hemoglobin electrophoresis is a test that can help identify the types and quantities of hemoglobin made by an individual. This test uses an electric field applied across a slab of gel-like material. Hemoglobins migrate through this gel at various rates and to specific locations, depending on their size, shape, and electrical charge. Isoelectric focusing and high-performance liquid chromatography (HPLC) use similar principles to separate hemoglobins. They can be used instead of or in various combinations with hemoglobin electrophoresis to determine the types and quantities of hemoglobin present. Hemoglobin electrophoresis results are usually within the normal range for all types of alpha thalassemia. Hemoglobin electrophoresis can also detect structurally

abnormal hemoglobins that may be co-inherited with a thalassemia trait. Sometimes DNA testing is needed in addition to the above screening tests. This test can be performed to help confirm the diagnosis and establish the exact genetic type of thalassemia.

Treatment

Because alpha thalassemia major is most often a fatal condition in the prenatal or newborn period, treatment has previously been focused on identifying affected pregnancies in order to provide appropriate management to reduce potential maternal complications. Pregnancy termination provides one form of management. Increased prenatal surveillance and early treatment of maternal complications is an approach that is appropriate for mothers who wish to continue their pregnancy with the knowledge that the baby will most likely not survive. In the last decade of the twentieth century and early 2000s, a handful of infants with this condition have survived long-term. Most of these infants received experimental treatment including transfusions before birth, early delivery, and bone marrow transplantation before birth, although the latter procedure had, as of 2004, not yet been successful. For those infants who survive to delivery, there seems to be an increased risk of developmental problems and physical effects, particularly heart and genital malformations. Otherwise, the medical outlook is similar to a child with beta thalassemia major, with the important exception that ongoing, lifelong blood transfusions begin at birth.

Beta thalassemia

Individuals with beta thalassemia major receive regular blood transfusions, usually on a monthly basis. This helps prevent severe anemia and allow for growth and development that is more normal. Transfusion therapy does have limitations, however. Individuals can develop reactions to certain proteins in the blood, called a transfusion reaction. Such a reaction can make locating appropriately matched donor blood more difficult. Although blood supplies in the United States are very safe, there remains an increased risk of exposure to such blood-borne infections as hepatitis.

An additional side effect of repeated transfusions is that the body is unable to get rid of the excess iron that accompanies each transfusion. A medication called desferoxamine is administered, usually five nights per week over a period of several hours, using an automatic pump that can be used during **sleep** or taken anywhere the person goes. This medication is able to bind to the excess iron, which can then be eliminated through urine.

If desferoxamine is not used regularly or is unavailable, iron overload can develop and cause tissue damage and organ damage and failure. The heart, liver, and endocrine organs are particularly vulnerable. Desferoxamine itself may produce on rare occasions allergic or toxic side effects, including hearing damage. Signs of desferoxamine toxicity are screened for and generally develop in individuals who overuse the medication when body iron levels are sufficiently low. Overall, however, transfusion and desferoxamine therapy have increased the life expectancy of individuals with the most severe types of beta thalassemia major to the fourth or fifth decade.

As of 2004, new treatments including medications that target the production of red blood cells (e.g. erythropoeitin) or fetal hemoglobin (e.g. hydroxyurea and butyrate) and bone marrow transplantation may offer more effective treatment of beta thalassemia major. Other possible treatments may include gene therapy techniques aimed at increasing the amount of normal hemoglobin the body is able to make.

Prognosis

Prognosis, as noted above, depends on the type and severity of the disease. Individuals with severe disease may be stillborn or die shortly after birth. On the other hand, some individuals with mild disease have a relatively normal life expectancy.

Prevention

Thalassemias are inherited diseases that cannot be prevented. It is, however, possible to identify carriers of the disease and provide them with genetic counseling and appropriate information concerning the chance of their offspring having thalassemia disease.

Individuals with hemoglobin H disease can reduce the likelihood of symptoms by avoiding infections and certain environmental triggers.

Parental concerns

If parents are thinking of having a child and believe they might be carriers of defective hemoglobin genes, they can be screened and receive genetic counseling so that they can assess their options.

Resources

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KEY TERMS

Anemia—A condition in which there is an abnormally low number of red blood cells in the bloodstream. It may be due to loss of blood, an increase in red blood cell destruction, or a decrease in red blood cell production. Major symptoms are paleness, shortness of breath, unusually fast or strong heart beats, and tiredness.

Bilirubin—A reddish yellow pigment formed from the breakdown of red blood cells, and metabolized by the liver. When levels are abnormally high, it causes the yellowish tint to eyes and skin known as jaundice. Levels of bilirubin in the blood increase in patients with liver disease, blockage of the bile ducts, and other conditions.

Bone marrow—The spongy tissue inside the large bones in the body that is responsible for making the red blood cells, most white blood cells, and platelets.

Bone marrow transplantation—A medical procedure in which a quantity of bone marrow is extracted through a needle from a donor, and then passed into a patient to replace the patient's diseased or absent bone marrow.

Desferoxamine—The primary drug used in iron chelation therapy. It aids in counteracting the life-threatening buildup of iron in the body associated with long-term blood transfusions.

Globin—One of the component protein molecules found in hemoglobin. Normal adult hemoglobin has a pair each of alpha-globin and beta-globin molecules.

Heme—The iron-containing molecule in hemoglobin that serves as the site for oxygen binding.

Hemoglobin—An iron-containing pigment of red blood cells composed of four amino acid chains (alpha, beta, gamma, delta) that delivers oxygen from the lungs to the cells of the body and carries carbon dioxide from the cells to the lungs.

Hemoglobin A—Normal adult hemoglobin that contains a heme molecule, two alpha-globin molecules, and two beta-globin molecules.

Hemoglobin electrophoresis—A laboratory test that separates molecules based on their size, shape, or electrical charge. It is used to identify abnormal hemoglobins in the blood.

Hydroxyurea—A drug that has been shown to induce production of fetal hemoglobin. Fetal hemoglobin has a pair of gamma-globin molecules in place of the typical beta-globins of adult hemoglobin. Higher-than-normal levels of fetal hemoglobin can ameliorate some of the symptoms of thalassemia.

Iron overload—A side effect of frequent blood transfusions in which the body accumulates abnormally high levels of iron. Iron deposits can form in organs, particularly the heart, and cause life-threatening damage.

Jaundice—A condition in which the skin and whites of the eyes take on a yellowish color due to an increase of bilirubin (a compound produced by the liver) in the blood. Also called icterus.

Mutation—A permanent change in the genetic material that may alter a trait or characteristic of an individual, or manifest as disease. This change can be transmitted to offspring.

Placenta—The organ that provides oxygen and nutrition from the mother to the unborn baby during pregnancy. The placenta is attached to the wall of the uterus and leads to the unborn baby via the umbilical cord.

Red blood cell—Cells that carry hemoglobin (the molecule that transports oxygen) and help remove wastes from tissues throughout the body.

Screening—A process through which carriers of a trait may be identified within a population.

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ORGANIZATIONS

Children's Blood Foundation. 333 East 38th St., Room 830, New York, NY 10016–2745. Web site: <www.childrensbloodfoundation.org>.

Cooley's Anemia Foundation Inc. 129–09 26th Ave. #203, Flushing, NY 11354. Web site: <www.thalassemia.org>.

March of Dimes Birth Defects Foundation. 1275 Mamaroneck Ave., White Plains, NY 10605. Web site: <www.modimes.org>.

National Organization for Rare Disorders (NORD). PO Box 8923, New Fairfield, CT 06812–8923. Web site: <www.rarediseases.org>.

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Thematic Apperception Test

Definition

The Thematic Apperception Test is a projective personality test.

Purpose

The Thematic Apperception Test (TAT) is widely used to research certain topics in psychology, such as dreams and fantasies, mate selection, the factors that motivate people's choice of occupations, and similar subjects. It is sometimes used in psychiatric evaluations to assess disordered thinking and in forensic examinations to evaluate crime suspects, even though it is not a diagnostic test. The TAT can be used to help people understand their own personality in greater depth and build on that knowledge in making important life decisions. Lastly, it is sometimes used as a screener in psychological evaluations of candidates for high-stress occupations (law enforcement, the military, religious ministry, for example).

Description

The TAT is a projective personality test that was designed at Harvard University in the 1930s by Christiana D. Morgan and Henry A. Murray. Along with the **Minnesota Multiphasic Personality Inventory** (MMPI) and the Rorschach inkblot test, the TAT is one of the most widely used **psychological tests**. A projective test is one in which a person's patterns of thought, attitudes, observational capacity, and emotional responses are evaluated on the basis of responses to ambiguous test materials. The TAT consists of 31 pictures that depict a variety of social and interpersonal situations. The subject is asked to tell a story to the examiner about each picture. Of the 31 pictures, ten are gender-specific while 21 others can be used with adults of either sex and with children.

There is no standardized procedure or set of cards for administering the TAT, except that it is a one-on-one test. It cannot be administered to groups. In one common method of administration, the examiner shows the subject only ten of the 31 cards at each of two sessions. The sessions are not timed, but average about an hour in length.

Precautions

The TAT has been criticized for its lack of a standardized method of administration as well as a lack of standard norms for interpretation. Studies of the interactions between examiners and test subjects have found that the race, sex, and social class of both participants influence both the stories that are told and the way the stories are interpreted by the examiner. Attempts have been made to design sets of TAT cards for African American and for elderly test subjects, but the results have not been encouraging. In addition, the 31 standard pictures have been criticized for being too gloomy or depressing; therefore, they may limit the range of personality characteristics that the test can assess.

Preparation

There is no specific preparation necessary before taking the TAT, although most examiners prefer to schedule sessions (if there is more than one) over two days.

Risks

The chief risks involved in taking the TAT are a bad "fit" between the examiner and the test subject and misuse of the results.

Parental concerns

The TAT does not yield a score, so its results can be difficult to interpret. It is important for parents to remember that the results of a single personality test may not accurately reflect their child's skills, talents, or problems and that there should not be too much emphasis placed upon the results of a single test.

Resources

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Rorschach—A projective test in which the participant is asked to interpret inkblots.

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ORGANIZATIONS

American Academy of Child and Adolescent Psychiatry. 3615 Wisconsin Avenue, NW, Washington, DC 20016–3007. Web site: <www.aacap.org>.

Helen Davidson
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Therapeutic baths

Definition

Bathing the skin in a variety of preparations in order to remove crusts, scales, and old medications or to relieve inflammation and **itching** is called taking a therapeutic bath. The term therapeutic bath is also used to refer to various types of warm-water soaks used to speed wound healing, to apply gentle heat to sore muscles or joints, to relieve emotional stress, or to treat a variety of physical disorders ranging from **sports injuries**, rheumatoid arthritis, and chronic **sinusitis** to painful **menstruation** and vascular disorders. Therapeutic baths are one form of hydrotherapy, which is a general term for the internal or external use of water for medical treatment.

Balneotherapy is the medical term for the use of baths or soaks to treat injuries or illnesses. It comes from the Latin word *balneum*, which means bath. Balneotherapy has been used for thousands of years to treat skin disorders, arthritis, paralysis, gynecological disorders, and depression and other emotional problems. The remains of ancient baths have been found in the Indus Valley in India, and the Romans discovered mineral springs in various parts of Europe that are still used for balneotherapy.

Purpose

Baths or soaks are an easy way to treat a variety of skin disorders involving large areas of the skin, injuries to or disorders of the muscles and joints, menstrual and menopausal discomfort, fatigue, or general stress and tension. They relieve general aches and pains and can ease dry or oily, inflamed or itchy skin. Hot baths are relaxing and stimulating; cool baths can reduce inflammation.

In children as well as adults therapeutic baths are useful for itchy skin, **hives**, **sunburn**, chafing, **poison ivy** and oak, eczema, skin irritation, and dry skin. They may also help to relieve emotional tension and stress. Warm-water soaks are recommended for speeding recovery from **sprains**, muscle aches and pains, and other athletic injuries.

Many **family** care physicians recommend warm-water therapeutic baths as a way to relieve labor pains during **childbirth** without administering drugs.

Therapeutic baths are used to treat a wider variety of disorders and injuries in Europe and the French-speaking parts of Canada than in the United States. In Eastern Europe and the countries of the former Soviet Union, therapeutic baths are used to treat children suffering from the aftereffects of head trauma as well as other physical injuries. One Italian spa lists recurrent earaches, sinus infections, and **acne** among the conditions that can be treated with therapeutic baths for children and adolescents as well as adults. European doctors often use mineral water in therapeutic baths or add seaweed, dried moss, mud, or various mineral salts to the bath water.

Description

For a therapeutic bath to treat eczema, the tub should be filled half-full with water at a comfortable temperature. The water should not be allowed to cool too much.

Different types of therapeutic baths are used for different skin conditions. The following are some examples:

- Colloidal oatmeal (oatmeal that has been ground into a fine powder, e.g. Aveeno) coats, soothes, and stops itching without drying out the skin.
- Potassium permanganate—a dark purple salt—makes a good disinfectant.
- Bath oils are used as an emollient to ease itchy skin and eczema. RoBathol and cottonseed oil are recommended for younger children.
- Cornstarch is a soothing, drying bath for itchy skin.
- Sodium bicarbonate can be cooling for sunburn or other hot, dry skin conditions.
- Saline (salt) water baths can be used to treat eczema in children. The recommended amount is one cup to a tubful of warm water.
- Chlorine bleach can be added to bath water for children who develop recurrent skin infections with eczema. The recommended amount is two teaspoons per gallon of water.

Therapeutic baths to treat **sports** injuries or relieve menstrual cramps may use slightly warmer water than is used to relieve skin disorders. Adolescents using therapeutic baths to relieve emotional stress may add a few drops of essential oils of lavender or other fragrant herbs to the bath water. Some people like to add eucalyptus oil to the bath water to relieve nasal congestion when they are recovering from colds or sinusitis.

Precautions

The temperature of the water for a therapeutic bath should feel comfortable to the hand. The bath should not last longer than 20 to 30 minutes because of the tendency of these soaks to soften and wear away the skin.

A bath mat should be placed in the tub before adding water, since medications may cause the floor of the tub to be slippery.

Eczema and other skin diseases can be treated with an ointment that contains a derivative of coal tar. Parts of the coal tar are volatile, so the bathroom should be well ventilated.

Parents should not leave small children alone in the bath because of the risk of drowning.

Essential flower or herb oils used to scent therapeutic baths should always be added to the water; they should never be applied directly to the skin.

Preparation

Parents should keep the room warm to minimize temperature fluctuations. This precaution is particularly important when bathing infants or younger children.

Parents should also take appropriate **safety** precautions, including removing hair dryers, electric shavers, or other small electrical appliances from the tub area. Another important safety precaution is to check the temperature setting on the hot water heater to make sure that it does not raise the temperature of the water to the scalding point. The standard factory setting on new household water heaters is 120°F (49°C), which is the highest setting considered to be safe. It is better for large families to purchase a larger hot water heater if there is a concern about the availability of hot water than to turn the thermostat on the heater higher than 120°F (49°C).

Aftercare

After the bath, the skin should be blotted (not rubbed) carefully with a towel. The patient should wear loose, light clothing after the bath. If the child or adolescent is being treated for eczema, an emollient should be applied within three minutes. Parents may use vegetable oil, petroleum jelly, or such commercial creams as Aveeno, Curel, Purpose, Dermasil, Neutrogena, DML Forte, and Eucerin. Some doctors may recommend preparations containing urea, lactic acid, or alpha-hydroxy acid.

Teenagers using therapeutic baths as part of rehabilitation after an athletic injury should follow the recommendations of their doctor or physical therapist regarding range-of-motion exercises or other treatments following the warm-water soak.

Risks

The most common risks associated with therapeutic baths are falls caused by loss of balance on a wet or slippery surface, electrocution caused by a hair dryer or other small appliance falling into the tub, scalding accidents from overheated water, and accidental drowning.

Some older children or adolescents may experience fatigue or a drop in blood pressure from long immersion in a therapeutic bath.

KEY TERMS

Balneotherapy—The medical term for the use of baths to treat disease.

Eczema—A superficial type of inflammation of the skin that may be very itchy and weeping in the early stages; later, the affected skin becomes crusted, scaly, and thick.

Hydrotherapy—The use of water (hot, cold, steam, or ice) to relieve discomfort and promote physical well-being. Also called water therapy.

Parental concerns

Therapeutic baths are a common and inexpensive treatment for a variety of skin disorders, menstrual cramps, and minor aches and pains. The chief concern of parents should be taking appropriate safety precautions regarding the hot water supply and the bathroom or tub area.

See also Atopic dermatitis; Dysmenorrhea; Sports injuries.

Resources

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American Academy of Dermatology (AAD). PO Box 4014, Schaumburg, IL 60168–4014. Web site: <www.aad.org>.

American Association of Naturopathic Physicians. 8201 Greensboro Drive, Suite 300, McLean, Virginia 22102. Web site: <http://naturopathic.org>.

Carol A. Turkington

Therapeutic school *see* **Alternative school**

Third and fourth pharyngeal pouch syndrome *see* **DiGeorge syndrome**

Throat culture

Definition

A throat culture is a microbiological procedure for identifying disease-causing bacterial organisms in material taken from the throat. A throat swab will capture the causative organism in most cases and the culture will allow the specific organism to be grown in the microbiology laboratory under certain conditions. The bacteria can then be identified, and results from antibiotic sensitivity tests on the bacteria will determine the appropriate treatment to be prescribed.

Purpose

The primary purpose of a throat culture is to identify the specific bacterial organisms that are causing a **sore throat** or throat infection, particularly to identify or to rule out the presence of group A, beta-hemolytic streptococci, the bacterial organisms that cause **strep throat**. Hemolytic means that these streptococci are capable of destroying red blood cells.

Since most sore throats are caused by viral infections rather than by strep organisms, a correct diagnosis is important to prevent unnecessary use of **antibiotics** for viruses that do not respond to them, and to begin effective treatment of strep or other throat infections as soon as possible. Throat cultures can also be used to identify other disease organisms that are present in the patient's throat and to identify people who are carriers of organisms that cause **meningitis** and **whooping cough**, among other diseases.

Besides the use of throat cultures in diagnosis, the bacteria identified are used to determine antibiotic sensitivity, allowing physicians to select the most appropriate and effective antibiotic to treat a specific infection. It is common for physicians to order culture and sensitivity tests at the same time.

Description

A throat culture will often be performed on an individual who has a severe sore throat or known symptoms of strep throat. These symptoms include a sore throat that may be accompanied by **fever**, body aches, and loss of appetite. The tonsils and the back of the throat may appear red, swollen, and streaked with pus. Symptoms usually appear one to three days after being exposed to the group A streptococcus *S. pyogenes*. Strep throat occurs more often among children than adults, with incidence at peak in fall and winter when school is in session and contact with other children is highest. Because strep

is highly contagious, **family** members and close contacts of individuals diagnosed with strep throat may also be advised to have throat cultures if they show signs of sore throat or other symptoms.

The specimen for throat culture is obtained by wiping the child's throat with a sterile cotton swab. The child is asked to tilt the head back and open the mouth wide. With the tongue depressed and the child saying "ah," the care provider wipes the back of the throat and the tonsils with the sterile swab, applying it to any area that appears either very red or is discharging pus. The swab is removed gently without touching the teeth, gums, or tongue. It is then placed in a sterile tube for immediate delivery to a laboratory. The swabbing procedure may cause gagging but is not painful. Obtaining the specimen takes less than 30 seconds. Laboratory results will be available as soon as bacteria grow in a special plate that has been streaked with the contaminated swab, usually within two to three days. Sometimes the organism cultured is not strep as suspected. The microbiology laboratory may use samples of the bacteria grown to perform other tests that will help identify the disease causing organism.

S. pyogenes is known to grow well in growth media such as rich broths or gels (agars) that are supplemented with blood. When strep is suspected, the throat material is cultured on blood agar that has been prepared as a broth and poured into petri dishes (plates) where it solidifies into a gel. Blood agar is usually made from the cell walls of red algae (also trypticase soy, heart infusion, or Todd-Hewitt agar) and sheep's blood. When the throat swab reaches the laboratory, the microbiologist uses it to make streaks directly across a blood agar plate. The covered plate is allowed to incubate at a specific temperature (35°–37°C) for 24 to 48 hours to foster the growth of bacteria. The bacteria will grow in clusters called colonies. If the organism is a group A hemolytic streptococcus, an area immediately around the bacterial colony will show hemolysis (the breaking up or lysing of red blood cells), leaving a clear zone surrounding the colony. This helps a technician identify a hemolytic strep organism visually. Other types of bacteria may grow in differently sized or shaped colonies, allowing the microbiologist to differentiate the bacteria. A sample of the bacterial colony may also be examined microscopically to evaluate bacterial type or morphology. Samples of the bacteria may be restreaked on another agar plate with small disks of specific antibiotics to see which antibiotics destroy the bacteria (sensitivity testing). The physician may then prescribe the most effective antibiotic.

When strep throat is suspected, it may be screened in a quick test in the doctor's office. These tests allow direct detection of streptococcal antigens in body fluids such as urine or blood serum or from a throat swab. The test uses a strip or disc that is chemically coated with an antibody specific for the strep antigen. If strep is present, a visible reaction occurs with the antibody on the strip when combined with material from the throat. Depending upon the manufacturer's method, results may be available in about ten to 30 minutes. These "instant" tests are not as definitive as cultures but their reliability has improved since they were first introduced. If an instant throat test is negative, however, a throat culture will still be performed to verify the negative results or to identify non-strep organisms.

Precautions

Gargling to clear the throat or treatment with antibiotics will affect culture results and may make identification of the bacteria impossible. The child should not gargle immediately before the culture.

The child's throat should be swabbed and the culture performed before any antibiotics are taken. The laboratory should be informed if the patient has recently taken antibiotics for the current infection or any other infection. After the culture, however, the physician may initiate early treatment by prescribing a broad spectrum antibiotic to be started before results of the culture are available. After the organism has been identified and sensitivity testing has indicated the most effective antibiotic, a different, more specific antibiotic can be prescribed.

The child's immunization history should be checked to evaluate the possibility that diseases other than strep are causing the sore throat. The care provider should wash his or her hands carefully after swabbing the throat and handling the specimen to prevent the spread of any infectious organisms. Hand washing should be done at home also to reduce contact with infective material. Spreading is usually from contact with droplets of material from the nose and throat of affected individuals.

Preparation

There is no special preparation involved before performing a throat culture. The individual does not need to avoid food or fluids before the test.

Aftercare

There are no special care recommendations after throat swab and culture have been performed. There are no unusual effects expected from having the throat swabbed, though the child may have a mild sensation of

something present in the throat for several hours after it has been swabbed.

Risks

Healthcare professionals, parents, or other contacts are at risk of exposure to the child's illness. Strep throat is highly contagious and easily spread through contact with droplets from the nose or throat.

Normal results

Normal results would include finding organisms that grow in healthy throat tissues (normal flora). These organisms include non-hemolytic and alpha-hemolytic streptococci, some *Neisseria* species, staphylococci, **diphtheria** and hemophilus organisms, pneumococci, yeasts, and Gram-negative rods.

Abnormal results

In addition to *S. pyogenes*, other disease agents may be identified in the throat culture. Besides other varieties of strep organisms, these organisms may include *Candida albicans*, which can cause thrush; *Corynebacterium diphtheriae*, which can cause diphtheria; and *Bordetella pertussis*, which can cause whooping **cough**. In addition, the appearance of a specific normal organism in very high numbers may also be regarded as an abnormal result.

Parental concerns

Parents may be concerned that effective treatment will be delayed because of waiting for the throat culture results, which can take up to 48 hours. Physicians may prescribe a broad spectrum antibiotic as initial treatment rather than waiting for culture results. When the culture results are available and sensitivity tests indicate a more effective antibiotic, the physician will likely prescribe a new antibiotic specific for the strep or other organism identified.

See also Strep throat.

Resources

ORGANIZATIONS

American Academy of Pediatrics. 141 Northwest Point Boulevard, Elk Grove Village, IL 60007–1098. Web site: <www.aap.org>.

Centers for Disease Control. 200 Independence Avenue, SW, Washington, DC, 20201. Web site: <www.cdc.gov>.

Centers for Disease Control and Prevention. 1600 Clifton Rd., NE, Atlanta, GA 30333. Web site: <www.cdc.gov>.

KEY TERMS

Agar—A gel made from red algae that is used to culture certain disease agents in the laboratory.

Antibiotics—Drugs that are designed to kill or inhibit the growth of the bacteria that cause infections.

Antigen—A substance (usually a protein) identified as foreign by the body's immune system, triggering the release of antibodies as part of the body's immune response.

Carrier—A person who possesses a gene for an abnormal trait without showing signs of the disorder. The person may pass the abnormal gene on to offspring. Also refers to a person who has a particular disease agent present within his/her body, and can pass this agent on to others, but who displays no symptoms of infection.

Diphtheria—A serious, frequently fatal, bacterial infection that affects the respiratory tract. Vaccinations given in childhood have made diphtheria very rare in the United States.

Hemolytic—Able to break down or dissolve red blood cells.

Morphology—Literally, the study of form. In medicine, morphology refers to size, shape, and structure rather than function.

Streptococcus—Plural, streptococci. Any of several species of spherical bacteria that form pairs or chains. They cause a wide variety of infections including scarlet fever, tonsillitis, and pneumonia.

Thrush—An infection of the mouth, caused by the yeast *Candida albicans* and characterized by a whitish growth and ulcers.

Whooping cough—An infectious disease of the respiratory tract caused by a bacterium, *Bordetella pertussis*. Also known as pertussis.

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Thrombocyte count *see* **Platelet count**

Thrush *see* **Candidiasis**

Thumb sucking

Definition

Thumb sucking is the childhood habit of putting the thumb in the mouth for comfort or to relieve stress.

Description

About half of all children suck their thumbs during infancy, with most starting in the first weeks of life. Ultrasound pictures of intrauterine life have even shown fetuses sucking their thumbs. One way that infants explore their world is by putting objects in their mouths and sucking on them. Thumb sucking appears to be a natural habit of children in all parts of the world. Sucking the thumb is soothing for a small child, and many children continue this habit for comfort and security into the early school years. Thumb sucking is most prevalent in children under two, and most children give up the habit on their own by age four.

Thumb sucking by itself is not a cause or symptom of physical or psychological problems. It is not known why some children suck their thumbs longer than others. More girls than boys suck their thumbs beyond age two. Researchers speculate that boys receive stronger negative messages from parents and peers that thumb sucking is infantile and not acceptable. Thumb sucking offers security to a child, but this behavior does not imply that the child is insecure. Most children have some sort of self-comforting ritual that may involve sucking the thumb, fingers, or a pacifier, pulling or twisting their hair, or stroking or sucking a soft toy or blanket. These are all normal habits of infancy that are eventually outgrown.

Some nineteenth-century physicians feared a variety of consequences from thumb sucking, such as weak moral character, and earlier generations of parents were advised to break this habit forcibly. Parents were sometimes told to place mechanical constraints on their children's hands to keep their thumbs out of their mouths. Children's thumbs were sometimes coated with a bitter substance, taped, or covered with gloves. It was also considered necessary to shame and humiliate the thumb sucker.

Modern doctors find few negative health effects of thumb sucking, even if prolonged, and parents are urged to let their children outgrow the habit on their own. Thumb sucking may be more of a problem for the parent than the child, if the parent is unsettled by the behavior. Weaning a young child from the habit before he or she is ready is usually difficult and may only prolong the thumb sucking.

Infancy

Some children suck their thumbs before they are even born, and others begin sucking their thumbs soon after birth. All or nearly all infants suck on their fingers, thumbs, or a pacifier. This is completely normal and very common.

Toddlerhood

Thumb sucking is most common in children who are younger than two years old. Many children stop sucking their thumbs by age three or four without any intervention.

Preschool

Preschool children may begin to become embarrassed by their thumb sucking if the children with whom they interact do not suck their thumbs and make fun of them. Most children in this age group who still suck their thumbs will stop on their own, and intervening may stress the child and make the problem worse. Even when they have stopped thumb sucking during the day, children may continue it as part of a nighttime falling **sleep** ritual.

School age

Most children have stopped sucking their thumbs before they begin school, or else stop sucking shortly thereafter, usually in response to **peer pressure**. If a school age child seems distressed about his or her thumb sucking, the parent may want to suggest ways in which they can work together to wean the child from the thumb. If the child does not want to give up thumb sucking, the dentist should be consulted to ensure that it is not doing any damage to the alignment of the teeth.

Common problems

There are a few cases where thumb sucking may become a problem. If a school-age child sucks his or her thumb and is teased by classmates, the child may wish to quit and need help either from parents or a counselor. Some dentists warn of misalignment of permanent teeth if a child of five or six sucks the thumb with a lot of

Young boy sucking his thumb. *(© Jennie Woodcock; Reflections Photolibrary/Corbis.)*

pressure on the teeth. Not all dentists agree, however, that thumb sucking is harmful to tooth development. If a child's dentist sees evidence that thumb sucking is causing a particular problem, the child may need to be urged to quit. If the child is having trouble quitting the habit, parents may be able to help with positive reinforcement. The child can be given a sticker or small reward for a day spent without thumb sucking. Parents can also help the child find something else to do with his or her hands when the child has the urge to suck the thumb. Parents should avoid negative pressure on children to stop sucking their thumbs; this habit is eventually outgrown by all children. In extreme cases, some dentists can prescribe an oral device to alter the shape of the roof of the child's mouth, so that it is unpleasant for the child to continue sucking. If a child has recently undergone any sort of trauma such as witnessing **divorce**, a pet's death, or **family** problems, treatment for thumb sucking should not be undertaken right away.

Parental concerns

Parents tend to be more concerned with thumb sucking than is actually warranted. Until the child is five or six, or there starts to be a problem with speech formation or teeth alignment, thumb sucking is not a problem.

When to call the doctor

If the child continues to suck his or her thumb after age five or six, or sucks it frequently or very hard after age three or four, the doctor may have helpful suggestions for the concerned parent. If the child's teeth are becoming misaligned because of thumb sucking the dentist should be consulted. If the thumb sucking is combined with other problems such as **anxiety** a doctor should be consulted.

Resources

BOOKS

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ORGANIZATIONS

International Association of Orofacial Myology. 970 Elizabeth Street, Denver, CO 80209. Web site: <www.iaom.com>.

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Tick bite *see* **Lyme disease**

Tics

Definition

A tic is a nonvoluntary body movement or vocal sound that is made repeatedly, rapidly, and suddenly. It has a stereotyped but nonrhythmic character. The child or adolescent with a tic experiences it as irresistible but can suppress the movement or noise for a period of time. Tics are categorized as motor or vocal, and as simple or complex. The word "tic" itself is French.

Tics are a type of dyskinesia, which is the general medical term given to impairments or distortions of voluntary movements. Although tics vary considerably in severity, they are associated with several neuropsychiatric disorders in children and adolescents. The American Psychiatric Association (APA) defined four tic disorders in the fourth edition of the *Diagnostic and Statistical Manual of Mental Disorders*, or *DSM-IV*. The disorders are distinguished from one another according to three criteria: the child's age at onset; the duration of the disorder; and the number and variety of tics.

- Transient tic disorder (also known as benign tic disorder of childhood): The criteria for transient tic disorder specify that the onset must occur before the age of 18 years; the tics must occur many times a day almost every day for at least four weeks but not longer than 12 months; and the child must not meet the criteria for **Tourette syndrome** or chronic tic disorder.
- Chronic motor or vocal tic disorder: To meet the diagnosis of chronic tic disorder, the child must be younger

than 18 years of age; the tics must have occurred nearly every day or intermittently for a period longer than a year, without a tic-free interval longer than three months; the tics must be either vocal or motor but not both; and the child must not meet the criteria for Tourette disorder.

- Tourette disorder (also known as Tourette syndrome, or TS): Tourette disorder is considered the most serious of the four tic disorders. The *DSM-IV* criteria for Tourette disorder specify that the child must be younger 18 years of age at onset; the tics must include multiple vocal as well as motor tics, although not necessarily at the same time; the tics must occur many times a day, nearly every day or at intervals over a period longer than a year, without symptom-free intervals longer than six months; there must be variations in the number, location, severity, complexity, and frequency of the tics over time; and the tics cannot be attributed to the effects of a substance (such as stimulants) or a disease of the central nervous system.
- Tic disorder not otherwise specified: This category includes all cases that do not meet the full criteria for any of the other tic disorders.

Description

Tics most commonly affect the child's face, neck, voice box, and upper torso but may involve almost any body part. The experience of having a tic is difficult to describe to those who have never been troubled by them. Having tics may be compared to having the sensation of having to **cough** because something is tickling one's throat or nose. The sensation is irresistible and immediate.

Simple tics

Simple tics involve only a few muscles or sounds that are not yet words. Examples of simple motor tics include nose wrinkling, facial grimaces, eye blinking, jerking the neck, shrugging the shoulders, or tensing the muscles of the abdomen. Simple vocal tics include grunting, clucking, sniffing, chirping, or throat-clearing noises. Simple tics rarely last longer than a few hundred milliseconds.

Complex tics

Complex tics involve multiple groups or muscles or complete words or sentences. Examples of complex motor tics include such gestures as jumping, squatting, making motions with the hands, twirling around when walking, touching or smelling an object repeatedly, and holding the body in an unusual position. Complex motor tics last longer than simple motor tics, usually several seconds or longer. Two specific types of complex motor tics that often cause parents concern are *copropraxia*, in which the tic involves a vulgar or obscene gesture, and *echopraxia*, in which the tic is a spontaneous imitation of someone else's movements.

Similarly, complex vocal tics involve full speech and language, which may range from the spontaneous utterance of individual words or phrases, such as "Stop," or "Oh boy," to speech blocking or meaningless changes in the pitch, volume, or rhythm of the child's voice. Specific types of complex vocal tics include *palilalia*, which refers to the child's repetition of his or her own words; *coprolalia*, which refers to the use of obscene words or abusive terms for certain racial or religious groups; and *echolalia*, in which the child repeats someone else's last word or phrase.

Sensory tics

Sensory tics are less common than either motor or vocal tics. The term refers to repeated unwanted or uncomfortable sensations, usually in the child's throat, eyes, or shoulders. The child may feel a sensation of tickling, warmth, cold, or pressure in the affected area.

Phantom tics

Phantom tics are the least common type of tic. A phantom tic is an out-of-body variation of a sensory tic in which the person feels a sensation in other people or objects. People with phantom tics experience temporary relief from the tic by touching or scratching the object involved.

Other features of tics

Tics typically occur in bouts or episodes alternating with periods of tic-free behavior lasting from several seconds to several hours. They generally diminish in severity when the child is involved in an absorbing activity such as reading or doing homework, and increase in frequency and severity when the child is tired, ill, or stressed. Some children have tics during the lighter stages of **sleep** or wake up during the night with a tic.

Severe complex motor tics carry the risk of physical injury, as the child may damage muscles or joints, fracture bones, or fall down during an episode of these tics. Some children harm themselves deliberately by self-cutting or self-hitting, while others hurt themselves unintentionally by touching or handling lighted matches, razor blades, or other dangerous objects. Severe complex vocal tics may interfere with breathing or swallowing.

Transmission

Tics as such are symptoms and are not transmitted directly from one person to another. Tic *disorders*, however, are known to run in families. In addition, some doctors think that tic disorders are more likely to develop in children who have had certain types of infections. These theories are discussed more fully below.

Demographics

Prevalence of tic disorders

The statistics given for tics and tic disorders vary from source to source, in part because tics vary considerably in severity, and many children with mild tics may never come to a doctor's attention. Estimates for the general North American population range from 3 to 20 percent for transient tics (particularly among children below the age of ten); 2–5 percent for chronic tic disorders; and 0.1–0.8 percent for Tourette syndrome. A Swedish study done in 2003 reported that 6.6 percent of a sample of Uppsala school children between the ages of 7 and 15 met *DSM-IV* criteria for tic disorders: 4.8 percent for transient tic disorder, 0.8 percent for chronic motor tic disorder, 0.5 percent for chronic vocal tic disorder, and 0.6 percent for Tourette syndrome. One study of American volunteers for military service reported a prevalence of 0.5 cases of TS per 1000 for males and 0.3 cases per 1000 for females. Tourette syndrome is known to be more common in males than in females, although the gender ratio is variously reported as 3: 1, 5: 1, or even 10: 1.

Little is known as of 2004 about the prevalence of tic disorders across racial or ethnic groups. One small study that was done in western North Carolina reported that Caucasian children were slightly more likely to have tic disorders than either African American or Native American children (2.1 percent to 1.5 percent and 1.5 percent respectively). The authors of the study cautioned, however, against applying their findings to larger groups of children in other parts of the United States.

Tic disorders and comorbid disorders

One important characteristic of tics and tic disorders is that they rarely occur by themselves. Tic disorders—particularly TS—have a high rate of comorbidity with other childhood disorders. The term *comorbid* is used to refer to a disease or disorder that occurs at the same time as another disorder. The frequencies of the most common disorders that may be comorbid with tic disorders and Tourette syndrome are as follows:

- attention-deficit/hyperactivity disorder (ADHD): 50 percent comorbidity with tic disorders, 90 percent comorbidity with TS
- **obsessive-compulsive disorder** (OCD): 11 percent and 80 percent respectively
- major depression: 40 percent and 44 percent respectively

Other psychiatric problems that often coexist with tics and tic disorders include **learning disorders**, **impulse control disorders**, school phobia, sensory hypersensitivity, and rage attacks.

Causes and symptoms

The causes of tics and tic disorders are not fully understood as of the early 2000s, but most researchers believe that they are multifactorial, or the end result of several causes. In the early twentieth century, many doctors influenced by Freud thought that tics were caused by hysteria or other emotional problems, and treated them with psychoanalysis. Psychoanalytic treatment, however, had a very low rate of success.

Since the 1970s, researchers have been looking at genetic factors in tic disorders and Tourette syndrome. With regard to TS, genetic factors are present in about 75 percent of children diagnosed with TS, with 25 percent having inherited genetic factors from both parents. The exact pattern of genetic transmission was not known as of 2004, however; autosomal dominant, autosomal recessive, and sex-linked inheritance patterns have all been studied and rejected. Some candidate genes for TS have also been tested and excluded. What is known is that the patient's environment and heredity play a significant part in the severity and course of TS.

Tic disorders as well as OCD sometimes develop after infections (usually **scarlet fever** or **strep throat**) caused by a group of bacteria known as group A beta-hemolytic streptococci, sometimes abbreviated as GABHS. These disorders are sometimes grouped together as PANDAS disorders, which stands for Pediatric Autoimmune Neuropsychiatric Disorders Associated with Streptococci. Some researchers think that the tics develop when antibodies in the child's blood produced in response to the bacteria cross-react with proteins in the brain tissue. The connection between **streptococcal infections** and tic disorders is questioned by some researchers, however, on the grounds that most children have a GABHS infection at some point in their early years, but the vast majority (95 percent) do not develop OCD or a tic disorder. There appears to be a closer connection between Sydenham's chorea, which is a movement disorder, and GABHS infections than between tic

disorders and these infections. One prospective study done at Yale reported in 2004 that new GABHS infections do *not* appear to cause a worsening of tics in children diagnosed with OCD or Tourette syndrome.

Neuroimaging studies have shown that tic disorders are related to abnormal levels of neurotransmitters known as dopamine, serotonin, and cyclic AMP in certain parts of the brain. A neurotransmitter is a chemical produced by the body that conveys nerve impulses across the gaps (synapses) between nerve cells. In addition to abnormalities in the production or absorption of these chemical messengers, imaging studies indicate that the blood flow and metabolism in a part of the brain called the basal ganglia are abnormally low. The basal ganglia are groups of nerve cells deep in the brain that control movement as well as emotion and certain aspects of thinking. In contrast to the low level of blood flow in the basal ganglia, the motor areas in the frontotemporal cortex of the brain show increased levels of activity.

The various types of tics themselves have already been described. Other symptoms that may be associated with tics and tic disorders include obsessive thoughts; difficulty concentrating or paying attention in school; forgetfulness; slowness in completing tasks; losing the thread of a conversation. These symptoms are usually regarded as side effects of interrupted thinking or behavior caused by the tics.

When to call the doctor

Most cases of mild tics do not require medical treatment and will clear up on their own over time. Doctors usually recommend that **family** members try to ignore simple tics, since teasing or other unwanted attention may make the tics worse. A visit to the doctor is recommended, however, under any of the following circumstances:

- The child is falling behind in school because of the tics.
- The child's relationships with peers and adults outside the family are affected by the tics.
- The child cannot carry out activities of daily living (self-feeding, bathing, getting dressed, etc.).
- The child has fallen, injured himself, or developed other physical problems because of the tics.
- Other family members have or have had tic disorders.
- The child has recently had an episode of strep throat or other streptococcal infection.
- The child has been diagnosed with OCD, ADHD, or depression.
- The tics have come on suddenly.

Diagnosis

Tic disorders are diagnosed by a process of excluding other possibilities; there are no definitive tests for these disorders as of the early 2000s. For this reason, the diagnosis of tic disorders is often delayed or sometimes missed altogether in milder cases. One study reported an average delay of five to 12 years between the initial symptoms and the correct diagnosis. In addition, diagnosis is complicated by the fact that children often learn to mask their tics by converting them to more socially acceptable or apparently voluntary movements or sounds.

History and physical examination

The first part of a medical workup for tics is the taking of a medical history and a general physical examination. The doctor will want to know whether there is a family history of tics or tic disorders, whether the child has been diagnosed with other childhood developmental or psychiatric disorders, and whether he or she has recently had strep throat or a similar infection.

The physical examination helps the doctor rule out such other possible diagnoses as Sydenham's chorea, a self-limited movement disorder that most commonly affects children between five and 15 years of age; other **movement disorders**; seizure disorders; **encephalitis**; neurosyphilis; Wilson's disease (a rare inherited disease that causes the body to retain copper); **schizophrenia**; **carbon monoxide poisoning**; cocaine intoxication; brain injuries caused by trauma; **cerebral palsy**; or the side effects of certain medications, particularly stimulants and antiepileptic drugs.

The doctor may not be able to observe the tic(s) during the child's first office visit, often because the child has learned to suppress or mask them. In some cases, a follow-up visit may be scheduled, or the doctor may refer the child to a child psychiatrist or neurologist for further observation. Another approach that can be used to confirm the diagnosis is to audiotape or videotape the child at home or in another less stressful setting.

Psychiatric inventories

Most child psychiatrists will administer the Yale Global Tic Severity Scale (YGTSS) during the intake interview and at follow-up visits in order to identify the particular tic disorder affecting the child, identify comorbid disorders if present, evaluate the severity of the tics, and monitor the child's response to treatment.

The YGTSS, which was first published in 1989, is a semi-structured interview that is widely used by researchers who study tic disorders. "Semi-structured"

means that it is an open-ended set of questions that allow the child's parents to describe the tics and other symptoms in detail rather than just answer brief yes-or-no questions.

Laboratory tests

As mentioned earlier, there are no laboratory tests to diagnose tics as such. In some cases, however, the doctor may order a blood test to rule out Wilson's disease or other metabolic disorders, or order a **throat culture** if the child has recently had strep throat. If the doctor suspects that the child has a PANDAS disorder, he or she may order a blood test to measure the level of antibodies against group A streptococci.

Imaging studies

As of 2004, imaging studies were not routinely performed on children or adolescents with tics unless the doctor suspects a brain injury, infection, or structural abnormality. **Magnetic resonance imaging** (MRIs), PET scans, and single-photon emission **computed tomography** (SPECT) scans have been used by researchers, however, to study the brains of patients diagnosed with Tourette syndrome.

In the summer of 2004, two engineers in Taiwan reported on the development of a computerized diagnostic system that will allow radiologists to use SPECT imaging to distinguish between chronic tic disorder and Tourette syndrome with a much higher degree of accuracy. The system appears to be potentially useful in speeding up the process of diagnosis and allowing earlier treatment of TS.

Treatment

After psychoanalysis was discredited in the 1970s as a treatment for tic disorders, some doctors urged using such antipsychotic drugs as haloperidol (Haldol) to treat TS by suppressing the tics. These drugs, which are sometimes called neuroleptics, have severe side effects and are likely to interact with other medications that the child may be taking. In addition, tics are increasingly recognized as complex phenomena that have an emotional as well as a physical dimension. As a result, the treatment of tic disorders has changed in the early 2000s in the direction of minimizing the use of medications in favor of a multidisciplinary approach.

The approach to assess a child with a tic disorder is as follows:

- Administer the YGTSS in order to evaluate the areas of the child's functioning that are most severely affected by the tics.
- Identify any comorbid disorders if present. In many cases, the tics do not interfere with the child's life as much as ADHD, OCD, or depression. ADHD should be the primary target of management in children diagnosed with a tic disorder and comorbid ADHD.
- Rank the symptoms in order of importance in order to focus treatment on the ones that are most significant to the child and the family.
- Emphasize controlling the tics and learning to live with them rather than trying to eliminate them with drugs.
- Use behavioral and psychotherapeutic approaches as well as medications.
- Involve the patient's teachers and other significant adults as well as parents in order to help monitor the child's symptoms and response to treatment.

Medications

There is no medication that can cure a tic disorder; all drugs that are used to treat these disorders as of the early 2000s are used only to manage tics. In general, doctors prefer to avoid medications in treating mild tics; start the treatment of moderate or severe tics with medications that have relatively few side effects, and prescribe stronger drugs only when necessary.

Children whose throat cultures or blood tests are positive for a GABHS infection are treated aggressively with **antibiotics**, most commonly penicillin V.

Psychotherapy

Psychotherapy for tics and tic disorders typically involves education about tic disorders and therapy for the family as well as individual treatment for the child. The American Academy of Child and Adolescent Psychiatry (AACAP) urges parents to avoid blaming or punishing the child for the tics, as shaming or harsh treatment increases the child's level of emotional stress and usually makes the tics worse.

Cognitive-behavioral approaches are the most common type of individual psychotherapy used to treat tics and tic disorders. Specific behavioral approaches include the following:

- Massed negative practice: In this form of behavioral treatment, the child is asked to perform the tic intentionally for specified periods of time interspersed with rest periods.

- Competing response training: This is a form of treatment of motor tics in which the child is taught to make the opposite movement to the tic.
- Self-monitoring: In awareness training, the child keeps a diary, small notebook, or wrist counter for recording tics. It is supposed to reduce the frequency of tic bouts by increasing the child's awareness of them.
- Contingency management: This approach works best in the home and is usually carried out by the parents. The child is praised or rewarded for not performing the tics and for replacing them with acceptable alternative behaviors.

As of the early 2000s, however, no controlled studies have been done comparing the effectiveness of these various behavioral approaches. At best, they appear to produce mixed results.

Surgery

Surgery is used very rarely to treat tic disorders; it is usually tried only if the tic has not responded to any medication and interferes significantly with the patient's life. Some patients with TS, however, have been successfully treated with stereotactic surgery involving high-frequency stimulation of the thalamus. Stereotactic surgery involves an approach that calculates angles and distances from the outside of the patient's skull to locate very small lesions or structures deep inside the brain. It allows the surgeon to remove tissue or treat injured areas through much smaller incisions.

Alternative treatments

The place of alternative or complementary therapies in treating tics is debated. One group of Chinese physicians reported successfully treating patients diagnosed with TS with acupuncture. However, a group of researchers studying traditional medicine in Bali found it ineffective in treating tic disorders, and a second group at Johns Hopkins reported that relaxation therapy did not have a statistically significant effect in treating children diagnosed with TS. There is also some evidence that gingko, ginseng, and some other herbs taken for their stimulant effects may increase the severity of tics in children and adolescents.

Nutritional concerns

Although some nutritionists have suggested a possible connection between sugar or food coloring and tic severity, no studies published as of 2004 had demonstrated such a connection. One study done at the University of Kansas did find a connection between **caffeine** (which is found in cola beverages and some other soft drinks as well as tea and coffee) consumption and tic severity in children. The study sample, however, was quite small.

Prognosis

The prognosis for most tics and tic disorders is quite good. In the majority of cases, the tics diminish in severity and eventually disappear as the child grows older. Even in Tourette syndrome, about 85 percent of children find that their tics diminish or go away entirely during or after **adolescence**. Tics that persist beyond the teenage years, however, usually become permanent.

Factors associated with a poorer prognosis for all tic disorders include the following:

- history of complications during the child's birth
- chronic physical illness in childhood
- physical or emotional abuse in the family or a history of family instability
- exposure to **anabolic steroids** or cocaine
- comorbid psychiatric or developmental disorders

Prevention

There are no known ways to prevent either tics or tic disorders.

Nutritional concerns

In some cases, parents may find it helpful to monitor the child's intake of cola, iced tea, other drinks containing caffeine, and certain herbal teas.

Parental concerns

Parental concerns related to tics and tic disorders are difficult to address in general terms, because tics can range in type and severity from simple noises or movements of short duration that do not attract much attention from others to complex tics of a physically harmful or socially embarrassing nature that attract a lot of attention. In addition, tics must often be managed in the context of another disorder affecting the child. Since the treatment of tics is individualized, it is best for parents to consult with the child's doctor(s) regarding special educational programs or settings, explaining the tics or tic disorder to others, dealing with the side effects of medications, and managing rage attacks or other symptoms that may be associated with the tics.

See also Movement disorders; Tourette syndrome.

KEY TERMS

Basal ganglia—Brain structure at the base of the cerebral hemispheres involved in controlling movement.

Chorea—Involuntary movements in which the arms or legs may jerk or flail uncontrollably.

Comorbidity—A disease or condition that coexists with the disease or condition for which the patient is being primarily treated.

Compulsion—A repetitive or ritualistic behavior that a person performs to reduce anxiety. Compulsions often develop as a way of controlling or "undoing" obsessive thoughts.

Coprolalia—The involuntary use of obscene language.

Copropraxia—The involuntary display of unacceptable/obscene gestures.

Dopamine—A neurotransmitter made in the brain that is involved in many brain activities, including movement and emotion.

Dyskinesia—Impaired ability to make voluntary movements.

Echolalia—Involuntary echoing of the last word, phrase, or sentence spoken by someone else.

Echopraxia—The imitation of the movement of another individual.

Multifactorial—Describes a disease that is the product of the interaction of multiple genetic and environmental factors.

Neuroleptic—Another name for the older type of antipsychotic medications, such as haloperidol and chlorpromazine, prescribed to treat psychotic conditions.

Neurotransmitter—A chemical messenger that transmits an impulse from one nerve cell to the next.

Palilalia—A complex vocal tic in which the child repeats his or her own words, songs, or other utterances.

PANDAS disorders—A group of childhood disorders associated with such streptococcal infections as scarlet fever and strep throat. The acronym stands for Pediatric Autoimmune Neuropsychiatric Disorders Associated with Streptococci.

Semi-structured interview—A psychiatric instrument characterized by open-ended questions for discussion rather than brief questions requiring yes or no answers.

Stereotactic technique—A technique used by neurosurgeons to pinpoint locations within the brain. It employs computer imaging to guide the surgeon to the exact location for the surgical procedure.

Stereotyped—Having a persistent, repetitive, and senseless quality. Tics are stereotyped movements or sounds.

Streptococcus—Plural, streptococci. Any of several species of spherical bacteria that form pairs or chains. They cause a wide variety of infections including scarlet fever, tonsillitis, and pneumonia.

Tic—A brief and intermittent involuntary movement or sound.

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American Academy of Child and Adolescent Psychiatry. 3615 Wisconsin Avenue, NW, Washington, DC 20016–3007. Web site: <www.aacap.org.>.

National Institute of Neurological Disorders and Stroke (NINDS). National Institutes of Health. 9000 Rockville Pike, Bethesda, MD 20892. Web site: <www.ninds.nih.gov>.

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Rebecca Frey, PhD

Time-out procedure

Definition

Time out is a technique in which a child is removed from activity and forced to sit alone for a few minutes in order to calm down.

Description

The time out has become an increasingly popular method of dealing with children's inappropriate behavior. If a child becomes too aggressive or angry, the parent or caregiver may remove the child from the upsetting situation. Parents may have a special place in the home for time outs: in the child's room, in a certain chair, or on a rug in an out-of-the-way place. The child may be allowed to end the time out when he or she is ready or told to stay in the time-out place for a specific length of time. The time should be very short—one guide suggests a minute for each year of the child's age—as most young children cannot easily comprehend longer time spans.

The time out is not used as a punishment so much as an opportunity for the child to try to regain control of emotions. Some children can accomplish this by themselves, and being removed from a stressful **play** situation is all that they need. Other children may not be able to recover their equilibrium without help from an adult. The parent or caregiver may ask the child to try to calm down alone in the time-out spot and then give attention only after the child has made some effort.

It should be clear to the child that the time out is not punitive, and a child should not feel humiliated for having a time out. The time-out area should not be a constraining or frightening place, such as a locked closet. The time out should serve to teach the child to manage strong feelings safely, and after he or she has done so, the child should be praised for calming down.

There may be other techniques parents or caregivers can use before a time out becomes necessary. If an activity is too stressful to one or more children, it may be better to end the activity. Changing the situation may restore tempers more readily than a spell of reflection. If children are fighting because they are hungry or tired, then that need should be addressed. Children may benefit most from a time out if the issues of aggression or out-of-control behavior have been discussed at a time when the child was not upset. Although the goal of time out may be to teach the child to take responsibility for controlling his or her own behavior, depending on the age and **temperament** of the child, this may not be possible without support and comfort from parents or other concerned adults.

Toddlerhood

Children under three may not be mature enough to comprehend a time out, although for some it may be an effective tool.

Preschool

Time outs are usually most effective with preschool-age children. If time outs are used in a **preschool** or day-care situation, parents may want to discuss this with the teachers or caregivers so that time outs can be used consistently in situations at home too. Time outs for this age group should be very brief.

School age

School age children may be more resistant to the concept of time out. It is usually possible to give these children a certain sense of autonomy by having them help choose the time out location (when they are not angry) and allowing them to take themselves there. If they do not comply, this age group often responds well to being grounded until they choose to complete the time out.

Common problems

Many times children will vocalize their distress while they are in time out. Not insisting that children maintain silence for the completion of time out can be helpful, because it allows children to vent their feelings and makes time out easier to complete successfully. Children who leave time out before time is up can be gently held in place or put in a room while the parent holds the door shut. The room should not contain anything valuable or fragile and should not contain bookshelves or other things that the child may be able to pull and injure him or her self with.

Parental concerns

Children are often very vocally adverse to time outs, but time outs have been found to be effective ways of changing behavior in many children. Helping the child understand what behavior caused the time out and what kind of behavior is considered acceptable will help the child change the behavior in the future. Children cannot change their behavior if they do not know what is expected of them. Children's angry behavior, especially during the beginning of a time out, can be embarrassing for the parent, so it may be helpful if the situation arises in a public place to have the time out in a bathroom or in the car while always keeping the child in sight.

When to call the doctor

The doctor should be consulted if the child is very violent to him or herself or others during time out, or if behavior does not improve after time outs have been regularly enforced over a few weeks.

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Tish Davidson, A.M.
A. Woodward

Toddler elbow *see* **Nursemaid's elbow**

TOF *see* **Tetralogy of Fallot**

Toilet training

Definition

Toilet training is the process of teaching a young child to control the bowel and bladder and use the bathroom for elimination. A child is considered to be toilet trained when he or she initiates going to the bathroom and can adjust clothing necessary to urinate or have a bowel movement. Toilet training is sometimes called toilet learning or potty training.

Description

The average age at which children complete toilet training in the United States is approximately three years old. In some cases children learn bladder control first; others learn bowel control before bladder control. Control is generally first achieved during the daytime, well before a child is able to stay dry at night.

Some children achieve some control over bladder and/or bowel movements as early as nine months of age and are able to cooperate in controlling themselves to some degree by the age of 12 to 15 months. Most experts agree, however, that toilet training should only be initiated when a child exhibits certain signs of readiness that usually appear between the ages of two and three years of age. Unlike infants, toddlers know when they are urinating or defecating and may assume certain postures or become quiet when they are about to move their

bowels. They have also learned the vocabulary their **family** uses for elimination. Another sign is a sense of fastidiousness and desire for order that appears at this stage of development. Children are likely to ask parents to change their dirty diapers right away, and they show a general interest in orderliness that can be harnessed for purposes of toilet training. A child this age also has a pronounced desire to imitate the parent of the same sex, a trait that can be used to advantage in enticing her to use the toilet. Lastly, the child will begin to show signs of being able to delay urination or bowel movements such as waking from **sleep** still dry or refraining from urinating or defecating for longer periods of time while not wearing a diaper.

Strategies

Child care experts generally recommend a strategy that uses praise as a motivator, has little pressure from the parents, and is fun for the child. It has been found that when parents wait until their toddler has attained the greatest possible degree of readiness, the process is easier, faster, and accompanied by fewer lapses. The emphasis is on letting the child proceed at his own pace, motivated by the desire to be a "big boy" or "big girl" and imitate his parents. Measures that may cause pressure and **anxiety** need to be avoided.

The first step in toilet training is to purchase a potty. There are different versions of potties, including ones that sit on the floor and are emptied after each use, ones that have cups to protect against splatters, and ones that sit on top of an adult toilet with or without a step stool for the child to climb up to it. The floor-level model is most often recommended for the first stages of toilet training. Some recommend taking the child to the store to help pick out his or her own potty, then helping to personalize it with a name, stickers, paint, etc., with the general idea of making the potty a prized possession of the child's, not something to be feared.

The child should first spend some time sitting on the potty, first while clothed and then with clothes removed, so that he or she is comfortable sitting on it. The connection between what she is doing on her small potty and what the adults and siblings do on the big potty should be emphasized. One suggestion is to bring the child to the potty with a dirty diaper and the contents placed in it so he or she can see that this is where they belong. Parents should watch for cues from the child that he or she may be about to urinate or have a bowel movement, such as a concentrated look, yanking at his or her diaper, squatting, or grunting. Often this behavior will happen first thing in the morning, right after a nap, or approximately 20 minutes after a meal. The child should be taken to the potty, his or her diaper should be removed, and the child encouraged to sit for at least one minute. Some children may enjoy reading a book or singing a song while waiting. Special read-aloud books about toilet training are popular. Parents should never strap a child into a potty or force him or her to sit on it. If the child has not used the potty after five minutes or so, he or he should be encouraged to get dressed and try again soon.

The general consensus from experts is that much encouragement and praise should be used when a child cooperates with toilet training and when he or she begins to urinate or defecate in the potty. Rewards such as hugs and kisses, verbal praise, stickers, stars, or favorite treats can be used when the child uses the potty or tells a parent he or she has to use it. Pull-up diapers or plastic training pants can be purchased so that the child can remove them him or herself. For many children, simply progressing from diapers to training pants and then to regular underpants is an incentive in itself. When accidents occur, they should be treated casually; punishment, teasing, or chastising should be avoided.

Nighttime training usually begins when a child can stay dry all day, for at least four to six hours. Girls usually reach this point before boys; some girls begin to stay dry at naptime and even occasionally at night before the age of two. After the age of two, dry nights become more frequent: 45 percent of girls and 35 percent of boys stay dry at night at the ages of two to three. With many children, nighttime training is not done until the age of three and, in many cases, not complete until four or five. The signal from the child's bladder has to be strong enough to wake him from sleep and get him to the bathroom at least once or twice a night. As many as 25 percent of children have relapses after they have been dry at night for six months or longer, usually due to a temporary stressor. In a minority of children, nighttime bladder control does not develop until after the age of five; this situation often occurs in families where there is a history of enuresis (bedwetting).

Common problems

In some cases a child may resist all toilet training efforts from the parents, some going so far as to resist sitting on the potty or even holding back bowel movements. Toilet training resistance may be the result of a parent over-admonishing the child when accidents are made or the child does not use the potty when directed. In some cases the child is simply not ready for toilet learning. More rarely, resistance can be caused by a condition that causes the child **pain** when he or she uses the potty, such as painful urination associated with a urinary tract infection. If a child is uncooperative during the toilet training

process, parents can try letting the child initiate the process when he or she is ready, using rewards and positive feedback each time the child is successful in using the potty or goes a whole day without soiling his or her pants, replacing the child's diaper or training pants with regular underwear or having the child change his or her own clothes when accidents occur.

One potential negative effect of resistance is that the child can hold back bowel movements, resulting in **constipation**. This in turn makes elimination uncomfortable and even painful, creating even greater reluctance and resistance on the part of the child. Severe cases of constipation can cause painful anal fissures, fecal soiling (**encopresis**), or rectal enlargement. Unusual delays in toilet training normal children or regressions to soiling generally indicate family stress and/or underlying emotional problems and may require counseling to be effectively resolved.

Parental concerns

Toilet training is often a dreaded and frustrating task for parents. The process can go more smoothly for parent and child if parents are educated on training techniques that emphasize waiting until a child shows signs of readiness before initiating training and taking a child-oriented approach.

When to call the doctor

Parents should contact a healthcare provider if their child exhibits any of the following behaviors:

- holding back bowel movements or constipation
- evidence of painful urination or defecation
- extended toilet training resistance (i.e. lasts several months)

Resources

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Stephanie Dionne Sherk

Tonsillitis

Definition

Tonsillitis is an infection and swelling of the tonsils, which are oval-shaped masses of lymph gland tissue located on both sides of the back of the throat.

Description

The tonsils normally help to prevent infections. They act like filters to trap bacteria and viruses entering the body through the mouth and sinuses. The tonsils also stimulate the immune system to produce antibodies that help fight infections. Anyone of any age can have tonsillitis; however, it is most common in children between the ages of five and 15 years.

Transmission

Tonsillitis is transmitted from one person to another in the same way that many common diseases are, such as by coughing and sneezing. It can also spread when a child touches his or her nose and then other children's **toys** or by children eating or drinking with the same utensils. Children with bacterial tonsillitis are usually no

longer contagious 24 hours after beginning a course of **antibiotics**.

Demographics

Tonsillitis is very common among children. Nearly all children will have some form of tonsillitis at least once.

Causes and symptoms

Tonsillitis is caused by viruses or bacteria that make the tonsils swell and become inflamed. Most cases of tonsillitis are caused by viruses, which cannot be treated with antibiotics. A mild or severe **sore throat** is one of the first symptoms of tonsillitis. Symptoms can also include **fever**, chills, tiredness, muscle aches, earache, **pain** or discomfort when swallowing, and swollen glands in the neck. Very young children may be fussy and stop eating. When a doctor or nurse looks into the mouth with a flashlight, the tonsils may appear swollen and red. Sometimes, the tonsils will have white or yellow spots or flecks. Symptoms usually last four to six days.

When to call the doctor

If the child is displaying the symptoms of tonsillitis and has had a sore throat for more than 48 hours, especially when accompanied by a fever, a doctor should be called. The doctor can determine if the child has tonsillitis, if it is bacterial or viral, and treat the problem accordingly. If the child cannot breathe or cannot swallow emergency medical attention should be sought.

Diagnosis

The diagnosis of tonsillitis is made from the visible symptoms and a physical examination of the patient. The doctor examines the eyes, ears, nose, and throat, looking at the tonsils for signs of swelling, redness, or discharge. A careful examination of the throat is necessary to rule out **diphtheria** and other conditions that may cause a sore throat. Since most sore throats in children are caused by viruses rather than bacteria, the doctor may take a **throat culture** in order to test for the presence of streptococcal bacteria. A throat culture is performed by wiping a cotton swab across the tonsils and back of the throat and sending the swab to a laboratory for culturing. Streptococcus pyogenes, the bacterium that causes "strep" throat, is the most common bacterial agent responsible for tonsillitis. Depending on what type of test is used for strep, the doctor may be able to determine within a few minutes if *S. pyogenes* is present. The quick tests for strep are not as reliable as a laboratory culture, which can take 24 to 48 hours. If the results of a quick test are positive, however, the doctor can prescribe antibiotics right away. If the quick test results are negative, the doctor can do a throat culture to verify the results and wait for the laboratory report before prescribing antibiotics. A blood test may also be done to rule out a more serious infection or condition and to check the white blood cell count to see if the body is responding to the infection. In some cases, the doctor may order blood tests for mononucleosis, since about one third of patients with mononucleosis develop **streptococcal infections** of the tonsils.

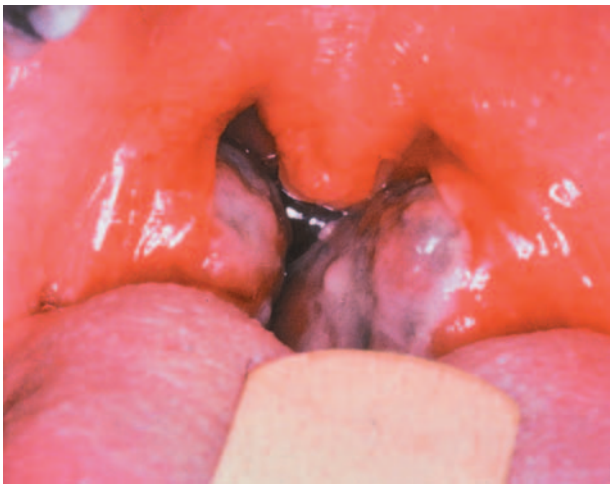
Treatment

Treatment of tonsillitis usually involves keeping the patient comfortable while the illness runs its course. This supportive care includes bed rest, drinking extra fluids, gargling with warm salt water, and taking pain relievers. Children under the age of 12 should not be given aspirin as a pain reliever because of the threat of **Reye's syndrome**. Frozen juice bars and cold fruit drinks can bring some temporary relief of sore throat pain. Drinking warm tea or broth can also be soothing. If the throat culture shows that *S. pyogenes* is present, penicillin or other antibiotics will be prescribed. An injection of benzathine or procaine penicillin may be most effective in treating the infection, but it is also painful. If an oral antibiotic is prescribed, it must be taken for the full course of treatment, usually 10 to 14 days, even if the symptoms are no longer present. If the child has several episodes of severe tonsillitis, the doctor may recommend a tonsillectomy, which is the surgical removal of the tonsils.

Alternative treatment

Strengthening the immune system is important whether tonsillitis is caused by bacteria or viruses. Naturopaths often recommend dietary supplements of vitamin C, bioflavonoids, and beta-carotenes, found naturally in fruits and vegetables, to ease inflammation and fight infection. A variety of herbal remedies also may be helpful in treating tonsillitis. Calendula (*Calendula officinalis*) and cleavers (*Galium aparine*) target the lymphatic system, while echinacea (*Echinacea spp.*) and astragalus (*Astragalus membranaceus*) stimulate the immune system. Goldenseal (*Hydrastis canadensis*), myrrh (*Commiphora molmol*), and bitter orange act as antibacterials. Lomatium dissectum and ligusticum porteri have an antiviral action. Some of the homeopathic medicines that may be used to treat symptoms of tonsillitis include:

- arsenicum
- belladonna
- hepar sulphuris



An examination of this patient's mouth reveals acute tonsillitis. *(© 1993 NMSB. Custom Medical Stock Photo, Inc.)*

KEY TERMS

Streptococcus pyogenes—A common bacterium that causes strep throat and can also cause tonsillitis.

Tonsillectomy—A surgical procedure to remove the tonsils. A tonsillectomy is performed if the patient has recurrent sore throats or throat infections, or if the tonsils have become so swollen that the patient has trouble breathing or swallowing.

Tonsils—Common name for the palatine tonsils, which are lymph masses in the back of the mouth, on either side of the tongue. Tonsils act like filters to trap bacteria and viruses.

- lachesis
- lycopodium
- mercurius
- phytolacca
- rhus toxicodendron

As with any condition, the treatment and dosage should be appropriate for the particular symptoms and age of the patient.

Prognosis

Tonsillitis usually resolves within a few days with rest and supportive care. Treating the symptoms of sore throat and fever will make the child more comfortable. If fever persists for more than 48 hours, however, or is higher than 102°F (38.9°C) the child should be seen by a doctor. If antibiotics are prescribed to treat an infection, they should be taken as directed for the complete course of treatment, even if the child starts to feel better in a few days. Prolonged symptoms may indicate that the child has other upper respiratory infections, most commonly in the ears or sinuses. An abscess behind the tonsil (a peritonsillar abscess) may also occur. In rare cases, a persistent sore throat may point to more serious conditions, such as **rheumatic fever** or **pneumonia**.

Prevention

The bacteria and viruses that cause tonsillitis are easily spread from person to person. It is not unusual for an entire **family** or several students in the same classroom to come down with similar symptoms, especially if *S. pyogenes* is the cause. The risk of transmission can be lowered by avoiding exposure to anyone who already has tonsillitis or a sore throat. Drinking glasses and eating utensils should not be shared and should be washed in hot, soapy water before reuse. Old toothbrushes should be replaced to prevent reinfection. People who are caring for someone with tonsillitis should wash their hands frequently to prevent spreading the infection to others.

Parental concerns

Tonsillitis usually has no long term effects if it is detected and treated promptly. If it is not treated it can lead to other medical conditions such as rheumatic fever, kidney inflammation, or abscesses that could block a child's breathing passage.

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Tish Davidson, A.M.

Tooth decay

Definition

Tooth decay, which is also called dental cavities or dental caries, is the destruction of the outer surface (enamel) of a tooth.

Description

Tooth decay results from the action of bacteria that live in plaque. Plaque is a sticky, whitish film formed by a protein in saliva (mucin) and sugary substances in the mouth. The plaque bacteria sticking to tooth enamel use the sugar and starch from food particles in the mouth to produce acid, which destroys the tooth's enamel.

Baby bottle tooth decay

Baby bottle tooth decay is a dental problem that develops in infants, especially infants that are put to bed with a bottle containing a sweet liquid. Baby bottle tooth decay is also called nursing-bottle caries and bottle-mouth syndrome. Bottles containing liquids such as milk, formula, fruit juices, sweetened drink mixes, and sugar water continuously bathe an infant's mouth with sugar. The bacteria in the mouth use this sugar to produce acid that destroys the child's teeth. The upper front teeth are typically the ones most severely damaged; the lower front teeth are protected to some degree by the tongue. Pacifiers dipped in sugar, honey, corn syrup, or other sweetened liquids also contribute to baby bottle tooth decay. The first signs of damage are chalky white spots or lines across the teeth. As decay progresses, the damage to the child's teeth becomes more obvious.

Demographics

Tooth decay is a common health problem, second in prevalence only to the **common cold**. It has been estimated that 90 percent of people in the United States have at least one cavity and that 75 percent of people had their first cavity by the age of five. Although anyone can have a problem with tooth decay, children are at particularly high risk. The good news is the number of children with cavities in the United States went down in the last few decades of the twentieth century. Some estimates are that as of the early 2000s cavities among adolescents have been reduced by nearly 40 percent. This rate decrease is explained in part by the fact that more areas have added fluoride to their drinking water and more children get regular, good dental care. However, children still drinking from a bottle anytime after their first birthday are more likely to have tooth decay.

Causes and symptoms

Tooth decay requires the simultaneous presence of three factors: plaque bacteria, sugar, and a vulnerable tooth surface. Although several microorganisms found in the mouth can cause tooth decay, the primary disease agent appears to be *Streptococcus mutans*. The simple sugars used by the bacteria are glucose, sucrose, and lactose. They are converted primarily into lactic acid. When this acid builds up on an unprotected tooth surface, it dissolves the **minerals** in the enamel, creating holes and weak spots (cavities). As the decay spreads inward into the middle layer (the dentin), the tooth becomes more sensitive to temperature and touch. When the decay reaches the center of the tooth (the pulp), the resulting inflammation (pulpitis) produces a **toothache**.

When to call the doctor

If a child complains of tooth or jaw **pain** and his or her cheek is swollen, and if he or she has a **fever** over 100°F (37.8°C), a dentist should be called right away. A dentist should be called during normal business hours if the child has tooth or jaw pain for more than a day, if white spots are noticed on an infant's teeth, or if there appear to be any other problems with the teeth or gums.

Diagnosis

Tooth decay develops at varying rates. It may be found during a routine six-month dental checkup before the individual is even aware of a problem. In other cases, the individual may experience common early symptoms, such as sensitivity to hot and cold liquids or localized discomfort after eating very sweet foods. The dentist or dental hygienist may suspect tooth decay if a dark spot or a pit is seen during a visual examination. Front teeth may be inspected for decay by shining a light from behind the tooth. This method is called transillumination. Areas of decay, especially between the teeth, will appear as noticeable shadows when the teeth are transilluminated. X rays may be taken to confirm the presence and extent of the decay. The dentist then makes the final clinical diagnosis by probing the enamel with a sharp instrument.

Tooth decay in pits and fissures may be differentiated from dark shadows in the crevices of the chewing surfaces by a dye that selectively stains parts of the tooth that have lost mineral content. A dentist can also use this dye to tell whether all tooth decay has been removed from a cavity before placing a filling.

Damage caused by baby bottle tooth decay is often not diagnosed until the child has a severe problem, because many parents do not schedule regular dental

exams for their small children. It is recommended that a child's first trip to the dentist be before one year of age and that trips to the dentist occur regularly every six months after that.

Treatment

To treat most cases of tooth decay in older children, the dentist removes all decayed tooth structure, shapes the sides of the cavity, and fills the cavity with an appropriate material, such as silver amalgam or composite resin. The filling is put in to restore and protect the tooth. If decay has attacked the pulp, the dentist or a specialist called an endodontist may perform root canal treatment and then cover the tooth with a crown.

In cases of baby bottle tooth decay, the dentist must assess the extent of the damage before deciding on the treatment method. If the problem is caught early, the teeth involved can be treated with fluoride, followed by changes in the infant's feeding habits and better **oral hygiene**. Primary teeth with obvious decay in the enamel that has not yet progressed to the pulp need to be protected with stainless steel crowns. Fillings are not usually an option in small children because of the small size of their teeth and the concern of recurrent decay. When the decay has advanced to the pulp, pulling the tooth is often the treatment of choice. Unfortunately, loss of primary teeth at this age may hinder the young child's ability to eat and speak. It may also have negative effects on the alignment and spacing of the permanent teeth when they come in.

Prognosis

With timely diagnosis and treatment, the progression of tooth decay can be stopped relatively painlessly. If the pulp of the tooth is infected, the infection may be treated with **antibiotics** prior to root canal treatment or extraction. The longer the decay goes untreated, however, the more destructive it becomes and the longer and more intensive the necessary treatment will be. In addition, an individual with two or more areas of tooth decay is at increased risk of developing additional cavities.

Prevention

It is easier and less expensive to prevent tooth decay than to treat it. The four major prevention strategies are proper oral hygiene, fluoride, sealants, and attention to diet.

Oral hygiene

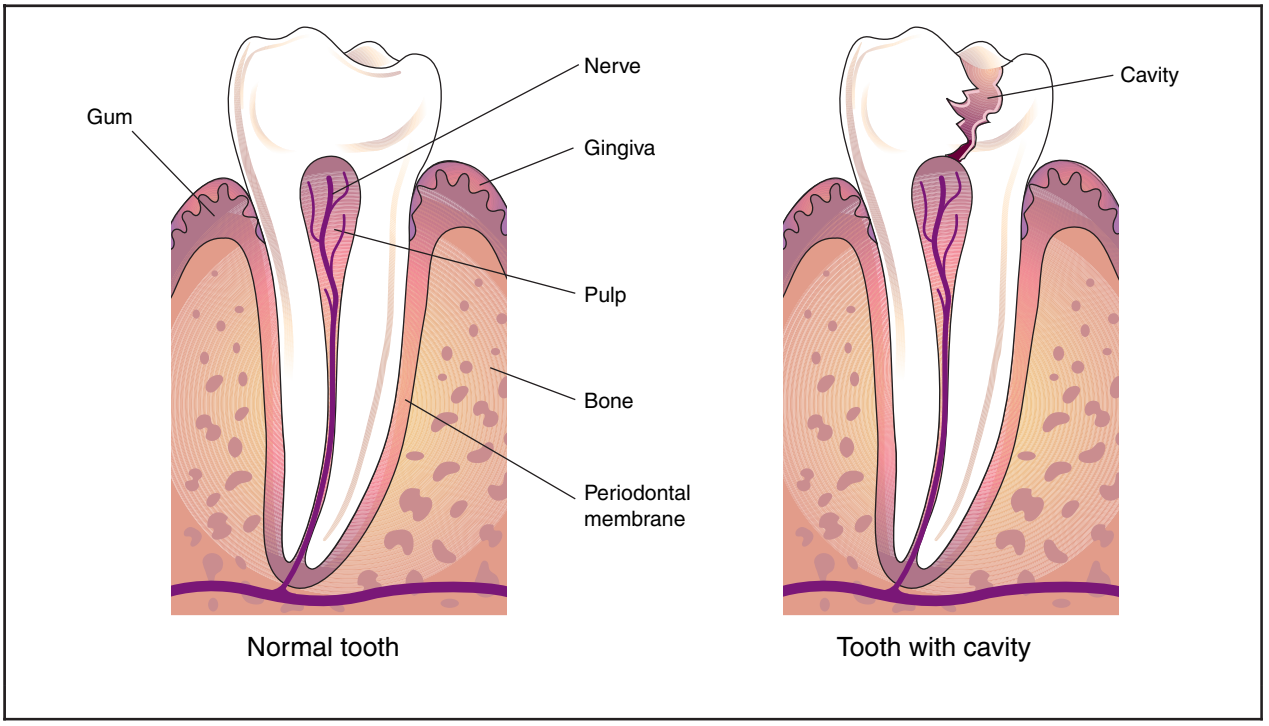
The best way to prevent tooth decay is to brush the teeth at least twice a day, preferably after every meal and snack, and floss daily. Cavities develop most easily in spaces that are hard to clean. These areas include surface grooves, spaces between teeth, and the area below the gum line. Effective brushing cleans each outer tooth surface, inner tooth surface, and the horizontal chewing surfaces of the back teeth, as well as the tongue. Flossing once a day helps prevent gum disease by removing food particles and plaque at and below the gum line, as well as between teeth. Patients should visit their dentists every six months for oral examination and professional cleaning.

Parents can easily prevent baby bottle tooth decay by not allowing a child to fall asleep with a bottle containing sweetened liquids. If a bottle is necessary when the child is falling asleep it should be filled only with plain, unsweetened water. The child should be introduced to drinking from a cup around six months of age and usually weaned from bottles by 12 months. If an infant seems to need oral comfort between feedings, a pacifier specially designed for the mouth may be used. Pacifiers, however, should never be dipped in honey, corn syrup, or other sweet liquids.

After the eruption of the first tooth, parents should begin routinely wiping the infant's teeth and gums with a moist piece of gauze or a soft cloth, especially right before bedtime. Parents may begin brushing a child's teeth with a small, soft toothbrush at about two years of age, when most of the primary teeth have come in. They should apply only a very small amount (the size of a pea) of toothpaste containing fluoride. Too much fluoride may cause spotting (fluorosis) of the tooth enamel. As the child grows, he or she will learn to handle the toothbrush, but parents should control the application of toothpaste and do the follow-up brushing until the child is about seven years old.

Fluoride application

Fluoride is a natural substance that slows the destruction of enamel and helps to repair minor tooth decay damage by remineralizing tooth structure. Toothpaste, mouthwash, fluoridated public drinking water, and vitamin supplements are all possible sources of fluoride. Children living in areas without fluoridated water should receive 0.25 mg/day of fluoride before age three, 0.5 mg/day of fluoride from three to six years of age, and 1 mg/day after age six. Sometimes children can also have their teeth treated with fluoride at the dentist's office.



Tooth decay is the destruction of the outer surface, or enamel, of a tooth. It is caused by acid buildup from plaque bacteria, which dissolve the minerals in the enamel and create cavities. *(Illustration by Electronic Illustrators Group.)*

Sealants

Because fluoride is most beneficial on the smooth surfaces of teeth, sealants were developed to protect the irregular surfaces of teeth. A sealant is a thin plastic coating that is painted over the grooves of chewing surfaces to prevent food and plaque from being trapped there. Sealant treatment is painless, because no part of the tooth is removed, although the tooth surface is etched with acid so that the plastic will adhere to the rough surface. Sealants are usually clear or tooth-colored, making them less noticeable than silver fillings. They cost less than fillings and can last up to 10 years, although they should be checked for wear at every dental visit. Children should get sealants on their first permanent "six-year" molars, which come in between the ages of five and seven, and on the second permanent "12-year" molars, which come in between the ages of 11 and 14. Sealants should be applied to the teeth shortly after they erupt, before decay can set in. Although sealants have been used in the United States for about 25 years, one survey by the National Institute of Dental Research reported that fewer than 8 percent of American children have them.

Diet

The risk of tooth decay can be lowered by choosing foods wisely and eating less often. Foods high in sugar and starch, especially when eaten between meals, increase the risk of cavities. The bacteria in the mouth use sugar and starch to produce the acid that destroys the enamel. The damage increases with more frequent eating and longer periods of eating. For better dental health, children should eat a variety of foods, limit the number of snacks, avoid sticky and overly sweetened foods, and brush often after eating. Drinking water is also beneficial for rinsing food particles from the mouth. Children can be taught to rinse their mouth out with water after eating if they are unable to brush after lunch at school.

Parental concerns

If tooth decay is not treated, it can result in other, more serious, problems involving the gums, cheeks, or jaw. Baby bottle tooth decay that is not treated quickly can result in the affected teeth being removed. Although the child will eventually develop adult teeth to replace the baby teeth, missing baby teeth can result in overcrowding when the adult teeth come in. Missing baby teeth can also result in the adult teeth coming in crooked, the child having to chew on one side of his or her mouth, and speech delays. As of 2004, most cavities could be fixed without much discomfort by a medical professional and without any serious longterm consequences if the cavities are found and treated early.

KEY TERMS

Amalgam—A mixture (alloy) of silver and several other metals used by dentists to make fillings for cavities.

Caries—The medical term for tooth decay.

Cavity—A hole or weak spot in the tooth surface caused by decay.

Dentin—The middle layer of a tooth, which makes up most of the tooth's mass.

Enamel—The hard, outermost surface of a tooth.

Fluoride—A chemical compound containing fluorine that is used to treat water or applied directly to teeth to prevent decay.

Mucin—A protein in saliva that combines with sugars in the mouth to form plaque.

Plaque—A sticky, colorless film of bacteria, sugars, and mucin that forms on teeth and causes tooth decay.

Pulp—The soft, innermost layer of a tooth that contains its blood vessels and nerves.

Sealant—A thin plastic substance that is painted over teeth as an anti-cavity measure to seal out food particles and acids produced by bacteria.

Transillumination—A technique of checking for tooth decay by shining a light behind the patient's teeth. Decayed areas show up as spots or shadows.

Resources

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ORGANIZATIONS

American Dental Association. 211 E. Chicago Ave., Chicago, IL 60611. Web site: <www.ada.org>.

American Dental Hygienists' Association. 444 North Michigan Ave., Chicago, IL 60611. Web site: <www.adha.org>.

National Institute of Dental Research. 31 Center Drive, MSC 2190, Building 31, Room 5B49, Bethesda, MD 20892–2190. Web site: <www.nidcr.nih.gov/>.

Tish Davidson, A.M.

Toothache

Definition

A toothache is any **pain** or soreness within or around a tooth, indicating inflammation and possible infection.

Description

A toothache may feel like a sharp pain or a dull ache. The tooth may be sensitive to pressure, heat, cold, or sweets. In cases of severe pain, identifying the problem tooth is often difficult. Any patient with a toothache should see a dentist for diagnosis and treatment. Most toothaches get worse if left untreated.

Demographics

Toothaches are common. Yet people have fewer cavities on average in the early 2000s than they did in the nineteenth or twentieth century, in part because in the late 1900s many towns added fluoride to the drinking water and many dentists began prescribing fluoride tablets as a preventative measure. Fewer cavities and better tooth health have reduced the number of toothaches.

Causes and symptoms

Toothaches may result from any of a number of causes:

- **tooth decay** (dental caries)
- inflammation of the tooth pulp (pulpitis)
- abscesses
- gum disease, including periodontitis
- loose or broken filling
- cracked or impacted tooth
- exposed tooth root
- food wedged between teeth or trapped below the gum line

- tooth nerve irritated by clenching or grinding of teeth (bruxism)
- pressure from congested sinuses
- traumatic injury

When to call the doctor

If the toothache lasts for more than 24 hours an appointment with the dentist should be made. If there is **fever**, swelling, intense pain, or bleeding in addition to the toothache the dentist should be seen right away.

Diagnosis

Diagnosis includes identifying the location of the toothache, as well as the cause. The dentist begins by asking the patient specific questions about the toothache, including the types of foods that make the pain worse, whether the tooth is sensitive to temperature or biting, and whether the pain is worse at night. The dentist then examines the patient's mouth for signs of swelling, redness, and obvious tooth damage. The presence of pus indicates an abscess or gum disease. The dentist may flush the sore area with warm water to dislodge any food particles and to test for sensitivity to heat. The dentist may then dry the area with gauze to determine sensitivity to touch and pressure. The dentist may probe tooth crevices and the edges of fillings with a sharp instrument, looking for areas of tooth decay. Finally, the dentist may take x rays, looking for evidence of decay between teeth, a cracked or impacted tooth, or a disorder of the underlying bone.

Treatment

Treatment depends on the underlying cause of the toothache. If the pain is due to tooth decay, the dentist will remove the decayed area and restore the tooth with a filling of silver amalgam or composite resin. Loose or broken fillings are removed, new decay cleaned out, and a new filling is placed. If the pulp of the tooth is damaged, root canal therapy is needed. The dentist or a specialist called an endodontist removes the decayed pulp, fills the space left behind with a soothing paste, and covers the tooth with a crown to protect and seal it. If the damage cannot be treated by these methods, or if the tooth is impacted, the tooth must be extracted. If the dentist finds an infection, **antibiotics** are given to treat it.

Toothaches should always be professionally treated by a dentist. Some methods of self-treatment, however, may help manage the pain until professional care is available:

- rinsing with warm salt water
- using dental floss to remove any food particles
- taking **acetaminophen** (Tylenol) or ibuprofen (Advil) to relieve pain (Aspirin is not recommended for children because of the risk of Reye's syndrome.)
- applying a cold compress against the outside of the cheek
- using clove oil (*Syzygium aromaticum*) to numb the gums (The oil may be rubbed directly on the sore area or used to soak a small piece of cotton and applied to the sore tooth. Clove oil should not be put on the tongue because it often burns or stings.)

Alternative treatment

Toothaches caused by infection or tooth decay must be treated by a dentist. Several alternative therapies may be helpful for pain relief until dental treatment is available. Clove oil (*Syzygium aromaticum*) may be rubbed on sensitive gums to numb them or added to a small cotton pellet that is then placed into or over a hole in the tooth. The herb corydalis (*Corydalis yanhusuo*) may also help relieve toothache pain. Pain also may be reduced by using acupressure, acupuncture, or reiki.

Prognosis

Prompt dental treatment provides a positive outcome for toothache. In the absence of active infection, fillings, root canal treatments, or extractions may be performed with minimal discomfort to the patient. When a toothache is left untreated, a severe infection may develop and spread to the sinuses or jawbone, and eventually cause blood poisoning.

Prevention

Maintaining proper **oral hygiene** is the key to preventing toothaches. The best way to prevent tooth decay is to brush at least twice a day, preferably after every meal and snack. Flossing once a day also helps prevent gum disease by removing food particles and bacteria at and below the gum line, as well as between teeth. Children should visit the dentist at least every six months for oral examinations and professional cleaning. Dentists often recommend that children see the dentist for the first time before they are one year old. Parents should help young children brush their teeth. Fluoride is also very helpful in preventing tooth decay. If the town's water is not fluoridated, the parent should ask the dentist for fluoride supplements.

KEY TERMS

Abscess—A localized collection of pus in the skin or other body tissue caused by infection.

Bruxism—Habitual clenching and grinding of the teeth, especially during sleep.

Cavity—A hole or weak spot in the tooth surface caused by decay.

Dental caries—A disease of the teeth in which microorganisms convert sugar in the mouth to an acid that erodes the tooth. Commonly called a cavity.

Enamel—The hard, outermost surface of a tooth.

Endodontist—A dentist who specializes in diagnosing and treating diseases of the pulp and other inner structures of the tooth.

Impacted tooth—Any tooth that is prevented from reaching its normal position in the mouth by another tooth, bone, or soft tissue.

Periodontitis—Inflammation of the periodontium, the tissues that support and anchor the teeth. Without treatment it can destroy the structures supporting the teeth, including bone.

Pulp—The soft, innermost layer of a tooth that contains its blood vessels and nerves.

Pulpitis—Inflammation of the pulp of a tooth that involves the blood vessels and nerves.

Parental concerns

Toothaches are most often caused by cavities. If not treated promptly an infection could begin or spread. If infection spreads to the blood, serious complications can result.

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Tish Davidson, A.M.
Bethany Thivierge

Topical antibiotics *see* **Antibiotics, topical**

TORCH test

Definition

The TORCH test, sometimes called the TORCH panel, belongs to a category of blood tests called infectious-disease antibody titers. A titer is the serial dilution of antibodies (protein molecules or immunoglobulins produced by the immune system in response to specific disease agents) found in blood serum that determines their level of concentration. Antibodies are proteins produced by the immune system in response to infectious agents that are foreign to the body, such as viruses, bacteria, parasites, or toxins. These infectious organisms have antigens on their surfaces that stimulate the immune system to produce corresponding antibodies. IgM antibodies are produced in response to viruses. The TORCH test screens for the presence of IgM antibodies, and the titer determines their concentration in the blood. The name of the test is an acronym derived from the initial letters of the five groups of chronic infections: **toxoplasmosis**, other viruses, **rubella**, cytomegalovirus (CMV), and **herpes simplex** virus (HSV). The "other viruses" usually include syphilis, **hepatitis B**, coxsackie virus, Epstein-Barr virus (mononucleosis), varicella-zoster virus, and human parvovirus. The test is performed by various methods in the clinical laboratory and may also be referred to as viral immunoglobulins testing. Methods used in the early 2000s are more sensitive and specific and can identify the specific virus.

Purpose

A TORCH test is performed to help screen for certain virus infections in infants who may have been exposed to a causative organism. The five groups of disease-causing organisms whose antibodies are measured by the TORCH test are grouped together because they can cause a cluster of symptomatic birth defects in newborns. This group of defects is sometimes called the TORCH syndrome. The pediatrician may order the TORCH test to be performed when a newborn has these symptoms, in order to determine if any of the five types of infection may be involved.

Symptoms of TORCH syndrome that may encourage testing include the following:

- Small size in proportion to length of the mother's pregnancy at time of delivery: Infants who are smaller than would be expected (below the tenth percentile) are referred to as small-for-gestational-age (SGA).
- Enlarged liver and spleen.
- Low level of platelets (tiny cellular elements in blood that are an important part of coagulation).
- Skin rash: The type of skin rash associated with the TORCH syndrome is usually reddish-purple or brown and is caused by the leakage of blood from broken capillaries into the baby's skin.
- Central nervous system impairment: This may include **encephalitis**, calcium deposits in brain tissue, or seizures.
- **Jaundice**: Yellow-stained skin and whites of the eyes due to elevated levels of bilirubin, a substance normally filtered out by the liver. Jaundice may indicate liver dysfunction, although it can also be a normal result of red cell turnover in the newborn.

Description

Besides general symptoms that may encourage a pediatrician to order the TORCH panel of tests, each of the TORCH infections has its own origins and may have a characteristic cluster of symptoms in newborns. These unique characteristics, the general condition and symptoms of the child, and the test results are studied in order for the physician to make a diagnosis.

Toxoplasmosis

Toxoplasmosis is caused by *Toxoplasma gondii*, a parasite that can be acquired by the mother from handling cat feces, drinking unpasteurized milk, or eating contaminated meat. The infection is carried to the infant through the mother's placenta and can cause impairment of the infant's eyes (opthalmic impairment) and central nervous system (neurological dysfunction). The organism can invade brain or muscle tissue and form cysts. Infection acquired by the mother later in pregnancy usually decreases the likelihood of infection in the infant at birth although eye problems may occur in **adolescence**. Toxoplasmosis early in pregnancy is more likely to cause miscarriage or serious birth defects. The incidence of toxoplasmosis in newborns is one in 1,000 live births.

Other viruses (syphilis)

Syphilis is caused by the spiral- or coil-shaped bacteria (spirochete), *Treponema pallidum*. It is transmitted among adults through sexual intercourse. About 2 to 5 percent of children born to mothers diagnosed with syphilis have the disease at birth. Syphilis was added to the TORCH panel because of an increase in reported cases after 1990. Syphilis can cause early delivery, miscarriage, and is a potentially life-threatening infection for an affected fetus, often resulting in stillbirth. The mortality rate in infants infected with syphilis is about 54 percent.

Rubella

Rubella is a virus that has a seasonal pattern, with epidemics most likely in the spring. Between 0.1 to 2 percent of newborns are infected with rubella. The rate of fetal infection varies according to the timing of the mother's infection during pregnancy. Birth defects, however, are most likely (85%) in infants infected during the first eight weeks of pregnancy. Infants born with rubella may already show signs of heart disease, retarded growth, hearing loss, blood disorders, vision problems, or **pneumonia**. They may also develop problems later in childhood, including **autism**, hearing loss, brain syndromes, immune system disorders, or thyroid disease.

Cytomegalovirus (CMV)

Cytomegalovirus belongs to the herpesvirus group of infections. It can be transmitted through body secretions, as well as by sexual contact; some newborns acquire CMV through the mother's breast milk. In adults, it produces symptoms resembling those of mononucleosis. About 1 to 2.2 percent of newborns in the United States are infected with CMV. Of this group, 10 percent have measurable symptoms. The mortality rate for these symptomatic newborns is 20 to 30 percent. Surviving infants with CMV may suffer from hearing problems (15%) or **mental retardation** (30%). Newborns who acquire CMV during the birth process or shortly after

birth may develop pneumonia, hepatitis, or various blood disorders.

Herpes simplex virus (HSV)

Herpesvirus infections are among the most common viral infections in humans. They are spread by oral or genital contact. It is estimated that between one in 1,000 and one in 5,000 infants are born with HSV infections. About 80 percent of these infections are acquired during the birth process itself; the virus enters the infant through its eyes, skin, mouth, and upper respiratory tract. Of infants born with HSV infection, about 20 percent have localized infections of the eyes, mouth, or skin. About 50 percent of infected infants will develop the disease throughout the body (disseminated) within nine to 11 days after birth. Disseminated herpes infections attack the liver and adrenal glands, as well as other body organs. Without treatment, the mortality rate is 80 percent. Even with antiviral medication, the mortality rate is still 15 to 20 percent, with 40 to 55 percent of the survivors having long-term damage to the central nervous system. In order to begin early, effective treatment, it is critical for pediatricians to diagnose HSV infection in newborns as soon as possible.

Performing the TORCH panel requires obtaining a sample of the infant's blood. Samples from infants are usually obtained by the heelstick procedure when only a small quantity of blood is needed. The baby's foot is wrapped in a warm cloth for five minutes to bring blood to the surface and help it to flow more easily. The foot is then sterilized with an alcohol swab and a lancet is used to puncture the baby's heel on one side, avoiding the center of the heel to prevent inflammation of the bone. The blood sample is drawn in tiny capillary tubes, properly labeled, and taken to the laboratory for testing. In rare instances, a phlebotomist is not able to draw sufficient blood from a heel puncture, and a physician may draw venous blood from a femoral vein in the groin area or another vein larger than veins in an infant's arms.

Since the TORCH test is a screening or first-level test, the pediatrician may order tests of other body fluids or tissues to confirm the diagnosis of a specific infection. In suspected cases of toxoplasmosis, rubella, or syphilis, cerebrospinal fluid may be obtained from the infant by spinal tap in order to confirm the diagnosis. A diagnosis of CMV is usually confirmed by culturing the virus in a sample of the infant's urine. In HSV infections, tissue culture is the best method to confirm the diagnosis.

Precautions

Because toxoplasmosis can be transmitted by handling cat feces, pregnant women should avoid cleaning cat boxes or handling cats. Any suspected infection should be reported to the obstetrician so that testing for the causative parasite in the mother can be performed.

Medical personnel and **family** members must be aware of the possible presence of infective organisms in the infant and proper precautions taken, such as hand washing, when the infant or the infant's body fluids (blood, urine, feces) are handled.

If the infant has had blood drawn often from the same site on the heel or heels, causing scarring, inflammation, or the accumulation of tissue fluid, it may cause inaccurate test results.

False negative and false positive results can occur with the TORCH test for immunoglobulins because of cross-reacting antibodies, especially among the different types of herpes viruses.

Preparation

No special preparation, other than sterile technique by medical personnel, is required.

Aftercare

The site from which blood is withdrawn must be kept clean after the procedure and must be checked regularly for bleeding. A small adhesive patch may be used to protect the site.

Risks

The performance of the TORCH test carries no significant risk. Drawing blood for the test may involve light bleeding or bruising at the site of puncture or blood may accumulate under the puncture site (hematoma), requiring that a new location be found for subsequent tests. The infant's heel may be at risk of scarring, infection of the bone, cellulitis (inflammation of cellular tissue), small lumpy calcium deposits.

Normal results

The normal result of a TORCH panel reveals normal levels of immunoglobulin M (IgM) antibody in the infant's blood. IgM is one of five types of antibodies (protein molecules) produced by the immune system and found in blood. IgM is a specific class of antibody that seeks out virus particles. It is the most common type of immunoglobulin in newborns and, therefore, the most useful indicator of the presence of one of the TORCH virus infections.

KEY TERMS

Antibody—A special protein made by the body's immune system as a defense against foreign material (bacteria, viruses, etc.) that enters the body. It is uniquely designed to attack and neutralize the specific antigen that triggered the immune response.

Antigen—A substance (usually a protein) identified as foreign by the body's immune system, triggering the release of antibodies as part of the body's immune response.

Bacteria—Singular, bacterium; tiny, one-celled forms of life that cause many diseases and infections.

Titer—The highest dilution of a material (e.g., serum or other body fluid) that produces a reaction in an immunologic test system. Also refers to the extent to which an antibody can be diluted before it will no longer react with a specific antigen. Also spelled titre.

Virus—A small infectious agent consisting of a core of genetic material (DNA or RNA) surrounded by a shell of protein. A virus needs a living cell to reproduce.

Abnormal results

The general abnormal or "positive" result reveals high levels of IgM antibody present in the infant's blood. The test can be refined further for antibodies specific to given disease agents. The TORCH screen, however, can produce both false-positive and false-negative findings. Doctors can measure IgM levels in the infant's cerebrospinal fluid, as well as in the blood, if confirmation is needed.

Parental concerns

Parents will necessarily be concerned about the possibility of infection in the child and the amount of testing that may have to be done. Awareness of the value of the TORCH panel of tests to help confirm the presence of an infective organism and its concentration in the blood is important, especially because confirmatory tests lead to faster, more effective treatment. Medical personnel can teach parents about safe practices for handling an infant with a virus infection that can possibly spread to family members.

See also Cytomegalovirus (CMV) infection; Infectious mononucleosis; Hepatitis B.

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Torticollis *see* **Wryneck**

Tourette syndrome

Definition

Tourette syndrome (TS) is an inherited disorder of the nervous system, characterized by a variable expression of unwanted movements and noises (**tics**).

Description

Tourette syndrome is also known as Gilles de la Tourette syndrome, named after Gilles de la Tourette, a French neurologist who first described the syndrome in 1885. Children with Tourette syndrome show symptoms before the age of 18, usually around age seven, and have symptoms that usually last into adulthood. The symptoms of Tourette syndrome are usually motor and/or vocal tics, although in some children other symptoms such as socially inappropriate comments, and socially inappropriate or self-injurious behaviors sometimes

occur. Children with Tourette syndrome are more likely to have **obsessive-compulsive disorder** (OCD), attention deficient disorder (ADD), and **attention deficit hyperactivity disorder** (ADHD). The symptoms of Tourette syndrome are extremely variable over time, with some symptoms beginning and some ceasing to be a problem as the child grows. Many people with Tourette syndrome experience a decrease in symptoms as they age, and some people see a complete disappearance of their symptoms.

Demographics

Tourette syndrome is found in all populations and all ethnic groups, but is three to four times more common in males than females. The exact frequency of Tourette syndrome is unknown, but estimates range from 0.05 percent to 2 percent. Estimates vary widely in part because many people with Tourette syndrome have very mild symptoms and may not seek medical attention. It is estimated that there are about 1,000 new cases of Tourette syndrome diagnosed in the United States every year.

Causes and symptoms

The causes of Tourette syndrome are not fully understood. Most studies agree that symptoms of Tourette syndrome involve the chemicals in the brain that help transmit information from one nerve cell in the brain to another. These chemicals are called neurotransmitters. Some studies suggest that the tics in Tourette syndrome are caused by an increased amount of a neurotransmitter called dopamine. Other studies suggest instead that there is a problem with a different neurotransmitter called serotonin. Still others believe the problem involves other chemicals required for normal functioning of the brain.

Most studies suggest that Tourette syndrome is an autosomal dominant disorder with decreased penetrance. An autosomal disorder is one that occurs because of an abnormal gene on a chromosome that is not a sex-linked chromosome. A dominant disorder means that it only takes one abnormal gene in a pair of genes to have the disorder. Parents each pass one copy of each gene to their child. Because in autosomal dominant disorders one gene is abnormal, people with this disorder have about a 50 percent chance of passing the abnormal gene to their offspring. Decreased penetrance means that not all people who inherit the abnormal gene develop symptoms. There is some evidence that females who inherit the Tourette syndrome gene have a lower probability of exhibiting symptoms than males who inherit the gene.

The principal symptoms of Tourette syndrome include simple and complex motor and vocal tics. Simple motor tics are characterized by brief muscle contractions of only one or a small number of muscle groups. An eye twitch is an example of a simple motor tic. Complex motor tics tend to appear more complicated and purposeful than simple tics and involve coordinated contractions of several muscle groups. Some examples of complex motor tics include the act of hitting oneself or jumping.

Vocal tics are actually manifestations of motor tics that involve the muscles required for producing sound. Simple vocal tics include **stuttering**, stammering, abnormal emphasis of part of a word or phrase, and inarticulate noises such as throat clearing, grunts, and high-pitched sounds. Complex vocal tics typically involve the involuntary expression of words. Perhaps the most striking example of this is coprolalia, the involuntary expression of obscene or socially inappropriate words or phrases, which occurs in fewer than one-third of people with Tourette syndrome. The involuntary echoing of the last word, phrase, sentence, or sound vocalized by oneself (phalilalia) or by another person or sound in the environment (echolalia) are also classified as complex tics.

The type, frequency, and severity of tics exhibited varies tremendously among individuals with Tourette syndrome. Tourette syndrome has a variable age of onset, and tics can start anytime between infancy and age 18. Initial symptoms usually occur before the early teens; the average age of onset for both males and females is approximately seven years. Most individuals with symptoms initially experience simple muscle tics involving the eyes and the head. These symptoms can progress to tics involving the upper torso, neck, arms, hands, and occasionally the legs and feet. Complex motor tics are usually the latest-onset motor tics. Vocal tics usually have a later onset than motor tics.

Not only is there extreme variability in symptoms among individuals with Tourette syndrome, but individuals commonly experience variability in type, frequency, and severity of symptoms over the course of their lifetime. Adolescents with Tourette syndrome often experience unpredictable and more severe than usual symptoms, which may be related to fluctuating hormone levels and decreased compliance in taking medications. Many people who as children have Tourette syndrome experience a decrease in symptoms or a complete end to symptoms in their adult years.

Several factors appear to affect the severity and frequency of tics. Stress appears to increase the frequency and severity of tics, while concentration on another part of the body that is not involved in a tic can result in the temporary alleviation of symptoms. Relaxation follow-

ing attempts to suppress the occurrence of tics may result in an increased frequency of tics. An increased frequency and severity of tics can also result from exposure to such drugs as steroids, cocaine, amphetamines, and **caffeine**. Hormonal changes, such as those that occur prior to the menstrual cycle, can also increase the severity of symptoms.

Other associated symptoms

People with Tourette syndrome are more likely to exhibit non-obscene, socially inappropriate behaviors such as expressing insulting or socially unacceptable comments or performing socially unacceptable actions. It is not known whether these symptoms stem from more general dysfunction of impulse control that might be part of Tourette syndrome.

Tourette syndrome appears to also be associated with attention deficit disorder (ADD), a disorder characterized by a short attention span and impulsivity, and in some cases hyperactivity. Researchers have found that 21 to 90 percent of individuals with Tourette syndrome also exhibit symptoms of ADD.

People with Tourette syndrome are also at higher risk for having symptoms of obsessive-compulsive disorder (OCD), a disorder characterized by persistent, intrusive, and senseless thoughts (obsessions) or compulsions to perform repetitive behaviors that interfere with normal functioning. A person with OCD, for example, may be obsessed with germs and may counteract this obsession with continual hand washing. Symptoms of OCD are present in 1.9 to 3 percent of the general population, whereas 28 to 50 percent of people with Tourette syndrome have symptoms of OCD.

Self-injurious behavior (SIB) is also seen more frequently in those with Tourette syndrome. Approximately 34 to 53 percent of individuals with Tourette syndrome exhibit some form of self-injuring behavior. The SIB is often related to OCD but can also occur in those with Tourette syndrome who do not have OCD.

Symptoms of **anxiety** and depression are also found more commonly in people with Tourette syndrome. It is not clear, however, whether these are symptoms of Tourette syndrome or occur as a result of having to deal with the symptoms of moderate to severe Tourette syndrome.

People with Tourette syndrome may also be at increased risk for having learning disabilities and **personality disorders** and may be more predisposed to such behaviors as aggression, antisocial behaviors, severe temper outbursts, and inappropriate sexual behavior.

When to call the doctor

Parents should call the doctor if they notice the symptoms of Tourette syndrome. The initial tics usually initially involve the face or head, but the doctor should be consulted if any uncontrolled repetitive behavior is observed.

Diagnosis

Tourette syndrome cannot be diagnosed through laboratory tests. Sometimes laboratory tests can be helpful, however, in ruling out other possible conditions. The diagnosis of Tourette syndrome is made by observing and interviewing the child, looking at the family's medical history, and talking to the child's **family** and sometimes to other caregivers. The diagnosis of Tourette syndrome is complicated by a variety of factors. The extreme range of symptoms of this disorder can make it difficult to differentiate Tourette syndrome from other disorders with similar symptoms. Diagnosis is further complicated by the fact that some tics appear to be within the range of normal behavior. For example, an individual who only exhibits such tics as throat clearing and sniffing may be misdiagnosed with a medical problem such as **allergies**. In addition, such bizarre and complex tics as coprolalia may be mistaken for psychotic or so-called bad behavior. Diagnosis is also made more difficult because often individuals attempt to control tics in public, and, therefore, the healthcare professional may have difficulty observing the symptoms firsthand. Although there is some disagreement over what criteria should be used to diagnose Tourette syndrome, the most common aid in the diagnosis is the *DSM-IV*. The *DSM-IV* outlines suggested diagnostic criteria for a variety of conditions, including Tourette syndrome.

DSM-IV criteria are:

- presence of both motor and vocal tics at some time during the course of the illness
- the occurrence of multiple tics nearly every day through a period of more than one year without a remission of tics for a period of greater than three consecutive months
- distress or impairment in functioning caused by symptoms
- onset occurs prior to age 18
- symptoms not due to medications or drugs and not related to another medical condition

Some physicians criticize the *DSM-IV* criteria, arguing that they do not include the full range of behaviors and symptoms seen in Tourette syndrome. Others

criticize the criteria because they limit the diagnosis to those who experience a significant impairment, which may exclude individuals who have the syndrome but exhibit milder symptoms. For these reasons many physicians use their clinical judgment as well as the *DSM-IV* criteria as a guide to diagnosing Tourette syndrome.

Treatment

There is no cure for Tourette syndrome. Treatment involves the control of symptoms through educational and psychological interventions and/or medications. The treatment and management of Tourette syndrome varies from patient to patient and should focus on the alleviation of the symptoms that are most bothersome to the individual or that cause the most interference with daily functioning.

Psychological and educational interventions

Psychological treatments such as counseling are not generally useful for the treatment of tics but can be beneficial in the treatment of associated symptoms such as obsessive-compulsive behavior and attention deficit disorder. Counseling may also help individuals to cope better with the symptoms of Tourette syndrome and to have more positive social interactions. Psychological interventions may also help people cope better with stressors that can normally trigger tics. The education of family members, teachers, and peers about Tourette syndrome can be helpful and may help to foster acceptance and prevent social isolation.

Medications

Many people with mild symptoms of Tourette syndrome never require medication. Those with more severe symptoms may require medication for all or part of their lifetime. As of 2004, the most effective treatment of tics associated with Tourette syndrome involved the use of drugs such as haloperidol, pimozide, sulpiride, and tiapride, which decrease the amount of dopamine in the body. Unfortunately, even at low dosages, these drugs bring a high incidence of side effects. The short-term side effects can include sedation, dysphoria, weight gain, movement abnormalities, depression, and poor school performance. Long-term side effects can include **phobias**, memory difficulties, and personality changes. These drugs are, therefore, better suited for short-term rather than long-term therapy.

In many cases, treatment of associated conditions such as ADD and OCD is considered more important than the tics themselves. Clonidine used in conjunction with such stimulants as Ritalin may be useful for treating people with Tourette syndrome who also have symptoms of ADD. Stimulants should be used with caution in individuals with Tourette syndrome, since they can sometimes increase the frequency and severity of tics. OCD symptoms in those with Tourette syndrome are often treated with such drugs as Prozac, Luvox, Paxil, and Zoloft.

In many cases the treatment of Tourette syndrome with medications can be discontinued after **adolescence**. Trials should be performed through the gradual tapering off of medications and should always be done under a doctor's supervision.

Prognosis

The prognosis for Tourette syndrome is fairly good. Although symptoms generally get worse during early adolescence, many people with Tourette syndrome experience a lessening of the severity of their symptoms during late adolescence and early adulthood. Approximately one third of children with Tourette syndrome will experience complete or nearly complete remission during their late adolescent and early adult years. Another third will experience a significant drop off in the severity and/or frequency of their symptoms during this time. It is difficult to tell how many children with Tourette syndrome experience complete remission over their entire adult lives, but it has been estimated to be about 8 percent. Many children who do not have complete and lasting remission will experience months or even years without significant symptoms. There does not appear to be a definite correlation between the type, frequency, and severity of symptoms and the eventual prognosis. People with Tourette syndrome who have other symptoms such as obsessive-compulsive disorder, attention deficit disorder, and self-injurious behavior usually have a poorer prognosis.

Prevention

There is no known way to prevent Tourette syndrome.

Parental concerns

Tourette syndrome does not, in itself, negatively affect **intelligence** or cognition. It is, however, often associated with other disorders such as obsessive-compulsive disorder and attention deficit disorder. It is also sometimes associated with learning and psychological disorders, many of which are often more debilitating than Tourette syndrome itself. Tourette syndrome does not reduce life expectancy. Children with Tourette syn-

KEY TERMS

Attention deficit disorder (ADD)—Disorder characterized by a short attention span, impulsivity, and in some cases hyperactivity.

Autosomal dominant—A pattern of inheritance in which only one of the two copies of an autosomal gene must be abnormal for a genetic condition or disease to occur. An autosomal gene is a gene that is located on one of the autosomes or non-sex chromosomes. A person with an autosomal dominant disorder has a 50% chance of passing it to each of their offspring.

Coprolalia—The involuntary use of obscene language.

Copropraxia—The involuntary display of unacceptable/obscene gestures.

Decreased penetrance—Individuals who inherit a changed disease gene but do not develop symptoms.

Dysphoria—Feelings of anxiety, restlessness, and dissatisfaction.

Echolalia—Involuntary echoing of the last word, phrase, or sentence spoken by someone else.

Echopraxia—The imitation of the movement of another individual.

Neurotransmitter—A chemical messenger that transmits an impulse from one nerve cell to the next.

Obsessive-compulsive disorder—An anxiety disorder marked by the recurrence of intrusive or disturbing thoughts, impulses, images, or ideas (obsessions) accompanied by repeated attempts to supress these thoughts through the performance of certain irrational and ritualistic behaviors or mental acts (compulsions).

Phalilalia—Involuntary echoing by an individual of the last word, phrase, sentence, or sound he/she vocalized.

Tic—A brief and intermittent involuntary movement or sound.

drome often have problems socializing because of embarrassment over uncontrollable tics and negative reactions from parents, teachers, and peers who do not understand the disorder. Children with Tourette syndrome may need special attention to help them cope with the social implications of their disorder.

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Tish Davidson, A.M.

Toxic shock syndrome

Definition

Toxic shock syndrome (TSS) is an uncommon, but potentially serious, illness that occurs when poisonous substances (toxins) produced by bacteria enter the bloodstream. The toxins cause a type of blood poisoning that results in high **fever**, symptoms of shock, and potentially organ failure.

Description

Initially toxic shock syndrome was associated with the use of ultra-absorbent tampons by menstruating girls and women. Between 1978 and 1980 thousands went to emergency rooms with high fever, **vomiting**, low blood pressure, **diarrhea**, and a rash resembling **sunburn**. Once ultra-absorbent tampons were taken off the market, the number of cases of toxic shock syndrome decreased substantially.

As of 2004, two different causes of toxic shock were recognized. Staphylococcal toxic shock syndrome (TSS) is caused by the bacteria *Staphylococcus aureus*. Streptococcal toxic shock syndrome (STSS, first described in

1987, is caused by *Streptococcus pyogenes* bacteria. Although both types of toxic shock are rare, STSS is more common and has a higher rate of serious complications and death.

Transmission

TSS is caused by a strain of *S. aureus* found in the nose, mouth, and occasionally the vagina. The bacteria produce a characteristic toxin. In large enough quantities, the toxin can enter the bloodstream, causing a potentially fatal reaction.

Although scientists still do not fully understand the link between TSS and tampons, most medical researchers suspect that tampons introduce oxygen into the vagina, which is normally an oxygen-free area of the body. Oxygen triggers bacterial growth, and the more absorbent the tampon, the longer it is left in place and the more toxin-producing bacteria it can harbor.

The streptococcal bacteria that cause STSS often enter the body through an infected wound in the skin, infection following surgery, postpartum or post abortion infection, or bone infection. STSS almost never develops following a simple **strep throat** infection.

Demographics

Although the majority of cases of TSS occur in menstruating girls and women, the disease may occur in people of any race and age, including children. STSS infection may occur in individuals who are weakened from surgery, injury, or disease that weakens the immune system. New mothers also are at higher risk for toxic shock syndrome, as are those who have recently had chicken pox. This disease is rare. Only about 100 cases of TSS and 300 cases of STSS were reported in the United States in 1996.

Causes and symptoms

Toxic shock syndrome begins suddenly about two days after infection occurs with a fever of 102°F (38.9°C) or above, vomiting and watery diarrhea, **headache**, and sunburn-like rash, together with a **sore throat** and body aches. Blood pressure may plummet a day or two after the first symptoms appear. When blood pressure drops, an individual may become disoriented or go into shock. The kidneys or liver may fail. After these developments, the skin on the hands and feet may peel. With STSS, flesh around the infected site may become damaged and die (become necrotic).

When to call the doctor

Toxic shock is a medical emergency that needs to be treated immediately in the hospital. Parents should go to the emergency room or call an ambulance if their child has a fast, weak pulse; cold hands and feet with pale moist skin; mental confusion or lethargy; abnormal breathing; a sunburn-like rash; high fever; or skin that is red, swollen, and infected.

Diagnosis

Diagnosis is made based on history, presenting symptoms, and culture of bacteria from the blood or wound. A rapid streptococcal test can be done with results available in 15 minutes. This test is positive in more than 85 percent of cases of toxic shock.

Treatment

Because toxic shock is a medical emergency, treatment is usually begun before laboratory results are available. The first line of treatment is to attempt to reverse the symptoms of shock. This process usually involves the administration of fluids intravenously. The site of infection is cleaned, and **antibiotics** are administered. If organ failure occurs, oxygen, the use of a respirator, or kidney dialysis may be necessary. It may also be necessary to surgically remove any infected and dying tissue.

Prognosis

Many otherwise healthy individuals recover from toxic shock in two to three weeks; however, the length of recovery is variable and depends on how early and how aggressively the disease is treated. About 3 percent of individuals with TSS die. The death rate with STSS can be as high as 30 to 70 percent.

Prevention

Women and girls who use tampons should always wash their hands before inserting a tampon and change the tampon every four to six hours. Skin **wounds** should be cleaned with an antiseptic and covered with a bandage.

Parental concerns

Although the risk of TSS is very low, parents may prefer that their daughters use pads rather than tampons when menstruating.

KEY TERMS

Shock—A medical emergency in which the organs and tissues of the body are not receiving an adequate flow of blood. This deprives the organs and tissues of oxygen and allows the build-up of waste products. Shock can be caused by certain diseases, serious injury, or blood loss.

Staphylococcal infection—Infection with one of several species of *Staphylococcus* bacteria. Staphylococcal infections can affect any part of the body and are characterized by the formation of abscesses. Also known popularly as a staph infection.

Streptococcus—Plural, streptococci. Any of several species of spherical bacteria that form pairs or chains. They cause a wide variety of infections including scarlet fever, tonsillitis, and pneumonia.

Toxin—A poisonous substance usually produced by a microorganism or plant.

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Tish Davidson, A.M.

Toxoplasmosis

Definition

Toxoplasmosis is an infectious disease caused by the one-celled parasitic organism *Toxoplasma gondii*. Although most individuals do not experience any symptoms, the disease can be very serious and even fatal in fetuses, newborns, and individuals with weakened immune systems.

Description

Toxoplasmosis is caused by a one-celled parasite *Toxoplasma gondii*. This parasite is found worldwide. It causes infections that can be either acute or chronic. In about 60 percent of healthy adults who become infected, the organism causes no symptoms (asymptomatic). Most of the remaining 40 percent experience mild, flu-like symptoms, low-grade **fever**, and fatigue that resolve without intervention in a few weeks. Once exposed, reinfection does not occur in healthy individuals. However, in immunocompromised individuals, such as those with HIV/AIDS, symptoms can be severe, life threatening, and recurring. *T. gondii* infection of a fetus or newborn can also cause severe neurological impairment, blindness, **mental retardation**, and death. When a fetus acquires the infection through its mother, this is called congenital toxoplasmosis.

Transmission

The organism that causes toxoplasmosis can be transmitted in four ways. The most common way is through contact with feces of an infected cat. Cats, the primary carriers of the organism, become infected by eating rodents and birds infected with *T. gondii*. Once ingested, the organism reproduces in the intestines of the cat, producing millions of eggs known as oocysts. These oocysts are excreted in cat feces daily for approximately two weeks. In the United States, approximately 50 percent of cats have been infected with *T. gondii*.

Oocysts are not capable of producing infection until approximately 24 hours after being excreted in warm climates and longer in cold climates. However, they remain infective in water or moist soil for about one year. Humans become infected when they come in contact with and accidentally ingest oocysts when changing cat litter, playing in contaminated sand, working in the garden or similar activities, or by eating unwashed vegetables and fruit irrigated with untreated water that has been contaminated with cat feces.

The second way humans become infected with *T. gondii* is through eating raw or undercooked meat. When cattle, sheep, or other livestock forage through areas contaminated with cat feces, these animals become carriers of the disease. The organism forms cysts in the muscle and brain of the livestock. When humans eat raw or undercooked infected meat, the walls of the cysts are broken down in the human digestive tract, and the individual becomes actively infected. The encysted

organism can be killed by freezing or cooking the meat well.

The only form of direct person-to-person transmission occurs from mother to fetus during pregnancy. This transmission occurs only if the mother is in the acute, or active, stage of infection when the organism is circulating in the mother's blood. It is estimated that about one third of women with active infections pass the infection along to their fetus. Women who have become infected six months or more before conception do not pass the infection on to their fetus, because the organism has become dormant (inactive) and formed thick-walled cysts in muscle and other tissues of the body. Reactivation of the infection in healthy individuals is extremely rare. Women who give birth to one infected child do not pass the infection to their fetus during subsequent pregnancies unless they are immunocompromised (for example, with **AIDS**) and the infection recurs.

Finally, individuals can also become infected through blood and organ transplant from an infected person.

Demographics

Men and women of all races are equally affected by *T. gondii,* however, except for immunocompromised individuals, the implications are more serious for women, as they can pass the infection on to their offspring. The rate of infection in the United States varies considerably with location. Studies have found that the infection rate in women of childbearing age ranges from 30 percent in Los Angeles to 3.3 percent in Denver. Varying sanitary conditions and culinary habits, such as eating raw meat, account for some of this variation. The rate of infection increases with the age of the individual. About 3,500 to 4,000 children are born in the United States each year with congenital toxoplasmosis. Outside the United States, fetal infection rates tend to be higher, although the number of babies born with congenital toxoplasmosis was as of 2004 declining worldwide.

Causes and symptoms

In fetuses, the severity of infection is dependent on the time of transmission. Fetuses who acquire the infection during the first trimester of pregnancy often are stillborn or die shortly after birth. Fetuses who acquire the infection late in pregnancy often show no symptoms when born.

Severe infections lead to seizure disorders, neurological disorders, abnormal muscle tone, deafness, partial or complete blindness caused by a condition called chorioretinitis, and mental retardation. These conditions may not be present at birth, especially if the infection occurred late in pregnancy. Vision deficits, especially, tend to show up later in life.

Young children can acquire toxoplasmosis in the same ways as adults. However, symptoms and complications when the disease is acquired after birth tend to be much milder than with congenital toxoplasmosis.

Children and adults with weakened immune systems have a high risk of developing serious symptoms, including cerebral toxoplasmosis, an inflammation of the brain (**encephalitis**), one-sided weakness or **numbness**, mood and personality changes, vision disturbances, **muscle spasms**, and severe headaches. If untreated, cerebral toxoplasmosis can lead to coma and death.

When to call the doctor

Women who believe they may have become infected shortly before conception or during pregnancy should call their doctor immediately. Treatment is possible during pregnancy. Symptoms in the newborn may be obvious during the newborn examination. If they are not, parents should consult their doctor if they feel their child has any neurological or vision complications or is not meeting appropriate developmental milestones.

Diagnosis

A diagnosis of toxoplasmosis is made based on clinical signs and supporting laboratory results, including visualization of the organism in body tissue or isolation in animals. Blood tests for toxoplasmosis are designed to detect increased amounts of a protein or antibody produced in response to infection with *T. gondii*. Antibody levels can be elevated for years, however, even when the disease is in a dormant state. **Amniocentesis** (sampling amniotic fluid) between 20 and 24 weeks of gestation can detect toxoplasmosis in the fetus.

Treatment

Most healthy individuals who contract toxoplasmosis do not require treatment, because the healthy immune system is able to control the disease. Symptoms are not usually present. Mild symptoms may be relieved by taking over-the-counter medications, such as **acetaminophen** (Tylenol) and ibuprofen (Motrin, Advil). **Sore throat** lozenges and rest may also ease the symptoms.

The benefits of treating women who contract toxoplasmosis during pregnancy almost always outweigh any risks involved. Treatment is with antibiotic and antimicrobial drugs. Transmission of toxoplasmosis from the

mother to the fetus may be prevented or reduced if the mother takes the antibiotic spiramycin. Later in a pregnancy, if the fetus has contracted the disease, treatment with the antibiotic pyrimethamine (Daraprim, Fansidar) and folinic acid (an active form of **folic acid**) may be effective. Babies born with toxoplasmosis who show symptoms of the disease may be treated with pyrimethamine, the sulfa drug sulfadiazine (Microsulfon), and folinic acid. Healthy children over the age of five usually do not require treatment. Infected individuals with weakened immune systems may require lifetime drug treatment to keep the infection from recurring.

Prognosis

The prognosis is poor when congenital toxoplasmosis is acquired during the first three months of pregnancy. Afflicted children die in infancy or suffer damage to their central nervous systems that can result in physical and mental retardation. Infection later in pregnancy often results in only mild symptoms, if any. The prognosis for acquired toxoplasmosis in adults with strong immune systems is excellent. The disease often disappears by itself after several weeks. However, the prognosis for immunodeficient patients is not as positive. These patients often relapse when treatment is stopped. The disease can be fatal to all immunocompromised patients, especially individuals with AIDS, and particularly if not treated.

Prevention

There are no drugs that can eliminate T. gondii cysts in animal or human tissues. Humans can reduce their risks of developing toxoplasmosis by practicing the following measures:

- freezing foods (to 10.4°F/–12°C) and cooking foods to an internal temperature of 152°F/67°C to kill the cysts
- practicing sanitary kitchen techniques, such as washing utensils and cutting boards that come into contact with raw meat
- keeping pregnant women and children away from household cats and cat litter
- disposing of cat feces daily because the oocysts do not become infective until after 24 hours
- helping cats to remain free of infection by feeding them dry, canned, or boiled food and by discouraging hunting and scavenging
- washing hands after outdoor activities involving soil contact and wearing gloves when gardening

KEY TERMS

Cyst—An abnormal sac or enclosed cavity in the body filled with liquid or partially solid material. Also refers to a protective, walled-off capsule in which an organism lies dormant.

Immunocompromised—A state in which the immune system is suppressed or not functioning properly.

Oocyst—A developmental stage of certain parasitic organisms, including those responsible for malaria and toxoplasmosis, in which the zygote of the organism is enclosed in a cyst.

Parental concerns

Fear of infection during pregnancy is the most common parental concern. When a fetus is found early in pregnancy to be infected, parents are faced with the decision of whether to continue the pregnancy given the likelihood of serious complications to the fetus.

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Tish Davidson, A.M.

Toys

Definition

Toys are physical items used in **play** by children.

Description

An estimated 2.6 billion toys, including electronic toys and **video games**, are sold in the United States each year, according to the Toy Industry Association, Inc. Toys can support cognitive growth, development of fine motor and **gross motor skills**, and improve problem solving and attention. Children may find extended periods of play with a toy, whether it was purchased in a store or found in the home (recycled plastic containers and empty spools of thread, for example). Most children will be happy to play with a few favorite toys—the size of the toy inventory is not critical to successful play. Parents and others who choose toys for children should take into account the following characteristics of the child for whom the toy is intended. These include the age and developmental stage, his or her interests, ease of use of the toy, the necessity for adult supervision, the presence of younger siblings for whom the toy could pose a hazard, and whether the toy is designed for independent play or group play.

While computer and video game sales have more than tripled in the past decade, to nearly $240 million last year from $65 million in 1996, toys were expected to have their third straight year of slight decline in 2004. Children are still riding bikes, sipping from play tea sets, and enjoying some of the same toys their parents did, including building blocks, erector sets, and Lincoln Logs. But the pressures bearing down on traditional toys are many. Kids are growing up faster and putting down Barbie dolls and G.I. Joe action figures at an earlier age, increasingly smitten by the grownup images of young celebrities such as high-profile athletes, movie and television stars, and recording artists. The prime audience for toys has shrunk as the children of the immense baby boom generation have grown into teenagers and beyond. And as in other industries, giant retailers have taken sales from specialized toy chains and squeezed some of the incentive to devise the next great toy.

Toy labeling

The U.S. Consumer Product Safety Commission has developed guidelines for age grading of toys and related products. Most toy manufacturers use these guidelines in labeling toys and games for age-appropriateness. Manufacturers also consider recommendations of experts in child development regarding the stages of physical, emotional, and intellectual development. Four main criteria are considered in establishing age guidelines:

- Physical skills: Can the child manipulate and play with the features of the toy as it was designed?
- Understanding: Can the child understand how to use the toy?
- Interest: Is the toy of interest to a child of a particular age?
- Safety: Is the toy safe for a child at this particular stage?

The Consumer Product Safety Commission (CPSC) has established a number of regulations related to toy safety. These are published by the American Society for Testing and Measurement (ASTM) under the safety standard known as ASTM F963. This standard is voluntary, but the majority of U.S. toy manufacturers comply with its guidelines. In fact, many incorporate a message about the toy's compliance with ASTM F963 on the toy packaging.

Infancy and toddlerhood

Toy manufacturers consider the size of toy parts—which are likely to be put into the mouth by an older infant or toddler—in designing toys. Anyone purchasing a toy for the youngest children must take the **choking** hazard seriously and make appropriate selections. When a new toy is brought into the home or child-care setting, all wrapping material should be promptly discarded. Plastic wrapping in particular may pose a suffocation hazard to the youngest children. The U.S. government maintains statistics on toy-related injuries and deaths. Many accidents involving toys are not caused by the toy itself; for example, a child may trip over a toy that was not put away after play. When an unsafe toy reaches the marketplace, U.S. government inspectors may discover it and order its recall; additionally, vigilant parents and caregivers can observations about toy safety to the CPSC. Manufacturers routinely cooperate with the CPSC in recalling products that are deemed unsafe or dangerous.

In 2002, the medical journal *Clinical Reference Systems* issued pediatric advisories on age-appropriate toys. Some of the recommendations follow. Suggested play things for infants include: interesting objects hung in view, such as brightly colored mobiles, crib decals, and colorful wall posters; sturdy rattles; large plastic rings; soft toys for throwing; colorful balls; light plastic blocks; cloth cubes; music boxes; teething toys; floating bath animals; washable squeak toys; nests of hollow blocks; and rough-smooth touching books; washable cloth picture books; and sturdy, colorful picture books.

Suggested toys for toddlers include: pyramid rings; large nesting blocks; large and small colored building blocks; cuddly stuffed animals; large, soft balls; washable, unbreakable dolls; push-pull toys with rounded handles; simple musical instruments; sand box and sand toys; water toys; transportation toys (trucks, cars, trains, and airplanes); objects to imitate adults such as plastic garden tools, toy telephones, and toy dishes or pots; and sturdy, colorful picture books.

Preschool and school age

U.S. law requires that toys and games for young children (ages three to six) carry a warning about choking hazards. If the toy or game includes small parts, marbles, or balloons, it must be marked that it is not appropriate for children under the age of three. Beyond toddlerhood, children begin to develop their own ideas about play activities and the toys that they want. They will be influenced by what they see advertised on television and by their peers. Toy fads and television show tie-ins can be powerfully persuasive to children. Parents may experience their first opportunities to teach about **peer pressure** and independent decision-making over toy requests. Toys should be selected to stimulate play and related cognitive and physical development; fad toys are less likely to sustain play activity and support development beyond the fad stage.

Toys and play items appropriate for preschool-age children include: large and small transportation toys; cuddly toy animals; simple musical instruments; farm and zoo animal sets; miniature circus; hospital, police, and fire station sets; bean bags; large balls; art materials such as paints, modeling clay, paste, and colored paper; wagons; tricycles; crawl-through play equipment; simple construction sets; nursery rhyme books; humorous and playful books; activity books; and books about familiar people and places.

For early school-age children, six to nine years old, suggested toys include: construction sets; art materials such as crayons, chalk, paint, modeling clay, and simple weaving materials; chalk, Velcro, or flannel boards; small bicycles; wagons; jump ropes; simple board games; playhouses; puzzles; kites; globes or planetarium sets; aquariums; terrariums; and books about jokes, riddles, tongue twisters, animals, insects, birds, reptiles, and children from other lands and cultures.

Appropriate toys for older school-age children, nine to 12 years, include: croquet, badminton, and shuffleboard sets; **sports** equipment (baseball, basketball, soccer, football, and tennis); skates and skateboards; aquariums and terrariums; craft sets; hobby sets; electric trains; radio-controlled vehicles; model kits; board games; microscopes; binoculars; compact disc players; camping and backpacking equipment; and books about adventure, science fiction, fantasy, science topics, simple biographies, and jokes, puzzles, riddles, and tongue twisters.

Parental concerns

Violent toys and video games

In addition to product safety, one of the biggest concerns of parents is the growing trend towards toys and video games that promote violence, crime, and war. In 2002, a national department store chain published an advertisement on its web site for a toy called "Forward Command Post" that featured an American soldier standing in a bombed-out house. It's an example of the growing collaboration, in recent years, between the toy and entertainment industry and the U.S. military, according to the activist group Worldwatch Institute. Video games with themes of terrorism and war in Middle-Eastern settings are selling well. In video games, kids can experience virtual combat, are exposed to exploding virtual body parts, and practice committing murder and theft to win games.

Traditionally, boys' and girls' toys have often been contrasted as being "rough-and-tumble" versus "nurturing." But that distinction may be disappearing, not only for healthy reasons of waning gender stereotyping, but for more questionable reasons such as the growing tolerance of—or obliviousness to—aggression and hostility in play by both sexes, according to an article in the May-June 2003 issue of *World Watch*, the institute's official publication.

Many parents are concerned about the growing number and level of acceptance of toys and video games that promote violence and war. However, not everyone agrees on what constitutes a violent toy. Most parents agree guns are symbols of violence. But at what point is the line drawn between a child perceiving a play gun as a toy or a device of violence? Many might agree a BB gun is violent since it can kill and wound small animals and birds and injure humans. But what about a squirt gun that shots water or a Nerf gun that shoots foam balls? To help answer these questions, Daphne White, founder of the Lion & Lamb Project, published a list of guidelines in the November-December 2004 issue of *Mothering* magazine. Lion & Lamb is an organization founded in 1995 to stop the marketing of violent toys and entertainment to children.

According to White's guidelines, violent toys and video games:

Age-appropriate toys

Age range	Toys
Birth to two months	Activity centers to look at and listen to; mobiles over cribs
Two to four months	Rattles, teethers, activity centers to hit or kick while on back.
Four to six months	Soft books, roly poly toys
Seven to nine months	Nesting cups, pop-up toys
Ten to twelve months	Push and pull toys, large blocks, board books, toys that require hand manipulation to "make something happen"
Thirteen to fifteen months	Toy telephone, walk-behind toys like doll stroller, soft dolls and animals, balls
Sixteen to eighteen months	Simple toy musical instruments, playing with water or sand, shape sorters
Nineteen months to two years	Rocking horse, easy puzzles, make-believe toys (plastic houses and people, toy cars and trucks, play food and dishes), crayons and paper
Two to three years	Tricycle, toy basketball hoop and balls, toy trains with tracks, dolls with bottles and other equipment, toy kitchen equipment, coloring books and crayons, books
Three to four years	Simple crafts (including scissors, glue, and paper), beginning board games, toys for imaginative play
Four to five years	Simple sports equipment, books, board and card games, computer games, collections, building blocks
Five to six years	Small blocks and building sets, art supplies, activity books, beginning reader books, games
Six years and up	Music, books, games, sports equipment. By this age, kids may get particular about their toys according to what is popular with their friends

(Table by GGS Information Services.)

- Promote violence and aggression as the best way to settle disputes.
- Depict violent actions as fun, harmless, and "cool."
- Encourage children to act out aggressive scenarios.
- Foster aggressive competition.
- Depend on "enemies" that must be "destroyed."

When to call the doctor

An inability to play with or lack of interest in toys at an early age may indicate a developmental problem in such areas as gross and **fine motor skills**. If this is suspected, a pediatrician, psychologist, or other specialist should be consulted.

See also Cognitive development; Fine motor skills; Gross motor skills.

KEY TERMS

Cognitive—The ability (or lack of) to think, learn, and memorize.

Fine motor skill—The abilities required to control the smaller muscles of the body for writing, playing an instrument, artistic expression and craft work. The muscles required to perform fine motor skills are generally found in the hands, feet and head.

Gross motor skills—The abilities required to control the large muscles of the body for walking, running, sitting, crawling, and other activities. The muscles required to perform gross motor skills are generally found in the arms, legs, back, abdomen and torso.

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Ken R. Wells

KEY TERMS

Endoscopy—Visual examination of an organ or body cavity using an endoscope, a thin, tubular instrument containing a camera and light source. Many endoscopes also allow the retrieval of a small sample (biopsy) of the area being examined, in order to more closely view the tissue under a microscope.

Esophageal atresia—Blockage or closure of the esophagus, the tube leading from the mouth to the stomach.

Gastrostomy tube—A tube that is inserted through a small incision in the abdominal wall and that extends through the stomach wall into the stomach for the purpose of introducing parenteral feedings. Also called a gastric tube, gastrointestinal tube, or stomach tube.

Tracheoesophageal fistula

Definition

Tracheoesophageal fistula (TEF) is a birth defect in which the trachea is connected to the esophagus. In most cases, the esophagus is discontinuous (an **esophageal atresia**), causing immediate feeding difficulties.

Description

The trachea, or windpipe, carries air to the lungs. The esophagus carries food to the stomach. Sometimes during development these two tubes do not separate completely but remain connected by a short passage. When this happens, air enters the gastrointestinal system, causing the bowels to distend, and mucus is breathed into the lungs causing aspiration **pneumonia** and breathing problems.

There are three main types of TEF. In 85 to 90 percent of tracheoesophageal fistulas, the top part of the esophagus ends in a blind sac, and the lower part inserts into the trachea. In the second type, the upper part of the esophagus is connected directly to the trachea, while the lower part ends in a pouch. In a rare type of fistula called an H type, both the esophagus and trachea are complete, but they are connected by a small passageway. This is the most difficult type of tracheoesophageal fistula to diagnose, because both eating and breathing are possible. TEFs often occur in babies with additional birth defects.

Demographics

TEFs occur in about one of every 3,000 live births. They are slightly more common in boys than in girls. Some studies suggest that the occurrence of TEFs increases with the age of the mother.

Causes and symptoms

Tracheoesophageal fistulas arise as a developmental abnormality. At birth, the infant has difficulty swallowing. Eating produces severe coughing spells that interfere with breathing. Aspiration pneumonia can develop from fluid breathed into the lungs.

Small H type fistulas may go undiagnosed until later in life. Symptoms of an H type fistula include frequent pulmonary infections and bouts of abdominal bloating.

When to call the doctor

TEFs are normally diagnosed within hours of birth, because feeding and breathing problems are immediately apparent. Some H type defects are not detected until later in life.

Diagnosis

Diagnosis that the esophagus is interrupted is confirmed by the inability to insert a nasogastric suction tube into the stomach. The exact type and location of the fistula can be determined using a radiopaque catheter, which allows pictures to be taken of the esophagus. X rays may show air in the bowels. Endoscopy often fails to locate the fistula if it is small.

Treatment

Babies with all but H type fistulas are unlikely to survive without surgical separation and repair of the trachea and the esophagus. Surgery is usually done at a hospital that has special facilities for treating seriously ill newborns. However, surgery cannot always be performed immediately because of **prematurity**, the presence of other birth defects, or complications from aspiration pneumonia.

While awaiting surgery, the infant's condition is stabilized. Preoperative care concentrates on avoiding aspiration pneumonia and includes the following:

- elevating the head to avoid reflux and aspiration of the stomach contents
- using a suction catheter to continuously remove mucus and saliva that could be inhaled
- when necessary, placement of a gastrostomy tube for feeding
- withholding feeding by mouth

When surgery is performed, the esophagus is reconnected to make it continuous and separate from the trachea. If the two ends of the esophagus are too far apart to be reattached, a piece of tissue from the large intestine is used to join the parts.

Prognosis

The survival rate of infants with tracheoesophageal fistulas improved dramatically toward the end of the twentieth century. In uncomplicated cases, the survival rate is close to 100 percent. However, often babies with TEFs have other birth defects that limit their recovery.

When the esophagus is successfully separated and reattached, many infants have difficulty swallowing, because the contractility of the esophagus is impaired. Infants may also have problems with gastroesophageal reflux, in which the acidic contents of the stomach back up into the bottom of the esophagus and cause ulcers and scarring. Long-term follow-up, however, finds that 80 to 90 percent of children who have repaired TEFs as infants eat normally by the time they are in elementary school. As more individuals with corrected TEFs reach adulthood, there is some evidence that suggests they are more susceptible to esophageal cancers. However, as of 2004, there was not enough data to confirm these findings.

Prevention

Tracheoesophageal fistulas are defects in development of the fetus that cannot be prevented.

Parental concerns

Parents often worry about the effect a TEF may have on their child's later ability to eat and participate in normal activities such as **sports**. In the absence of other birth defects, almost all children have no restrictions on their eating and activities by the time they start school.

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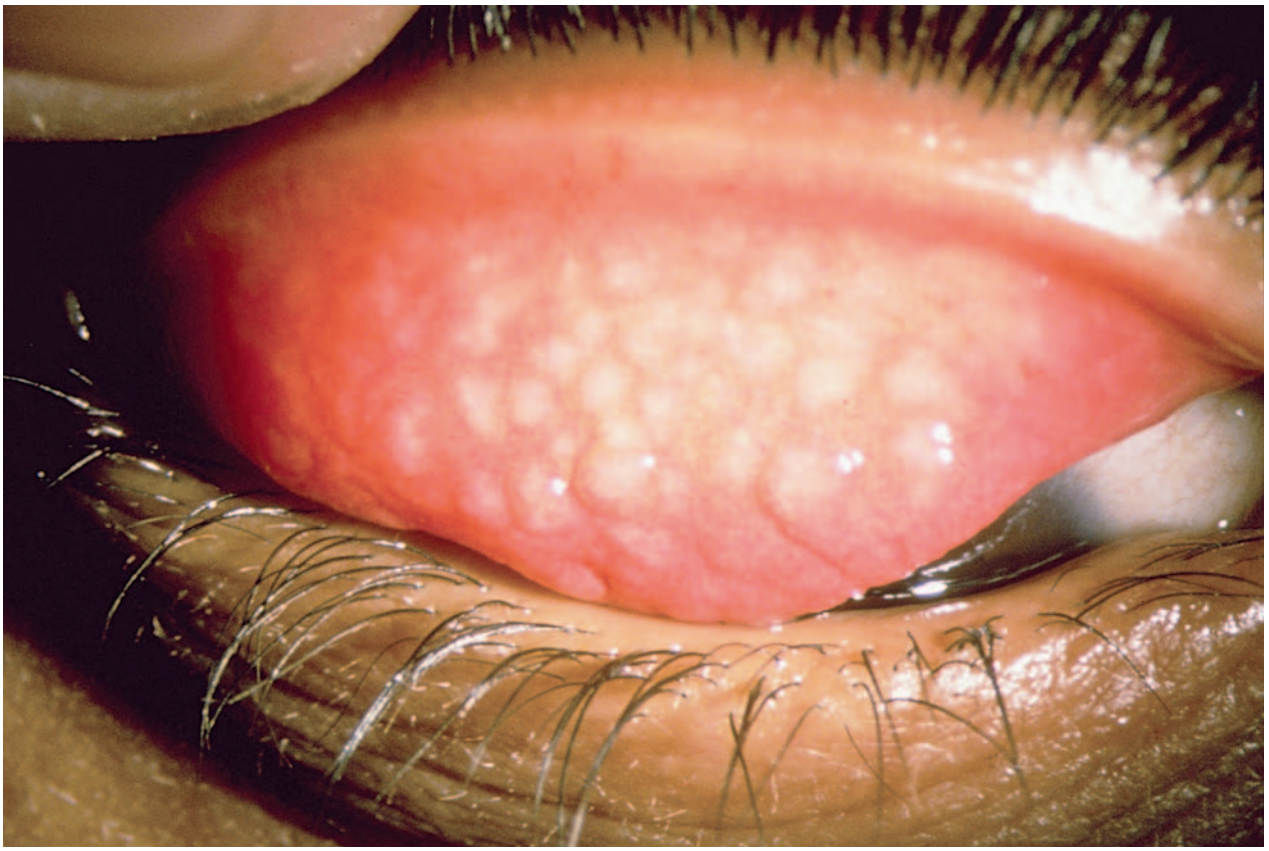
Trachoma

Definition

Trachoma, also called granular **conjunctivitis** or Egyptian ophthalmia, is a contagious, chronic inflammation of the mucous membranes of the eyes, caused by the bacterium *Chlamydia trachomatis*. It is characterized by swelling of the eyelids, sensitivity to light, and eventual scarring of the conjunctiva and cornea of the eye.

Description

Trachoma is a disease associated with poverty and unhygienic conditions. It is most common in hot, dry, dusty climates in the developing world where water is



A close-up of a human eye with trachoma. Trachoma is caused by *Chlamydia trachomatis* and commonly results in blindness if left untreated. *(Custom Medical Stock Photo Inc.)*

scarce and sanitation is poor. Trachoma is the most common infectious cause of blindness in the world. It has two stages. The first stage is active infection of the conjunctiva by the bacterium *C. trachomatis*. The conjunctiva is the clear mucous membrane that lines the inside of the eyelid and covers the white part (sclera) of the eye. This stage is highly contagious.

Acquiring trachoma does not provide immunity against re-infection, so repeat infections are the norm in many communities where the disease circulates continuously among **family** members. The frequency of active infection peaks in children ages three to five. In some communities, as many as 90 percent of children under age five are actively infected.

The second stage involves damage to the cornea, the transparent covering of the front of the eye. After repeated infections, the eyelids swell and the eyelashes begin to turn inward so that they scratch the cornea every time the individual blinks. This scratching is painful, and it scars the cornea, eventually resulting in the cornea becoming opaque. Individuals are often blind by middle age. Repeated, extended, untreated periods of infection are required for blindness to occur. An occasional, treated infection does not result in blindness.

Transmission

C. trachomatis is spread through direct contact. Infected young children serve as a reservoir of infection. The bacteria are then transmitted by close physical contact with family members and other caregivers. The bacteria are also spread through shared blankets, pillows, and towels. The bazaar fly *Musca sorbens* lays its eggs in human feces that can be contaminated with trachoma bacteria. These flies pick up bacteria on their bodies and can transmit them to humans.

Certain conditions promote the spread of trachoma bacteria. These include:

- poor personal hygiene
- poor body waste and trash disposal
- insufficient water supply for washing
- shared sleeping space
- close association with domestic animals

KEY TERMS

Conjunctivitis—Inflammation of the conjunctiva, the mucous membrane covering the white part of the eye (sclera) and lining the inside of the eyelids also called pinkeye.

Cornea—The clear, dome-shaped outer covering of the eye that lies in front of the iris and pupil. The cornea lets light into the eye.

Demographics

Trachoma is widespread and present in a high percentage of the population in many parts of Africa, Iraq, Afghanistan, Burma, Thailand, and Viet Nam. Pockets of high trachoma infection also exist in southern Mexico, eastern Brazil, Ecuador, North Africa, India, China, Siberia, Indonesia, New Guinea, Borneo, and in Aboriginal communities in central Australia. Although trachoma is rare in developed countries, it is occasionally found in the United States in some Native American communities and in parts of Appalachia.

The greatest risk for contracting trachoma is having a family member with the disease. Although the disease shows no gender preference, two to three times more women eventually become blind than men, probably because they are the primary caretakers of small children who are infected. The active stage of the disease is most prevalent in children ages three to five. Blindness is most common in middle age. The World Health Organization (WHO) estimates that as of the early 2000s, between 360 and 500 million people are affected by trachoma worldwide and that six million people are blind because of the disease. In some heavily infected areas, up to 25 percent of the population becomes blind from this infection.

Causes and symptoms

The early symptoms of trachoma include the development of follicles (small sacs) on the conjunctivae of the upper eyelids; **pain**; swollen eyelids; discharge; tearing; and sensitivity to light. If the infection is not treated, the follicles develop into large yellow or gray pimples, and small blood vessels develop inside the cornea. In most cases, both eyes are infected. The incubation period is about one week.

Repeated infections eventually lead to contraction and turning-in of the eyelids. The eyelashes then scratch the corneas and conjunctivae, every time the individual blinks. This scratching leads to scarring of the cornea, eventual blockage of the tear ducts, and blindness.

When to call the doctor

U.S. parents should call the doctor if they notice any discomfort or discharge from their child's eye, especially if they have recently traveled in areas where trachoma is common.

Diagnosis

Diagnosis is based on a combination of the patient's history (especially living or traveling in areas with high rates of trachoma) and examination of the eyes. The doctor looks for the presence of follicles or scarring. In developed countries where laboratory facilities are available, the doctor takes a small sample of cells from the child's conjunctivae and examines it, following a procedure called Giemsa staining, to confirm the diagnosis. In underdeveloped countries where medical resources are scarce, diagnosis is made based on an examination only.

Treatment

The preferred treatment is the oral antibiotic azithromycin (Zithromax). This medicine has replaced treatment with other **antibiotics** (usually **tetracyclines**), because only a single dose of azithromycin is required to clear the infection. Oral single dose treatment increases compliance. Everyone in the family should be treated at the same time, whether they show clinical signs of the disease or not, because transmission among family members is so common.

Individuals with complications from untreated or repeated infections require surgery. Surgery can be used for corneal transplantation or to correct eyelid deformities. It does not, however, prevent re-infection.

Prognosis

The prognosis for full recovery is excellent if the individual is treated promptly. If the infection has progressed to the stage of follicle development, prevention of blindness depends on the size of the follicles, the presence of additional bacterial infections, and the development of scarring. The longer the period of infection, the greater the risk of corneal scarring and blindness.

Prevention

Trachoma is a preventable disease. Prevention depends upon good hygiene and public health. The

WHO has developed a program called SAFE, which aims to prevent blindness caused by trachoma. The elements of the program are surgery, antibiotic treatment, facial cleansing and improved personal hygiene, and environmental improvements. Despite this prevention program, permanent gains in controlling trachoma have been elusive.

Parental concerns

In the United States, parents should check with the Centers for Disease Control (available online at <www.cdc.gov>) for advisories about the prevalence of trachoma if they are planning to travel with their children to underdeveloped countries.

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Sight Savers International. c/o Chapel & York, PMB #293, 601 Pennsylvania Avenue, NW, Suite 900, South Building, Washington, DC 20004. Web site: <www.sightsavers.org>.

Tish Davidson, A.M.

Traction *see* **Immobilization**

Transposition of the great arteries

Definition

Transposition of the great arteries (TGA) is a birth defect causing a fatal condition in which there is a reversal, or switch, in the primary connections of the two main (great) blood vessels to the heart, the aorta and pulmonary artery.

Description

There are two great arteries that transport blood away from the heart, the pulmonary artery and the aorta. Normally, the pulmonary artery carries blood from the right ventricle to the lungs. The aorta carries blood from the left ventricle to the vessels of the rest of the body.

Ordinarily, blood returning to the heart is depleted in oxygen. It goes first to the right atrium of the heart and then to the right ventricle where it is pumped to the lungs. While in the lungs, the blood picks up more oxygen. After the lungs, the blood flows to the left atrium, then the left ventricle pumps the blood out through the aorta to the rest of the body, thereby supplying the body with oxygenated blood.

In children with transposition of the great arteries, the connection of the two great arteries is reversed. This condition causes oxygen depleted blood to be circulated to the body because the aorta is connected to the right ventricle. Blood returning to the heart goes to the right atrium and ventricle, and then it goes into the aorta for distribution throughout the body instead of to the lungs to be oxygenated. At the same time, blood in the lungs goes to the left atrium, the left ventricle, but then back to the lungs rather than going to the body because the pulmonary artery is connected to the left ventricle. The result is that highly oxygenated blood keeps recycling through the lungs, while oxygen-depleted blood recycles through the body without going through the lungs to reoxygenate. The body cannot survive without oxygenated blood.

This condition occurs during the fetal development and must be treated promptly after birth if the newborn is to survive. The newborn can survive for a few days while the foramen ovale, a small hole in the septum that separates the two atria, is open, allowing some oxygenated blood to escape and mix into the blood that is being pumped throughout the body. However, within a few days after birth, the foramen ovale normally closes, and no oxygenated blood is available for the body.

Demographics

Transposition of the great arteries affects 20 to 30 of every 100,000 live births each year. It is the most common reason for cyanotic heart disease in newborns accounting for 5 to 7 percent of all infants with **congenital heart disease**. Transposition of the great arteries is most often an isolated defect and is not associated with other congenital syndromes. It affects males more than females with 60 to 70 percent of all cases occurring in males. It does not affect any race or nationality more than another.

Causes and symptoms

Transposition of the great arteries is a birth defect that occurs during fetal development. There is no identifiable disease or cause. The main symptom is a cyanotic or blue baby appearance, caused by a general lack of oxygen in the body's tissues.

Diagnosis

Diagnosis is made immediately after birth, when it is observed that the newborn has a bluish color. A definite diagnosis is made by x ray, electrocardiography (ECG), and echocardiography.

Treatment

Transposition of the great arteries may be treated by the use of medications called prostaglandins which keep the ductus arteriousus open. A procedure called a cardiac catheterization can then be performed during which a small thin tube (catheter) with a balloon tip, may be used to enlarge the opening between the two atria until surgery can be performed. However, both procedures are temporary treatments that help prolong the infant's life, in some cases allowing him or her to gain strength, until surgery can be performed. The only permanent solution for this condition is open-heart surgery. In transposition of the great arteries repair surgery, the infant's heart is stopped, and blood is circulated through the body using a mechanical heart-lung machine. The two great arteries are reconnected to their proper destination. This correction restores the normal blood flow pattern. The coronary arteries are also reconnected, so that they can supply blood to the heart itself.

Prognosis

Left untreated, this disease is fatal within the first weeks of life. After surgical repair, the survival rate is 90 percent, and most individuals grow and develop normally.

KEY TERMS

Aorta—The main artery located above the heart that pumps oxygenated blood out into the body. The aorta is the largest artery in the body.

Atrial—Referring to the upper chambers of the heart.

Cyanosis—A bluish tinge to the skin that can occur when the blood oxygen level drops too low.

Echocardiography—A non-invasive technique, using ultrasound waves, used to look at the various structures and functions of the heart.

Electrocardiagram (ECG, EKG)—A record of the electrical activity of the heart, with each wave being labeled as P, Q, R, S, and T waves. It is often used in the diagnosis of cases of abnormal cardiac rhythm and myocardial damage.

Foramen ovale—The foramen ovale is a fetal cardiac structure that allows the blood in both upper chambers (atria) of the heart to mix. After birth, the pressure rises in the left atrium pushing this opening closed, allowing the heart to function in a two-sided fashion: the right side carries the unoxygenated blood to the lungs, and the left side pumps the oxygenated blood out into the body.

Oxygenate—To supply with oxygen.

Pulmonary artery—An artery that carries blood from the heart to the lungs.

Ventricles—The lower pumping chambers of the heart. The ventricles push blood to the lungs and the rest of the body.

Prevention

Because there is no identifiable cause, there is no way to prevent this condition.

Parental concerns

Transposition of the great arteries is a complex congenital malformation; however, open heart surgery is highly effective in correcting it. Most children have an excellent outcome and a normal healthy life. For most children, activity level, appetite, and growth eventually return to normal. Open heart surgery to repair transposi-

tion of the great arteries is usually performed within the first days or weeks of life.

After the child's heart surgery parents should follow all instructions given by the healthcare team. Most children continue to be seen by a team of doctors including the pediatrician, cardiologist, and pediatric cardiac surgeon.

When to call the doctor

Following open heart surgery, parents should call the doctor if any of the following occurs:

- fever of 101.5 (38.6°C) or higher
- swelling or puffiness around the child's eyes, arms, or legs
- redness or swelling, cloudy yellow drainage, or an opening at the incision site
- rapid breathing
- increased fatigue or tiredness
- dry **cough** that was not present before surgery
- decreased appetite or refusal to eat
- increased pain

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Traumatic amputations

Definition

Traumatic amputation is the accidental severing of some or all of a body part.

Description

Traumatic amputation most often affects limbs and appendages such as the arms, ears, feet, fingers, hands, legs, and nose. Amputations may be partial (some tissue connects the amputated part to the body) or complete (the amputated part is completely severed from the body).

Demographics

Trauma is the second leading cause of amputation in the United States. About 30,000 traumatic amputations occur in United States each year. Four of every five traumatic amputation victims are male, and most of them are between the ages of 15 and 30.

Causes and symptoms

Some of the more common causes of pediatric traumatic amputations are accidents with lawnmowers, automobiles, motorcycles, power tools, and farm equipment. Amputations may be caused by sharp objects such as knives or blades ("guillotine" amputation) or by heavy objects or mechanisms (crushing amputation). Crushing injuries are the more common cause of traumatic amputations.

Blood loss may be massive or minimal, depending on the nature of the injury and the site of the amputation. Patients who lose little blood and have less severe injuries sometimes feel more **pain** than patients who bleed heavily and whose injuries are life-threatening.

Phantom pain

About 80 percent of all amputees over the age of four experience tingling, **itching**, **numbness**, or pain in the place where the amputated part used to be. About 30 percent of amputees experience a sensation of the amputated part "telescoping" or shrinking into the viable part of the limb. Phantom sensations may begin immediately after the amputation, or they may develop months or years later. They often occur after an injury to the site of the amputation.

These intermittent feelings may have the following characteristics:

- occur frequently or only once in a while
- be mild or intense
- last for a few minutes or several hours
- help patients adjust more readily to an artificial limb (prosthesis)

When to call the doctor

A partial or complete amputation is a medical emergency and as such, the affected child (and amputated body part, if possible) should be transported to an emergency center immediately.

Diagnosis

When the patient and the amputated part(s) reach the hospital, a physician will assess the probability that the severed tissue can be successfully reattached (called replantation). The Mangled Extremity Severity Score (MESS) is a diagnostic tool used to assess the probability of successful replantation and assigns numerical values to such factors as body temperature, circulation, numbness, paralysis, tissue health, and the patient's age and general health. The total score is doubled if blood supply to the amputated part has been absent or diminished for more than six hours.

A general, emergency, or orthopedic surgeon makes the final determination about whether surgery should be performed to reattach the amputated part(s). The surgeon also considers the wishes and lifestyle of the child and parents. Additional concerns are how and to what extent the amputation will affect the child's quality of life and ability to perform everyday activities.

Treatment

First aid or emergency care given immediately after the amputation has a critical impact on both the physicians' ability to salvage and reattach the severed part(s) and the patient's ability to regain feeling and function. Muscle tissue dies quickly, but a well-preserved part can be successfully reattached as much as 24 hours after the amputation occurs. Tissue that has not been preserved will not survive for more than six hours.

Initial response

The most important steps to take when a traumatic amputation occurs are:

- Contact the nearest emergency services provider, clearly describe what has happened, and follow any instructions given.
- Make sure the victim can breathe; administer CPR if necessary.
- Control bleeding using direct pressure; minimize or avoid contact with blood and other body fluids.
- Patients should not be moved if back, head, leg, or neck injuries are suspected or if motion causes pain. If none is found by a trained professional, position the victim flat, with the feet raised 12 inches above the surface.
- Cover the victim with a coat or blanket to prevent shock.

The injured site should be cleansed with a sterile solution and wrapped in a clean towel or other thick material that will protect the wound from further injury. Tissue that is still attached to the body should not be forced back into place. If it cannot be gently replaced, it should be held in its normal position and supported until additional care is available.

Saving the patient's life is always more important than recovering the amputated part(s). Transporting the patient to a hospital or emergency center should never be delayed until missing pieces are located.

Preserving tissue

No amputated body part is too small to be salvaged. Debris or other contaminating material should be removed, but the tissue should not be allowed to get wet. An amputated body part should be wrapped in bandages, towels, or other clean, protective material and sealed in a plastic bag. Placing the sealed bag in a cooler or in a container that is inside a second container filled with cold water or ice will help prevent tissue deterioration.

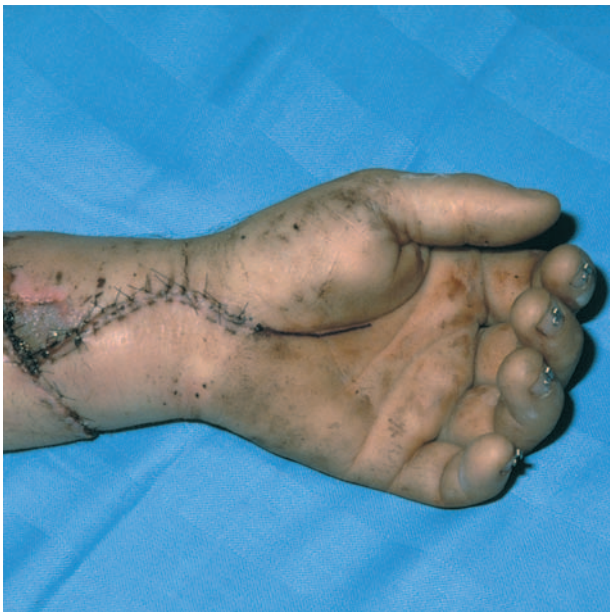
Replantation

A number of factors influence whether an amputated part can be successfully reattached. These include:

- age of the patient (younger patients tend to heal better and faster)
- location of amputation (replantations of the upper extremities are more successful than those of the lower extremities)
- type of wound (sharp **wounds** are repaired more successfully than crushing injuries)
- health of the patient (e.g. if he or she is able to withstand prolonged surgery)
- amount of contamination to the wound (a grossly contaminated part has a much lower chance of successfully being reattached)
- length of time the amputated part was detached from the body (chance of successful replantation decreases after six hours)

Post-care

Techniques such as biofeedback, cognitive-behavioral **pain management**, hypnosis, acupuncture, ultra-



This man's hand was surgically reattached following a traumatic amputation. *(Photograph by Michael English. Custom Medical Stock Photo, Inc.)*

KEY TERMS

Phantom pain—Pain, tingling, itching, or numbness in the place where the amputated part used to be.

Pulmonary embolism—Blockage of an artery in the lungs by foreign matter such as fat, tumor tissue, or a clot originating from a vein. A pulmonary embolism can be a very serious, and in some cases fatal, condition.

Shock—A medical emergency in which the organs and tissues of the body are not receiving an adequate flow of blood. This deprives the organs and tissues of oxygen and allows the build-up of waste products. Shock can be caused by certain diseases, serious injury, or blood loss.

sound, and physical therapy have all been used to treat post-amputation and phantom pain.

Nutritional concerns

Proper **nutrition** is essential to optimize healing after an amputation or reattachment surgery. A well-balanced diet rich in **vitamins** and with adequate caloric value is recommended to promote healing.

Prognosis

Possible complications of traumatic amputation include:

- excessive bleeding and shock
- infection
- muscle shortening
- pulmonary embolism
- death

Improved medical and surgical care and rehabilitation have improved the long-term outlook for such patients. Children tend to heal faster than adults and adapt more quickly to disability.

Prevention

The best way to prevent traumatic amputation is to observe precautions such as using seat belts and obeying speed limits and other traffic regulations. It is important to take special precautions when using potentially dangerous equipment and make sure machinery is turned off and disconnected before attempting to service or repair it. Appropriate protective clothing should be worn at all times. Children should be closely monitored when in the vicinity of lawnmowers, power tools, farm equipment, or other machinery that can cause serious injury.

Parental concerns

Parents of child amputees are faced with difficult decisions such as whether to get a limb prosthesis for their child and how to handle issues with negative body image. Parents will be encouraged to work with their child's rehabilitation team, which may include physicians, prosthetists, physical therapists, occupational therapists, psychologists, and/or teachers, to help the child adjust to the traumatic loss of a limb. Tools such as amputee dolls may be helpful in explaining how a prosthetic limb will be worn and to encourage positive body image.

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National Amputation Foundation. 40 Church St., Malverne, NY 11565. Web site: <www.nationalamputation.org>.

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Treponema infection *see* **Pinta**

Trichotillomania

Definition

Trichotillomania is a psychiatric condition in which an individual has an uncontrollable desire to pull out his own body hair. It is generally considered to be an impulse control disorder but is sometimes classified as either a subtype or variant of **obsessive-compulsive disorder** (OCD).

Description

Trichotillomania is the most common cause of hair loss in children. First described in 1889, trichotillomania is a psychiatric disorder, the result of which is **alopecia** or hair loss, caused by repeated pulling of one's hair from, most often the head, followed by the eyelashes and eye brows. But the hair of any part of the body may be pulled and multiple sites may be involved. The individual with trichotillomania will have bald spots on the head or missing eyelashes or eyebrows.

There is an immense amount of embarrassment and denial associated with trichotillomania. It is common for individuals with this disorder to deny their behavior and attempt to hide their hair loss. The hair loss may be disguised by wearing wigs, hats, scarves or hair clips, or by applying make-up or even by tattooing. The act of hair pulling is a private one. Rarely does the hair pulling occur in the presence of another, except for close **family** members. Because of this fact, social alienation is common in trichotillomania.

The hair pulling may occur either when the individual is relaxed or under stress. For some individuals with trichotillomania, certain situations, such as watching TV, lying in bed, or talking on the phone, will trigger the behavior. The individual either may focus intensely on the hair pulling or the pulling may be done unconsciously. Immediately before pulling hair, the individual with trichotillomania feels a mounting tension. This tension is relieved as a hair root is successfully pulled. Since a tingling sensation is felt upon successfully pulling a hair follicle completely from its root, a neurodermatologic connection may reinforce hair pulling as a means of tension relief. When the hair root remains intact and the hair shaft is broken, this sensation is not felt and the patient may repetitively pull hairs until successful. After pulling the hair, patient may carefully examine the hair root, and the hair bulb may be rubbed along the lips for further stimulation. The hairs may be ingested by some patients.

The amount of time each day that the patient engages in hair pulling may consist of either several brief periods, or a longer intense period. The typical trichotillomania patient will spend one to three hours daily pulling hairs. The urge to pull can be so intense that the individual with trichotillomania cannot think of anything except hair pulling. Thus, social life and work production often suffer with trichotillomania.

The act of hair pulling in trichotillomania is often ritualistic. The necessary implements, such as tweezers, are collected, the location where this is to be performed is determined, the preferred texture or color to be pulled may be planned as well as disposal of the hairs.

Rarely, the individual with trichotillomania may attempt to pull the hairs of others. The hairs of a pet or doll or the fibers of an inanimate object, such as sweater, may be pulled as well. In addition to hair pulling, the hair may be bitten off or twisted or twirled.

Co-existing psychiatric diagnoses such as **anxiety**, depression, and addictive disorders are common in trichotillomania. **Tics**, borderline **personality disorders**, and OCD are all more prevalent in trichotillomania than in the general population. The hair pulling in trichotillomania can be differentiated from that in OCD in that the hair pulling in trichotillomania is an impulse behavior where in OCD it is a repetitive act performed as part of an obsession. The individual with OCD is aware of his or her actions, while the individual with trichotillomania is not always conscious that he or she is pulling hairs.

Trichotillomania is not the underlying cause of hair pulling if there is a medical reason for the hair loss or if another co-existing psychiatric disorder such as hallucination provokes the hair pulling.

Demographics

Trichotillomania usually begins in the preteens but has been reported in children as young as one year old and has been seen first in adults over 50 years old. Patients in their seventies may suffer from trichotillomania. The mean age of onset is 12 years of age in girls and eight years of age in boys. This condition is seven times more common in children than in adults. Among young children there is no gender preference. But among adults, it is reported up to 10 times more often in females than in males. This may be skewed because females are more likely to seek attention for a medical problem, and because it is easier for males to disguise their compulsions, e.g. by shaving or because of social acceptance of male pattern hair loss.

The total number of Americans who pull their own hair at some point in their lifetime may be as high as 11 million. The prevalence of trichotillomania has been estimated to be as high as 2 percent of the general population. Among college students surveyed, more than 10 percent of college students pull their hair at some point, although only 1 percent meets the criteria for trichotillomania.

Causes and symptoms

There is no clear cause of trichotillomania, but there are psychoanalytical, behavioral, or biological theories for this disorder. Some of the more commonly accepted theories for trichotillomania are:

- childhood trauma
- stressful events
- neurochemical imbalance

The psychoanalytic model purports that trichotillomania occurs in an attempt to resolve a childhood trauma, the most common of which is sexual abuse. According to this model an unconscious unresolved past conflict triggers hair pulling.

The behavioral theory for trichotillomania states that a stressful event, such as moving or the loss of a loved one, or a family conflict precedes the onset of hair pulling and that hair pulling begins in an attempt to relieve tension caused by a stressful event. This behavior continues beyond the initial stimulus and eventually becomes habitual. Later the patient may not be aware of this initial trigger. For a child, the stressor may not be just a single event, but may occur in response to what a child may perceive as excessive demands from an authoritarian or an overbearing parent.

Biological theories for trichotillomania include a neurochemical imbalance, such as a serotonin imbalance. Drugs that correct for serotonin imbalance improve symptoms in many with this disorder. Altered dopamine levels may also play a role in trichotillomania. It is not clear if genetic factors are involved in the development of trichotillomania, although some studies report an increased percentage of relatives with various psychiatric disorders.

The most common symptom of trichotillomania is hair loss. The pattern of alopecia in trichotillomania varies among patients and the degree of hair loss will range from a barely noticeable thinning to total loss of hair. Some patients pull out hairs without regard for symmetry, while others will attempt to follow a pattern or pull out hairs in an effort to maintain symmetry of appearance. Usually, the hair loss on the head is patchy or poorly defined. There are neither scars nor any inflammation in the area of scalp hair loss. The top is the most affected region of the head. Tonsure trichotillomania is a pattern hair loss of the scalp in which hair is present only at the nape and on the outer edge of the scalp. The eyelashes and eyebrows may be plucked off, and hair loss may be noted on the arms, legs, and body. Pubic hair may be sparse.

When to call the doctor

Any continuous pulling of hair or hair loss should be reported to a medical professional, as there are medical causes for hair pulling and hair loss, and if trichotillomania is the underlying cause for this problem, then medical and psychiatric treatment needs to be initiated as soon as possible, since the earlier the intervention, the greater the likelihood that the behavior can be controlled. It is important to realize that the occasional or infrequent twisting, pulling, or chewing of hair in a child does not constitute trichotillomania and does not require medical attention.

Diagnosis

The diagnosis of trichotillomania is made by history and interview, along with histological examination of the hairs in the area of hair loss as well as skin tissue in the area. All other medical causes of hair loss must be eliminated. Since patients are adept at disguising and denying the symptoms of trichotillomania, the condition may go on for years without detection or treatment. Most patients are embarrassed to admit to hair pulling and the resultant sequelae, and elicitation of this behavior is difficult. The patient will not usually report **pain**. All of this makes the diagnosis of trichotillomania difficult. The

patient must be made to feel comfortable admitting to and then discussing the behavior.

The clinician may use rating scales to assist in the diagnosis of trichotillomania and to assess the degree to which a patient has trichotillomania. These scales include the Psychiatric Institute Trichotillomania Scale, National Institutive of Mental Health-Trichotillomania Severity Scale, Yale-Brown Obsessive Scale modified for Trichotillomania, the National Institute of Mental Health-Trichotillomania Impairment Scale, and the Minnesota Trichotillomania Assessment Inventory.

According to the American Psychiatric Association there are five criteria which must be met in order for trichotillomania to be diagnosed. They are as follows:

- The hair pulling is recurrent and a noticeable pattern of hair loss is observed.
- The patient feels increased tension prior to the hair pulling.
- This tension is relieved upon pulling hairs.
- The pulling is not associated with another mental condition, and there is no medical cause for the hair pulling.
- The behavior interferes with or disrupts the patient's social and work activities.

There is a subgroup of hair-pullers who do not meet the second and third criteria listed above. These individuals are less likely to hide their behavior and do not suffer from low **self-esteem** as frequently as those who meet all of the above criteria. There is some debate about whether these people have trichotillomania and about whether these criteria for diagnosis of trichotillomania are too restrictive.

Histological examination of hair follicles and skin biopsies also help in the diagnosis of trichotillomania. In the areas of hair loss in trichotillomania there will be a mixture of short and longer hairs in the area of hair loss. Trichomalacia or distortion of the hair follicles is often present in trichotillomania.

Trichotillomania must be differentiated from medical causes of hair loss and these include: skin conditions such as **psoriasis**; trauma, such as that from radiation; endocrine disorders such as **hypothyroidism**; infectious diseases such as herpes zoster; inflammation such that of the lids margins, called blepharitis; and tinea capitis, a fungal infection of the scalp. Other psychiatric disorders, such as **schizophrenia**, must also be ruled out.

Treatment

Usually, the patient with trichotillomania does not present for treatment until, on average, two years after the hair pulling has begun. Traditional treatment for trichotillomania involves psychological or behavioral therapy, or medication. Behavior modification, especially with children, helps the child to increase his or her awareness of the hair pulling. Behavioral therapy may be as simple as acknowledging the problem and instituting a plan for desensitization of the behavior.

Habit reversal training (HRT), a cognitive behavioral therapy, has been successfully used in the treatment of trichotillomania. Under HRT treatment the patient acquires increased awareness of his or her actions and learns alternative behavior to the hair pulling. HRT has been employed in group therapy. Addressing the behavior of trichotillomania in a group setting is helpful so the patients realize that they are not the only ones with this problem. This experience also improves social interaction, as isolation is common among patients with trichotillomania.

Medication to correct biochemical imbalances in the brain is a common component of trichotillomania treatment. But since drug trials in children and adolescents have been limited, behavioral therapy is often instituted alone first, prior to using medication. But for some with trichotillomania, behavioral therapy is more successful when drug therapy helps reduce the urge to pull hair. For these individuals, relapses are more frequent when pharmacotherapy is reduced or discontinued.

There are no FDA drugs which specifically treat trichotillomania. The drugs used to treat this disorder have been developed for treatment of other psychiatric problems. The drug which has been the most successful in treatment of trichotillomania is clomipramine (Anafranil), a tricyclic antidepressant.

Since it is hypothesized that serotonin activity is abnormal in trichotillomania, selective serotonin reuptake inhibitors (SSRIs) are commonly given to improve symptoms. Prozac is a common SSRI. Drugs in this class given to treat trichotillomania in children include sertraline (Zoloft), fluvoxamine (Luvox), and clomioramine. The effectiveness of a given drug varies considerably from person to person. If one SSRI drug is not successful in controlling trichotillomania in a given individual, another drug in this class may work. Risperdone and clonazepam, which address a dopamine imbalance, can be added to SSRIs if an SSRI drug does not satisfactorily control symptoms. But these drugs, called neuroleptics, have more side effects in children than in adults.

Since those with trichotillomania do not report pain, drugs to decrease pain thresholds have been tried as well. Other drugs that are given to treat this disorder include buspirone, lithium (Lithobid), naltrexone, paroxetine (Paxil), valproate, and the antipsychotic drug, quetiapine.

Treatment of the resultant medical complications of hair pulling must be addressed. Carpal tunnel can develop from repetitive pulling. Infections at the site of the hair pulling and blepharitis at the eyelid margins can occur, both of which are treated with **topical antibiotics** and corticosteroids. If there is significant eyelash and eyebrow loss, called madarosis, blepheropigmentation or surgical tattooing can be performed. Although not done often, transplantation of hairs to these areas is possible.

Topical application of colladion can help with regrowth of hair but will not be successful long term unless the underlying behavior is controlled. If the hair pulling continues for long periods without treatment, the alopecia may be permanent.

Anemia, **malnutrition**, and digestive disorders, including bowel obstructions, can develop, if trichotillomania develops into trichotillophagia or eating of the hairs. Trichobezoars, or hairballs, can form when the hair is bitten off and ingested.

For many with trichotillomania, hair pulling is not an activity that can be stopped at will. For some, however, the suppression of hair pulling may be possible, even if the underlying urge persists. The family needs to be a part of therapy since familial stressors may have triggered trichotillomania.

Because of the shame involved with hair pulling the patient may have other medical problems which go untreated because he or she will not seek any medical care at all, for **fear** that hair pulling and its associated stigmata will be uncovered. Thus, it is important that once trichotillomania is diagnosed that the healthcare provider inquire into any other medical concerns that the patient may have.

Alternative treatment

Hypnosis has been used in treatment of childhood trichotillomania. The Erickson approach of hypnosis helps the child to substitute hair pulling for a stroking behavior. Other approaches to hypnosis in trichotillomania teach the child that he or she has control over events in his or her life, including hair pulling. There are other hypnotic techniques that employ adverse conditioning, so that the hair pulling becomes associated with pain instead of pleasure.

Other techniques, consider alternative, used to trichotillomania include biofeedback, **yoga**, and **exercise**.

Prognosis

When trichotillomania appears in early childhood, the duration of time during which the child is afflicted, is limited. The remission rate for children diagnosed before age six is high. For many children with trichotillomania, the condition resolves by adulthood.

The prognosis is much more difficult for those who develop trichotillomania after age 13. These children have a higher rate of other co-existing psychiatric disorders. Unfortunately, among those individuals who need long-term treatment for trichotillomania, as is the case when the initial presentation occurs in late childhood or as in **adolescence** or in adulthood, there is a high relapse rate in spite of intervention. A lack of definitive cause for trichotillomania makes treatment difficult, and the prognosis for a total recovery is poor, although the behavior may be satisfactorily controlled with therapy.

Prevention

Since, as of 2004, the actual cause of trichotillomania was not known, there is no known means of prevention.

Parental concerns

Parents must realize that the earlier the treatment for trichotillomania is begun, the more likely that the hair pulling can be controlled. When trichotillomania strikes the adolescent it is especially important that the behavior be addressed and treated promptly. Adolescence is a time when self-esteem and independence are developing. If the adolescent does not have a positive body image, then fear or ridicule from family and peers can affect his or her ability to interact with others. Development of normal healthy relationships as an adult may be impaired if the family and such support mechanisms as therapy are not in place.

Since often the family dynamics provoke this behavior, parental involvement in therapy is essential. If necessary, the parents must be open to establishing new boundaries within the parent-child relationship.

It is important that parents to realize that trichotillomania is a complex and not completely understood behavior. But it is increasingly believed that trichotillomania has a biological basis, and thus parents must understand that they did not cause it and that they are not the only parents with a child who has trichotillomania. Support for trichotillomania may be found through the Trichotil-

lomania Learning Center (available online at <www.trich.org>). Many larger cities may have local support groups. Healthcare providers may help with location of such groups locally.

See also Alopecia; Obsessive-compulsive disorders.

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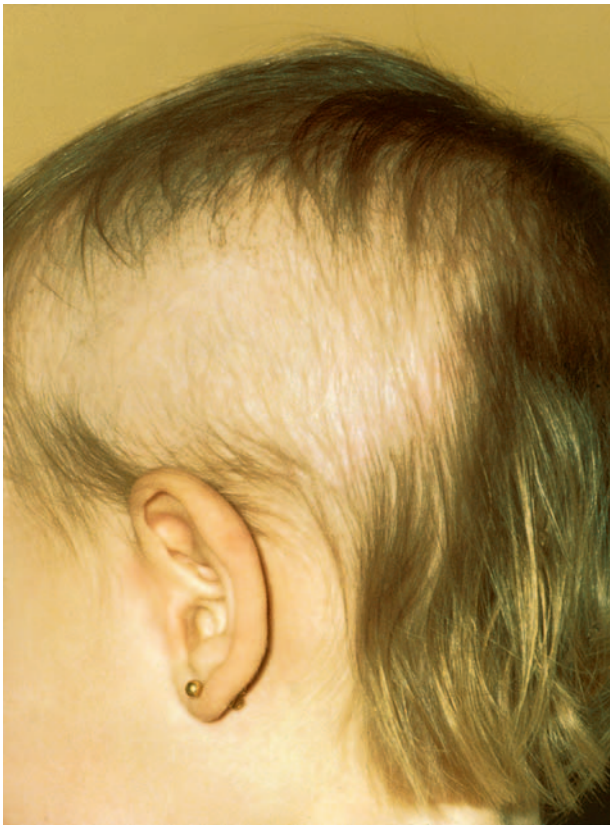

Child's scalp showing hair loss from trichotillomania. *(© NMSB/Custom Medical Stock Photo.)*

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KEY TERMS

Alopecia—The loss of hair, or baldness.

Dopamine—A neurotransmitter made in the brain that is involved in many brain activities, including movement and emotion.

Histology—The study of tissue structure.

Hypnosis—The technique by which a trained professional induces a trance-like state of extreme relaxation and suggestibility in a patient. Hypnosis is used to treat amnesia and identity disturbances that occur in dissociative disorders.

Obsessive-compulsive disorder—An anxiety disorder marked by the recurrence of intrusive or disturbing thoughts, impulses, images, or ideas (obsessions) accompanied by repeated attempts to supress these thoughts through the performance of certain irrational and ritualistic behaviors or mental acts (compulsions).

Remission—A disappearance of a disease and its symptoms. Complete remission means that all disease is gone. Partial remission means that the disease is significantly improved, but residual traces of the disease are still present. A remission may be due to treatment or may be spontaneous.

Serotonin—A widely distributed neurotransmitter that is found in blood platelets, the lining of the digestive tract, and the brain, and that works in combination with norepinephrine. It causes very powerful contractions of smooth muscle and is associated with mood, attention, emotions, and sleep. Low levels of serotonin are associated with depression.

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Martha Reilly, OD

Triple antibiotic ointment *see* **Antiobiotics, topical**
Trisomy 13 *see* **Patau syndrome**
Trisomy 18 *see* **Edwards' syndrome**
Trisomy 21 *see* **Down syndrome**

Truancy

Definition

Truancy is unapproved absence from school, usually without a parent's knowledge.

Description

Truancy is a serious problem in many communities in the United States. All states have laws governing compulsory education. Noncompliance results in penalties for the parent(s) or guardian of the truant student. The majority of the states require that students attend school until at least age 16. Those students who do not attend school regularly are often taking the first step toward a lifetime of problems. Most experts believe that truancy is a powerful and accurate predictor of involvement in crime and violence. The United States Department of Justice reports that 80 percent of those in prison were at one time truants. The percent of juvenile offenders who started as truants is even higher, approaching 95 percent. Truancy is different from school phobia, in which a child fails to attend school because of **anxiety**.

As of 2004, no national database existed to define the number children who are truant, partly because there is no uniform definition of truancy. Some districts consider children truant only if they miss a half or full day of school, while others consider missing a single scheduled class period as truancy. The Los Angeles School District has estimated that 10 percent of its students are absent each day and that only 5 percent return with written notes from home excusing the absence. Pittsburgh, Pennsylvania, schools reported 3,500 students, or 12 percent of all students, were absent on an average school day; 70 percent of those were unexcused. Milwaukee, Wisconsin, reported 4,000 unexcused absences on an average school day. Miami, Florida, reported that over 70 percent of 13- to 16-year-olds prosecuted for crimes were truant. The No Child Left Behind Act of the early 2000s requires school districts to report truancy, so national numbers were expected to become available. Boys and girls are equally likely to be truant. The average age of truant students is 15 years, but some children begin skipping school as young as 10.

Why children are truant

According the United States Department of Education's 1996 *Manual to Combat Truancy*, skipping school is a cry for help and a signal that the child is in trouble. Psychiatrists consider truancy one of many symptoms of **oppositional defiant disorder** or the more serious

diagnosis of **conduct disorder**, especially when truancy begins before age 13.

There are many reasons why children become truant. These include:

- lack of interest in education and alienation from school
- falling behind academically in school
- **fear** of violence on the way to school or at school
- alienation from authority
- lax parental supervision
- lack of parental support for education
- drug and alcohol abuse
- working long hours while attending school, resulting in chronic exhaustion
- lack of significant consequences for failure to attend school
- problems at home that require supervising younger children or helping dysfunctional adults

Truancy as a predictor of behavior

Truancy is a strong and reliable predictor of delinquent behavior, especially among males. Children who are habitual truants are more likely to engage in undesirable and antisocial behaviors such as gang membership, marijuana use, alcohol use, inhalant and hard drug use, high-risk sexual behavior, cigarette **smoking**, suicidal behaviors, theft, and vandalism. Truant girls are more likely to become pregnant and drop out of school. Most habitual truants eventually enter the juvenile court system. As adults, habitual truants have more employment and marital problems and are jailed far more often than nontruants.

Truancy is a gateway to serious violent and nonviolent crime. Law enforcement agencies have linked high rates of truancy to high rates of daytime burglary and vandalism. In addition, they have found habitual truants are more likely to belong to **gangs** and participate in violent crimes and assaults.

Combating truancy

Communities in which anti-truancy programs have been successful use a combination of incentives and sanctions to keep students in school. In the *Manual to Combat Truancy*, five key points are defined for minimizing truancy. The first step is to involve parents in all aspects of truancy prevention. To stop truants, the school must be able to provide parents with notification of their child's absence on the day the absence occurs. Schools are advised to create an efficient attendance-tracking system and to communicate students' absences to parents immediately.

Second, schools must have firm policies on the consequences for truancy, and all students should be aware of the sanctions that will be imposed if they are absent without an excuse. Some states have found that linking truancy to the ability to obtain a driver's license effectively reduces unexcused absences. Others have invoked a daytime curfew, allowing police to question any young person not in school during school hours.

Third, parents must take responsibility for keeping their children in school. Most state laws impose fines or jail terms on parents of truants. School districts vary in how aggressive they are about holding parents accountable; however, more are becoming tougher. For example, in 2003, the Upper Darby School District in suburban Philadelphia had 14,000 students. This school system sends 10 to 12 parents to jail each year for their children's failures to attend school.

Alternately, some states are investigating ways to use incentives such as linking eligibility for public assistance to truancy as an effective way to capture parents' interest in keeping their children in school. Another positive incentive provides increased eligibility for services to families whose children attend school regularly. Many communities also offer effective parenting courses, **family** counseling, and mediation for returning the student to school.

Fourth, root causes of truancy must be addressed. The root causes of truancy are complex and varied and can include drug use, membership in a peer group of truants or gangs, lack of direction in education, poor academic performance, and violence at or near school. By analyzing the reasons students are truant, the school administration may be able to correct or improve the problem and reduce truancy. For example, if students stay away from school because of inadequate academic skills, special tutoring programs may be initiated. If students have concerns about violence near the school, the administration may request increased security from the police for the surrounding neighborhood. Local businesses can be enlisted to support school-to-work programs to help students make the transition to employment.

Finally, a close link between the school, law enforcement, juvenile court, family court officials, and social service agencies may lead to solutions for truancy. Some communities have authorized the police to patrol neighborhoods where truant youth are likely to spend the school hours. Daytime curfews are also effective in some cities, where school age children can be questioned if they are on the streets during school hours.

Common problems

Truancy is not normally an isolated problem in a child's life. The following comparisons from a 2003 study published in the *Journal of the American Academy of Child and Adolescent Psychiatry* highlight the problem. The first percentage given is for truant children. The percentage of each activity in nontruant children is given in parentheses for comparison.

- all psychiatric disorders: 25.4 percent (6.8 percent)
- oppositional defiant disorder: 9.7 percent (2.3 percent)
- conduct disorder: 14.8 percent (1.6 percent)
- depression: 7.5 percent (1.6 percent)
- conflictual relationships with peers: 16.2 percent (8.7 percent)
- living in poverty: 31.3 percent (19.1 percent)
- single-parent household: 45.9 percent (21.8 percent)
- lax parental supervision: 31.5 percent (6.7 percent)
- mother currently diagnosed as depressed: 11.9 percent (5.5 percent)
- parents teenagers at time of birth: 15.3 percent (8.4 percent)

Parental concerns

Almost half of all truants live in single-parent households, usually headed by women. Parents are concerned that they have lost control of their children and fear legal sanctions if their child skips school. They also are concerned about their child dropping out of school and becoming involved in crime and the criminal justice system. Parents may also fail to understand the attendance laws or have cultural biases against the education system.

When to get help

Truancy is a symptom that things are out of control in a child's life. Parents need to seek help from the school, social service agencies, and mental health professionals at the first sign that their child is skipping school.

See also Conduct disorder.

Resources

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National Association of School Psychologists. 4340 East West Highway, Suite 402, Bethesda, MD 20814. Web site: <www.nasponline.org>.

Tish Davidson, A.M.

TSS *see* **Toxic shock syndrome**

Tuberculosis

Definition

Tuberculosis is a chronic, infectious disease that primarily attacks the lungs.

Description

Tuberculosis (TB) is caused by a bacteria that primarily attacks the lungs. An individual may be "TB infected," meaning the bacteria are in the body but are in an inactive state, walled off behind scab-like structures that are the body's defense mechanism, or have "TB disease," when the bacteria actively spread throughout the body and can cause damage to the lungs or other organs. The severity of the attack depends on whether the bacteria spread from the lungs to other parts of the body. TB infection in the blood, the meninges (membranes around the brain and spinal cord), or the kidneys are the most serious. Children between the ages of six and 24 months are the most susceptible to **meningitis**; it is the chief cause of tuberculin death among children.

Transmission

The bacteria that causes TB, *Mycobacterium tuberculosis*, is transmitted by droplets when an infected per-

son coughs or sneezes. It is not spread through kissing or other physical contact. Children nearly always contract the disease from an infected adult.

Demographics

In 2003, the Centers for Disease Control and Prevention (CDC) reported 14,874 cases of tuberculosis in the United States, or 5.1 cases per 100,000 population. The actual number of TB infections, however, is estimated to be much higher, as high as ten million. In 2002, there were 802 tuberculosis-related deaths. The District of Columbia had the highest rates of TB, with 14 cases per 100,000 people in 2003; Montana and Wyoming had the lowest rate, with 0.8 cases per 100,000 population. Children less than 15 years of age represented 6 percent of reported TB cases, and 15–24-year-olds represented 11 percent of all cases. Worldwide, TB cases are the rise, with nearly 8.8 million new cases a year being estimated by the World Health Organization (WHO).

Causes and symptoms

Mycobacterium tuberculosis is a microscopic, rod-shaped bacterium. The majority of individuals who are infected with TB do not go on to have active disease. Active TB can be triggered when a person's immune system is weakened, such as from human **immunodeficiency** virus (HIV), **malnutrition**, or alcohol abuse.

Early symptoms of TB include unusual fatigue, **fever**, loss of weight, **headache**, coughing, and irritability. An infected child may have night sweats and **cough** up blood. In advanced stages, the patient will suffer persistent coughing, breathlessness, and fever. Many times TB is not diagnosed and becomes dormant; this is known as initial tuberculosis. In severe cases among young children between the ages of two and four, initial TB can be fatal. The disease can reoccur, or reactivate, during **adolescence** when resistance is low, and may disappear on its own or develop into serious lung disease.

When to call the doctor

Parents should contact their child's doctor if the child has been in contact with someone who has been diagnosed with or is suspected to have tuberculosis, or if the child exhibits the symptoms of the disease, particularly persistent fever, night sweats, and cough.

Diagnosis

Tuberculosis is nearly always diagnosed by tuberculin skin tests, although one can also be diagnosed by chest **x rays** and analysis of sputum (matter from the respiratory tract) smears and cultures. The most common tuberculin skin test is the Mantoux test, which consists of injecting a small amount of protein from the TB bacillus into the forearm. A reddening and swelling of the area after 24–72 hours signals the presence of TB. A negative result, however, may not necessarily exclude a diagnosis of TB.

Treatment

The disease is treated with a regimen of strong **antibiotics** such as Rifampin and Isoniazid for six months to two years. Because some strains of the disease are unusually drug-resistant, cultures are grown from the patient's bacteria and tested with a variety of drugs to determine the most effective treatment. In cases of strong drug-resistant strains, the child may undergo surgery to remove the infected areas.

Infants with TB are usually hospitalized but children and teenagers can generally lead active lives within two weeks of beginning medication. It is imperative that the mediation prescribed be taken faithfully.

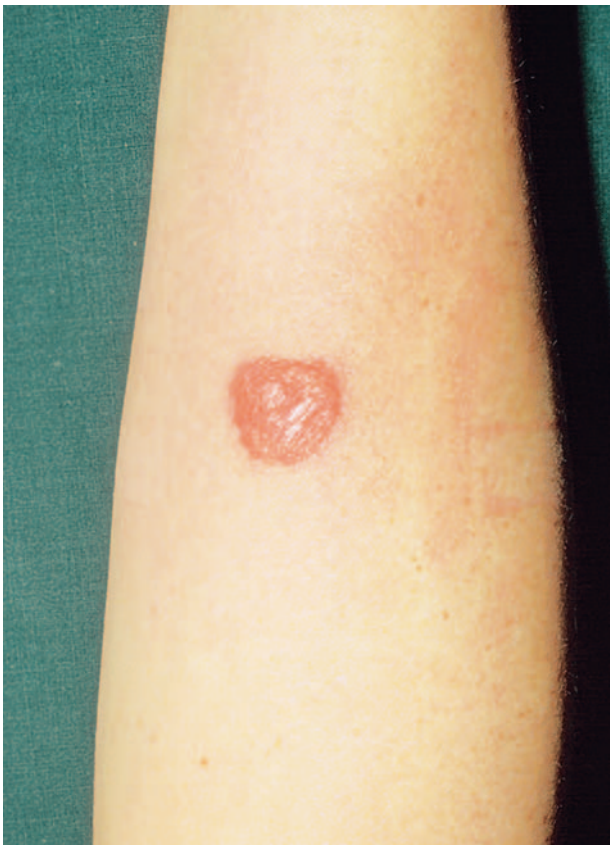
Prognosis

With treatment, TB infection that is not drug resistant can nearly always be cured as long as patients are consistent with their medications and considerable lung damage as not already occurred. Drug-resistant TB has a lower cure rate. Without treatment, the disease will continue to progress; approximately one-half of untreated TB patients will die of the disease.

Prevention

Stopping the spread of tuberculosis is the most effective way of preventing its incidence among children. All adults who work with children should be screened regularly. In many communities, children are tested when they reach their first birthday and then at one-to-three year intervals throughout the school years. The medical profession is divided on the issue of screening; some physicians believe that the screening should be focused in areas of common occurrence or within high-risk populations such as foreign-born children. The practice of relying on parents to report results of the skin testing has also come under criticism from some members of the medical community.

While a vaccine for TB does exist (Bacille Calmette-Guerin or BCG vaccine), it is not widely available in the United States and has had conflicting reports about its efficacy. Being inoculated with BCG vaccine does not always prevent infection with the disease. The vaccine is only recommended for children in the United



Lesion on the arm of a child infected with tuberculosis. *(© Mediscan/Visuals Unlimited.)*

States if they live with someone who has active TB that cannot be treated or is drug-resistant.

Nutritional concerns

Poor **nutrition** is closely related active tuberculosis; children with adequate nutrition are more resistant to the disease than those who suffer from malnutrition.

Parental concerns

If a child has been infected with TB and is prescribed drug therapy to treat the disease, it is imperative that parents closely monitor their child to ensure that the medication is taken as prescribed; if the medication is not taken frequently enough or until it is no longer needed, drug-resistant TB can arise.

Resources

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"TB on the Rise." *Patient Care* 38, no. 6 (June 2004): 9-10.

KEY TERMS

Mantoux test—A tuberculin skin test. Also called the PPD (purified protein derivative) test.

Meningitis—An infection or inflammation of the membranes that cover the brain and spinal cord. It is usually caused by bacteria or a virus.

Sputum—The substance that is coughed up from the lungs and spit out through the mouth. It is usually a mixture of saliva and mucus, but may contain blood or pus in patients with lung abscess or other diseases of the lungs.

Young, Douglas B. and Brian D. Robertson. "TB Vaccines: Global Solutions for Global Problems." *Science* 284, no. 5419 (May 28, 1999): 1479.

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American Lung Association. 61 Broadway, 6th Floor, New York, NY 10006. (800) 548-8252. Web site: <www.lungusa.com>.

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Mary McNulty
Stephanie Dionne Sherk

Tuberous sclerosis

Definition

Tuberous sclerosis is a genetic disorder in which noncancerous (benign) tumors grow on the brain, skin, kidneys, eyes, heart, and lungs.

Description

The name tuberous sclerosis refers to characteristics of the benign tumors that grow within the brain. The tumors have root-like or tuberous appendages. Over time, the tumors undergo sclerosis, meaning they calcify and grow hard.

Symptoms of tuberous sclerosis may be identifiable at birth or may develop over time.

Demographics

In the United States, as of the early 2000s, there are between 25,000 and 40,000 individuals with tuberous sclerosis. Globally, about 1 to 2 million individuals have the disease. The disease occurs in about one out of every 6,000 newborns. There is no gender, racial, or ethnic predilection.

Causes and symptoms

Tuberous sclerosis occurs when at least one of two genes (either TSC-1 on chromosome 9 or TSC-2 on chromosome 16) is defective. Normally, the two genes produce proteins called hamartin and tuberin, respectively. These proteins seem to serve as inhibitors of tumor growth. When the TS genes are defective or absent, the proteins are either absent or deficient, which allows tumor growth.

Most cases of tuberous sclerosis occur due to spontaneous mutations. This means that the disease does not occur due to the inheritance of an abnormal gene, but rather because the baby's gene is defective for some reason other than inheritance.

Symptoms

The tumors of tuberous sclerosis occur throughout the body, including the brain, heart, lungs, kidneys, eyes, and skin. Other symptoms include seizures, **developmental delay**, behavior problems, and skin problems.

KIDNEYS Cysts on the kidneys tend to appear during the second or third decade of life. In most cases, they do not interfere with kidney functioning. Rarely, there are so many cysts that the kidneys functioning is impaired, or the cysts bleed, resulting in anemia. Fatty growths within the kidneys (called angiolipomas) may grow so large that they cause **pain** and/or kidney failure. Rarely, malignant tumors of the kidney (renal cell carcinoma) occur within an existing angiolipoma.

BRAIN Several types of brain tumors can grow, resulting in blockage of the flow of cerebrospinal fluid, fluid backup, headaches, and visual disturbances.

HEART Benign tumors in the heart (rhabdomyomas) may block circulation or may exist uneventfully.

EYES White areas in the retina, called phakomas, are characteristic of the disease (and may aid in diagnosis) but do not result in visual disturbances.

SKIN A variety of skin disorders are noted in tuberous sclerosis, including areas of under-pigmented skin (hypomelanic macules); reddish bumps on the face (facial angiofibromas); raised patches on the forehead (called forehead plaques); areas of rough, thickened skin on the neck or back (shagreen patches); tiny fleshy bumps around or under the toe- or fingernails (ungula or subungual fibromas); skin tags (molluscum fibrosum); flat brown patches.

BEHAVIOR About 33 to 50 percent of all tuberous sclerosis patients have problems such as learning disabilities, severe **mental retardation**, attention deficit disorder, **obsessive-compulsive disorder**, **autism**, aggression, rage, or self-harming behavior.

Diagnosis

Tuberous sclerosis is diagnosed when the characteristic tumors are noted in the skin, heart, brain, or kidneys. Many patients come to the healthcare provider's attention after they have begun to have seizures. Further examination with CT and/or MRI scans, ultrasound, and Wood's lamps to view the eyes will reveal the presence of the characteristic tumors of tuberous sclerosis.

Treatment

As of 2004, no cure was available for tuberous sclerosis. Antiseizure medications may be prescribed, as well as medications to treat attention deficit disorder and obsessive-compulsive disorder. Skin lesions may be removed or reduced via dermabrasive or laser procedures. Surgery may be performed to remove enlarging kidney tumors, to avoid the advent of kidney failure.

Prognosis

Most individuals with tuberous sclerosis have a normal lifespan. The prognosis for their quality of life depends on the severity of their behavioral and cognitive symptoms. Individuals whose symptomatology is confined to kidneys or skin (as opposed to having multiple behavioral symptoms) may do very well.

Prevention

As of 2004, there was no way to prevent tuberous sclerosis.

Parental concerns

Parents of child with tuberous sclerosis should be prepared to answer any questions their child or the child's siblings may have about the disease. Siblings may **fear** they will catch the disease or perhaps caused it, and may need to reassured that they are not at fault.

Resources

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Rosalyn Carson-DeWitt, MD

Turner syndrome

Definition

Turner syndrome is a birth defect caused by the absence of an X chromosome in some or all cells of a female, which inhibits sexual development and usually causes infertility.

Description

Chromosomes are structures in the nucleus of every cell in the human body that contain the genetic information necessary to direct the growth and normal functioning of all cells and systems of the body. A normal individual has a total of 46 chromosomes in each cell, two of which are responsible for determining gender. Normally, females have two X chromosomes, and males have one X and one Y chromosome.

In Turner syndrome, an error occurring very early in development results in an abnormal number and arrangement of chromosomes. Most commonly, an individual with Turner syndrome will be born with 45 chromosomes in each cell rather than 46. The missing chromosome is an X chromosome. The affected person is always female.

Turner syndrome may result in a wide spectrum of symptoms, from major heart defects to minor cosmetic issues. Some individuals with Turner syndrome may only have a few symptoms while others may have many. Almost all girls with Turner syndrome have short stature and loss of ovarian function, but the severity of the symptoms varies among individuals.

Turner syndrome is also referred to as Bonnevie-Ullrich syndrome, gonadal dysgenesis, and monosomy X.

Demographics

The prevalence of Turner syndrome is widely reported as being approximately one per 2,500 live female births, although researchers have reported prevalence rates that range from one in 3,125 to one in 5,000 live female births. About 1 to 2 percent of all female conceptions have a missing X chromosome. Of these, the majority (99%) spontaneously abort, usually during the first trimester of pregnancy.

Causes and symptoms

Turner syndrome usually occurs sporadically, which means that the mutation occurs during fetal development and is not inherited from either parent. In rare cases, a parent may carry rearranged chromosomes that can result in Turner syndrome in a daughter, which is the only situation in which the Turner syndrome is inherited.

More than half of all girls with Turner syndrome are mosaics, which means that the mutation occurs in some but not all cells of their body. Therefore, Turner syndrome can vary in severity. The fewer the affected cells, the milder the disease.

Symptoms of a girl with Turner syndrome include:

- short stature
- webbed skin of the neck
- abnormal eye features (drooping eyelids)
- abnormal bone development, such as a "shield-shaped," broad flat chest
- absent or retarded development of secondary sexual characteristics that normally appear at **puberty**, including sparse pubic hair and small breasts

- coarctation (narrowing) of the aorta
- bicuspid aortic valve
- infertility
- dry eyes
- absence of **menstruation**

Growth in children with Turner syndrome is characterized by a slight **intrauterine growth retardation**, relatively normal growth rates for the first several years of life, a progressive deceleration of growth later in childhood, and the lack of a pubertal growth spurt. The average height of Turner women is 147 cm (57.8 inches), varying between 135 (53 inches) and 163 cm (64 inches). This is about 20 cm (7.8 inches) shorter than the height of women with normal chromosomes.

Normal pubertal development and spontaneous menstrual periods do not occur in the majority of children with Turner syndrome. Most girls with Turner syndrome do not have ovaries with healthy oocytes capable of fertilization and embryo formation. However, it is estimated that 3 to 8 percent of girls with a single X chromosome and 12 to 21 percent of females with sex chromosome mosaicism may have normal pubertal development and spontaneous menstrual periods. A few pregnancies have been reported in women with Turner syndrome.

Individuals with Turner syndrome report an increased incidence of **fractures** in childhood and osteoporotic fractures in adulthood. The primary cause of osteoporosis may be inadequate levels of estrogen circulating in the body; however, defects in bone structure or strength may also be related to the loss of unknown X-chromosome genes.

The incidence of type II diabetes, also known as insulin resistant diabetes (glucose intolerance), has been reported to be increased in Turner syndrome, with individuals having twice the risk of the general population for developing this disease. The muscles of many persons with Turner syndrome fail to use glucose efficiently, which may contribute to the development of high blood sugar.

Many women with Turner syndrome have high blood pressure, which may even occur during childhood. High blood pressure may be due to aortic constriction or to kidney abnormalities; however, in a majority of cases, no specific cause for high blood pressure can be identified.

Kidney problems are present in about one third of girls with Turner syndrome and may contribute to high blood pressure. Three types of kidney problems have been reported: the presence of a single horse-shoe shaped kidney (normally two distinct, bean-shaped structures are present); an abnormal urine-collecting system; or an abnormal artery supply to the kidneys.

From 5 to 10 percent of girls with Turner syndrome have a severe constriction of the major blood vessel coming from the heart (**coarctation of the aorta**). This defect is thought to be a result of an obstructed lymphatic system compressing the aorta during fetal development. Other major defects and its major vessels are reported to a lesser degree. As many as 15 percent of children with Turner syndrome have bicuspid aortic valves, where the major blood vessel from the heart has only two rather than three components to the valve regulating blood flow.

Juvenile rheumatoid arthritis, an autoimmune condition, has been associated with Turner syndrome. The prevalence seems to be at least six times greater than would be expected if the two conditions were only randomly associated. Girls with Turner syndrome have an elevated prevalence rate of dental caries and such other periodontal conditions as gum disease and plaque.

Approximately one-third of girls with Turner syndrome have a thyroid disorder, usually **hypothyroidism**. Symptoms of this condition include decreased energy, dry skin, cold-intolerance, and poor growth.

Contrary to earlier reports, most individuals with Turner syndrome are not mentally retarded. They may have some learning disabilities, particularly with regard to spatial perception, visual-motor coordination, and mathematics. This specific learning problem is referred to as Turner neurocognitive phenotype and appears to be due to loss of X chromosome genes important for selected aspects of nervous system development. The verbal skills of girls with Turner syndrome are usually normal. Some girls with Turner syndrome may also have difficulties with memory and motor coordination, which may be related to estrogen deficiency.

When to call the doctor

Parents should call their healthcare provider if their infant has symptoms of this disorder or if an adolescent girl's sexual development appears to be delayed.

Diagnosis

Turner syndrome is either diagnosed at birth because of associated anomalies or at puberty when there is absent or delayed menses and delayed development of normal secondary sexual characteristics. During a physical examination, the doctor looks for underdeveloped breasts and genitalia, webbed neck, short stature,

low hairline in back, simian crease (a single crease in the palm), and abnormal development of the chest. An ultrasound may reveal small or undeveloped female reproductive organs while a gynecologic examination may reveal a dry vaginal lining. A kidney ultrasound can be used to evaluate abnormalities of the kidneys. After diagnosis, echocardiogram (heart ultrasound) and an MRI of the chest are performed to evaluate possible cardiac defects.

Hands and feet of infants with Turner syndrome may be swollen or puffy at birth; there may be swelling at the nape of the neck. These babies often have soft nails that turn upwards on the ends when they are older. These features appear to be due to obstruction of the lymphatic system during fetal development. Another characteristic cosmetic feature is the presence of multiple pigmented nevi (colored spots on the skin).

Turner syndrome is confirmed on the basis of genetic analysis of chromosomes, which can be done prior to birth. However, the predictive value of **amniocentesis** in diagnosing Turner syndrome varies from 21 to 67 percent. There is no significant relation between the mother's age and risk of Turner syndrome.

Treatment

Most individuals with Turner syndrome require female hormone therapy to promote development of secondary sexual characteristics and menstruation. The time of beginning therapy varies with individuals. Experts recommend that therapy begin when a woman expresses concern about her onset of puberty or by the age of 15. Girls and women with Turner syndrome should be treated with estrogen/progesterone to maintain their secondary sexual development and to protect their bones from osteoporosis until at the least the usual age of menopause (50 years). The use of estrogen therapy may also improve memory and motor coordination problems associated with estrogen deficiency. Assisted reproductive technology may allow for women with Turner syndrome to become pregnant with donated ooctyes.

All women receiving long-term, female hormone therapy require periodic gynecological examinations, because those with Turner syndrome have an increased risk of developing neoplasms, such as gonadoblastoma and dysgerminoma, which arise from their rudimentary streak gonads (a condition in which germ cells are absent and the ovary is replaced by a fibrous streak).

Because it is so dangerous, experts suggest early screening and surgery for aortic coarctation of the artery in girls with Turner syndrome. Bicuspid aortic valves can deteriorate or become infected, so it is advised that all girls with Turner syndrome undergo annual cardiac evaluations. Kidney problems may also be corrected surgically, but there still may be a tendency for high blood pressure and infections. Diabetes type II can be controlled through careful monitoring of blood-sugar levels, diet, **exercise**, regular health care, and medication if necessary. Hypothryoidism can be easily treated with thyroid hormone supplements.

A low hairline at the back of the neck is one of several characteristics of Turner syndrome. *(NMSB/Custom Medical Stock Photo, Inc.)*

Plastic surgery to correct webbing of the neck should be considered at an early age (before entering school) for girls with Turner syndrome.

Final adult height in individuals with Turner syndrome can be increased if growth hormone (GH) treatment is given relatively early in childhood. However, not all individuals get a good growth response to GH.

Prognosis

Most children with Turner syndrome can live relatively normal lives. The prognosis for a person with Turner syndrome is dependent on the other associated conditions that may be present. Care must be taken to regularly monitor patients for the health problems that are associated with Turner syndrome. For example, heart or kidney defects may significantly impact their quality of life. Without these types of conditions, however, their life expectancy is normal. Support will be necessary to help an adolescent girl cope with body image issues and to help some women accept the fact that they will never be able to have children.

Parental concerns

Families may wish to seek counseling regarding the effects of the syndrome on relationships within the

KEY TERMS

Bicuspid aortic valve—A condition in which the major blood vessel from the heart has only two rather than three components to the valve regulating blood flow.

Coarctation of the aorta—A congenital defect in which severe narrowing or constriction of the aorta obstructs the flow of blood.

Mosaic—A term referring to a genetic situation in which an individual's cells do not have the exact same composition of chromosomes. In Down syndrome, this may mean that some of the individual's cells have a normal 46 chromosomes, while other cells have an abnormal 47 chromosomes.

Ovary—One of the two almond-shaped glands in the female reproductive system responsible for producing eggs and the sex hormones estrogen and progesterone.

family. Many people respond with guilt, **fear**, or blame when a genetic disorder is diagnosed in the family, or they may overprotect the affected member. Support groups are often good sources of information about Turner syndrome; they can offer helpful suggestions about living with it as well as emotional support.

Resources

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Turner Syndrome Society of the United States. 14450 TC Jester, Houston, TX 77014. Web site: <www.turner-syndrome-us.org>.

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Judith Sims, MS
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Twin pregnancy *see* **Multiple pregnancy**

Twins

Definition

Twins occur when two babies are born at the same birth.

Description

Identical, or monozygotic, twins are of the same sex and are genetically identical and physically similar, because they both come from one ovum (egg), which, after fertilization, divides in two and develops into two separate fetuses. Fraternal, or dizygotic, twins occur when the mother produces two eggs in one monthly cycle and both eggs are fertilized. The conceptions may take place on two separate occasions and could involve different fathers.

Fertilized egg division which produces twins can either happen early or late in development. In the case of early separation, the two fetuses either share an amniotic sac or each has a separate amniotic sac. If the fetuses share an amniotic sac, they also share a placenta. If the two fetuses have separate amniotic sacs, they can either share a placenta or have two separate placentas. Twins can also result from a fertilized egg that divides slightly later in development. In this case, the twins share an amniotic sac and a placenta. It is from these cases of late separation that conjoined (Siamese) twins sometimes develop.

Fraternal twins, who are no more genetically alike than ordinary siblings, may be of the same or different sex and may bear some similarity of appearance. Fraternal twinning appears to be passed on by the female members of a **family**. If the mother is a fraternal twin herself, has fraternal twin siblings, or fraternal twin relatives on her side of the family, she is more likely to give birth to fraternal twins. If she has already given birth to fraternal twins, her chances of giving birth to fraternal twins again

are four times greater than those of a woman who has not had fraternal twins. In vitro fertilization increases a woman's chances for having multiple birth.

The number of twins born in the United States rose between the early 1980s and the early 2000s. In 1980, there were 69,339 sets of twins born, and in 2002 there were 125,134 sets of twins born in the United States. According to data gathered by the Centers for Disease Control (CDC), there is considerable variation among the states in number and rate of twin births. In 1994, for example, the twin birth rate ranged from 19.8 per 1,000 live births in Idaho and New Mexico to 27.7 per thousand in Connecticut and Massachusetts. One factor that may influence the distribution of multiple births is whether the state provides insurance coverage for procedures such as in vitro fertilization (IVF) and other treatments to improve fertility. These procedures increase the chance of multiple births.

Ethnicity is another factor that may correlate to the twin birth rate. For 1994, the twin birth rate among non-Hispanic white mothers was 24.3 per 1000 live births; among non-Hispanic black mothers, 28.3 per 1000; and among Hispanic mothers, 18.6 per 1000. There are also significant differences internationally in the number of twins born with the rate in Belgium almost six times the rate in China.

The CDC also studies whether maternal age has any correlation with the rate of twin births. The data seem to suggest that mothers in states with rates of twin births higher than the overall rate for the United States are older on average, and mothers in states with rates of twin births lower than the overall rate for the United States are younger. Again, as in vitro fertilization is more widely done, the incidence of multiple births will increase.

Infancy

Parents should avoid giving twins very similar names. Twins should be treated as two individuals and not as a package. They may need to be fed at different times and may develop skills at different rates. It is important to spend time with each twin separately so that they become used to being separated from each other for short times and know that they are each valued as individuals.

Toddlerhood

To help twins understand who they are as individuals, parents should avoid dressing both twins the same. It is preferable that each child receive **toys** that are geared towards their individual interests rather than each receiving the same toy.

School age

Sibling rivalry can be more intense in twins than in siblings of different ages. This is not unusual, because teachers, coaches, and even parents tend to compare twins. All children compare themselves to their siblings, and having others do this regularly can add to the pressure and stress of being a twin. Parents should consider arranging to have the twins put in different classes in school to help foster individuality. Each twin will probably have different skills, interests, and friends, and they should be encouraged to peruse activities separately if their interests diverge. Helping teachers, coaches, **babysitters**, and friends understand that it is important to treat the twins as two separate people can be very important. Friends should be encouraged to give separate gifts for birthdays and holidays, taking each child's special interests and talents into account.

Common problems

Twins often have a harder time developing their own independent identities than other children. Twins are more likely to have low birth weights or be delivered prematurely than single babies.

Parental concerns

Raising twins can be more challenging than raising two single children. The children may need to eat, **sleep**, and be changed at different times when they are infants. It can also be more expensive, because things like car seats and cribs must be purchased at the same time instead of reused for the second child. Some stores have special discounts for parents of twins.

When to call the doctor

Parents should call the doctor if one or both of their children seems ill, just as they would for any other child or children.

Resources

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Twin girls. Although many twins like to dress and act alike, especially at a young age, others try to differentiate themselves from each other, particularly in the teen years. *(© Dennis Degnan/Corbis.)*

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KEY TERMS

Dizygotic—From two zygotes, as in non-identical, or fraternal twins. The zygote is the first cell formed by the union of sperm and egg.

Monozygotic—From one zygote, as in identical twins. The zygote is the first cell formed by the union of sperm and egg.

Placenta—The organ that provides oxygen and nutrition from the mother to the unborn baby during pregnancy. The placenta is attached to the wall of the uterus and leads to the unborn baby via the umbilical cord.

ORGANIZATIONS

National Organization of Mothers of Twins Clubs. PO Box 438, Thompsons Station, TN 37179–0438. Web site: <www.nomotc.org>.

Tish Davidson, A.M.

Tympanometry *see* **Audiometry**

U

Undescended testes

Definition

Also known as cryptorchidism, undescended testes is a congenital condition characterized by testicles that do not follow the normal developmental pattern of moving into the scrotum before birth.

Description

In the fetus, the testes are in the abdomen. As development progresses, they migrate downward through the groin and into the scrotum. This event takes place late in fetal development, during the eighth month of gestation. In some newborn boys the testes are not present in the scrotum, either because the testes did not descend or because the testes never developed in the fetus.

Demographics

Eighty percent of all undescended testes cases naturally correct themselves during the first year of life. Only 3 to 4 percent of full-term baby boys have undescended testes, and half of those complete the journey by the age of three months. Up to 30 percent of boys born prematurely have testes that have not yet made the full descent. In 5 percent of cases of undescended testes, the testis on one side is completely absent. In 10 percent of cases, both testes are completely absent.

Causes and symptoms

There are many different and complex reasons why one or both testes may not descend. Sometimes the failure is due to problems that occur during pregnancy with the tissues as they are developing or with hormone levels in the developing fetus. If the testes did not descend because they are absent, then the likely cause is different than for testes that are present but did not descend. In the case of absence, it is possible that the testes never developed at all because the blood flow was cut off to them as they were developing, preventing their formation. One or both of the testicles can be undescended; therefore, the scrotum can appear to be either missing or lopsided.

When to call the doctor

The doctor will check for the testes in the scrotum during the normal newborn examination. If the parent notices that their male infant's testes do not appear normal or do not appear to be present at all, the parent should alert the doctor. If the testes have not descended by the time the child is six months of age, the parent should call the doctor to begin discussing possible treatment options.

Diagnosis

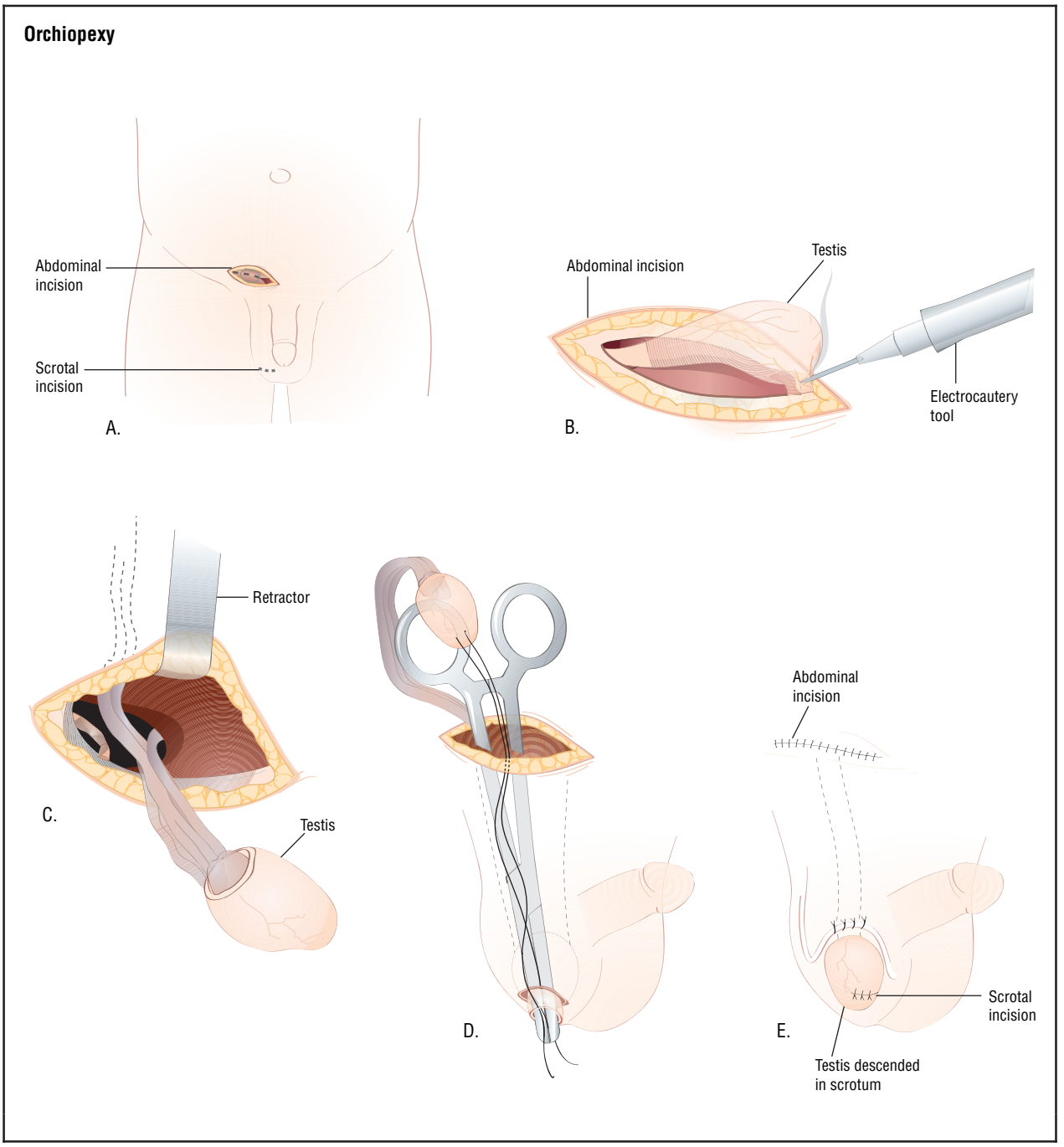
The newborn examination always checks for testes in the scrotum. It they are not found, a search will be conducted, but not necessarily right away. If the testes are present at all, they can be anywhere within a couple inches of the appropriate spot. In most cases, the testes will drop into place later. In 5 percent of cases, one testis is completely absent. In 10 percent of cases, the condition occurs on both sides. Presence of undescended testes is differentiated from absence of testicles by measuring the amount of gonadotropin hormone in the blood.

Treatment

Once it is determined that the testes will not naturally descend, treatment options must be considered. Hormone therapy is a possible treatment but does not have a very high success rate. Another treatment option is surgery. The procedure is called an orchidopexy and is relatively simple once the testes are located. The surgery is usually performed when the boy is between one and two years old.

Prognosis

Of full-term baby boys who have undescended testes, half will descend on their own without intervention



An orchiopexy is used to repair an undescended testicle in childhood. An incision is made into the abdomen, the site of the undescended testicle, and another is made in the scrotum (A). The testis is detached from surrounding tissues (B) and pulled out of the abdominal incision attached to the spermatic cord (C). The testis is then pulled down into the scrotum (D) and stitched into place (E). *(Illustration by Argosy, Inc.)*

by the age of three months. Eighty percent of all undescended testes cases naturally correct themselves during the first year of life. Of those cases that do not correct themselves naturally, intervention is very important, because undescended testes increase the likelihood of sterility and testicular **cancer**. Undescended testes are twice as likely to develop cancer as normally descended testes. Ten percent of all testicular cancers are in undescended testes. An adult man is three to 17 times more likely to develop testicular cancer if he has had a testis

KEY TERMS

Cryptorchidism—Undescended testes, a condition in which a boy is born with one or both testicles in the lower abdomen rather than the scrotum.

Embryonic—Early stages of life in the uterus.

Fetal— Refers to the fetus. In humans, the fetal period extend from the end of the eight week of pregnancy to birth.

Orchiopexy—A surgical procedure that places an undescended testicle in the scrotum and/or attaches a testicle to the scrotum.

that did not descend naturally. Surgery done to move the testis into the scrotum does not reduce the likelihood of malignancy but allows accessibility of the testes to screen for masses which will allow early treatment. The incidence of testicular cancer in men who did not have both testes descend normally is about 1 in 2000.

Many children who have undescended testes have reduced fertility as adults. It is thought that as many as 50 to 75 percent of children with undescended testes have problems with fertility as adults. Children with undescended testes are also more likely to develop hernias and have problems with their urinary tract.

Prevention

There is no known way to prevent undescended testes.

Parental concerns

Undescended testes are of concern because, although they are not known to be a threat to the child's immediate health, they are associated with an increased likelihood of negative outcomes later in life, including an increased likelihood of sterility and an increased incidence of testicular cancer.

Resources

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ORGANIZATIONS

American Urological Association. 1000 Corporate Blvd., Linthicum, MD 21090. Web site: <www.urologyhealth.org>.

Tish Davidson, A.M.

Ureter anomalies, congenital *see* **Congenital ureter anomalies**

Urinary reflux *see* **Vesicoureteral reflux**

Urinary tract infections *see* **Cystitis**

Urticaria *see* **Hives**

V

Vaccination

Definition

Vaccination introduces a vaccine into the body to produce immunity and prevent specific diseases.

Description

Many diseases that once caused widespread illness, disability, and death are now prevented by vaccines in developed countries. Vaccines are medicines that contain weakened or dead bacteria or viruses. When a child receives a vaccine, his or her immune system responds by producing antibodies, substances that weaken or destroy disease-causing organisms. When the child comes in contact with live bacteria or viruses of the same kind that are in the vaccine, the antibodies prevent those organisms from making the child sick. Vaccines also stimulate the cellular immune system. In other words, the child becomes immune to the disease the organisms normally cause. Building immunity by using a vaccine is called immunization. Childhood immunizations are safe and remain the most effective way to prevent disease.

Vaccines contain antigens (weakened or dead viruses, bacteria, and fungi that cause disease and infection). When introduced into the body, the antigens stimulate the immune system response by instructing B cells to produce antibodies, with assistance from T-cells. The antibodies are produced to fight the weakened or dead viruses in the vaccine. The antibodies ''practice'' on the weakened viruses, preparing the immune system to destroy real and stronger viruses in the future. When new antigens enter the body, white blood cells (called macrophages) engulf them, process the information contained in the antigens, and send it to the T-cells so that an immune system response can be mobilized.

General use

In the early 2000s, children in the United States and in other developed countries routinely have a series of vaccinations that begins at birth. Vaccinations in children began about 1900 with the smallpox vaccine. In 1960 there were only five vaccines in eight shots. The number of vaccinations children receive has steadily increased since that time. As of 2004, children receive 11 different vaccines given in up to 20 shots by age two years. Given according to a specific schedule, these vaccinations protect against **hepatitis B**; **diphtheria**, **tetanus**, pertussis (**whooping cough**) (DTP); **measles**, **mumps**, **rubella** (German measles); varicella (**chickenpox**); **polio**; pneumococcus; and *Haemophilus influenza* type B (Hib disease, a major cause of spinal **meningitis**) and, in some states, **hepatitis A**. This series of vaccinations is recommended by the American Academy of Family Physicians, the American Academy of Pediatrics, and the Centers for Disease Control and Prevention and is a requirement in all states before children can enter school. States make exceptions for children who have medical conditions such as **cancer** that prevent them from having vaccinations, and some states also make exceptions for children whose parents object for religious or other reasons.

Several vaccines are delivered in one injection, such as the measles-mumps-rubella (MMR) and diphtheria-tetanus-pertussis (DTP) combinations.

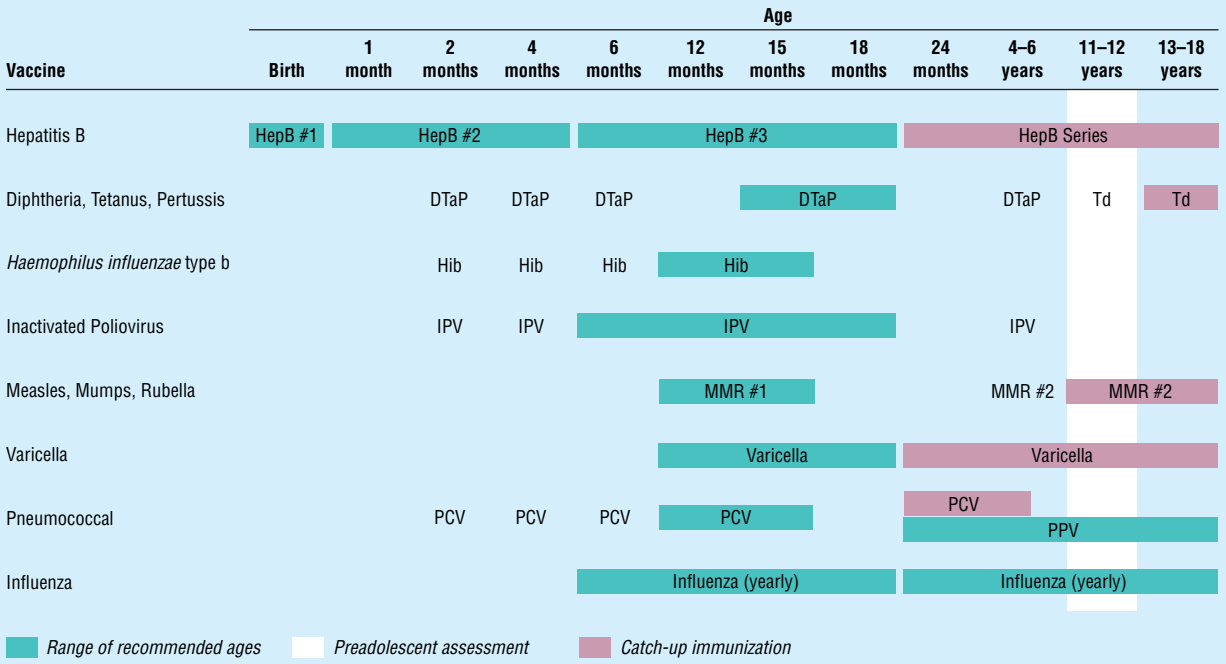
Vaccines are used in several ways. Some vaccines, such as the **rabies vaccine**, are given only when a child comes in contact with the virus that causes the disease, such as through a dog bite.

Recommendations for other vaccines and immunobiologic medicines depend on the child's health status or area of world where the family might travel. Such treatments are vaccine or immune globulin for hepatitis A, typhoid, meningitis, Japanese **encephalitis**, and **rabies**.

In addition the uses discussed above, vaccines are available for preventing anthrax, cholera, plague, **tuberculosis**, and yellow fever. Most vaccines are given as injections, but a few are taken orally.

The administration of vaccines to meet travel requirements should not interfere with or postpone any

Recommended childhood and adolescent immunization schedule, United States



SOURCE: Department of Human Services Centers for Disease Control and Prevention, 2005.

(Graph by GGS Information Services.)

of the routine childhood immunizations. If necessary, the routine immunization schedule can be accelerated to give as many vaccines as possible before departure. Decisions about vaccinations for children with chronic illnesses are made with the child's doctor.

Parents who are planning to travel with children to another country should find out what vaccinations are needed. Some vaccinations may be needed 12 weeks before the trip, so getting this information early is important. Many major hospitals and medical centers have travel clinics that provide this information. The traveler's health section of the Centers for Disease Control and Prevention also has information on vaccination requirements.

A vaccination health record helps parents and healthcare providers keep track of a child's vaccinations. The record should start when the child has his or her first vaccination and should be kept up-to-date with each added vaccination. While most doctors follow the recommended vaccination schedule, some flexibility is allowed. For example, vaccinations scheduled for age two months may be given anytime between six to ten weeks. Slight departures from the schedule do not keep the child from developing immunity, as long as all the vaccinations are received close to the right times.

Precautions

Vaccines are not always effective, and there is no way to predict whether a vaccine will "take" in any particular child. To be most effective, vaccination programs depend on the whole community participating. An increase in the number of vaccines given to children and the increased percentage of children receiving vaccines has resulted in a dramatic decrease in the number of vaccine-preventable diseases. In the United States, most young parents as of 2004 had never seen many of diseases that vaccines prevent. Even people who do not develop immunity through vaccination are safer because their friends, neighbors, children, and coworkers are immunized.

Factors influencing recommendations for childhood vaccination include age-specific risks of disease and

complications, the ability of a given age group to respond to the vaccine, and the potential interference with the immune response to transferred maternal antibody. There are vaccines for the youngest age group at risk for developing the disease and known to develop a satisfactory antibody response to the vaccination.

Like most medical procedures, vaccination has risks as well as great benefits. When children receive a vaccine, parents should be told about both. Questions or concerns should be discussed with a doctor or other healthcare provider. The Centers for Disease Control and Prevention, located in Atlanta, Georgia, is also a good resource for information.

Vaccines may cause problems for children with certain **allergies**. Children who are allergic to the **antibiotics** neomycin or polymyxin B should not take rubella vaccine, measles vaccine, mumps vaccine, or the combined measles-mumps-rubella (MMR) vaccine. Children who have had a severe allergic reaction to baker's yeast should not take the **hepatitis B vaccine**. Patients who are allergic to antibiotics such as gentamicin sulfate, streptomycin sulfate, or other amino glycosides should check with their doctors before the taking influenza vaccine, as some influenza vaccines contain small amounts of these drugs. Also, some vaccines, including those for influenza, measles, and mumps, are grown in the laboratory in fluids of chick embryos, and should not be given to children who are allergic to eggs. In general, parents of children who have had an unusual reaction to a vaccine in the past should report the reaction to the doctor before taking the same vaccine again. Doctors need to know about allergies to foods, medicines, preservatives, or other substances.

Children with other medical conditions should be given vaccines with caution. Influenza vaccine may reactivate Guillain-Barre syndrome (GBS) in patients who have had it before. This vaccine also may worsen illnesses that involve the lungs, such as **bronchitis** or **pneumonia**. Vaccines that cause fever as a side effect may trigger seizures in people who have a history of seizures caused by fever.

Certain vaccines are not recommended during pregnancy. However, women who are at risk of getting specific disease such as polio may receive the vaccine to prevent medical problems in their babies. Vaccinating a pregnant woman with tetanus toxoid can prevent tetanus in the baby at birth.

Women should avoid becoming pregnant for three months after taking rubella vaccine, measles vaccine, mumps vaccine, or the combined measles-mumps-rubella (MMR) as these vaccines may cause problems in the unborn baby.

Women who are breastfeeding should check with their doctors before taking any vaccine.

Side effects

Most side effects from vaccines are minor and easily treated. The most common are **pain**, redness, and swelling at the injection site. Some children may also develop a fever or a rash. Rarely, vaccines may cause severe allergic reactions, swelling of the brain, or seizures. Unusual reaction after receiving a vaccine should be reported to the doctor right away.

Interactions

Vaccines may interact with other medicines and medical treatments. When this happens, the effects of the vaccine or the other medicine may change or the risk of side effects may be greater. Radiation therapy and cancer drugs may reduce the effectiveness of many vaccines or may increase the chance of side effects. Parents should let the doctor know of all medicines taken by the child and learn whether the possible interactions could interfere with the therapeutic effects of the vaccine or the other medicines.

Parental concerns

All vaccines used for routine child vaccinations in the United States may be given simultaneously. There is no evidence that simultaneous administration of vaccines either reduces vaccine effectiveness or increases the risk of adverse events. The only vaccines which should not be given at the same time are cholera and yellow fever vaccines.

Some vaccines are mixed in one solution, such as measles-mumps-rubella (MMR) and diphtheria-tetanus-pertussis (DTP) combination. A survey of the literature as of 2004 indicated no evidence supporting the idea that multiple vaccines in any way overwhelm or weaken the immune system. Most young infants have strong immune systems that are capable of responding to all the recommended vaccines. The protection from bacterial and viral infections provided by vaccines preserves the infant's immune systems to fight off other infections.

Most doctors follow the recommended vaccination schedule, with some flexibility. For example, vaccinations that are scheduled for age two months may be given anytime between six to 10 weeks. Slight departures from the schedule will not stop the child from developing immunity, as long as the child gets all the vaccinations close the right times.

KEY TERMS

Anthrax—A bacterial infection, primarily of livestock, that can be spread to humans. In humans it affects the skin, intestines, or lungs.

Antibody—A special protein made by the body's immune system as a defense against foreign material (bacteria, viruses, etc.) that enters the body. It is uniquely designed to attack and neutralize the specific antigen that triggered the immune response.

Bacteria—Singluar, bacterium; tiny, one-celled forms of life that cause many diseases and infections.

Cholera—An infection of the small intestine caused by a type of bacterium. The disease is spread by drinking water or eating foods that have been contaminated with the feces of infected people. It occurs in parts of Asia, Africa, Latin America, India, and the Middle East. Symptoms include watery diarrhea and exhaustion.

Encephalitis—Inflammation of the brain, usually caused by a virus. The inflammation may interfere with normal brain function and may cause seizures, sleepiness, confusion, personality changes, weakness in one or more parts of the body, and even coma.

Feces—The solid waste, also called stool, that is left after food is digested. Feces form in the intestines and pass out of the body through the anus.

Guillain-Barré syndrome—Progressive and usually reversible paralysis or weakness of multiple muscles usually starting in the lower extremities and often ascending to the muscles involved in respiration. The syndrome is due to inflammation and loss of the myelin covering of the nerve fibers, often associated with an acute infection. Also called acute idiopathic polyneuritis.

Immune system—The system of specialized organs, lymph nodes, and blood cells throughout the body that work together to defend the body against foreign invaders (bacteria, viruses, fungi, etc.).

Immunization—A process or procedure that protects the body against an infectious disease by stimulating the production of antibodies. A vaccination is a type of immunization.

Inflammation—Pain, redness, swelling, and heat that develop in response to tissue irritation or injury. It usually is caused by the immune system's response to the body's contact with a foreign substance, such as an allergen or pathogen.

Meningitis—An infection or inflammation of the membranes that cover the brain and spinal cord. It is usually caused by bacteria or a virus.

Microorganism—An organism that is too small to be seen with the naked eye, such as a bacterium, virus, or fungus.

Organism—A single, independent unit of life, such as a bacterium, a plant, or an animal.

Plague—A serious, potentially life-threatening infectious disease caused by the bacterium *Yersinia pestis*. The disease is usually transmitted to humans by the bites of infected rodent fleas. There are three major types: bubonic, pneumonic, and septicemic.

Seizure—A sudden attack, spasm, or convulsion.

Tuberculosis—Tuberculosis (TB) is a potentially fatal contagious disease that can affect almost any part of the body, but is mainly an infection of the lungs. It is caused by a bacterial microorganism, the tubercle bacillus or *Mycobacterium tuberculosis*. Symptoms include fever, weight loss, and coughing up blood.

Typhoid fever—A severe infection caused by a bacterium, *Salmonella typhi*. People with this disease have a lingering fever and feel depressed and exhausted. Diarrhea and rose-colored spots on the chest and abdomen are other symptoms. The disease is spread through poor sanitation.

Virus—A small infectious agent consisting of a core of genetic material (DNA or RNA) surrounded by a shell of protein. A virus needs a living cell to reproduce.

Yellow fever—An infectious disease caused by a virus. The disease, which is spread by mosquitoes, is most common in Central and South America and Central Africa. Symptoms include high fever, jaundice (yellow eyes and skin) and dark-colored vomit, a sign of internal bleeding. Yellow fever can be fatal.

Immunizations are not given when a child has signs of an acute illness. An interrupted primary series of immunizations need not started again but may simply continue after the child recovers. The child's doctor is the best person to decide when each vaccination should be given.

The eventual goal in child care is to reduce stress. Parents should try to increase the child's feeling of security and well-being by close involvement with the immunization process. Providing explanations of the immunization plan, special tests, and procedures suitable to the child's age is helpful. Infants and toddlers are not likely to understand verbal explanations, but they have a strong parental attachment and need affection to ease fears. Small children also have an urgent need for their mothers to defend them during medical treatments. Older children may even protest or despair in getting an injection but are usually accepting of reasonable explanations.

The health-care professional reviews the immunization record and the health status of the child at each visit. If necessary the nurse or doctor helps the parent correctly position the child and exposure of the injection site. Parents should hold a small child on their laps securely for the injection; older children may be put on the examination table in the doctor's office. After the injection, parents can give the child immediate comfort to control crying and then leave the treatment room.

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Varicella zoster *see* **Chickenpox**

Varicella zoster vaccine (VZV) *see* **Chickenpox vaccine**

Vasculitides

Definition

Vasculitides is the plural of the word vasculitis, which may be used to describe any disorder characterized by inflammation of the blood or lymph vessels. Vasculitis is not a distinctive disease in its own right, but rather a symptom or characteristic of a number of different diseases. It can affect any type or size of blood vessel—large arteries and veins as well as arterioles, venules, or capillaries. The term juvenile vasculitides is sometimes used to refer to a group of disorders that primarily affect children and adolescents. These disorders vary widely in their severity as well as the specific blood vessels and organs affected. Some are mild and may resolve even without treatment, while others are potentially life-threatening. The most common childhood vasculitides are **Kawasaki syndrome** (sometimes called Kawasaki disease) and Henoch-Schönlein purpura.

The most widely used classification scheme for the vasculitides was first proposed at an international conference in 1994. It classifies these disorders according to the size of the blood vessels involved:

- Large-sized vessel vasculitis. This category includes two disorders, only one of which—Takayasu arteritis (TA)—is found in children and adolescents.
- Medium-sized vessel vasculitis. This category includes infantile polyarteritis nodosa (IPAN) and Kawasaki disease.
- Small-sized vessel vasculitis. The disorders in this category include Henoch-Schönlein purpura (HSP) and Wegener's granulomatosis.

Description

Vasculitis may damage blood vessels in two different ways. In some cases the inflamed tissue becomes weakened and stretches, producing a bulge in the wall of the vessel known as an aneurysm. The aneurysm may eventually rupture or burst, allowing blood to escape into nearby tissues. In other cases, the inflammation causes the blood vessel to narrow, sometimes to the point that blood can no longer flow through the vessel. When enough of the larger vessels supplying a specific organ

or other part of the body are closed by inflammation, the tissue that is starved for blood may die. The area of dead tissue is called an infarction or infarct.

The early symptoms of vasculitis frequently include **fever**, weakness, loss of appetite, weight loss, tiring easily, pains in the muscles or joints, and swollen joints. Some of the childhood vasculitides affect the skin, producing **rashes**, ulcers, or reddish-purple spots known as purpura. Others affect the lungs, digestive tract, kidneys, liver, nervous system, eyes, or brain, resulting in symptoms ranging from **pain** in the abdomen, **diarrhea**, coughing, or high blood pressure to shortness of breath, visual disturbances, **headache** or fainting, and **numbness** in the limbs. The specific symptoms of the more common childhood vasculitides are described in more detail below.

Transmission

Some of the childhood vasculitides may be preceded and possibly triggered by infectious diseases. In addition, Kawasaki disease sometimes occurs in epidemics, such as those reported in Japan in 1979, 1982, and 1985. No epidemics, however, have been reported since 1985.

Demographics

Most vasculitides are relatively rare disorders; one source estimates that about 100,000 persons (including adults as well as children and adolescents) are hospitalized each year in the United States for treatment of vasculitis. Although this number is small compared to the number of those treated for **cancer** or diabetes, the vasculitides can nonetheless have a significant financial and emotional impact on the families of children diagnosed with them.

The demographics of specific childhood vasculitides are as follows:

- Henoch-Schönlein purpura (HSP). HSP most commonly affects children between the ages of three and 12 years. The sex ratio is about 1.5–2 males for every one female. The disease is more common in North America between November and January; it is estimated to affect 14 or 15 children per 100,000. About 50–60 percent of children with HSP were diagnosed with **strep throat** or another upper respiratory infection two to three weeks before the onset of the vasculitis.
- Kawasaki disease. Kawasaki disease affects between one and three children per 10,000 in the United States each year. It is much more common in Japan, striking one child per thousand below the age of five. In the United States, Kawasaki disease is more common among children of Japanese descent than among children from other racial or ethnic backgrounds. It is also more likely to affect African Americans than Caucasians. The sex ratio is about 1.5 males for every one female. Kawasaki disease is primarily a disease of younger children; the average age at onset is 18 months, with 80 percent of cases found in children younger than five years.
- Infantile polyarteritis nodosa (IPAN). IPAN is a rare disease, and is sometimes described as a severe variant of Kawasaki disease. The incidence of IPAN in the United States is uncertain as of the early 2000s, primarily because of disagreements among doctors about the classification of childhood vasculitides; the most common figure given is 0.7 cases per 100,000 children. The first known case of IPAN was reported in London in 1870, although polyarteritis nodosa in adults was first described in 1852. Like Kawasaki disease, IPAN is more common in children of Asian descent. The male to female ratio is not known for certain, but is thought to be about two to one.
- Takayasu arteritis (TA). TA, which was first described by a Japanese ophthalmologist in 1908, is primarily a disease of adolescent and young adult women, although it has been diagnosed in children as young as six months. TA is relatively rare, affecting about 2.6 individuals per million. In the United States it is most common in young women of Japanese descent, with a male to female ratio of one to eight. In India, however, TA is more commonly associated with **tuberculosis**, and the sex ratio is two females for every one male.
- Wegener's granulomatosis. Wegener's granulomatosis is also a rare disease, diagnosed in one to three individuals per 100,000, with only 3 percent of these cases found in people below 20 years of age. It is, however, one of the most serious vasculitides. The male to female ratio is two to one.

Causes and symptoms

Causes

There is no single disease process that underlies all the childhood vasculitides. Various causes have been proposed for specific disorders.

- Henoch-Schönlein purpura. Although the ultimate cause of HSP was unknown as of 2004, the disease is preceded by an acute upper respiratory infection in at least half the children diagnosed with it. In other cases HSP appears to be triggered by an immune complex reaction to certain vaccines (most commonly vaccines for typhoid, **measles**, yellow fever, or cholera) or

medications (most commonly penicillin, erythromycin, quinidine, or quinine). A characteristic finding in children diagnosed with HSP is higher levels of immunoglobulin A (IgA) in the blood and deposits of IgA on the walls of the child's blood vessels.

- Kawasaki disease. It is thought that an infectious organism of some kind is the cause of Kawasaki disease, although no specific virus or bacterium has been identified as of 2004. The disease has been linked to a variety of disease agents, including parvovirus B19, **HIV infection**, measles, **influenza** viruses, rotaviruses, adenoviruses, *Klebsiella pneumoniae*, and *Mycoplasma pneumoniae*. Some doctors think that genetic and immunologic factors are involved as well as an infectious organism.
- Infantile polyarteritis nodosa. As with Kawasaki disease, various infectious organisms have been proposed as the cause of IPAN, including **hepatitis B** virus, Epstein-Barr virus (EBV), various retroviruses, streptococci, and even a virus usually found in cats. None of these viruses or bacteria has been found in all patients with IPAN, however. Another theory is that IPAN is an immune complex disease, but its trigger had not been identified as of 2004.
- Takayasu arteritis. The cause of TA is unknown as of the early 2000s but may involve genetic factors, as the disease has been reported in identical twins.
- Wegener's granulomatosis. The cause of Wegener's granulomatosis was not known as of 2004. As with other childhood vasculitides, various disease organisms (including fungi as well as bacteria or viruses) have been suggested as the cause, but none have been definitely identified. A genetic cause seems unlikely, as it is unusual for two people in the same **family** to develop the disease.

Symptoms

The early symptoms of the childhood vasculitides are often difficult to distinguish from those of other illnesses. This section will focus on the symptoms specific to each disease.

- Henoch-Schönlein purpura (HSP). HSP is an acute but self-limited illness characterized by a low-grade fever (around 100.4°F [38°C]), purpura, joint pains (usually in the ankles and knees), abdominal pain, bleeding in the digestive tract, and inflammation of the kidneys. Boys with HSP often have inflammation of the testicles.
- Kawasaki disease. Kawasaki disease has three stages: an acute stage lasting about 11 days, characterized by a high fever (over 104°F [40°C]), strawberry tongue and cracked lips, **conjunctivitis**, involvement of the liver, kidneys, and digestive tract, and inflammation of the heart muscle; a subacute phase lasting about three weeks, characterized by irritability, loss of appetite, the peeling of skin from the finger tips, and the development (in about 20 percent of patients) of aneurysms in the coronary artery; and a convalescent phase marked by expansion of the aneurysms and possible heart attack. As of the early 2000s, Kawasaki disease is the leading cause of acquired heart problems for children in the developed countries.
- Infantile polyarteritis nodosa. The early symptoms of IPAN are nonspecific, usually including fever, loss of appetite, weight loss, and pain in the abdomen. The disease is most likely to affect the kidneys, heart, or liver. Depending on the organ(s) involved, the child may develop aneurysms in the arteries supplying the kidneys, kidney failure, aneurysms in the coronary artery, congestive heart failure, massive bleeding in the digestive tract, aneurysms in the arteries supplying the brain, and **stroke**. About half of patients with IPAN develop pains in the joints or skin rashes; boys frequently have pain in the testicles.
- Takayasu arteritis. Takayasu arteritis is a chronic inflammatory disorder that affects the aorta (the large artery that leaves the heart) and its major branches. Its early symptoms include fever, weight loss, and a general feeling of tiredness. The disease may not be diagnosed for months or even years, however. The inflammation of the aorta eventually leads either to the formation of aneurysms or the narrowing or complete blocking of the blood vessels. The patient may feel aching or pain in parts of the body affected by inadequate blood supply, such as aching in the legs while walking or cramping sensations in the abdomen after meals. In rare cases, the patient may suffer a heart attack or stroke. The patient may develop high blood pressure if the blood supply to the kidneys is affected. TA is sometimes called pulseless disease because the doctor may not be able to detect the pulse on one side of the patient's body. Another diagnostic clue is a significant difference (greater than 30 mm Hg) in the blood pressure on the right and left sides of the body.
- Wegener's granulomatosis. Wegener's most commonly affects the upper respiratory tract, the eyes, ears, kidneys, and skin. The disease is called a granulomatosis because it is characterized by the formation of granulomas, which are small lumps or nodules of inflammatory cells in the patient's tissues. The patient may have recurrent ear infections that are slow to heal, inflammation of the tissues inside the eye, inflamed sinuses, nosebleeds, coughing up blood, narrowing of the windpipe, and saddle nose, which is a deformity caused by the collapse of cartilage inside the nose. The

patient may also have joint pains, loss of appetite, skin lesions, and fever. Vasculitis associated with Wegener's may lead to a heart attack. If untreated, the disease eventually progresses to kidney failure and death.

When to call the doctor

Although not all children who are eventually diagnosed with vasculitis will have all of the following signs and symptoms, parents should consult the doctor if most are present:

- The child's symptoms are constitutional; that is, they affect his or her overall physical health on a basic level. Malaise (a general feeling of physical discomfort), loss of appetite, fever, and loss of energy are examples of constitutional symptoms.
- The symptoms involve more than one organ or organ system.
- The child has noticeable purpura.
- The child has **tingling** or other unusual sensations followed by numbness in certain parts of the body.

Diagnosis

The diagnosis of vasculitis is complicated by several factors. To begin with, many of the early symptoms of the childhood vasculitides are not specific to these disorders and may have a wide range of other possible causes. In many cases the doctor may not be able to make the diagnosis until specific organs or organ systems are affected. The doctor will begin by ruling out such other possible diagnoses as bacterial or viral infections, collagen vascular disease, hypersensitivity reactions, and malignant tumors.

Another factor that complicates diagnosis is that the various childhood vasculitides have overlapping symptom profiles. Although lists of diagnostic criteria have been drawn up for the various disorders in this group, some patients do not meet the full criteria of any one disorder.

The first step in diagnosis is taking a careful history. The child's doctor may be able to narrow the diagnostic possibilities on the basis of the patient's age, sex, ethnicity, and a history of recent vaccinations or upper respiratory infections. The doctor will also ask whether the child is experiencing fever, abdominal cramping, diarrhea, or pains in the muscles and joints. The second step is a general physical examination. Several of these disorders affect the circulation or eyes as well as the skin. For example, Takayasu arteritis affects the patient's pulse and blood pressure, as well as producing small hemorrhages in the retina of the eye, while Kawasaki disease is characterized by conjunctivitis (inflammation of the tissues lining the eyelids). The doctor will examine the child's skin for purpura, other skin rashes or ulcers, reddening or swelling of the skin, and will note the locations of these abnormalities. In most cases the doctor will refer the child to a specialist for further evaluation. The specialist may be a pediatric rheumatologist, cardiologist, neurologist, or specialist in infectious diseases.

Laboratory tests for vasculitis include blood and urine tests. The blood tests include a complete blood count, a blood serum chemistry panel, erythrocyte sedimentation rate (ESR), tests for rheumatoid factor and circulating immune complexes, and tests for antineutrophil cytoplasmic antibodies (c-ANCA and p-ANCA). Urinalysis is done to evaluate kidney function. The doctor may also order skin, muscle, lung, or sinus biopsies in order to distinguish among the various childhood vasculitides.

Imaging studies that are used to diagnose the various childhood vasculitides include chest **x rays** or CT scans of the sinuses (Wegener's granulomatosis); CT scan of the aorta, angiography or ultrasonography (Takayasu's arteritis); arteriography or echocardiography (IPAN); chest x ray and echocardiography (Kawasaki disease); chest x ray, abdominal ultrasound, or barium contrast study of the digestive tract (Henoch-Schönlein purpura).

Treatment

The treatment of children with one of these disorders is highly individualized; it is tailored to the specific organs affected and the child's overall condition. Henoch-Schönlein purpura usually resolves on its own without any specific therapy. The general goals of treatment for vasculitis are to reduce inflammation in the affected blood vessels; maintain an adequate blood supply to the vital organs and skin; and monitor the side effects of the medications given to treat vasculitis.

Medications

Most patients with vasculitis will be given corticosteroids (usually prednisone) to reduce joint pain if present and inflammation in the blood vessels. Other types of drugs that are commonly used are the immunosuppressants (usually cyclophosphamide, methotrexate, azathioprine, or etanercept) and anticoagulants (usually heparin). Immunosuppressants are drugs that are given to treat inflammation by lowering the intensity of the body's reaction to allergens and other trig-

gers, while anticoagulants are given to prevent blood clots from forming and blocking blood vessels that have already been narrowed by inflammation. Children with muscle or joint pain may be given **non-steroidal anti-inflammatory drugs**, or NSAIDs, many of which are available without a prescription. An antibiotic (usually trimethoprim-sulfamethoxazole) is sometimes given to children with Wegener's granulomatosis to control flares, or recurrences of the disease.

Surgery

Patients with Takayasu arteritis often require surgical repair of damaged arteries. The most common procedures used are balloon angioplasty or stenting. Stents are small metal tubes or wires that are inserted into damaged blood vessels to hold them open. In severe cases, the damaged section of the artery may have to be removed completely and replaced with a graft made from an artificial material.

Alternative treatment

There is little information about the use of alternative treatments for vasculitides in children, most likely because the disorders in this category are relatively uncommon and vary widely with regard to the organ systems affected, symptom severity, and prognosis. One Chinese medical journal has reported on the benefits of treating children with Henoch-Schönlein purpura with a remedy made from colquhounia root, while a team of Dutch researchers has observed that acupuncture appears to be effective in reducing the inflammation associated with vasculitis. The researchers noted, however, that large randomized trials comparing acupuncture with mainstream treatments had not been undertaken as of 2004.

Some herbal preparations have been associated with harmful effects on the heart and circulatory system; however, the cases that have been reported mostly involve either contaminated or adulterated herbal products, or interactions between prescription medications and herbal preparations. The herbs most frequently mentioned in these case reports are aconite, ephedra, and licorice. The extent of the problem is not known as of the early 2000s because no large-scale analyses have been done. In any event, however, parents should *never* give a child a herbal remedy without first consulting the child's doctor, whether or not the child is taking prescription drugs.

Nutritional concerns

Children who develop high blood pressure with one of these disorders are usually placed on a low-sodium diet.

Prognosis

The prognoses for the childhood vasculitides vary widely depending on the disease and the extent of organ involvement. In general, children whose lungs or kidneys are affected have poorer outcomes.

- Henoch-Schönlein purpura. The prognosis for children with HSP is generally good, as the disease usually goes away by itself even without treatment; however, about a third of patients have recurrences.
- Kawasaki disease. Patients who are treated promptly have a good prognosis. The mortality rate in the early 2000s is estimated at 0.1–2 percent.
- Infantile polyarteritis nodosa. IPAN has a poor prognosis even when treated aggressively; the 10-year survival rate for this vasculitis is only 20 percent.
- Takayasu arteritis. TA is a chronic disorder with a high rate of relapse; the 15-year survival rate is about 95 percent.
- Wegener's granulomatosis. Untreated Wegener's is fatal, usually within five months. With treatment, about 87 percent of patients have a remission of the disease, but 53 percent have recurrences.

Prevention

Apart from minimizing a child's exposure to strep throat and similar upper respiratory infections, there is nothing that parents can do to prevent vasculitis in children, in that the cause(s) of these disorders are still unknown.

Parental concerns

The impact of childhood vasculitis on a child's family varies widely, depending on the child's age at onset, the specific symptoms of the disorder, its severity, the types of medications or other treatments that are needed, and the prognosis. Parents should work closely with the child's pediatrician and other specialists who may be involved to monitor the child's progress through regular follow-up appointments. The child's primary doctor can usually provide advice regarding such concerns as medication side effects, limitations on the child's activities if any, and explaining the disease to the child and other family members.

See also Conjunctivitis; Kawasaki syndrome.

KEY TERMS

Aneurysm—A weakened area in the wall of a blood vessel which causes an outpouching or bulge. Aneurysms may be fatal if these weak areas burst, resulting in uncontrollable bleeding.

Aorta—The main artery located above the heart that pumps oxygenated blood out into the body. The aorta is the largest artery in the body.

Conjunctivitis—Inflammation of the conjunctiva, the mucous membrane covering the white part of the eye (sclera) and lining the inside of the eyelids also called pinkeye.

Flare—A sudden worsening or recurrence of a disease.

Granuloma—An inflammatory swelling or growth composed of granulation tissue

Henoch-Schönlein purpura—A syndrome sometimes classified as a hypersensitivity vasculitis, associated with a variety of digestive symptoms, pain in the joints, and kidney involvement. Purpura comes from the Latin word for "purple" and refers to the reddish-purple spots on the skin caused by leakage of blood from inflamed capillaries.

Infarct—An area of dead tissue caused by inadequate blood supply.

Kawasaki syndrome—A syndrome of unknown origin that affects the skin, mucous membranes, and the immune system of infants and young children. It is named for the Japanese pediatrician who first identified it in 1967.

Malaise—The medical term for a general condition of unease, discomfort, or weakness.

Rheumatologist—A doctor who specializes in the diagnosis and treatment of disorders affecting the joints and connective tissues of the body.

Saddle nose—A sunken nasal bridge.

Stent—A slender hollow catheter or rod placed within a vessel or duct to provide support or to keep it open.

Strawberry tongue—A sign of scarlet fever in which the tongue appears to have a red coating with large raised bumps.

Takayasu arteritis—A disease in which the aorta and its major branches become inflamed. It is often accompanied by high blood pressure, an abnormal pulse, and visual symptoms.

Vasculopathy—Any disease or disorder that affects the blood vessels.

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American College of Rheumatology (ACR). 1800 Century Placce, Suite 250, Atlanta, GA 30345-4300. (404) 633-3777. Fax: (404) 633-1870. <http://www.rheumatology.org>.

Arthritis Foundation. P. O. Box 7669, Atlanta, GA 30357-0669. (800) 283-7800. Web site: <http://www.arthritis.org>.

National Institute of Arthritis and Musculoskeletal and Skin Diseases (NIAMS). 1 AMS Circle, Bethesda, MD 20892-3675. (301) 495-4484 or (877) 22-NIAMS. Fax: (301) 718-6366. Web site: <http://www.niams.nih.gov>.

National Institute of Neurological Disorders and Stroke (NINDS). National Institutes of Health. 9000 Rockville Pike, Bethesda, MD 20892. (301) 496-5751. Web site: <http://www.ninds.nih.gov>.

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Rebecca Frey, PhD

Vegetarianism

Definition

Vegetarianism is the voluntary abstinence from eating meat. Vegetarians refrain from eating meat for various reasons, including religious, health, and ethical ones. Lacto-ovo vegetarians supplement their diet with dairy (lactose) products and eggs (ovo). Vegans (pronounced vee-guns) do not eat any animal-derived products at all.

Description

Vegetarianism has been steadily gaining acceptance as an alternative to the meat-and-potatoes bias of the traditional American diet. Several factors contribute to the interest in vegetarianism in the United States. Outbreaks of **food poisoning** from meat products, as well as increased concern over the additives in meat such as hormones and **antibiotics**, have led some people and professionals to question meat's safety. There is also an increased awareness of the questionable treatment of farm animals in factory farming.

But the growing health consciousness of Americans is probably the major reason for the surge in interest in vegetarianism. **Nutrition** experts have built up convincing evidence that there are major problems with the conventional American diet, which is centered on meat products that are high in cholesterol and saturated fat and low in fiber. Heart disease, **cancer**, and diabetes, which cause 68 percent of all deaths in America, are all believed to be influenced by this diet.

A vegetarian diet has many well-documented health benefits. It has been shown that vegetarians have a longer life expectancy than those who eat a meat-centered diet. The U.S. Food and Drug Administration (FDA) has stated that data has shown vegetarians to have a strong or significant probability against contracting **obesity**, heart disease, lung cancer, colon cancer, **alcoholism**, **hypertension**, diabetes, gallstones, gout, kidney stones, and ulcers. However, the FDA also points out that vegetarians tend to have healthy lifestyle habits, so other factors may contribute to their increased health besides diet alone.

Vegetarians have a huge number of statistics in their favor when it comes to presenting persuasive arguments in favor of their eating habits. Vegetarians claim that a vegetarian diet is a major step in improving the health of citizens and the environment. Americans eat over 200 pounds (91 kilograms) of meat per person per year. The incidence of heart disease, cancer, diabetes, and other

diseases has increased along with the dramatic increase in meat consumption during the twentieth century.

Many statistics show significantly smaller risks for vegetarians contracting certain conditions. The risks of women getting breast cancer and men contracting prostrate cancer are nearly four times as high for frequent meat eaters as for those who eat meat sparingly or not at all. For heart attacks, American men have a 50 percent risk of having one, but the risk drops to 15 percent for lacto-ovo vegetarians, and to only 4 percent for vegans. For cancer, studies of populations around the world have implied that plant-based diets have lower associated risks for certain types of cancer.

Nutritionists have repeatedly shown in studies that a healthy diet consists of plenty of fresh vegetables and fruits, complex carbohydrates such as whole grains, and foods that are high in fiber and low in cholesterol and saturated fat. Vegetarianism, a diet that fulfills all these criteria, has become part of many healthy lifestyles.

Some nutritionists have designed transition diets to help people become vegetarian in stages. Many Americans eat meat products at nearly every meal, and the first stage of a transition diet is to substitute just a few meals a week with wholly vegetarian foods. Then, particular meat products can be slowly reduced and eliminated from the diet and replaced with vegetarian foods. Red meat can be reduced and then eliminated, followed by pork, poultry, and fish. For those wishing to become pure vegetarians or vegans, the final step is to choose other nutrient-rich foods in order to eliminate eggs and dairy products. Individuals should be willing to experiment with transition diets and should have patience when learning how combine vegetarianism with social activities such as dining out.

The transition to vegetarianism can be smoother for adolescents who make informed choices with dietary practices. Sound nutritional guidelines include decreasing the intake of fat, increasing fiber, and emphasizing fresh fruits, fresh vegetables, beans and lentils, and whole grains in the diet while avoiding processed foods and sugar.

Thanks to the growing interest in vegetarianism, many meat substitutes are now readily available. Tofu and tempeh are made from soybeans that are high in protein, calcium, and other nutrients. There are ''veggieburgers'' that can be grilled like hamburgers, and vegetarian substitutes for hot dogs, corn dogs, chicken, turkey, ham, bologna, pastrami, and sausage with surprisingly authentic textures and taste. Major vegetarian meat substitute brands include Morningstar Farms, Boca, Gardenburger, and Lightlife. There are many vegetarian cookbooks on the market as well as magazines such as *Vegetarian Times*, *Veggie Life*, and *Vegetarian Journal*.

Famous vegetarians, past and present, include Leonardo da Vinci, Sir Isaac Newton, Leo Tolstoy, Ralph Waldo Emerson, Gandhi, physician Albert Schweitzer, writer George Bernard Shaw, champion tri-athlete Dave Scott, and musicians Paul McCartney, George Harrison, John Lennon, Yoko Ono, Alanis Morissette, Bob Dylan, and Bruce Springsteen.

Infancy, toddlerhood, and preschool

Babies, toddlers, and preschoolers can do well on a vegetarian diet, especially one that includes eggs and dairy products. If they are not included, the young child may suffer from shortages of **vitamins** B12, B2, and D; protein; calcium; and zinc. The child may also need iron supplements because iron in plant food is not absorbed well.

Infants and toddlers require many calories in order to grow at the normal rate. At about seven to eight months of age, babies are ready to start eating protein-rich foods. Instead of pureed meats, vegetarian infants should be given protein alternatives such as pureed peas, beans, and lentils, cottage cheese, pureed tofu, and yogurt.

It is important that toddlers eat high-calorie vegetarian foods such as diced nuts, olives, dates, and avocados so they get enough calories. Most importantly, parents should make sure a vegetarian child eats a wide variety of foods, according to a 2002 advisory from the journal *Clinical Reference Systems*.

Parents must take care to insure the child gets enough food for growth, since a vegetarian diet relies heavily on bulk foods that are filling but usually short of calories. Parents who are vegetarians and want their baby to be one should discuss the topic with a pediatrician. Young children who are vegetarians should be monitored regularly to make sure their weight and height are appropriate for their age.

School age

About 2 percent of Americans age six to 17 (about 1 million) are vegetarian, the same percentage as among American adults, and 0.5 percent are vegan, according to a 2002 survey by the Vegetarian Resource Group (VRG). Six percent of six to 17 year olds do not eat meat but do eat fish and/or poultry.

Teens who follow a vegetarian diet are more likely to meet recommendations for total fat, saturated fat, and number of servings of fruits and vegetables as compared

to non-vegetarians. They also have higher intakes of iron, vitamin A, fiber, and diet soda, and lower intakes of vitamin B12, cholesterol, and fast food. Most teens, whether they were vegetarian or not, do not meet recommendations for calcium, according to the VRG survey.

The survey concluded that rather than viewing adolescent vegetarianism as a phase or fad, the diet could be viewed as a healthy alternative to the traditional American meat-based diet. The survey also stated that vegetarian diets in **adolescence** could lead to lifelong health-promoting dietary practices. The survey was reported in the July-August 2002 issue of the VRG publication *Vegetarian Journal*.

Common problems

In general, a well-planned vegetarian diet is healthy and safe. However, vegetarians, and particularly vegans who eat no animal products, need to be aware of particular nutrients that may be lacking in non-animal diets. These are amino acids, vitamin B12, vitamin D, calcium, iron, zinc, and essential fatty acids. Infants and growing children have higher requirements for these nutrients.

Vegetarians should be aware of getting complete protein in their diets. A complete protein contains all of the essential amino acids, which are the building blocks for protein essential to the diet because the body cannot make them. Meat and dairy products generally contain complete proteins, but most vegetarian foods such as grains and beans contain incomplete proteins, lacking one or more of the essential amino acids. However, vegetarians can easily overcome this by combining particular foods in order to create complete proteins. For instance, beans are high in the amino acid lysine but low in tryptophan and methionine, but rice is low in lysine and high in tryptophan and methionine. Thus, combining rice and beans makes a complete protein.

Eating dairy products or nuts with grains also makes proteins complete. Oatmeal with milk on it is complete, as is peanut butter on whole wheat bread. Proteins do not necessarily need to be combined in the same meal, but generally within four hours.

Getting enough vitamin B12 may be an issue for some vegetarians, particularly vegans, because meat and dairy products are the main sources. Vitamin supplements that contain vitamin B12 are recommended. Spirulina, a nutritional supplement made from algae, is also a vegetarian source, as are fortified soy products and nutritional yeast.

Vitamin D can be obtained by vitamins, fortified foods, and sunshine. Calcium can be obtained in enriched tofu, seeds, nuts, beans, dairy products, and dark green

KEY TERMS

Amino acid—An organic compound composed of both an amino group and an acidic carboxyl group. Amino acids are the basic building blocks of proteins. There are 20 types of amino acids (eight are "essential amino acids" which the body cannot make and must therefore be obtained from food).

Cholesterol—A steroid fat found in animal foods that is also produced in the human body from saturated fat. Cholesterol is used to form cell membranes and process hormones and vitamin D. High cholesterol levels contribute to the development of atherosclerosis.

Essential fatty acid (EFA)—A fatty acid that the body requires but cannot make. It must be obtained from the diet. EFAs include omega-6 fatty acids found in primrose and safflower oils, and omega-3 fatty acids oils found in fatty fish and flaxseed, canola, soybean, and walnuts.

Gout—A metabolic disorder characterized by sudden recurring attacks of arthritis caused by deposits of crystals that build up in the joints due to abnormally high uric acid blood levels. In gout, uric acid may be overproduced, underexcreted, or both.

Hypertension—Abnormally high arterial blood pressure, which if left untreated can lead to heart disease and stroke.

Lacto-ovo vegetarian—People who do not eat meat, but do include dairy products and eggs in their diets.

Lysine—A crystalline basic amino acid essential to nutrition.

Methionine—An amino acid that, when not metabolized properly, allows homocysteine to build up in the blood. Folic acid aids methionine metabolism.

Spirulina—A genus of blue-green algae that is sometimes added to food to increase its nutrient value.

Tryptophan—An essential amino acid that has to consumed in the diet because it cannot be manufactured by the body. Tryptophan is converted by the body to niacin, one of the B vitamins, and serotonin, a neurotransmitter.

Vegan—A vegetarian who does not eat eggs or dairy products.

vegetables, including broccoli, kale, spinach, and collard greens. Iron is found in raisins, figs, beans, tofu, whole grains, potatoes, and dark green leafy vegetables. Iron is absorbed more efficiently by the body when iron-containing foods are eaten with foods that contain vitamin C, such as fruits, tomatoes, and green vegetables. Zinc is abundant in nuts, pumpkin seeds, beans, whole grains, and tofu.

For vegetarians who do not eat fish, getting enough omega-3 essential fatty acids may be an issue, and supplements such as flaxseed oil should be considered, as well as consumption of walnuts and canola oil. Another essential fatty acid, omega-6, found in fish, can be obtained from borage oil or evening primrose oil supplements.

Vegetarians do not necessarily have healthier diets. Some studies have shown that some vegetarians consume large amounts of cholesterol and saturated fat. It is quite possible to be a vegetarian yet eat an unhealthy fast-food or junk food diet. Eggs and dairy products contain cholesterol and saturated fat, while nuts, oils, and avocados are vegetable sources of saturated fat. To reap the full benefits of a vegetarian diet, vegetarians should be conscious of cholesterol and saturated fat intake.

Parental concerns

Parents should closely monitor their vegetarian child's height, weight, and general health. A child who is not getting enough vitamins, **minerals**, and other nutrients may have symptoms such as skin **rashes**, fatigue, a painful and swollen tongue, irritability, pale skin, mental slowness, or difficulty breathing. The diets of vegetarian adolescents should be monitored closely to make sure they are eating a variety of foods, including fruits, vegetables, beans, whole grains, and non-meat protein sources.

When to call the doctor

Parents should consult their child's pediatrician or physician if they are unsure the child's vegetarian diet is nutritionally adequate. A doctor should also be consulted if a child's weight or height is not appropriate for their age.

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The Vegetarian Resource Group. PO Box 1463, Baltimore, MD 21203. Web site: <www.vrg.org>.

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Venereal diseases *see* **Sexually transmitted diseases**

Vertigo *see* **Dizziness**

Vesicoureteral reflux

Definition

Vesicoureteral reflux (VUR) is a condition in which urine flows from the bladder, back up the ureter, and back into the kidneys.

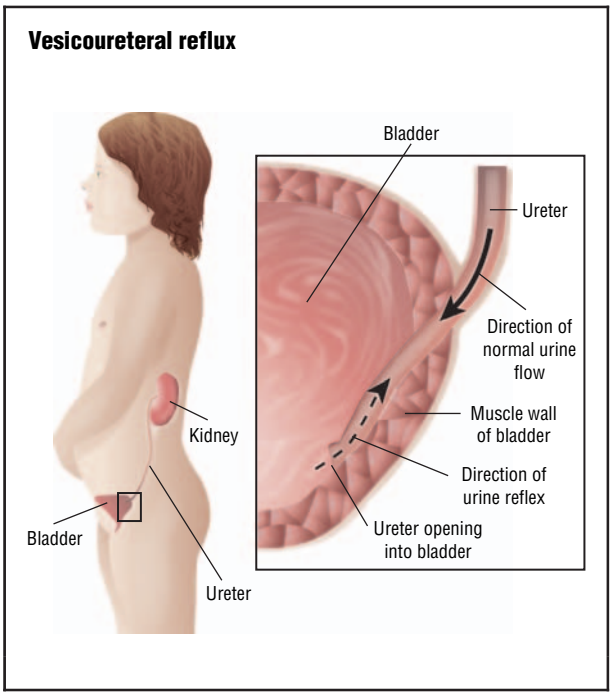

Illustration of vesicoureteral reflux in a child, a condition in which urine abnormally flows back up into the ureters, causing repeated urinary tract infections. *(Illustration by GGS Information Services.)*

Description

The normal flow of urine begins in the collecting system of each kidney. Urine then flows out of each kidney and into a tube called the ureter. Each ureter leads into the bladder, where the urine collects until it is passed out of the body. Normally, urine flows only in this direction. In vesicoureteral reflux, however, urine that has already collected in the bladder is able to flow backwards from the bladder, up the ureter, and back into the collecting system of the kidney. VUR may be present in either one or both ureters.

Vesicoureteral reflux causes damage to the kidneys in two ways. The kidney is not designed to withstand very much pressure. When VUR is present, backpressure of the urine on the kidney is significant. This can damage the kidney. Second, the kidney is usually sterile, meaning that no bacteria are normally present within it. In VUR, bacteria that enter through the urinary tract may be carried back up the ureter with the urine. These bacteria can enter the kidney, causing severe infection.

Demographics

VUR mostly occurs in the prenatal stage and may be observed at birth (congenital), although it may not be detected until an infection heralds its presence. VUR may run in families. The condition affects about 1 percent of all children. More boys than girls have VUR.

Causes and symptoms

Most cases of VUR are due to a defect in the way the ureter is implanted into the bladder. The angle may be wrong or the valve (which should allow urine only one-way entrance into the bladder) may be weak. Structural defects of the urinary system may also cause VUR. These include a situation in which two ureters leave a kidney, instead of the usual one (duplicated ureters) and in which the ureter is greatly enlarged at the end leading into the bladder (ureterocele).

VUR alone does not usually cause symptoms. Symptoms develop when an infection has set in. The usual symptoms of infection are frequent need to urinate, **pain** or burning with urination, and blood or pus in the urine. Occasionally, VUR is suspected when a child has a difficult time becoming toilet trained. In these cases, the bladder may become irritable and spastic, because it is never totally empty of urine. When the kidneys have been damaged, high blood pressure may develop. Over time, severe damage and scarring of the kidneys leads to kidney failure.

Diagnosis

Urinary tract infections are diagnosed through laboratory examination of urine samples. Kidney size and scarring can be assessed through ultrasound examination of the kidneys.

VUR itself is diagnosed by a test called a voiding cystourethrogram. This test involves inserting a small tube (catheter) into the bladder. The bladder is then filled with a dye solution, which lights up on the x-ray picture. A series of pictures are taken immediately, followed by x rays taken while the patient is urinating. This tracking allows reflux to be demonstrated and also reveals whether the level of reflux increases when pressure increases during urination. Reflux is then graded as follows based on the height and effects of the VUR:

- Grade I: VUR enters just the portion of the ureter closest to the bladder. The ureter appears normal in size.
- Grade II: VUR enters the entire ureter and goes up into the collecting system of the kidney. The ureter and the collecting system appear normal in size and structure.
- Grade III: VUR enters the entire ureter and kidney collecting system. Either the ureter or the collecting system is abnormal in size or shape.

KEY TERMS

Bladder—The muscular sac which receives urine from the kidneys, stores it, and ultimately works to remove it from the body during urination.

Reflux—The backward flow of a body fluid or secretion. Indigestion is sometimes caused by the reflux of stomach acid into the esophagus.

Ureter—The tube that carries urine from the kidney to the bladder; each kidney has one ureter.

- Grade IV: Similar to grade III, but the ureter is greatly enlarged.
- Grade V: Similar to grade IV, but the ureter is also abnormally twisted/curved, and the collecting system is greatly enlarged, with absence of the usual structural details.

Once VUR has been diagnosed, its progress may be followed with a nuclear scintigram, in which a radioactive substance is put into the bladder via catheter, and a gamma camera takes images that reveal the presence and degree of VUR. This test exposes the child to less radiation than does a standard VCUG. Doppler ultrasound techniques were as of 2004 under study as a radiation exposure-free alternative to VCUG.

Treatment

Treatment depends on the grade that is diagnosed. In grades I and II, the usual treatment involves long-term use of a small daily dose of **antibiotics** to prevent the development of infections. The urine is tested regularly to make sure that no infection occurs. The kidneys are evaluated regularly via ultrasound and VCUG (every 12 to 18 months) to make sure that they are growing normally and that no new scarring has occurred. Grades III, IV, and V VUR can be treated with antibiotics and careful monitoring. New infections, scarring, or stunting of kidney growth may result in the need for surgery. Grades IV and V are extremely likely to require surgery.

Surgery for VUR consists of reimplanting the ureters into the bladder at a more normal angle. This adjustment usually improves the functioning of the valve leading into the bladder. When structural defects of the urinary system are present, surgery will almost always be required to repair these defects.

Prognosis

Prognosis is dependent on the grade of VUR. About 80 percent of children with grades I and II VUR simply grow out of the problem. As they grow, the ureter lengthens, changing its angle of entry into the bladder and resolving the reflux. The average age of VUR resolution is about six to seven years. About 50 percent of children with grade III VUR require surgery. Nearly all children with grades IV and V VUR require surgery. In these cases, it is usually best to perform surgery when the patient is relatively young, in order to avoid damage and scarring to the kidneys.

Prevention

While as of 2004 there was no known method of preventing VUR, it is important to note that a high number of the siblings of children with VUR also have VUR. Many of these siblings (about 36%) have no symptoms but are discovered through routine examinations prompted by their brother's or sister's problems. It is important to identify these children, so that antibiotic treatment can be used to prevent the development of infection and kidney damage.

Parental concerns

It is important that parents of children with VUR understand the importance of following the instructions for antibiotic administration. Although their child may not appear at all ill, the antibiotics are crucial to protecting the health and development of their child's kidneys. Children with VUR should also be monitored for the development of **constipation**, which can complicate the VUR. Problems with bladder emptying can make toilet teaching a slower process in children with VUR.

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Video games

Definition

Video games are electronic, interactive games known for their vibrant colors, sound effects, and complex graphics.

Description

First mass-marketed in the 1970s, video games are played by installing cartridges into a game box connected by wire to a television set. The child then manipulates a joystick or controller to control the actions of a character or series of characters as the characters face obstacles displayed on the screen. Video games, designed chiefly to appeal to children and adolescents, can also be played in arcades, on computers, and on small, hand-held screens.

As of 2004 nearly every home in the United States with children had one or more of the most popular game systems, for example, Nintendo GameCube, Sony Playstation2, or Microsoft Xbox. Few children have not been exposed to some form of video game, and access to the games is readily available to children from all walks of life.

Video games for home use proved popular from the start. Children are particularly attracted to them for a variety of reasons. Fantasy characters and situations appeal to young imaginations and provide an escape from everyday routine and the stresses presented by parents, friends, and school. In addition, the games give children a level of control that they do not experience in real life, as the characters on the screen respond to the children's commands. Players also receive immediate rewards for making the right moves. Most games can be played at a variety of skill levels so that every player can be challenged.

The popularity of video games has been matched by the controversy they have sparked among parents, psychologists, and educators. The most prevalent objection results from the violent themes and characters that dominate in most video games. A 1989 study by the National Coalition on Television Violence (NCTV) found that, of the 95 most popular home video games, 58 percent were war games and 83 percent featured violent themes. As technology has improved to allow the games to show situations and characters that are more realistic, debate has escalated about the potential effects of video games on children's behavior. One NCTV study that monitored the playground behavior of eight- to ten-year-olds immediately after playing a laser-weapon game found an 80 percent increase in fighting. There is also added concern that repeated exposure to violence desensitizes children to its effects. Other experts and video game manufacturers contend that negative effects have not been proven adequately, and, in fact, playing such games gives players an avenue for the harmless release of stress and aggression.

Public pressure prompted some video game manufacturers in the early 1990s to begin labeling games with warnings about violent or sexually explicit content. In 1994, in response to considerable political pressure and the possibility of a federal rating agency, the industry created its own rating system, overseen by the Entertainment Software Rating Board (ESRB). Ratings are assigned based on the games' suitability for various age groups. An "Early Childhood" designation on a game box indicates that the game is suitable for players ages three and older, and there is no violence, sexual content, or profanity. The designation "Everyone" indicates the game is for players ages six and older and may contain minimal violence or crude language. A "Teen" game for ages 13 and up may contain violence, profanity, and mild sexual themes. A "Mature" rating is considered suitable only for ages 17 and older and may include more intense violence, profanity, and mature sexual themes. "Adults Only" games are not intended for people under 18 and may include graphic depictions of sex and violence. The ratings system, however, is just a guide, and parents still need to oversee which video games their children buy and **play**.

In the past, the issue of gender bias in video games was another area of considerable debate. Not only were most video games male-oriented **sports** and combat games, female characters in the games were portrayed as victims to be rescued by the male hero or objects of violence or sexual desire. In the early 2000s, however, an increasing number of games had girl-oriented themes and an increasing number of gender neutral games became available.

Besides the socialization concerns presented by video games, medical concerns were also raised in the early 1990s, when video games were linked to epileptic seizures experienced by some 50 children. About one third of the children had experienced previous seizures, and there were questions about whether the seizures they experienced were related to playing or watching a video game. Two large studies later reported that the children who experienced video game-related seizures (VGRS) were particularly sensitive to light and that video games with flashing lights merely precipitated, rather than caused, the seizures. Sitting too close to the screen could exacerbate the effects of the light sensitivity, as could the increasingly complex graphic technology featured in

games. Individuals with epilepsy are not thought to be particularly susceptible to VGRS, and no lasting neurological damage had as of 2004 been linked to these seizures.

Despite the controversy surrounding video games, benefits have also been noted: development of **hand-eye coordination**, increases in concentration, logical thinking skills, and healthy competition among children, as well as socialization skills gained from sharing strategies and the heightened **self-esteem** resulting from successful performances. One research study even found that doctors who had played more video games had better surgical skills.

Toddlerhood

There are a number of specialized video and computer games that are designed to be educational for toddlers. Many use familiar characters to teach basic things such as shape matching, the alphabet, and counting.

Preschool

Children in **preschool** can be exposed to video and computer games that reinforce the basic skills that they are learning, such as phonetics, shapes, colors, and basic addition.

Elementary school

School-age children can be encouraged to play educational games that reinforce what they are learning in the classroom. Parents should research the games that their children want to buy to ensure appropriate content for the child's age group. In the early 2000s marketers have developed increasing numbers of educational games that are also adventurous and exciting. Children, especially young ones, should be encouraged to play these instead of more violent games.

Middle and high school

The effect of violent games on behavior and social development is an especially important concern for older children. These children often spend much of their time playing video games when their parents are not present to supervise the content. Also, many teens buy video games with money earned from allowances or part time jobs, making it harder for parents to control which titles are purchased.

Studies have begun to find significant correlations between violence in video games and violence in real life. One study done on eighth and ninth graders compared teens who generally had personalities considered non-aggressive but who played violent games to those teens who had aggressive personalities but did not play violent games. The researchers found that the non-aggressive, video game playing teens were actually more likely to get in physical fights than the teens considered aggressive but who did not play video games.

KEY TERMS

Entertainment Software Rating Board (ESRB)—The industry board that rates video games.

Video game related seizures (VGRS)—Seizures thought to be brought on by the flashing lights and complex graphics of a video game.

Some states are trying to pass laws that would make it illegal to sell video games with certain ratings to people under the age for which the games are intended. Even if laws are created to try to prevent underage sale of very violent video games, parents should still be alert to what their teen is playing. Making the teen play video games in a common area and not in his or her bedroom with the door closed can be an important first step in regulating game play and facilitating discussion.

Common problems

Children often become very involved in video games and do not want to stop playing them. Setting concrete limits about the amount of time that can be spent playing games and then enforcing these limits is essential. Even educational games should not be played to excess, because playing video games is not a substitute for positive social interaction or traditional learning. Children can also be encouraged to play the games with other children, because discussing strategies and problem solving in a group is a positive social activity.

Parental concerns

The amount and degree of violence in video games is an important concern for parents. Monitoring the games that a child buys or rents and plays is an important way to help deal with this problem. If a child plays a violent video game at an arcade or another child's house, it can be helpful to discuss the difference between games and reality and to discuss what the real life repercussions of the actions taken in the game would be.

When to seek help

If a child has violent or **aggressive behavior** or a tendency to mimic the negative actions taken by charac-

ters in a video game it may be helpful to consult a mental health professional to discuss possible solutions.

See also Television habits.

Resources

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Violence and violent behavior *see* **Aggressive behavior**

Vision development *see* **Eye and vision development**

Vitamin D deficiency

Definition

Vitamin D deficiency exists when the concentration of 25-hydroxy-vitamin D (25-OH-D) in the blood serum occurs at 12 nanograms/milliliter (ng/ml) or less. This is one-half to one-fourth the amount normally present. When vitamin D deficiency continues for many months in growing children, the disease commonly referred to as rickets occurs.

Description

Vitamin D is a fat-soluble vitamin, meaning it can be dissolved in fat. While some vitamin D is supplied by the diet, most of it is made in the body. To make vitamin D, cholesterol, a substance widely distributed in animal tissues, the yolk of eggs, and various oils and fats, is necessary. Once cholesterol enters the body, a slight alteration in the cholesterol molecule occurs, with one change taking place in the skin. This alteration requires ultraviolet light, a component of sunlight. Vitamin D deficiency and rickets tend to occur in children who do not get enough sunlight and who do not eat foods that are rich in vitamin D.

Once consumed or made in the body, vitamin D is further altered to produce a substance called 1,25-dihydroxy-vitamin D (1,25-diOH-D). The conversion of vitamin D to 1,25-diOH-D occurs in the liver and kidney. The role of 1,25-diOH-D in the body is to keep the concentration of calcium at a constant level in the bloodstream. Maintaining calcium at a constant level is absolutely required for human life, since dissolved calcium is required for nerves and muscles to work. One of the ways in which 1,25-diOH-D accomplishes this is by stimulating the absorption of dietary calcium by the intestines.

The sequence of events that can lead to vitamin D deficiency and later to bone disease, is as follows: a lack of vitamin D in the body creates an inability to manufacture 1,25-diOH-D. This results in decreased absorption of dietary calcium and an increased loss of calcium in the feces. When this happens, the bones are affected. Vitamin D deficiency results in a lack of bone mineralization (calcification) in growing children.

Demographics

Vitamin D deficiency is not common in the United States and other industrialized countries because of the wide availability of vitamin D fortified infant formulas and milks. It is somewhat more common in northern areas where there is not as much sunlight present during many parts of the year. Vitamin D deficiency is also slightly more common in inner city areas, because environmental factors, such as smog, can block the necessary ultraviolet (UV) component of sunlight. Children with darkly pigmented skin are more likely to be vitamin D deficient than light skinned children. Children who are exclusively breast-fed without vitamin D supplementation, particularly if they are not exposed to sunlight, are at higher risk of vitamin D deficiency.

Causes and symptoms

Vitamin D deficiency can be caused by conditions that result in little exposure to sunlight. These conditions include: living in northern regions, having dark skin, and having little chance to go outside. Children whose faces and bodies remain covered when outside can develop vitamin D deficiency even while living in a sunny

climate. In-born errors of vitamin D metabolism can also cause vitamin D deficiency and rickets; these children cannot convert inactive vitamin D to active vitamin D and suffer the same symptoms as children with a nutritional deficiency.

Most foods contain little or no vitamin D. As a result, sunshine is often a deciding factor in whether vitamin D deficiency occurs. Although fortified milk and fortified infant formula contain high levels of vitamin D, human breast milk is rather low in the vitamin. (The term fortified means that **vitamins** are added to the food by the manufacturer.)

The Recommended Dietary Allowance (RDA) of vitamin D for both children and adults is 200 International Units (IU) per day. Saltwater fish such as salmon, herring, and sardines are naturally rich in vitamin D. Vitamin D fortified milk contains 400 IU per quart (liter), so half a quart (liter) of milk provides the RDA. For comparison, human breast milk contains only 4 to 60 IU per quart.

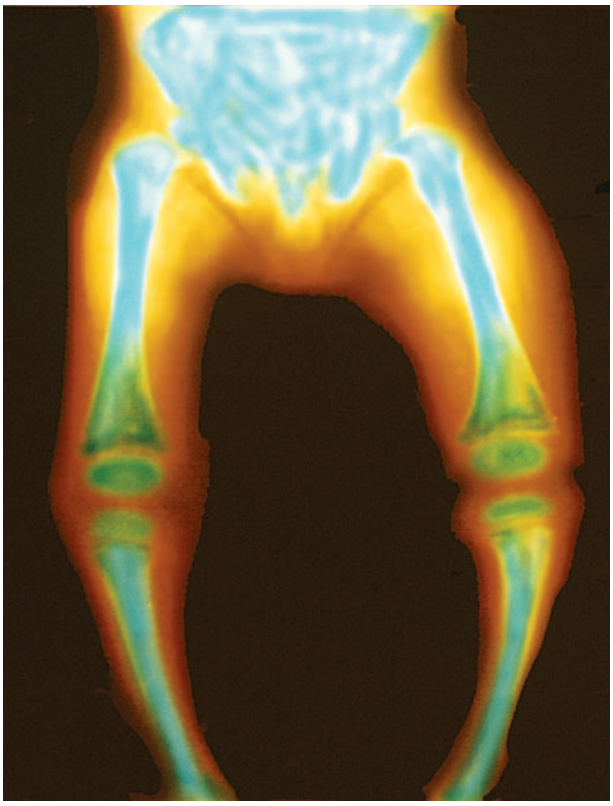
No harm is likely to result from vitamin D deficiency that occurs only a few days a year. If the deficiency occurs for a period of many months or years, however, rickets may develop. The symptoms of rickets include bowed legs and bowed arms. The bowed appearance is due to the softening of bones, and their bending if the bones are weight-bearing. Bone growth occurs through the creation of new cartilage, a soft substance at the ends of bones. When the mineral calcium phosphate is deposited onto the cartilage, a hard structure is created. In vitamin D deficiency, though, calcium is not available to create hardened bone, and the result is soft bone. Other symptoms of rickets include bony bumps on the ribs called rachitic rosary (beadlike prominences at the junction of the ribs with their cartilages) and knock-knees. Seizures may also occasionally occur in a child with rickets, because of reduced levels of dissolved calcium in the bloodstream.

When to call the doctor

The doctor should be called if the parent notices that the child has any signs of vitamin D deficiency or rickets. Such signs include skeletal **pain**, bowed limbs, and impaired growth. If there are lifestyle factors that make the child at risk for vitamin D deficiency, such as low milk or formula intake, a doctor should be consulted about the possibility of using vitamin D supplements.

Diagnosis

Vitamin D deficiency is diagnosed by measuring the level of 25-hydroxy-vitamin D in the blood serum. The normal concentration of this form of vitamin D ranges from 25 to 50 ng/ml. Deficiency occurs when this level decreases to about 12 ng/ml or less.



X ray of a child's lower body affected by rickets, a result of a vitamin D deficiency. *(© Dr. LR/Photo Researchers, Inc.)*

Rickets is diagnosed by x-ray examination of the leg bones. A distinct pattern of irregularities, abnormalities, and a coarse appearance can be clearly seen if a child has rickets. Measurements of blood plasma 25-OH-D, blood plasma calcium, and blood plasma parathyroid hormone must also be obtained for the diagnosis of this disease. Parathyroid hormone and 1,25-diOH-D work together in the body to regulate the levels of calcium in the blood.

Treatment

Rickets heals promptly with large doses vitamin D administered orally each day for approximately one month. During this treatment, the doctor should monitor the levels of 25-OH-D in the plasma to make sure that they are raised to a normal level. The bone abnormalities (visible by x ray) generally disappear gradually over a period of three to nine months. Parents are instructed to take their infants outdoors for approximately 20 minutes per day with their faces exposed. Children should be

KEY TERMS

25-hydroxy-vitamin D—The form of vitamin D that is measured in order to assess vitamin D deficiency.

Cholesterol—A steroid fat found in animal foods that is also produced in the human body from saturated fat. Cholesterol is used to form cell membranes and process hormones and vitamin D. High cholesterol levels contribute to the development of atherosclerosis.

Fat-soluble vitamin—A vitamin that dissolves easily in fat or oil, but not in water. The fat-soluble vitamins are vitamins D, E, A, and K.

International unit (IU)—A measurement of biological activity in which one IU is equal to one mg (milligram).

Rachitic rosary—Beadlike bumps present at the junction of the ribs with their cartilages. It is often seen in children with rickets.

Recommended Dietary Allowance (RDA)—The Recommended Dietary Allowances (RDAs) are quantities of nutrients in the diet that are required to maintain good health in people. RDAs are established by the Food and Nutrition Board of the National Academy of Sciences, and may be revised every few years. A separate RDA value exists for each nutrient. The RDA values refer to the amount of nutrient expected to maintain good health in people. The actual amounts of each nutrient required to maintain good health in specific individuals differ from person to person.

Rickets—A condition caused by the dietary deficiency of vitamin D, calcium, and usually phosphorus, seen primarily in infancy and childhood, and characterized by abnormal bone formation.

encouraged to **play** outside and to eat foods that are good sources of vitamin D. These foods include cod liver oil, egg yolks, butter, oily fish and also foods, including milk and breakfast cereals, that are fortified with synthetic vitamin D.

Care must be taken in treating vitamin D deficiency, since high doses of vitamin D are toxic (poisonous) and can result in the permanent deposit of **minerals** in the heart, lungs, and kidneys. Symptoms of toxicity are **nausea**, **vomiting**, pain in the joints, and lack of interest in eating food. In adults, vitamin D toxicity occurs with eating 50,000 IU or more per day. In infants, toxicity occurs with 1,000 IU per day. The continued intake of toxic doses results in death.

Rickets are usually treated with oral supplements of vitamin D, with the recommendation to acquire daily exposure to direct sunlight. An alternative to sunlight is the use of an ultraviolet lamp. When people use UV lamps, they need to cover their eyes to protect them against damage. Many types of sunglasses allow UV light to pass through, so only those that are opaque to UV light should be used. Attempts to acquire sunlight through glass windows fail to help the body make vitamin D because UV light does not pass through window glass.

Rickets may also occur with calcium deficiency, even when a child is regularly exposed to sunshine. This type of rickets has been found in various parts of Africa. The bone deformities are similar to, or are the same as, those that occur in typical rickets; however, calcium deficiency rickets is treated by increasing the amount of calcium in the diet. No amount of vitamin D can cure the rickets of a child with a diet that is extremely low in calcium. For this reason, it is recommended that calcium be given in conjunction with vitamin D supplementation.

Prognosis

The prognosis for correcting vitamin D deficiency and rickets is excellent. Vitamin D treatment results in the return of bone mineralization to a normal rate, the correction of low plasma calcium levels, the prevention of seizures, and a recovery from bone pain. On the other hand, already established deformities such as bowed legs and the rachitic rosary persist throughout adult life.

Prevention

Vitamin D deficiency is a very preventable. Eating foods that are high in vitamin D or foods that have been fortified with additional vitamins in combination with getting moderate amounts of exposure to direct sunlight, are usually enough to prevent vitamin D deficiency.

Some authorities still recommend exposure to sunshine as a way to prevent vitamin D deficiency, but early exposure to direct sunlight may be linked to a higher incidence of skin **cancer** later in life, so other experts recommend that infants not be taken into direct sunlight without protective coverings or sunscreen until at least six months of age. These experts recommend that supplemental drops or fortified formulas instead of direct sunlight provide infants' daily requirements of Vitamin D. Children playing in the sunlight with sunscreen on is

not an effective way for them to get vitamin D because the sunscreen inhibits its production in the skin.

Nutritional concerns

Vitamin D deficiency is caused by the child not getting enough vitamin D through **nutrition** and exposure to sunshine. Even after a case of vitamin D deficiency has successfully been resolved special care should be taken with the child's diet, as vitamin D deficiency can reoccur.

Parental concerns

Vitamin D deficiency can cause rickets, which can lead to permanently stunted or irregular growth. Vitamin D deficiency can usually be easily corrected if it is noticed early, and if so the symptoms often resolve themselves. However, negative effects such as short stature and pelvic deformations can be permanent.

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Vitamins

Definition

Vitamins are organic components in food that are needed in very small amounts for growth and for maintaining good health. The vitamins include vitamins D, E, A, and K (fat-soluble vitamins), and folate (**folic acid**), vitamin B_{12}, biotin, vitamin B_6, niacin, thiamin, riboflavin, pantothenic acid, and vitamin C (ascorbic acid) (water-soluble vitamins). Vitamins are required in the diet in only tiny amounts, in contrast to the energy components of the diet. The energy components of the diet are sugars, starches, fats, and oils, and these occur in relatively large amounts in the diet.

Most of the vitamins are closely associated with a corresponding vitamin deficiency disease. **Vitamin D deficiency** causes rickets, a disease of the bones. Vitamin E deficiency occurs only very rarely and causes nerve damage. Vitamin A deficiency, common throughout the poorer parts of the world, causes night blindness. Severe vitamin A deficiency can result in xerophthalmia, a disease that, if left untreated, results in total blindness. Vitamin K deficiency results in spontaneous bleeding. Mild or moderate folate deficiency, common throughout the world, can result from the failure to eat green, leafy vegetables or fruits and fruit juices. Folate deficiency causes megaloblastic anemia, which is characterized by the presence of large abnormal cells called megaloblasts in the circulating blood. The symptoms of megaloblastic anemia are tiredness and weakness. Vitamin B_{12} deficiency occurs with the failure to consume meat, milk, or other dairy products. Vitamin B_{12} deficiency causes megaloblastic anemia and, if severe enough, can result in irreversible nerve damage. Niacin deficiency results in pellagra, which involves skin **rashes** and scabs, **diarrhea**, and mental depression. Thiamin deficiency results in beriberi, a disease resulting in atrophy, weakness of the legs, nerve damage, and heart failure. Vitamin C deficiency results in scurvy, a disease that involves bleeding. Diseases associated with deficiencies in vitamin B_6, riboflavin, or pantothenic acid have not been found in the humans, though persons who have been starving or consuming poor diets for several months, might be expected to be deficient in most of the nutrients, including vitamin B_6, riboflavin, and pantothenic acid. Rarely, deficiency in B_6 results in neurologic problems. Issues of toxicity are connected to the over consumptions of vitamins, particularly E, K, and B. Also, lack of regulation in the vitamin industry means consumers ought only to buy well-known brands.

Some of the vitamins serve only one function in the body, while other vitamins serve a variety of unrelated functions. Hence, some vitamin deficiencies tend to result in one type of defect, while other deficiencies result in a variety of problems.

Description

Vitamin treatment is usually done in three ways: by replacing a poor diet with one that supplies the recommended dietary allowance, by consuming oral supplements, or by injections. Injections are useful for persons with diseases that prevent absorption of fat-soluble vitamins. Oral vitamin supplements are especially useful for persons who otherwise cannot or will not consume food that is a good vitamin source, such as meat, milk, or other dairy products. For example, a vegetarian who will

not consume meat may be encouraged to consume oral supplements of vitamin B_{12}.

Treatment of genetic diseases which impair the absorption or utilization of specific vitamins may require megadoses of the vitamin throughout one's lifetime. Megadose means a level of about 10 to 1,000 times greater than the RDA. Pernicious anemia, homocystinuria, and biotinidase deficiency are three examples of genetic diseases which are treated with megadoses of vitamins.

General use

People are treated with vitamins for three reasons. The primary reason is to relieve a vitamin deficiency, when one has been detected. Chemical tests suitable for the detection of all vitamin deficiencies are available. The diagnosis of vitamin deficiency is often aided by visual tests, such as the examination of blood cells with a microscope, the x-ray examination of bones, or a visual examination of the eyes or skin.

A second reason for vitamin treatment is to prevent the development of an expected deficiency. Here, vitamins are administered even with no test for possible deficiency. One example is vitamin K treatment of newborn infants to prevent bleeding. Food supplementation is another form of vitamin treatment. The vitamin D added to foods serves the purpose of preventing the deficiency from occurring in persons who may not be exposed much to sunlight and who fail to consume foods that are fortified with vitamin D, such as milk. Niacin supplementation prevents pellagra, a disease that occurs in people who rely heavily on corn as the main source of food and who do not eat much meat or milk. In general, the American food supply is fortified with niacin.

A third reason for vitamin treatment is to reduce the risk for diseases that may occur even when vitamin deficiency cannot be detected by chemical tests. One example is folate deficiency. The risk for cardiovascular disease can be slightly reduced for a large fraction of the population by folic acid supplements. And the risk for certain birth defects can be sharply reduced in certain women by folic acid supplements.

Vitamin treatment is important during specific diseases in which the body's normal processing of a vitamin is impaired. In these cases, high doses of the needed vitamin can force the body to process or use it in the normal manner. One example is pernicious anemia, a disease that tends to occur in middle age or old age and impairs the absorption of vitamin B_{12}. Surveys have revealed that about 0.1 percent of the general population, and 2–3 percent of the elderly, may have the disease. If left untreated, pernicious anemia leads to nervous system damage. The disease can easily be treated with large oral daily doses of vitamin B_{12} (hydroxocobalamin) or with monthly injections of the vitamin.

Vitamin supplements are widely available as over-the-counter products. But whether they work to prevent or curtail certain illnesses, particularly in people with a balanced diet, is in the early 2000s a matter of debate and ongoing research. For example, vitamin C is not proven to prevent the **common cold**. Yet millions of Americans take it for that reason. Consumers should ask a physician or pharmacist for more information on the appropriate use of multivitamin supplements.

The diagnosis of a vitamin deficiency usually involves a blood test. An overnight fast is usually recommended as preparation prior to withdrawal of the blood test so that vitamin-fortified foods do not affect the test results.

KEY TERMS

Genetic condition—A condition that is passed from one generation to the next but does not necessarily appear in each generation. Examples of genetic conditions include Down syndrome, Tay-Sach's disease, sickle cell disease, and hemophilia.

Plasma—A watery fluid containing proteins, salts, and other substances that carries red blood cells, white blood cells, and platelets throughout the body. Plasma makes up 50 percent of human blood.

Recommended dietary allowance (RDA)—The Recommended Dietary Allowances (RDAs) are quantities of nutrients in the diet that are required to maintain good health in people. RDAs are established by the Food and Nutrition Board of the National Academy of Sciences, and may be revised every few years. A separate RDA value exists for each nutrient. The RDA values refer to the amount of nutrient expected to maintain good health in people. The actual amounts of each nutrient required to maintain good health in specific individuals differ from person to person.

Serum—The fluid part of the blood that remains after blood cells, platelets, and fibrogen have been removed. Also called blood serum.

Vitamin status—The state of vitamin sufficiency or deficiency of any person. For example, a test may reveal that a patient's folate status is sufficient, borderline, or severely inadequate.

The response to vitamin treatment can be monitored by chemical tests, by an examination of red blood cells or white blood cells, or by physiological tests, depending on the exact vitamin deficiency.

Precautions

Vitamin A and vitamin D can be toxic in high doses. Side effects range from **dizziness** to kidney failure. Consumers should ask a physician or pharmacist about the correct use of a multivitamin supplement that contains these vitamins.

Side effects

Few side effects are associated with vitamin treatment if vitamins are taken within the prescribed dosages. Excessive intake of some B vitamins may impart a greenish color to urine. Any possible risks depend on the vitamin and the reason why it was prescribed. Consumers should ask a physician or pharmacist about how and when to take vitamin supplements, particularly those that have not been prescribed by a physician.

Parental concerns

The dosage of vitamin supplements should not exceed the recommended daily allowance without a recommendation by a physician. Recommended dosages vary with age, so parents should be should to give vitamins to children that are specially formulated for children. Vitamin bottles will list recommended doses for different age groups. Infants and toddlers may also benefit from vitamin supplements if they do not eat a variety of foods. Liquid vitamin supplements are available commercially for these young children.

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ORGANIZATIONS

American Academy of Family Physicians. 11400 Tomahawk Creek Parkway, Leawood, KS 66211–2672. Web site: <www.aafp.org/>.

American Academy of Pediatrics. 141 Northwest Point Boulevard, Elk Grove Village, IL 60007–1098. Web site: <www.aap.org/default.htm>.

American Association of Naturopathic Physicians. 8201 Greensboro Drive, Suite 300, McLean, VA 22102. Web site: <http://naturopathic.org/>.

American College of Obstetricians and Gynecologists. 409 12th St., SW, PO Box 96920, Washington, DC 20090–6920. Web site: <www.acog.org/>.

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Vocal cord dysfunction

Definition

Vocal cord dysfunction (VCD) is a disorder that occurs when the vocal cords move toward each other when a person breathes, narrowing the airway and causing wheezing and difficulty breathing. VCD is also called paradoxical vocal cord motion (PVCM).

Description

Normally when an individual breathes in (inhales) or out (exhales) the vocal cords are drawn apart by the muscles of the larynx (voice box) to make a wider opening for air to move into or out of the lungs. In an individual with vocal cord dysfunction, instead of being drawn apart, the vocal cords move together, narrowing and partially blocking the airway. This is called adduction of the vocal cords. Adduction of the vocal cords happens most commonly during inhalation, although it can also happen during exhalation. As a result of the narrowed airways, the individual may **cough**, wheeze, feel short of breath, or make a high-pitched, harsh sound (called **stridor**) with each breath.

VCD is often misdiagnosed as either **asthma** or exercise-induced bronchospasm. As a result, many individuals with VCD are treated with inhalers and steroids for asthma, which do not help control VCD and which have potentially harmful side effects.

Demographics

The number of people with VCD in the general U.S. population is unknown. The disorder often occurs in conjunction with asthma and is frequently a missed diagnosis. Several small studies have found that about 40 percent of individuals who have VCD also have asthma and that about 10 to 15 percent of individuals whose asthma does not respond to aggressive treatment (refractory asthma) actually have VCD.

VCD has been found in individuals as young as three and as old as 82. However, in adults it most often occurs between the ages of 20 and 40. In children it appears most often about age 14 or 15. VCD is much more common in females than in males. In children under 18, about 85 percent of individuals diagnosed with VCD are girls. In children, the disorder has a strong association with competitive **sports** and **family** orientation toward high achievement. In adults it has a strong association with **anxiety** and stress. This association with stress is present, but less frequent in children.

Causes and symptoms

VCD was first recognized in 1842, when it was thought that hysteria, a common designation at that time for several psychological conditions, brought about spasm of the muscles of the larynx. By 1900, it was generally accepted that VCD was the physical expression of stress or other psychological conditions. It was not until the 1980s that physicians began to revisit the assumptions about the disorder and examine more closely its physical causes. As of 2004, the causes of VCD was not completely clear.

In the early 2000s, it is thought that the disorder may have multiple causes and that some of the triggers may be different in children and adults. VCD appears to be associated with the following:

- injury to the brain cortex
- brainstem compression (mainly in children)
- Arnold-Chiari syndrome (mainly in children)
- gastroesophageal reflux disease (GERD; in children and adults)
- chronic sinus infection/postnasal drip
- strenuous **exercise** (often in children)
- exposure to inhaled irritants (smoke, toxic chemicals; mainly in adults)
- psychological causes (most obvious in adults)
- nerve injury during **congenital heart disease** surgery or other chest surgeries
- failure to respond to asthma treatments

VCD usually comes on suddenly. Between attacks, the individual can breathe normally. The symptoms of a VCD attack are varied, but most strongly imitate those of asthma. Its similarity to asthma, along with the fact that some people with VCD actually also have asthma, complicates diagnosis. Common signs and symptoms include the following:

- coughing (about 75% of individuals)
- wheezing
- stridor
- voice changes during an attack
- difficulty inhaling (most common)
- difficulty exhaling (less common; usually irritant-induced)
- panic, anxiety, **fear** of suffocating
- insufficient oxygen in the blood (hypoxia)
- chest tightness

- panting in short shallow breaths
- feeling like something is stuck in the throat
- skin turning blue

When to call the doctor

Immediate emergency medical assistance is essential whenever there are any signs of breathing difficulty.

Diagnosis

Diagnosis of VCD is quite difficult. VCD can mimic the symptoms of severe asthma, allergic reactions (**anaphylaxis**), spasm of the larynx (laryngospasm), or a foreign object lodged in the throat. VCD is often a diagnosis of exclusion, which means that other possibilities are considered first, and when these are eliminated, VCD is considered. This may require a lot of testing.

The best way to determine if an individual has VCD is by doing a laryngoscopy. In a laryngoscopy, a slender, flexible tube containing a fiber optic camera is inserted through the nose and down the throat to the larynx. This examination allows the doctor to see the vocal cords and watch how and when they move.

Since between attacks the vocal cords appear to move normally, it is necessary to trigger an attack. Individuals cannot voluntarily produce symptoms of VCD, so they are usually exposed to an irritant or undergo an exercise stress test in order to bring on a VCD attack. The doctor then watches the vocal cords move. A classic finding is that the vocal cords move toward each other when the individual inhales, leaving a small triangular hole or chink at the back of the larynx. Individuals with asthma do not show this triangular chink.

Most people go through a series of other tests and often get other diagnoses, most commonly refractory (unresponsive) asthma, before they have a laryngoscopy and receive a definite diagnosis of VCD. Other tests that are frequently done to pinpoint or eliminate certain respiratory disorders include arterial blood gas values (to measure oxygen in the blood), **pulmonary function tests** (to measure lung capacity), with flow-volume loops (to measure the rate of air flow at different points in the breathing process). A methacholine provocation test, which stimulates a response in asthmatics, but not in persons with VCD, also helps narrow the diagnosis.

Treatment

Treatment consists of two phases, immediate (acute) and long term. Acute care often occurs in a hospital emergency room. The most important aspect of acute care is to see that the individual is breathing and getting enough oxygen. Sometimes heliox therapy is given. Heliox is a mixture of 20 to 30 percent oxygen and 70 to 80 percent helium. Because this mixture is less dense and more oxygen-rich than regular air, it is easier to inhale. If the individual is still not getting enough oxygen, it may be necessary to perform a tracheotomy. In this operation, a tube is inserted in the larynx so that air can bypass the blockage.

Long-term therapy begins by stopping any treatments for other diagnoses such as asthma, and treating any underlying conditions, such as brainstem compression or GERD, affecting the disorder. Airborne irritants are removed from the individual's environment as much as possible. Speech therapy and teaching abdominal breathing techniques have been quite successful in preventing VCD attacks. If an individual does not respond adequately to speech therapy, psychotherapy is recommended, as in many people anxiety and stress are linked to VCD attacks. People can learn relaxation techniques and work through problems causing stress and anxiety. Occasionally anti-anxiety drugs are prescribed.

In an experimental procedure, botulinum toxin (Botox) may be injected into the larynx. The drug paralyzes the muscle, making it impossible for the vocal cords to move across the airway. This technique appears

KEY TERMS

Arnold-Chiari syndrome—A congenital malformation of the base of the brain.

Asthma—A disease in which the air passages of the lungs become inflamed and narrowed, causing wheezing, coughing, and shortness of breath.

Exercise-induced bronchospasm—A sudden contraction in the lower airway that causes breathing problems and is brought about by heavy exercise.

Gastroesophageal reflux disease (GERD)—A disorder of the lower end of the esophagus in which the lower esophageal sphincter does not open and close normally. As a result the acidic contents of the stomach can flow backward into the esophagus and irritate the tissues.

Laryngoscope—An endoscope that is used to examine the interior of the larynx.

Stridor—A term used to describe noisy breathing in general and to refer specifically to a high-pitched crowing sound associated with croup, respiratory infection, and airway obstruction.

to be successful but may require repeated injections as the toxin wears off. Another experimental device is a facemask that provides resistance when the individual inhales but not during exhalation. The resistance forces the person to breathe in more slowly and reduces stridor.

Alternative treatment

Some individuals have found biofeedback very helpful in controlling or moderating VCD attacks. Others have benefited from relaxation and mind control techniques.

Prognosis

The long-term outcome for VCD is not known and probably varies among individuals depends on the underlying cause of the disorder. Only a handful of people with VCD have been followed for 10 or more years, and all of them continued to have symptoms of the disorder. However, speech therapy and psychotherapy are often successful in reducing the number of attacks.

Prevention

Although the physical conditions that cause VCD cannot be prevented, individuals can be educated not to panic and to use certain breathing techniques when they begin to feel symptoms of VCD. In addition, airborne pollutants should be eliminated from the individual's environment. These steps can be somewhat successful in minimizing attacks.

Parental concerns

Parents have obvious reason to be concerned when their child has sudden breathing problems. Many children with VDC make multiple trips to the emergency room before the condition is correctly diagnosed. Many medical professionals are only marginally familiar with VCD, because this problem is much less common than asthma. Parents may want to suggest additional testing for VCD if their child is being treated for asthma without success.

See also Asthma; Stridor.

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Volvulus *see* **Intestinal obstructions**

Vomiting *see* **Nausea and vomiting**

Von Willebrand disease

Definition

Von Willebrand disease is caused by a deficiency or an abnormality in a protein called von Willebrand factor and is characterized by prolonged bleeding.

Description

The Finnish physician Erik von Willebrand was the first to describe von Willebrand disease (VWD). In 1926, von Willebrand noticed that many male and female members of a large **family** from the Aland Islands had increased bruising (bleeding into the skin) and prolonged episodes of bleeding. The severity of the bleeding varied among family members and ranged from mild to severe and typically involved the mouth, nose, genital and urinary tracts, and occasionally the intestinal tract. Some women in the family also experienced excessive menstrual bleeding. What differentiated this

bleeding disorder from classical **hemophilia** was that it appeared not to be associated with muscle and joint bleeding and affected women and men rather than just men. Dr. von Willebrand named this disorder hereditary pseudohemophilia.

Pseudohemophilia, or von Willebrand disease (VWD) as it is called in the twenty-first century, occurs when the body does not produce enough of a protein, called von Willebrand factor(vWF), or produces abnormal vWF. vWF is involved in the process of blood clotting (coagulation). Blood clotting is necessary to heal an injury to a blood vessel. When a blood vessel is injured, vWF enables blood cells called platelets to bind to the injured area and form a temporary plug to seal the hole and stop the bleeding. vWF is secreted by platelets and by the cells that line the inner wall of the blood vessels (endothelial cells). The platelets stimulate the release other chemicals, called factors, which help form a strong permanent clot. vWF binds to and stabilizes factor VIII, one of the factors involved in forming the permanent clot.

A deficiency or abnormality in vWF can interfere with the formation of the temporary platelet plug and affect the normal survival of factor VIII. This indirectly interferes with the production of the permanent clot. Individuals with VWD, therefore, have difficulty in forming blood clots, and as a result, they may bleed for a longer time. In most cases the bleeding is due to an obvious injury, although it sometimes occurs spontaneously.

VWD is classified into three basic types: type 1, 2, and 3 based on the amount and type of vWF that is produced. Type 1 is the most common and mildest form and results when the body produces slightly decreased amounts of normal vWF. Type 2 can be classified into four subtypes (A, B, M, N) and results when the body produces an abnormal type of vWF. Type 3 is the rarest and most severe form and results when the body does not produce any detectable vWF.

Demographics

Approximately one out of 100 people are affected with VWD, making it the most common inherited bleeding disorder. VWD affects people of all ethnic backgrounds. Approximately 70 to 80 percent of people with VWD have type 1, and close to 20 to 30 percent have type 2. Type 3 is very rare and occurs in less than 1 percent of people with VWD. Type 3 occurs in about one out of every million people.

Causes and symptoms

The complex genetics of VWD involve a gene found on chromosome 12. Different types of changes in the vWF gene can affect the production of vWF. Some changes cause the vWF gene to produce decreased amounts of normal vWF, while other changes cause the gene to produce abnormal vWF. Each individual inherits two copies of each gene, one from the mother and one from the father. Most of the vWF gene changes are significant enough that a change in only one vWF gene is sufficient to cause VWD. However, some types of gene changes only cause VWD if both genes are changed, which often leads to more severe symptoms.

Type 1 VWD is called an autosomal dominant condition since it is caused by a change in only one vWF gene. Since type 1 VWD results in only a slight decrease in the amount of vWF produced, the symptoms are often mild and not apparent in some individuals. Most cases of type 2 VWD are autosomal dominant since a change in only one vWF gene results in the production of an abnormal form of vWF. An autosomal dominant form of VWD can be inherited from either parent or can occur as a spontaneous gene mutation (change) in the embryo that is formed when the egg and sperm cells come together during fertilization.

Some cases of type 2 VWD and all cases of type 3 VWD are autosomal recessive, since they are caused only by changes in both vWF genes. A person with an autosomal recessive form of VWD has inherited both a changed gene from the mother and a changed gene from the father. Parents who have a child with an autosomal recessive form of VWD are called carriers, since they each possess at least one changed vWF gene. Many carriers for the autosomal recessive forms of type 2 VWD and type 3 VWD do not have any symptoms. Each child born to parents who both have one changed gene has a 25 percent chance of having VWD, a 50 percent chance of being a carrier, and a 25 percent chance of not being and not having VWD disease. A person with an autosomal dominant form of VWD has a 50 percent chance of passing the changed gene on to his or her children who may or may not have symptoms.

VWD is usually a relatively mild disorder characterized by easy bruising, recurrent nosebleeds, heavy menstrual periods, and extended bleeding after surgeries and invasive dental work. There is a great deal of variability in the severity of symptoms, which can range from clinically insignificant to life threatening. Even children within the same family who are affected with the same type of VWD may exhibit different symptoms. A child with VWD may exhibit a range of symptoms over the course of his or her lifetime and may experience an

improvement in symptoms with age. The severity of the disease is partially related to the amount and type of vWF that the body produces, but it is also influenced by other genetic and non-genetic factors.

Type 1

Type 1, the mildest form of VWD, is usually associated with easy bruising, recurrent nosebleeds, heavy menstrual periods, and prolonged bleeding after surgeries and invasive work. Many people with type 1 VWD do not have any noticeable symptoms or only have prolonged bleeding after surgery or significant trauma. The amount of vWF produced by the body increases during pregnancy, so prolonged bleeding during delivery is uncommon in people with type 1 VWD.

Type 2

Children with type 2 VWD usually have symptoms from early childhood. Symptoms may even be present at birth. These children usually experience prolonged bleeding from cuts, easy bruising, nosebleeds, skin hematomas, and prolonged bleeding from the gums following tooth extraction and minor trauma. Gastrointestinal bleeding is rare but can be life-threatening. More than 50 percent of women with type 2 VWD experience heavy menstrual periods that may require a blood transfusion. Some women with type 2 VWD exhibit prolonged bleeding during delivery.

Type 3

Type 3 VWD can be quite severe and is associated with bruising and bleeding from the mouth, nose, and from the intestinal, genital, and urinary tracts. Type 3 is also associated with spontaneous bleeding into the muscles and joints, which can result in joint deformities. Some women with type 3 VWD experience prolonged bleeding during delivery.

When to call the doctor

If a child frequently experiences significant bleeding, takes longer than normal to stop bleeding, or experiences easy bruising, then the doctor should be consulted.

Diagnosis

Many children with VWD have mild symptoms or symptoms that can be confused with other bleeding disorders, making it difficult for a doctor to diagnose VWD based on clinical symptoms. VWD should be suspected in any child with a normal number of platelets in the blood and bleeding from the mucous membranes such as the nose, gums, and gastrointestinal tract. Testing for a child with suspected VWD often includes the measurement of the following:

- how long it takes for the bleeding to stop after a tiny cut is made in the skin (bleeding time)
- the amount of vWF (vWF antigen measurement)
- the activity of vWF (ristocetin co-factor activity)
- the amount of factor VIII (factor VIII antigen measurement)
- activity of factor VIII

Children with type 1 VWD usually have an increased bleeding time, but they may have an intermittently normal time. They also have a decreased amount of vWF and decreased vWF activity and usually have slightly decreased factor VIII levels and activity. Children with type 2 VWD have a prolonged bleeding time and decreased activity of vWF; they may also have decreased amounts of vWF and factor VIII and decreased factor VIII activity. Type 3 children have undetectable amounts of vWF, negligible vWF activity, factor VIII levels of less than 5 to 10 percent, and significantly reduced factor VIII activity. The activity of vWF is reduced for all types of VWD, making it the most sensitive means of identifying all three types. Individuals with borderline results should be tested two to three times over a three-month period.

Once a person is diagnosed with VWD, further testing such as vWF multimer analysis and ristocetin-induced platelet aggregation (RIPA) should be performed to determine the subtype. Multimer analysis evaluates the structure of the vWF, and RIPA measures how much ristocetin is required to cause the clumping of platelets in a blood sample. The vWF multimer analysis is able to differentiate children with a structurally normal vWF (Type 1) from children with a structurally abnormal vWF (Type 2) and is often able to identify the subtype of patients with Type 2 VWD. Children with type 1 VWD usually have normal to decreased RIPA concentrations. Depending on the subtype, patients with type 2 VWD either have increased or decreased RIPA. RIPA is usually absent and the multimer analysis shows undetectable vWF in children with type 3 VWD.

In some cases, DNA testing can be a valuable adjunct to biochemical testing. The detection of gene alteration(s) can confirm a diagnosis and can determine the type and subtype of VWD. It can also help to facilitate prenatal testing and testing of other family members. Unfortunately many people with VWD possess DNA changes that are not detectable through DNA testing. A child who has a mother, father, or sibling diagnosed with VWD should undergo biochemical testing for VWD. If

the relative with VWD possesses a detectable gene change, then DNA testing should be considered.

Prenatal testing

If one parent has been diagnosed with an autosomal dominant form of VWD or both parents are carriers for an autosomal recessive form of VWD, then prenatal testing should be considered. DNA testing can be performed through **amniocentesis** or chorionic villus sampling. If the DNA change in the parent(s) is unknown, then prenatal testing can sometimes be performed through biochemical testing of blood obtained from the umbilical cord. However this procedure is less accurate and is associated with a higher risk of pregnancy loss.

Treatment

VWD is most commonly treated by replacement of vWF through the administration of blood products that contain vWF or through treatment with desmopressin (DDAVP, 1-deamino-8-D-arginine vasopressin). DDAVP functions by increasing the amount of factor VIII and vWF in the circulating blood. Treatment with blood products or DDAVP may be started in response to uncontrollable bleeding or may be administered before procedures such as surgeries or dental work. The type of treatment chosen depends on the type of VWD and a patient's response to a preliminary treatment trial.

Treatment with desmopressin (DDAVP)

DDAVP is the most common treatment for people with type 1 VWD. About 80 percent of people with type 1 VWD respond to DDAVP therapy. Treatment with DDAVP can also be used to treat some people with type 2 VWD. Patients with type 2B VWD should not be treated with this medication, since DDAVP can induce dangerous platelet clumping. Type 3 VWD should not be treated with DDAVP, since this medication does not increase the level of vWF in type 3 patients. DDAVP should only be used in people who have been shown to be responsive through a pre-treatment trial transfusion with this medication.

DDAVP can be administered intravenously or through a nasal inhaler. DDAVP has relatively few side effects although some people may experience facial flushing, tingling sensations, and headaches after treatment with this medication. Often treatment with this medication is only required prior to invasive surgeries or dental procedures.

Treatment with blood products

Patients who are unable to tolerate or are unresponsive to drug-based treatments are treated with concentrated factor VIII obtained from blood products. Not all factor VIII concentrates can be used, since some do not contain enough vWF. The concentrate is treated to kill most viruses, although caution should be used since not all types of viruses are destroyed. If the factor VIII concentrates are unable to manage a severe bleeding episode, then blood products called cryoprecipitates, which contain concentrated amounts of vWF, or platelet concentrates should be considered. Caution should be used when treating with these blood products since they are not treated to kill viruses.

Other treatments and precautions

Medications called fibrinolytic inhibitors can be helpful in controlling intestinal, mouth, and nose bleeding. Estrogens, such as are found in **oral contraceptives**, increase the synthesis of vWF and can sometimes be used in the long-term treatment of women with mild to moderate VWD. Estrogens are also sometimes used before surgery in women with type 1 VWD. Some topical agents are available to treat nose and mouth bleeds. Patients with VWD should avoid taking aspirin, which can increase their susceptibility to bleeding. Children with severe forms of VWD should avoid activities that increase their risk of injury such as contact **sports**.

Prognosis

The prognosis for VWD is generally good, and most individuals have a normal lifespan. The prognosis can depend, however, on accurate diagnosis and appropriate medical treatment.

Prevention

There is no known way to prevent VWD. If an individual planning to become a parent believes he or she may be a carrier of VWD, genetic counseling is suggested so that options may be explored.

Parental concerns

VWD is usually very mild and does not cause unusual bleeding except after trauma or surgery. Children with moderate or severe VWD may need to be discouraged from playing contact sports or participating in other activities where injury is likely. Special care should be taken before surgical or dental procedures to ensure that severe bleeding does not occur.

KEY TERMS

Amniocentesis—A procedure performed at 16–18 weeks of pregnancy in which a needle is inserted through a woman's abdomen into her uterus to draw out a small sample of the amniotic fluid from around the baby for analysis. Either the fluid itself or cells from the fluid can be used for a variety of tests to obtain information about genetic disorders and other medical conditions in the fetus.

Autosomal dominant—A pattern of inheritance in which only one of the two copies of an autosomal gene must be abnormal for a genetic condition or disease to occur. An autosomal gene is a gene that is located on one of the autosomes or non-sex chromosomes. A person with an autosomal dominant disorder has a 50% chance of passing it to each of their offspring.

Autosomal recessive—A pattern of inheritance in which both copies of an autosomal gene must be abnormal for a genetic condition or disease to occur. An autosomal gene is a gene that is located on one of the autosomes or non-sex chromosomes. When both parents have one abnormal copy of the same gene, they have a 25% chance with each pregnancy that their offspring will have the disorder.

Biochemical testing—Measuring the amount or activity of a particular enzyme or protein in a sample of blood or urine or other tissue from the body.

Carrier—A person who possesses a gene for an abnormal trait without showing signs of the disorder. The person may pass the abnormal gene on to offspring. Also refers to a person who has a particular disease agent present within his/her body, and can pass this agent on to others, but who displays no symptoms of infection.

Chorionic villus sampling—A procedure used for prenatal diagnosis at 10–12 weeks gestation. Under ultrasound guidance a needle is inserted either through the mother's vagina or abdominal wall and a sample of the chorionic membrane. These cells are then tested for chromosome abnormalities or other genetic diseases.

Chromosome—A microscopic thread-like structure found within each cell of the human body and consisting of a complex of proteins and DNA. Humans have 46 chromosomes arranged into 23 pairs. Chromosomes contain the genetic information necessary to direct the development and functioning of all cells and systems in the body. They pass on hereditary traits from parents to child (like eye color) and determine whether the child will be male or female.

Desmopressin acetate (DDAVP)—A drug used to regulate urine production.

DNA—Deoxyribonucleic acid; the genetic material in cells that holds the inherited instructions for growth, development, and cellular functioning.

DNA testing—Analysis of DNA (the genetic component of cells) in order to determine changes in genes that may indicate a specific disorder.

Endothelial cells—The cells lining the inner walls of a body cavity or the cardiovascular system. Also known as endothelium.

Factor VIII—A protein involved in blood clotting that requires von Willebrand factor for stability and long-term survival in the bloodstream.

Gene—A building block of inheritance, which contains the instructions for the production of a particular protein, and is made up of a molecular sequence found on a section of DNA. Each gene is found on a precise location on a chromosome.

Mutation—A permanent change in the genetic material that may alter a trait or characteristic of an individual, or manifest as disease. This change can be transmitted to offspring.

Platelet—A cell-like particle in the blood that plays an important role in blood clotting. Platelets are activated when an injury causes a blood vessel to break. They change shape from round to spiny, "sticking" to the broken vessel wall and to each other to begin the clotting process. In addition to physically plugging breaks in blood vessel walls, platelets also release chemicals that promote clotting.

Prenatal testing—Testing for a disease, such as a genetic condition, in an unborn baby.

Protein—An important building blocks of the body, a protein is a large, complex organic molecule composed of amino acids. It is involved in the formation of body structures and in controlling the basic functions of the human body.

Skin hematoma—Blood from a broken blood vessel that has accumulated under the skin.

von Willebrand factor (vWF)—A protein found in the blood that is involved in the process of blood clotting.

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W

Walleye *see* **Strabismus**

Warts

Definition

Warts are small, benign growths caused by a viral infection of the skin or mucous membrane. The virus infects the surface layer. The viruses that cause warts are members of the human papilloma virus (HPV) family. Warts are not cancerous, but some strains of HPV, usually not associated with warts, have been linked with **cancer** formation. Warts are contagious from person to person and from one area of the body to another on the same person.

Description

There are approximately 60 types of HPV that cause warts, each preferring a specific bodily location. For instance, some types of HPV cause warts to grow on the skin, others cause them to grow inside the mouth, while still others cause them to grow on the genital and rectal areas. However, most can be active anywhere on the body. The virus enters through the skin and produces new warts after an incubation period of one to eight months. Warts are usually skin-colored and feel rough to the touch, but they also can be dark, flat, and smooth.

Warts are passed from person to person, directly and indirectly. Some people are continually susceptible to warts, while others are more resistant to HPV and seldom get them. The virus takes hold more readily when the skin has been damaged in some way, which may explain why children who bite their nails tend to have warts located on their fingers. People who take a medication to suppress their immune system or are on long-term steroid use are also prone to a wart virus infection. This tendency is seen in people with **AIDS**.

Demographics

Particularly common among children, young adults, and women, warts are a problem for 7–10 percent of the population.

Causes and symptoms

The more common types of warts include the following:

- common hand warts
- foot warts
- flat warts
- genital warts

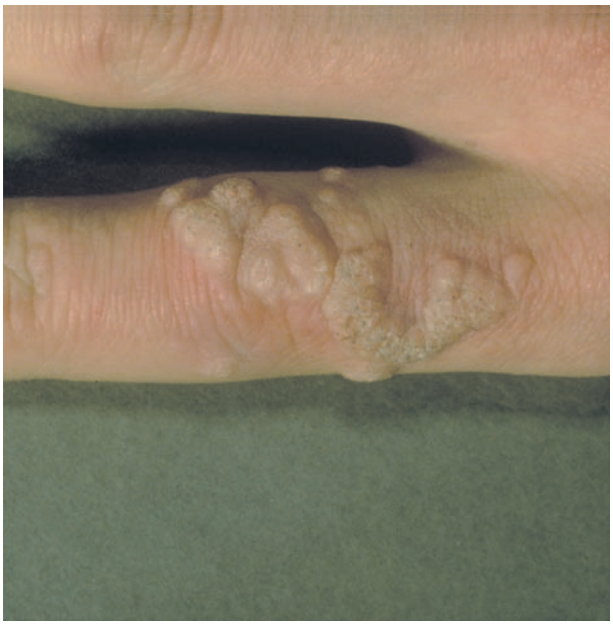
Hand warts

Common hand warts grow around the nails, on the fingers, and on the backs of hands. They appear more frequently where skin is broken, such as in areas where fingernails are bitten or hangnails are picked.

Foot warts

Foot warts are called plantar warts because the word plantar is the medical term for the sole of the foot, the area where the wart usually appears as a single lesion or as a cluster. Plantar warts, however, do not stick up above the surface like common warts. The ball of the foot, the heel, and the plantar part of the toes are the most likely locations for the warts because the skin in those areas is subject to the most weight, pressure, and irritation, making a small break or crack more likely.

Plantar warts are familiar to all ages groups, appearing frequently in children between the ages of 12 and 16. Adolescents often come into contact with a wart virus in a locker room, swimming pool area, or by walking barefooted on dirty surfaces. The blood vessels feeding them are the black dots that are visible on the wart. If left untreated, these warts can grow to an inch or more in circumference and spread into clusters of several warts. They are known to be very painful at times, the **pain**



Cluster of warts on a finger. *(Custom Medical Stock Photo Inc.)*

usually compared to the feeling of a permanent stone in the shoe particularly if the wart is on a pressure point of the foot. People with **diabetes mellitus** are prone to complications from plantar warts related to the development of sores or ulceration and the poor healing potential associated with diabetes.

Flat warts

Flat warts tend to grow in great numbers and are smaller and smoother than other warts. They can erupt anywhere, appearing more frequently on the legs of women, the faces of children, and on the areas of the face that are shaved by young adult males.

Genital warts

Genital warts, also called condyloma acuminata or venereal warts, are one of the most common forms of sexually transmitted disease (STD) in this country. Most experts contend that they are contracted by sexual contact with an infected person who carries HPV and are more contagious than other warts. It is estimated that two-thirds of the people who have sexual contact with a partner with genital warts will develop the disease within three months of contact. As a result, about 1 million new cases of genital warts are diagnosed in the United States each year.

Genital warts tend to be small flat bumps but they may be thin and tall. They are usually soft and not scaly like other warts. In women, genital warts appear on the genitalia, within the vagina, on the cervix, and around the anus or within the rectum. In men, genital warts usually appear on the tip of the penis but may also be found on the scrotum or around the anus. Genital warts can also develop in the mouth of a person who has had oral sexual contact with an infected person.

When to call the doctor

Individuals who notice warts in their genital area should see a doctor. A physician should be consulted for warts that bleed, are particularly painful, or that do not disappear after six to nine months.

Diagnosis

A physician may be able to diagnose warts with a simple examination. If the warts are small, the doctor may put a vinegar-like liquid on the skin, which makes the warts turn white and easier to see, and then use a magnifying glass to look for them.

Treatment

Most people attempt to treat warts themselves. Professional treatment is usually sought after self-treatment has been unsuccessful.

Home/self treatment

Many of the nonprescription wart remedies available at drug stores will remove simple warts from hands and fingers. These medications may be lotions, ointments, or plasters and work by chemically removing the skin that was affected by the wart virus. The chemicals are strong, however, and should be used with care since they can remove healthy as well as infected skin. These solutions should be avoided by diabetics and those with cardiovascular or other circulatory disorders whose skin may be insensitive and not appreciate irritation.

Flat warts are best treated with topical retinoides (retinoic acid) or a gel containing salicylic acid. The acid does not actually kill the wart virus, but waterlogs the skin so that the surface layer, with the virus, peels off. These products can take up to three months of treatment depending on the size and depth of the wart. Patches are also good to use. Rather than applying drops, a small pad is placed on the wart and left for 48 hours and then replaced with a new one. The patch usually contains a higher concentration of salicylic acid and may irritate the surrounding skin. If this occurs, people should switch to a gel or stop medication for a period. To help the healing process for flat facial warts, men should shave with an electric shaver or temporarily grow a beard. Women

with flat warts on areas that are shaved should use other methods to remove hair such as depilatory cream or wax.

Professional treatment

Physicians should be consulted if there are no signs of progress after a month of self treatment. Doctors have many ways of removing warts, including using stronger topically applied chemicals than those available in pharmacies. Some of these solutions include podofilox, topical podophyllum, and trichloracetic acid (TCA). Some burning and discomfort for one or more days following treatment can be expected. Although these chemicals are effective, they may not completely destroy all warts. A second method of removal is freezing or cryosurgery on the wart using liquid nitrogen. Cryotherapy is relatively inexpensive, does not require anesthesia, and usually does not result in scarring. Although temporarily uncomfortable, it provides an effective and safe way to deliver freezing temperatures to a particular area on the skin, and healing is usually quick. Physicians may also choose to burn the wart with liquid nitrogen or numb the skin and then scrape off the wart. Another removal process is electrocautery (electric burning), destroying the wart by burning it with an electric needle. Laser surgery is also an option for removing warts.

Genital warts are the most difficult to treat. They can be removed, but the viral infection itself cannot be cured. Often, because the warts are so small, more than one treatment may be needed. The virus continues to live in the deeper skin, which is why warts often return after they have been removed. Strong chemicals may be applied as well as surgical excision with or without electrocautery. This therapy requires a small operative procedure and a local anesthetic. Laser therapy, although more expensive, is often used for treating venereal warts that are more extensive. The use of lasers, which vaporize the lesion, can theoretically transmit the HPV. It is not at all clear, however, if this occurs.

There is no single recommended method for eliminating plantar warts. If detected early, cryotherapy is usually enough. However, they can be very resilient, requiring repeated treatment over several months. Treatment ranges from the conservative approach of applying chemical solutions to the more aggressive option of surgery. Persons with diabetes or vascular disease are usually treated with the more conservative methods.

Alternative treatment

There are a variety of alternative approaches to the treatment of warts. The following suggestions apply to common warts and plantar warts. They are not recommended for genital or cervical warts. Since genital and cervical warts are transmitted sexually, they should be treated by a physician.

For the treatment of common or plantar warts, practitioners may recommend the following remedies:

- Apply a paste made of vitamin C powder to the wart for one to two weeks.
- Place a crushed or sliced garlic clove over the wart for seven consecutive nights while sleeping.
- Soak the wart in water, put cross-hatches over it with a sterile needle, and apply drops of thuja (*Thuja occidentalis*) tincture onto the wart. Repeat the cross-hatching and tincture application until the wart is saturated with the tincture. Repeat several times each day for one to two weeks. (A tincture is an herbal extract made with alcohol.)
- Tape a piece of banana peel, smooth side down, over the wart and leave it on overnight. Repeat nightly for one to two weeks.

Prognosis

Even though genital warts may be removed, the virus itself continues to live. The HPV can cause tissue changes in the cervix of women with cervical infection. The general recommendation for women who have a history of genital warts is to see their doctors every six months for Pap smears to monitor any changes that may occur.

For plantar warts, the treatment goal is to destroy the wart and its virus without causing much damage to healthy skin. It is not unusual for treatment to cause pain until the foot heals because of the weight put on the foot.

Prevention

Plantar warts can be prevented by wearing shoes, changing shoes daily, keeping feet clean and dry, and not ignoring skin growths and changes in the skin. Genital warts can be prevented by using condoms and avoiding unprotected sex. Barrier protection will not, however, prevent the spread of wart-causing HPV to uncovered areas such as the pubis and upper thighs.

Nutritional concerns

Because warts are caused by a virus, general immune system support can be effective in helping to keep warts from coming back after treatment or to keep them from multiplying or growing. Eating a well balanced diet high in sources of **vitamins** A, C, and E can help strengthen the immune system. Avoiding stress,

KEY TERMS

Condyloma acuminata—Another term for a genital wart.

Cryotherapy—The use of a very low-temperature probe to freeze and thereby destroy tissue. Cryotherapy is used in the treatment skin lesions, Parkinson's disease, some cancers, retinal detachment, and cataracts. Also called cryosurgery.

Endometritis—Inflammation of the endometrium or mucous membrane of the uterus.

Epidermis—The outermost layer of the human skin.

Human papilloma virus (HPV)—A virus that causes common warts of the hands and feet, as well as lesions in the genital and vaginal area. More than 50 types of HPV have been identified, some of which are linked to cancerous and precancerous conditions, including cancer of the cervix.

Retinoic acid—Vitamin A_1 acid which is used topically to treat acne.

Salicylic acid—An agent prescribed to treat a variety of skin disorders, such as acne, dandruff, psoriasis, seborrheic dermatitis, calluses, corns, and warts.

which is believed to compromise the immune system, may also be helpful.

Parental concerns

Parents can help to prevent plantar warts by urging their children to wear shoes, change their shoes daily, and keep their feet clean and dry. Parents should also pay attention to growths and other changes in their child's skin. Instructing children in **condom** usage is a personal, parental decision. However, parents should tell their children that genital warts can be prevented by using condoms and avoiding unprotected sex. Barrier protection will not, however, prevent the spread of wart-causing HPV to uncovered areas such as the pubis and upper thighs.

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American Academy of Family Physicians. 11400 Tomahawk Creek Parkway, Leawood, KS 66211–2672. Web site: <www.aafp.org/>.

American Academy of Pediatrics. 141 Northwest Point Boulevard, Elk Grove Village, IL 60007–1098. Web site: <www.aap.org/default.htm>.

American Association of Naturopathic Physicians. 8201 Greensboro Drive, Suite 300, McLean, VA 22102. Web site: <http://naturopathic.org/>.

American College of Physicians. 190 N Independence Mall West, Philadelphia, PA 19106–1572. Web site: <www.acponline.org/>.

American Podiatric Medical Association. 9312 Old Georgetown Road Bethesda, MD 20814–1698. Web site: <www.apma.org/>.

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Water on the brain *see* **Hydrocephalus**

Webbed fingers and toes *see* **Polydactyly and syndactyly**

Wechsler intelligence test

Definition

The Wechsler **intelligence** tests are a widely used series of intelligence tests developed by clinical psychologist David Wechsler.

Purpose

The Wechsler Intelligence Scales for Children (regular, revised, and third edition) and Wechsler **Preschool** and Primary Scale of Intelligence are used as tools in school placement, in determining the presence of a learning disability or a **developmental delay**, in identifying giftedness, and in tracking intellectual development. They are often included in neuropsychological testing to assess the brain function of individuals with neurological impairments.

Description

The most distinctive feature of the Wechsler tests is their division into a verbal section and a nonverbal (or performance) section, with separate scores available for each subsection. All of the Wechsler scales are divided into six verbal and five performance subtests. The complete test takes 60 to 90 minutes to administer. Verbal intelligence, the component most often associated with academic success, implies the ability to think in abstract terms using either words or mathematical symbols. Performance intelligence suggests the ability to perceive relationships and fit separate parts together logically into a whole. The inclusion of the performance section in the Wechsler scales is especially helpful in assessing the cognitive ability of non-native speakers and children with speech and **language disorders**. The test can be of particular value to school psychologists screening for specific learning disabilities because of the number of specific subtests that make up each section.

The Wechsler Preschool and Primary Scales of Intelligence (WPPSI) have traditionally been geared toward children ages four to six years old, although the newest version of the test extends the age range down to three years and upward to seven years three months. The verbal section covers the following areas: general information (food, money, the body, etc.), vocabulary (definitions of increasing difficulty), comprehension (responses to questions), arithmetic (adding, subtracting, counting), sentences (repeating progressively longer sentences), and similarities (responding to questions such as "How are a pen and pencil alike?"). The performance section includes picture completion, copying geometric designs, using blocks to reproduce designs, working through a maze, and building an animal house from a model.

The Wechsler Intelligence Scale for Children (WISC) is designed for children and adolescents ages six to 16. Its makeup is similar to that of the Preschool Scale. Differences include the following: geometric designs are replaced by assembly of three-dimensional objects; children arrange groups of pictures to tell simple stories; they are asked to remember and repeat lists of digits; a coding exercise is performed in place of the animal house; mazes are a subtest. For all of the Wechsler scales (which also include the Wechsler Adult Intelligence Scale, or WAIS), separate verbal and performance scores, as well as a total score, are computed. These are then converted using a scale divided into categories (such as average and superior), and the final score is generally given as one of these categories rather than as a number or percentile ranking.

The Wechsler Intelligence Scales are standardized tests, meaning that as part of the test design, they were administered to a large representative sample of the target population, and norms were determined from the results. The scales have a mean, or average, standard score of 100 and a standard deviation of 15. The standard

KEY TERMS

Norms—A fixed or ideal standard; a normative or mean score for a particular age group.

Representative sample—A random sample of people that adequately represents the test-taking population in age, gender, race, and socioeconomic standing.

Standard deviation—A measure of the distribution of scores around the average (mean). In a normal distribution, two standard deviations above and below the mean includes about 95% of all samples.

Standardization—The process of determining established norms and procedures for a test to act as a standard reference point for future test results.

deviation indicates how far above or below the norm the subject's score is. For example, a ten-year-old is assessed with the WISC-III scale and achieves a full-scale IQ score of 85. The mean score of 100 is the average level at which all 10-year-olds in the representative sample performed. This child's score would be one standard deviation below that norm.

While the full-scale IQ score provides a reference point for evaluation, it is only an average of a variety of skill areas. A trained psychologist evaluates and interprets an child's performance on the scale's subtests to discover their strengths and weaknesses and offer recommendations based upon these findings.

Risks

The only known risk of the Wechsler intelligence tests is that the results are misused or are given undue weight.

Parental concerns

Results of intelligence tests should not be considered a complete indication of a child's future path. They are most useful in determining children who may need special attention, either because of disability or because of giftedness. Parents should consider the possible consequences carefully if they are considering telling their child the outcome of this or any other intelligence test.

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Weight issues *see* **Obesity**

Well-baby examination

Definition

Well-baby examinations are scheduled regularly during the first two years of life due to the rapid growth and change that occurs during infancy. During each visit the pediatrician monitors, advises, and answers questions on a baby's growth and development.

Purpose

The American Academy of Pediatrics recommends the newborn infant see a physician for a check-up at birth, two weeks, two months, four months, six months, nine months, 12 months, 15 months, 18 months, and 24 months, and annually thereafter. Most pediatricians follow this schedule, or some variation of it, in prescribing a check-up regimen for their patients. A well-baby exam consists of questions the parents will be asked about the baby's general health and development, followed by a physical exam. The exam includes measurements of length and height, weight and head circumference (the distance around the head), vital signs, and a general physical examination. Special attention is paid to whether

the baby has met normal developmental milestones. The physician will question parents or guardians about the activities of the baby to help assess developmental issues that are not observable by an office visit.

Description

A pediatrician performs a thorough physical exam at birth to determine the physical status of the newborn. This exam includes assessing size, weight, head circumference, chest circumference, genitalia, physical mobility, eyes, ears, nose, mouth, lungs, heart, elimination, presence of **neonatal reflexes**, and much more. If the hospital pediatrician is not the same as the one used for follow-up exams, it is important to obtain the birth records to bring to the first office visit. Because the majority of states have laws governing newborn testing, most hospitals do a hearing screen, metabolic screen to assess thyroid activity, and screen for **phenylketonuria** (PKU), a genetic disorder than can be easily corrected by diet.

The first well-baby visit occurs at two weeks, and a **family** medical history is usually taken at this time. The baby's height, weight, and head circumference will be measured. (Head circumference is an indirect measure of brain growth.) Abnormally slow or fast growth may indicate a problem that needs investigation. The health-care provider can show parents a graph that indicates where the baby's measurements are on a standard growth curve. The trend in growth over time is more important than what a baby's weight and height are at any particular visit. A complete head-to-toe exam will be performed, during which the parent may want to ask questions related to birth marks or anything that is perceived as unusual.

In addition to the physical exam, the physician will ask questions related to what the baby can do physically, i.e., lift the head briefly, respond to loud sounds, etc. These are developmental milestones that represent a normal progression of physical and mental maturity. Although each baby develops differently, these milestones indicate a child's progress over time. The physician may want to observe development if possible. The physician may provide guidance related to possible dangers in the home, such as the importance of installing and maintaining smoke detectors, keeping a baby away from plastic bags, and never leaving the baby unattended while on a changing table. During this visit, the parent will be asked about the stress of having a new baby and the situation at home. It is a provider's responsibility to evaluate every child for abuse, and this questioning should not be taken personally. Finally, if the **hepatitis B vaccine** was not given in the hospital, the first shot may be given at this visit. All other vaccines begin at the two-month visit.

The two-month visit will be a repeat of the two week visit with a physical exam, developmental and behavioral **assessment**, guidance for upcoming developmental changes, and immunizations. During the visit, a parent should never hesitate to ask any question that will assure them the baby is healthy and progressing normally. It is a good idea to make a list of questions before the office visit, because many parents inevitably forget what they wanted to ask. Many parents inquire about what could be given to the baby if there is a reaction to the injections. The immunizations received at this time include:

- DTP vaccine (or DtaP, **diphtheria**, **tetanus**, pertussis)—DTP (or DtaP) injections are given as a series of five injections and usually at ages two months, four months, six months, 15–18 months, and four to six years of age. At age 11 or 12, Td vaccine (tetanus and diphtheria) should be given if at least five years have elapsed since the last dose of DTaP. Td boosters are recommended every 10 years.
- Hib vaccine (*Haemophilus influenzae* type B)—Hib is given as a series of four injections at ages two months, four months, and six months, with a booster dose at 12–15 months.
- **Polio vaccine** (IPV, or inactivated poliovirus vaccine)—This is usually given in a series of five vaccines, at ages two months, four months, six to 18 months, and four to six years.
- Hep B (**hepatitis B** vaccine)—Hep B is given as a series of three injections. The first is given soon after birth and sometimes before hospital discharge. It the mother of a newborn carries the hepatitis B virus (HBV) in her blood, the baby needs to receive the first shot within 12 hours of birth. If the mother shows no evidence of HBV in her blood, the first dose may be deferred to the two-month exam. If the first shot was given in the hospital, the second shot is given at two months and the third at six months. If the first shot was given at two months, the second is given at three to four months, and the third at six to 18 months.
- PCV (Pneumococcal vaccine)—The newest addition to the immunization schedule, these vaccinations are often given as a series of four injections at two months, four months, six months, and 12–15 months of age.

The four-month exam proceeds in the same manner as the previous two—a physical exam, developmental and behavioral assessment with questions about what has been observed at home, and more immunizations. At this period, the baby should be babbling and making noises,

turning over, and trying to put everything in the mouth. Parents and the physician may discuss adding solid foods to the baby's diet, usually in the form of cereal. The immunizations given will depend on how and when the series was started.

The six-month exam is again similar. Generally the baby may be able to sit alone by this stage and may be ready to add pureed food to the diet. Once more the required immunizations will depend on the baby's history and previous injections. In October 2003, the Advisory Committee on Immunization Practices (ACIP) recommended universal influenza immunization of all children six through 23 months of age. They also recommend influenza immunization of household members and out-of-home caregivers of children younger than 24 months. Children under eight years of age who are receiving the **flu vaccine** for the first time should receive two doses separated by at least six weeks. Children under five years of age should not be vaccinated with the nasal-spray flu vaccine (LAIV).

The nine-month exam represents quite a change in baby from birth. The parent usually has many questions by this time regarding the baby's **sleep** habits, feeding patterns, teething, standing up, and so on. Again, a list is helpful to remind the parent of their own questions. The physical exam is performed, plotted on the standard growth curve, and any deviations are noted. Developmental assessment is commonly done by questioning. Does he/she pay attention to small objects and try to pick them up using his/her index finger and thumb? Can he/she locate sounds? Does he/she sit by himself/herself? Does she/he transfer objects from one hand to another? Does she/he show **stranger anxiety**? Guidance of what to expect over the next three months will again be provided. For example, the baby may begin to walk alone, make sounds, say the beginnings of words, or play peek-a-boo. The physician may discuss ways to keep a baby safe, including placing gates at the top and bottom of stairs; never leaving the baby alone in the bathtub; keeping the baby rear-facing in the car seat until 20 lbs (9 kg) and one year of age; and monitoring the temperature of the hot water heater to prevent **burns**. If the hepatitis B injection was not completed at the six month visit, it will be given at this exam.

Reaching the one-year exam is a big event in itself. The baby may be walking (assisted or unassisted) and talking a bit at this stage. The pediatrician will continue in the same manner as before—doing a physical exam and noting changes, asking questions about development, and inquiring about feeding and sleeping habits. A blood test for anemia may be performed at this visit if it was not done at the nine-month exam. Formula-fed babies are more at risk for iron deficiency than breast-fed babies. If there is a risk of lead paint exposure, a test for this may be done as well. The parent may have more questions relating to physical changes or developmental changes, because the baby is now on the verge of toddlerhood. Immunizations due at this time include:

- Measles, **mumps**, and **rubella** (MMR vaccine)—These are given by injection in two doses. The first is given at 12–15 months and the second is usually given before four to six years of age.
- Varicella (**chickenpox** vaccine)—Given by injection between the ages of 12–18 months or later for children who have not had chickenpox. Susceptible teens over 13 years of age should receive two doses given at least four weeks apart.
- Flu vaccine—For influenza, if needed.

Parents who may have to move during this first year or in any subsequent years should have the child's immunization and health record with them for a new provider to review.

The 15-month visit is very comparable to the previous visits but it does mark a few milestones in the child's health. It is a time when the little boy or girl that was in the baby you have known for the last 15 months can be seen. It is usually the last time immunizations are given before the pre-kindergarten shots. The typical physical exam and developmental evaluation will be performed and guidance on future development will be given. It is important to now be certain that doors and cabinets have locks, electrical sockets are covered, and objects on which the child can choke are removed from reach. The immunizations given at this visit will depend on those given at the prior visit.

The next exam will be at 18 months and will the same as the 15-month exam. If any immunizations were missed, they can be caught-up at this time. The same is true for the two-year check-up. Many pediatricians order various tests during the first two years depending on the family's history and the child's symptoms, i.e., urinalysis, tuberculin test, and blood tests. The American Academy of Pediatrics recommends cholesterol screening of children over age two whose parents have a history of cardiovascular disease before age 55, or have blood cholesterol levels above 240mg/dl.

Precautions

There are essentially no precautions to take for a visit. However, parents who may have a history of autoimmune disorders in their family should be aware that a preservative, thimersal, which contains mercury and is used in vaccines, has a possible link to **autism** and auto-

immune disorders. Many pharmaceutical companies now use a safer preservative called 2-phenoxy ethanol.

Preparation

The primary preparation for a well-baby exam involves the parent or guardian making a list of questions for the pediatrician.

Aftercare

The only aftercare necessary is when an infant has a slight reaction to the immunizations. The provider needs to inform the parent what to expect and what can be done to alleviate symptoms. **Pain** at the immunization size and a slight **fever** are often easily treated with **acetaminophen**.

Risks

There are few risks associated with well-baby visits. The risks with the preservative, thimersal, which is used in vaccines are mentioned above. Serious reactions to vaccines are extremely rare. More common problems associated with doctor visits are dealing with fears babies have of strangers touching them, and managing the child's pain from vaccinations.

Parental concerns

Concerns of many parents revolve around developmental delays and what could be done to assist advancement through these milestones. The parent needs to remember that all babies and children advance at their own pace and should never be compared to other children but only to the progress made individually. Of course, some children do have conditions that preclude normal development, and any significant lag should be monitored and investigated by the physician.

Developmental milestones that usually occur within the first year period are:

- Month one: lift head; move head from side to side; prefers the human face over shapes; turns toward familiar sounds; blinks at bright lights; focuses on items 8–12 inches (20–30 cm) away; has strong reflexes.
- Month two: smiles; tracks objects with eyes; makes noises other than crying; may make sounds that resemble vowels, as "ah" or "ooh."
- Months three and four: tracks moving objects; grasps items with hands and reaches for dangling objects; controls head; may begin trying to sit alone; recognizes people or familiar objects; develops a social smile; babbles and amuses self; responds to colors and shades; explores objects with mouth; recognizes breast or bottle; communicates pain, loneliness, or discomfort through crying; responds to rattle or bell.
- Months five and six: begins teething process; uses hands in a raking fashion to get **toys** closer; experiments with cause and effect; sits by self with minimal support; opens mouth for spoon; rolls over and back; copies facial expressions; makes two-syllable sounds.
- Months seven and eight: can self-feed some finger foods; turns in direction of voice; plays peek-a-boo; imitates many sounds; distinguishes emotions by tone of voice; responds to name; experiments with gravity by dropping things; has different reactions for different family members; gets into **crawling** position; shows some **anxiety** when removed from parent.
- Months nine and 10: picks up tiny objects; begins to identify self in mirror; drops objects and looks for them; starts to understand object permanence; goes from

KEY TERMS

Diphtheria—A serious, frequently fatal, bacterial infection that affects the respiratory tract. Vaccinations given in childhood have made diphtheria very rare in the United States.

Haemophilus influenzae—An anaerobic bacteria associated with human respiratory infections, conjunctivitis, and meningitis.

Hepatitis B—An infection of the liver that is caused by a DNA virus, is transmitted by contaminated blood or blood derivatives in transfusions, by sexual contact with an infected person, or by the use of contaminated needles and instruments.

Pertussis—Whooping cough, a highly contagious disease of the respiratory system, usually affecting children, that is caused by the bacterium Bordetella pertussis and is characterized in its advanced stage by spasms of coughing interspersed with deep, noisy inspirations.

Polio—Poliomyelitis, an acute viral disease marked by inflammation of nerve cells of the brain stem and spinal cord and can cause paralysis.

Tetanus—A potentially fatal infection caused by a toxin produced by the bacterium *Clostridium tetani*. The bacteria usually enter the body through a wound and the toxin they produce affects the central nervous system causing painful and often violent muscular contractions. Commonly called lockjaw.

tummy to sitting by self; pulls to standing; transfers object from hand to hand; gets upset if toy removed.

- Months 11 and 12: says "ma-ma" and "da-da" discrimately; understands "no" claps hands; waves bye-bye; triples birth weight and is 29–32 inches (75–81 cm) long; puts objects into containers and pulls them out; crawls well; shakes head no; afraid of strangers; interested in books; identifies self in mirror; shares toys but wants them back.

See also Cognitive development; Fine motor skills; Gross motor skills.

Resources

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ORGANIZATIONS

American Academy of Pediatrics. 141 Northwest Point Blvd., Elk Grove Village, IL, 60007. (847) 434-4000. Web site: <www.aap.org>.

WEB SITES

Center of Disease Control and Prevention. 1600 Clifton Rd., Atlanta, GA 30333. *2004 Childhood and Adolescent Immunization Schedule*. Available online at: <www.cdc.gov/nip/recs/child-schedule.htm#Printable>.

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Wermer's syndrome *see* **Multiple endocrine neoplasia syndromes**

Werndig-Hoffman disease *see* **Spinal muscular atrophy**

Wheezing *see* **Stridor**

Whooping cough

Definition

Whooping cough, also known as pertussis, is a highly contagious disease which causes classic spasms (paroxysms) of uncontrollable coughing, followed by a sharp, high-pitched intake of air which creates the characteristic whoop that is reflected in the disease's name.

Description

Whooping cough is caused by a bacteria called *Bordetella pertussis*. *B. pertussis* causes its most severe symptoms by attaching itself to those cells in the respiratory tract that have cilia. Cilia are small, hair-like projections that beat continuously and serve to constantly sweep the respiratory tract clean of such debris as mucus, bacteria, viruses, and dead cells. When *B. pertussis* interferes with this normal, cleansing function, mucus and cellular debris accumulate and cause constant irritation to the respiratory tract, triggering coughing and increasing further mucus production.

Whooping cough exists throughout the world. While people of any age can contract the disease, children under the age of two are at the highest risk for both the disease and for serious complications and death. Apparently, exposure to *B. pertussis* bacteria earlier in life gives individuals some immunity against infection with it later on. Subsequent infections resemble the **common cold**.

Demographics

According to the Centers for Disease Control and Prevention, since 1990, the reported incidence of pertussis has increased in the United States. Peaks occur at three to four year intervals. Since 1990, 14 states reported the number of cases of whooping cough to be more than two per 100,000 in the population. A high proportion of those cases occurred in persons aged ten years or older.

Nearly 75 percent of pertussis cases reported worldwide are in children; half of those children affected require **hospitalization**. Prior to effective immunization programs in the United States, pertussis was the major cause of death from infectious disease among individuals under the age of 14. Because developing countries as of 2004 did not have widespread immunization available, there continue to be about 50 million cases of pertussis every year across the globe, with 300,000 leading to death. About 38 percent of all hospitalizations from pertussis are in babies under the age of six months.

Causes and symptoms

Whooping cough has four somewhat overlapping stages: incubation, catarrhal stage, paroxysmal stage, and convalescent stage.

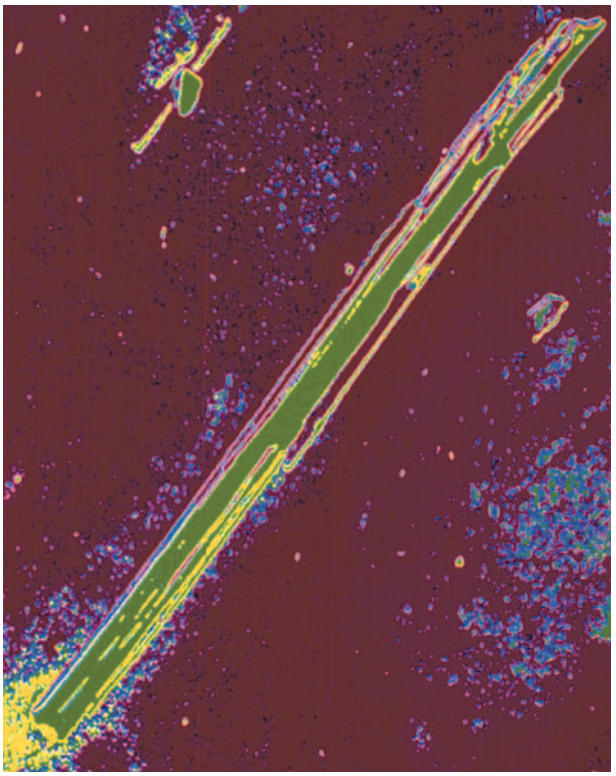
An individual usually acquires *B. pertussis* by inhaling droplets infected with the bacteria coughed into the air by someone already suffering with the infection. Incubation is the asymptomatic period (time when no evidence of disease is present) of seven to 14 days after breathing in the *B. pertussis* bacteria, during which the bacteria multiply and penetrate the lining tissues of the entire respiratory tract.

The catarrhal stage is often mistaken for an exceedingly heavy cold. People have teary eyes, sneezing, fatigue, poor appetite, and an extremely runny nose (rhinorrhea). This stage lasts approximately ten days to two weeks.

The paroxysmal stage, lasting two to four weeks, begins with the development of the characteristic whooping cough. Spasms of uncontrollable coughing, the whooping sound of the sharp inspiration of air, and **vomiting** are all hallmarks of this stage. The whoop is believed to occur due to inflammation and increased mucus, which narrow the breathing tubes, causing people to struggle to get air into their lungs; the effort results in intense exhaustion. The paroxysms (spasms) can be induced by over activity, feeding, crying, or even overhearing someone else **cough**.

The mucus that is produced during the paroxysmal stage is thicker and more difficult to clear than the more watery mucus of the catarrhal stage. Affected persons become increasingly exhausted when attempting to clear the respiratory tract through coughing. Severely ill children may have great difficulty maintaining the normal level of oxygen in their systems and may appear somewhat blue after a paroxysm of coughing, due to the low oxygen content of their blood. Such children may also suffer from swelling and degeneration of the brain (encephalopathy), which is believed to be caused both by lack of oxygen to the brain during paroxysms and by bleeding into the brain caused by increased pressure during coughing. Seizures may result from decreased oxygen to the brain. Some children have such greatly increased abdominal pressure during coughing that hernias result. Another complicating factor during this phase is the development of **pneumonia** from infection with another agent. The second pathogen successfully invades due to the person's already-weakened condition.

If individuals survive the paroxysmal stage, recovery occurs gradually during the convalescent stage, usually taking about three to four weeks. However, spasms of coughing may continue to occur over a period of months, especially when a person contracts a cold, or other respiratory infection.



A magnified image of a pertussis toxin crystal that causes whooping cough. *(National Institutes of Health/Custom Medical Stock Photo.)*

When to call the doctor

A physician or other healthcare professional should be contacted during the first two months of life to arrange for immunization. Otherwise, a person with a cough that lasts for more than a few days should be seen by a healthcare professional.

Diagnosis

A diagnosis that is based solely on a person's symptoms is not particularly accurate, as the catarrhal stage may appear to be a heavy cold, a case of the flu, or a case of **bronchitis**. Other viruses and **tuberculosis** infections can cause symptoms similar to those found during the paroxysmal stage. The presence of a pertussis-like cough along with an increase of certain specific white blood cells (lymphocytes) is suggestive of pertussis (whooping cough). However, cough can occur from other pertussis-like viruses. The most accurate method of diagnosis is to culture (grow on a laboratory plate) the organisms

obtained from swabbing mucus out of the nasopharynx (the breathing tube continuous with the nose). *B. pertussis* can then be identified by examining the culture under a microscope.

Treatment

Treatment with the antibiotic erythromycin is helpful only at very early stages of whooping cough, during incubation and early in the catarrhal stage. After the cilia and the cells bearing those cilia, are damaged, the process cannot be reversed. Such a person experiences the full progression of whooping cough symptoms; symptoms only improve when the old, damaged lining cells of the respiratory tract are replaced over time with new, healthy, cilia-bearing cells. However, treatment with erythromycin is still recommended, to decrease the likelihood of *B. pertussis* spreading. In fact, all members of the household in which an individual with whooping cough lives should be treated with erythromycin to prevent the spread of *B. pertussis* throughout the community. The only other treatment is supportive and involves careful monitoring of fluids to prevent **dehydration**, rest in a quiet, dark room to decrease paroxysms, and suctioning of mucus from the lungs.

Prognosis

Just under 1 percent of all cases of whooping cough in the United States result in death. Children who die of whooping cough usually have one or more of the following three conditions present:

- severe pneumonia, perhaps with accompanying encephalopathy
- extreme weight loss, weakness, and metabolic abnormalities due to persistent vomiting during paroxysms of coughing
- other pre-existing conditions, so that the person is already in a relatively weak, vulnerable state (such conditions may include low birth weight babies, poor **nutrition**, infection with the **measles** virus, presence of other respiratory or gastrointestinal infections or diseases)

Prevention

The mainstay of prevention lies in programs similar to the mass immunization program in the United States that begins immunization inoculations when infants are two months old. The pertussis vaccine, most often given as one immunization together with **diphtheria** and **tetanus**, has greatly reduced the incidence of whooping cough. Three injections (a primary and two booster shots) during early infancy confer complete immunity. Unfortunately, in the 1990s, there has been some concern about serious neurologic side effects from the vaccine itself. This concern led significant numbers of parents in England, Japan, and Sweden to avoid immunizing their children. Such actions led to major epidemics of the disease in those countries. However, several carefully constructed research studies disproved the idea that the pertussis vaccine is the cause of neurologic damage. Furthermore, a subsequent formulation of the pertussis vaccine became available. Unlike the former whole cell pertussis vaccine, which was composed of the entire bacterial cell that has been deactivated (and therefore unable to cause infection), the subsequent acellular pertussis vaccine does not use a whole cell of the bacteria but is comprised of between two and five chemical components of the *B. pertussis* bacteria. The acellular pertussis vaccine appears to greatly reduce the risk of unpleasant reactions to the vaccine, including high **fever** and discomfort following **vaccination**.

Nutritional concerns

Persons with whooping cough should be given adequate nutrition to assist their bodies in recovering from the infective agent.

Parental concerns

Parents should ensure that their children receive a complete series of immunizations (three injections) against whooping cough. Children who are suspected of having whooping cough should be seen by a healthcare professional. Early treatment is essential to limit the progression of the disease.

KEY TERMS

Cilia—Tiny hairlike projections on certain cells within the body. Cilia produce lashing or whipping movements to direct or cause motion of substances or fluids within the body. Within the respiratory tract, the cilia act to move mucus along, in an effort to continually flush out and clean the respiratory tract.

Encephalopathy—Any abnormality in the structure or function of brain tissues.

Hernia—A rupture in the wall of a body cavity, through which an organ may protrude.

Resources

BOOKS

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Williams syndrome

Definition

A rare congenital (present from birth) genetic disorder that results in physical and developmental delays and problems.

Description

Williams syndrome (WS) is sometimes also referred to as Williams-Beuren syndrome. The disorder was first described by J. C. P. Williams of New Zealand in 1961. WS is a genetic disorder that can be inherited but often arises through spontaneous change in a chromosome (mutation). Children with WS usually have a variety of physical problems, especially problems with hearts defects. They have "elfin" faces and usually are of short stature. Children with WS are often overfriendly and have varying intellectual disabilities, with relatively good skills in music and language.

Demographics

WS is estimated to occur in about one in 20,000 births. It affects about the same number of boys and girls.

Causes and symptoms

WS is thought to be caused by a deletion of genetic information on chromosome 7. WS can be passed down from parent to child, but it often arises spontaneously. The way in which WS spontaneously arises is not clear.

Physical characteristics typical of Williams syndrome include a broad forehead, puffiness around the eyes, starburst eye pattern (usually in green or blue-eyed children), upturned nose, depressed nasal bridge, full lips, widely spaced teeth, and small chin. In addition, a child with Williams syndrome often exhibits sloping shoulders or an elongated neck. Many individuals with Williams syndrome have heart disorders, typically supravalvular aortic stenosis (SVAS), which is a narrowing of the aorta. Kidney and bladder problems are also common. Poor muscle tone and problems with the skeletal joints become evident as a child with Williams syndrome moves into **adolescence**. As the child gets older **hypertension** often becomes a problem.

Williams syndrome babies typically have a low birth weight and are often diagnosed as failing to thrive.

Elevated levels of calcium in the blood (hypercalcemia) may develop in infancy, but this usually resolves without intervention in the first two years. Digestive system symptoms such as **vomiting**, **constipation**, and feeding difficulties may occur. The infant may not be able to settle into a normal **sleep** pattern and may seem to be extremely sensitive to noise, exhibiting agitation or distress when exposed to high-pitched sounds, such as electrical appliances, motors, and loud bangs.

By the time a child with Williams syndrome is ready to enter school, mild to severe learning difficulties may appear, including impulsiveness and poor concentration. Contributing to classroom difficulties are problems with vision and spatial relations. Concepts involving numbers—especially math and time—appear to be more difficult for children with WS. In the later elementary school years, a child with Williams syndrome may be more adept at producing language than at comprehending it. Poor muscle tone and physical development continue to contribute to difficulties with gross and **fine motor skills**. The child with WS may have difficulty forming relationships with peers, preferring the company of younger children or adults. Throughout childhood, the child with Williams syndrome may exhibit deficits in the ability to reason and in self-help skills.

Children with WS are overly social and outgoing, inappropriately friendly to adults and unwary of strangers. They are usually talkative, with intense enthusiasm bordering on obsession for topics that interest them.

Special care needs to be taken when children with Williams syndrome are given anesthesia.

When to call the doctor

If a parent notices that a child has the symptoms of WS the doctor should be consulted.

Diagnosis

Williams syndrome is present from birth, although it often remains undiagnosed until a later stage of development. After a child has missed several developmental milestones, the pediatrician may refer the child to a specialist for diagnosis. Developmental delays that are typical include delay in sitting or walking. Also commonly observed are poor fine motor coordination and delayed development in language (although individuals with WS go on to develop excellent language skills). After reviewing the child's medical and **family** history, physical condition, and observing the child's behavior, a specialist in birth defects may identify Williams syndrome. In many cases, a heart murmur or suspected heart disorder may lead a cardiologist to suspect Williams syndrome, since an estimated 70 to 75 percent of people with WS have mild to severe cardiovascular problems.

Until the early 2000s, the diagnosis of WS made based on the child having a certain number of the symptoms of the disease. As of 2004, it was possible to test a child's genes for the deletion that causes WS. A test technique known as fluorescent in situ hybridization (FISH) may be used to detect this deletion. This test is only done when it is considered very likely that a child has WS because many of the clinical features are present.

Treatment

Williams syndrome cannot be cured, but the ensuing symptoms, developmental delays, learning problems, and behaviors can be treated. Many different experts work together to help develop a comprehensive treatment plan that is geared to the needs of a specific child. Children need to be monitored regularly by a doctor to ensure that problems, especially cardiac problems and hypertension, do not arise. If such problems do arise, they need to be treated promptly. Non-physical treatment often involves teaching children life skills that will eventually allow them to live on their own or with minimal care and to hold jobs. Specialists who can be helpful in treating Williams syndrome include the following:

- cardiologist, to diagnose and prescribe treatment for heart or circulatory problems
- endocrinologist, to prescribe treatment if elevated calcium levels are detected in infancy
- pediatric radiologist, to conduct diagnostic renal and bladder ultrasound tests to diagnose and prescribe treatment for any abnormalities present
- occupational therapist, to assess development delays and prescribe a plan for therapy to acquire skills necessary for daily living

Prognosis

In most cases, the child with WS will require multidisciplinary care throughout adult life, with continued medical **assessment** to diagnose and treat medical complications early. The ability to live independently and to work are usually not limited by the physical problems, which are treated successfully in the majority of cases. Rather, psychological characteristics and the inability to behave appropriately in social settings are more likely to prevent the individual from living and functioning completely on his or her own. However, each year more individuals with William syndrome are able to live independently in supervised apartment settings.

KEY TERMS

Fluorescence in situ hybridization (FISH)—A technique for diagnosing genetic disorders before birth by analyzing cells obtained by amniocentesis with DNA probes.

Hypercalcemia—A condition marked by abnormally high levels of calcium in the blood.

Supravalvular aortic stenosis (SVAS)—A narrowing of the aorta.

Prevention

There was as of 2004 no known way to prevent Williams syndrome.

Parental concerns

Children with Williams syndrome usually grow up physically healthy as long as they receive treatment for any problems, especially cardiac problems, that arise. The amount of independence that a child with Williams syndrome will eventually be able to achieve usually depends on the particular symptoms of that child.

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Williams Syndrome Foundation. Williams Syndrome Foundation, University of California, Irvine, CA 926972300. Web site: <www.wsf.org>

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Willi-Prader syndrome *see* **Prader-Willi syndrome**

Wilms' tumor

Definition

Wilms' tumor is a cancerous tumor of the kidney that usually occurs in young children.

Description

When an unborn baby is developing, the kidneys are formed from primitive cells. Over time, these cells become more specialized. The cells mature and organize into the normal kidney structure. Sometimes, clumps of these cells remain in their original, primitive form. If these more primitive cells begin to multiply after birth, they may ultimately form a large mass of abnormal cells. This is known as a Wilms' tumor. Wilms' tumor may occur in only one or in both kidneys. About 7 percent of all cases of Wilms' tumor occur bilaterally (in both kidneys simultaneously).

Wilms' tumor is a type of malignant tumor. This means that it is made up of cells that are significantly immature and abnormal. These cells are also capable of invading nearby structures within the kidney and traveling out of the kidney into other structures. Malignant cells can even travel through the body to invade other organ systems, most commonly the lungs and brain. These features of Wilms' tumor make it a type of **cancer** that, without treatment, would eventually cause death. However, advances in medicine between the 1980s and the early 2000s have made Wilms' tumor a very treatable form of cancer.

Wilms' tumor occurs almost exclusively in young children. The average patient is about three years old. Females are only slightly more likely than males to develop Wilms' tumors. In the United States, Wilms' tumor occurs in about eight children per million in white children under the age of 15 years. Wilms' tumor makes up about 6 percent of all childhood cancers and ranks as the second most frequent cancerous abdominal tumor in children. The rate is higher among African Americans

and lower among Asian Americans. Wilms' tumors are found more commonly in patients with other types of birth defects. These defects include the following:

- absence of the colored part (the iris) of the eye (aniridia)
- enlargement of one arm, one leg, or half of the face (hemihypertrophy)
- certain birth defects of the urinary system or genitals
- certain genetic syndromes (WAGR syndrome, Denys-Drash syndrome, and Beckwith-Wiedemann syndrome)

Causes and symptoms

The cause of Wilms' tumor is not as of 2004 completely understood. Because 15 percent of all patients with this type of tumor have other inherited defects, it seems clear that at least some cases of Wilms' tumor may be due to an inherited alteration. It appears that the tendency to develop a Wilms' tumor can run in families. In fact, about 1.5 percent of all children with a Wilms' tumor have **family** members who have also had a Wilms' tumor. The genetic mechanisms associated with the disease are unusually complex.

Some patients with Wilms' tumor experience abdominal **pain**, **nausea**, **vomiting**, high blood pressure, or blood in the urine. However, the parents of many children with this type of tumor are the first to notice a firm, rounded mass in their child's abdomen. This discovery is often made while bathing or dressing the child and frequently occurs before any other symptoms appear. Rarely, a Wilms' tumor is diagnosed after there has been bleeding into the tumor, resulting in sudden swelling of the abdomen and a low red blood cell count (anemia).

About 5 percent of Wilms' tumor cases involve both kidneys during the initial evaluation. The tumor appears on either side equally. When pathologists look at these tumor cells under the microscope, they see great diversity in the types of cells. Some types of cells are associated with a more favorable outcome in the patient than others. In about 15 percent of cases, physicians find some degree of cancer spread (metastasis). The most common sites in the body where metastasis occurs are the liver and lungs.

Researchers have found evidence that certain types of lesions occur before the development of the Wilms' tumor. These lesions usually appear in the form of stromal, tubule, or blastemal cells.

Diagnosis

Children with Wilms' tumor generally first present to physicians with a swollen abdomen or with an obvious abdominal mass. The physician may also find that the child has **fever**, bloody urine, or abdominal pain. The physician will order a variety of tests before imaging is performed. These tests mostly involve blood analysis in the form of a white blood cell count, complete blood count, **platelet count**, and serum calcium evaluation. Liver and kidney function testing will also be performed as well as a urinalysis.

Initial diagnosis of Wilms' tumor is made by looking at the tumor using various imaging techniques. Ultrasound, **computed tomography** scans (CT scans) and **magnetic resonance imaging** (MRI scans) are helpful in diagnosing Wilms' tumor. Intravenous pyelography, in which a dye injected into a vein helps show the structures of the kidney, can also be used in diagnosing this type of tumor. Final diagnosis, however, depends on obtaining a tissue sample from the mass (biopsy) and examining it under a microscope in order to verify that it has the characteristics of a Wilms' tumor. This biopsy is usually done during surgery to remove or decrease the size of the tumor. Other studies (chest x rays, CT scan of the lungs, bone marrow biopsy) may also be done in order to see if the tumor has spread to other locations.

Treatment

Treatment for Wilms' tumor almost always begins with surgery to remove or decrease the size of the kidney tumor. Except in patients who have tumors in both kidneys, this surgery usually requires complete removal of the affected kidney. During surgery, the surrounding lymph nodes, the area around the kidneys, and the entire abdomen will also be examined. While the tumor can spread to these surrounding areas, it is less likely to do so compared to other types of cancer. In cases where the tumor affects both kidneys, surgeons will try to preserve the kidney with the smaller tumor by removing only a portion of the kidney, if possible. Additional biopsies of these areas may be done to see if the cancer has spread. The next treatment steps depend on whether the cancer has spread and if it has what other sites are involved. Samples of the tumor are also examined under a microscope to determine particular characteristics of the cells making up the tumor.

Information about the tumor cell type and the spread of the tumor is used in deciding the best kind of treatment for a particular patient. Treatment is usually a combination of surgery, medications used to kill cancer cells (**chemotherapy**), and x rays or other

high-energy rays used to kill cancer cells (radiation therapy). These therapies are called adjuvant therapies, and this type of combination therapy has been shown to substantially improve outcome in patients with Wilms' tumor. It has long been known that Wilms' tumors respond to radiation therapy. Likewise, some types of chemotherapy have been found to be effective in treating Wilms' tumor. These effective drugs include dactinomycin, doxorubicin, vincristine, and cyclophosphamide. In rare cases, bone marrow transplantation may be used.

The National Wilms' Tumor Study Group developed a staging system to describe Wilms' tumors. All of the stages assume that surgical removal of the tumor has occurred. Stage I involves favorable Wilms' tumor cells and is usually treated successfully with combination chemotherapy involving dactinomycin and vincristine and without abdominal radiation therapy. Stage II tumors involving a favorable histology (cell characteristics) are usually treated with the same therapy as Stage I. Stage III tumors with favorable histology are usually treated with a combination chemotherapy with doxorubicin, dactinomycin, and vincristine along with radiation therapy to the abdomen. Stage IV disease with a favorable histology is generally treated with combination chemotherapy with dactinomycin, doxorubicin, and vincristine. These patients usually receive abdominal radiation therapy and lung radiation therapy if the tumor has spread to the lungs.

In the case of Stage II through IV tumors with unfavorable, or anaplastic, cells, then the previously-mentioned combination chemotherapy is used along with the drug cyclophosphamide. These patients also receive lung radiation therapy if the tumor has spread to the lungs. Another type of tumor cell can be present in Stages I through IV. This cell type is called clear cell sarcoma of the kidney. If this type of cell is present, then patients receive combination therapy with vincristine, doxorubicin, and dactinomycin. All of these patients receive abdominal radiation therapy and lung radiation therapy if the tumor has spread to the lungs.

Prognosis

The prognosis for patients with Wilms' tumor is quite good, compared to the prognosis for most types of cancer. The patients who have the best prognosis are usually those who have a small-sized tumor, a favorable cell type, are young (especially under two years of age), and have an early stage of cancer that has not spread. Modern treatments have been especially effective in the treatment of this cancer. Patients with the favorable type of cell have a long-term survival rate of 93 percent, whereas those with anaplasia have a long-term survival rate of 43 percent and those with the sarcoma form have a survival rate of 36 percent.

KEY TERMS

Biopsy—The surgical removal and microscopic examination of living tissue for diagnostic purposes or to follow the course of a disease. Most commonly the term refers to the collection and analysis of tissue from a suspected tumor to establish malignancy.

Blastemal—An immature material from which cells and tissues develop.

Cancer—A disease caused by uncontrolled growth of the body's cells.

Malignant—Cells that have been altered such that they have lost normal control mechanisms and are capable of local invasion and spread to other areas of the body. Often used to describe a cancer.

Sarcoma—A type of cancer that originates from connective tissue such as bone or muscle.

Stromal—Pertaining to the type of tissue that is associated with the support of an organ.

Tubule—Tissues and cells associated with the structures that connect the renal pelvis to the glomeruli.

Prevention

There are no known ways as of 2004 to prevent a Wilms' tumor, although it is important that children with birth defects associated with Wilms' tumor be carefully monitored.

Parental concerns

Clearly, a child who is undergoing the rigors of treatment for Wilms' tumor is going to have some very difficult times. Feeling ill may cause more irritability than usual. Parents will want to consult a dietician for advice on how to provide the best possible **nutrition** for their child, who may have a hard time eating due to nausea from treatment. The child's pediatrician can help provide some guidelines to help the family understand how the child's development may be affected by the illness and treatment. Support groups can be very helpful for families who are facing cancer and cancer treatment.

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ORGANIZATIONS

American Cancer Society. 1515 Clifton Rd. NE, Atlanta, GA 30329. Web site: <www.cancer.org>.

March of Dimes Birth Defects Foundation. 1275 Mamaroneck Ave., White Plains, NY 10605. Web site: <www.modimes.org>.

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Wiskott-Aldrich syndrome

Definition

Wiskott-Aldrich syndrome (WAS) is a rare inherited disorder marked by a low level of blood platelets, eczema, recurrent infections, and a high risk of leukemia or lymph node tumors.

Description

Wiskott-Aldrich syndrome (WAS) was named for the two physicians who reported the disorder. In 1937, A. Wiskott, a physician working in Munich, described two affected boys of German ancestry who had repeated infections, a skin rash, and poor blood-clotting ability. Nearly twenty years later, R. A. Aldrich reported similar symptoms in members of an American **family** of Dutch ancestry.

WAS is inherited as an X-linked genetic disorder and thus only affects males. The gene responsible for WAS is located on the short arm of the X chromosome. Since males have only one X chromosome they only have one copy of the gene. If that copy carries the abnormal gene, they have WAS. In contrast, females have two X chromosomes. They have a normal copy of the gene on one chromosome even if an abnormal gene is on the other because the abnormal gene is very rare. The normal copy on one X chromosome is usually sufficient to prevent females from having WAS. However, women who have one abnormal copy of the WAS gene are designated as carriers. While they will not have WAS, they have a 50 percent risk of passing the gene to each of their sons who will have WAS. Carrier females also have a 50 percent risk of passing the defective copy of the gene to their daughters who also become carriers.

Researchers identified the gene for WAS in 1994 and pinpointed its location on the short arm of the X chromosome. As of 2000, over 100 different mutations had been found in the gene among WAS patients. The fact that there are many mutations explains some of the variability of symptoms among boys with WAS. However, even within the same family, affected individuals with the identical WAS gene mutation may have different degrees of severity of the disease. The mild form, X-linked thrombocytopenia, is also caused by mutations in this same gene.

Demographics

The WAS syndrome affects one in every 250,000 male children and occurs worldwide. In the year 2000, scientists estimated that about 500 Americans had WAS.

Causes and symptoms

The syndrome is caused by a defect (mutation) in a specific gene called the WAS gene that normally codes for the protein named Wiskott-Aldrich syndrome protein (WASP). This vital protein is a component of cells that are important in the body's defense against infection (lymphocytes). The same protein also functions in the cells that help prevent bleeding (platelets). A less severe form of the disease, X-linked thrombocytopenia, affects mainly the platelets.

Increased susceptibility to infections, eczema, and excessive bleeding and bruising are the hallmarks of WAS, although the symptoms can vary significantly from one patient to another. The immune system of patients with WAS produces too few B and T cells. B cells are the cells in the body that make antibodies. There are many types of T cells. Both B and T cells are needed to defend the body against infection. Because both types of cells are affected, WAS patients are subject to repeated infections from bacteria, fungi, and viruses. Ear infections, **meningitis**, and **pneumonia** are common in boys with WAS.

WAS patients also have abnormal platelets, the specialized blood cells that help to form blood clots and control bleeding. In WAS, the platelets are often too few (called thrombocytopenia) and too small. Some of the earliest symptoms of the syndrome may be noted during early infancy, including excessive bleeding after a **cir-**

cumcision, bloody **diarrhea**, and a tendency to bruise very easily.

Some patients also have too few red blood cells (anemia) and an enlarged spleen (splenomegaly). About 10 percent of patients develop malignancies, usually leukemia or tumors in the lymph nodes (non-Hodgkin's lymphoma).

Diagnosis

The diagnosis of WAS is usually suspected in male infants who have excessive bleeding, eczema, and frequent bacterial or viral infections. Special blood tests can then be ordered to confirm WAS. The blood of Wiskott-Aldrich patients shows a low **platelet count** and a weak immune (antibody) response. Blood is analyzed to determine the quantity of immunoglobulins in the blood as well as the ability of the immune system to mount an antibody response against common pathogens. It is also possible to confirm the diagnosis by obtaining a small sample of the patient's blood and analyzing the DNA for a mutation in the WAS gene. Information about the exact mutation and the quantity of WAS protein the defective gene can produce may help predict the severity of the individual's condition.

Carrier testing

If the specific WAS gene mutation is identified in an affected child, that child's mother can then be tested to confirm that she carries the gene. Other members of the mother's family may also want to consider testing to find out if they carry the same gene mutation. The first step in studying other family members is for a geneticist or genetic counselor to obtain a detailed family history and construct a pedigree (family tree) to determine which family members should be offered testing.

Prenatal diagnosis

In families in which there one child has been born with WAS, prenatal testing should be offered in subsequent pregnancies. When the mother is a carrier, there is a 50 percent chance with each subsequent pregnancy that the new baby will receive the abnormal copy of the gene. The key is to first identify the particular WAS gene mutation in the child with WAS. Then, early in a pregnancy, cells can be obtained from the developing fetus by chorionic villus sampling or **amniocentesis** and checked for the same mutation. Women who carry the abnormal WAS gene and are considering prenatal diagnosis should discuss the risks and benefits of this type of testing with a geneticist or genetic counselor.

Treatment

Standard treatments for individuals with WAS include **antibiotics** for infections and platelet and red blood cell transfusions. Corticosteroids and immune globulin may be given in an attempt to improve thrombocytopenia. Eczema can be treated with corticosteroid creams applied directly to the skin. The spleen is sometimes removed to improve thrombocytopenia. In individuals with WAS, however, removal of the spleen also increases the risk of certain types of infections. About 50 percent of individuals with WAS are helped by treatment with transfer factor, which is a substance derived from the T cells of a healthy person. Transfer factor is given to improve both blood clotting and immune functions. Bone marrow transplantation has been successful in a number of cases. It has been most successful in boys under five years of age when the donor is a sibling whose tissue type closely matches that of the individual with WAS. As of 2000, attempts were also being made to treat individuals with WAS with umbilical cord blood from unrelated newborns in cases in which the individual diagnosed with WAS has no matched sibling donor.

Prognosis

The prognosis for males diagnosed with Wiskott-Adrich syndrome is poor. The average individual lives about eight years. Death usually occurs due to severe bleeding or overwhelming infection. Those who survive into **adolescence** often develop leukemia, lymphoma, or autoimmune diseases such as vasculitis, arthritis, inflammatory bowel disease, and kidney disease.

Prevention

Although there are no available treatments to prevent the development of WAS in an individual who receives the defective gene, prenatal genetic counseling can help couples determine their risk of having a baby with WAS.

Parental concerns

Caring for a baby or child with WAS is a highly stressful task. The child's healthcare provider should help the parents decide what steps will be necessary in order to decrease the child's risk of infection. Excellent hand washing and careful food handling should always be followed, but the healthcare provider should also provide guidance about other ways to avoid exposure to infectious disease. The parents will need to balance their child's need for a normal life with peer interaction and the desire to reduce the chance of exposure to serious

KEY TERMS

Amniocentesis—A procedure performed at 16–18 weeks of pregnancy in which a needle is inserted through a woman's abdomen into her uterus to draw out a small sample of the amniotic fluid from around the baby for analysis. Either the fluid itself or cells from the fluid can be used for a variety of tests to obtain information about genetic disorders and other medical conditions in the fetus.

Anemia—A condition in which there is an abnormally low number of red blood cells in the bloodstream. It may be due to loss of blood, an increase in red blood cell destruction, or a decrease in red blood cell production. Major symptoms are paleness, shortness of breath, unusually fast or strong heart beats, and tiredness.

Chorionic villus sampling—A procedure performed at 10 to 12 weeks of pregnancy in which a needle is inserted either through the mother's vagina or abdominal wall into the placenta to withdraw a small amount of chorionic membrane from around the early embryo. The amniotic fluid can be examined for signs of chromosome abnormalities or other genetic diseases.

Eczema—A superficial type of inflammation of the skin that may be very itchy and weeping in the early stages; later, the affected skin becomes crusted, scaly, and thick.

Immune system—The system of specialized organs, lymph nodes, and blood cells throughout the body that work together to defend the body against foreign invaders (bacteria, viruses, fungi, etc.).

Mutation—A permanent change in the genetic material that may alter a trait or characteristic of an individual, or manifest as disease. This change can be transmitted to offspring.

Platelet—A cell-like particle in the blood that plays an important role in blood clotting. Platelets are activated when an injury causes a blood vessel to break. They change shape from round to spiny, "sticking" to the broken vessel wall and to each other to begin the clotting process. In addition to physically plugging breaks in blood vessel walls, platelets also release chemicals that promote clotting.

Prenatal diagnosis—The determination of whether a fetus possesses a disease or disorder while it is still in the womb.

Syndrome—A group of signs and symptoms that collectively characterize a disease or disorder.

Thrombocytopenia—A persistent decrease in the number of blood platelets usually associated with hemorrhaging.

X-linked—A gene carried on the X chromosome, one of the two sex chromosomes.

infection. Furthermore, parents of children with WAS need to maintain a high level of suspicion; when a child with WAS begins to act ill or develop a **fever**, it may be necessary to immediately begin antibiotic treatment in order to avoid a more serious infection.

Resources

ORGANIZATIONS

Immune Deficiency Foundation. 25 W. Chesapeake Ave., Suite 206, Towson, MD 21204. Web site: <www.primaryimmune.org/inside.htm>.

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NORD—National Organization for Rare Disorders Inc. Available online at <www.rarediseases.org> (accessed January 9, 2005).

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Working mothers

Definition

Working mothers, as a label, refers to women who are mothers and who work outside the home for income in addition to the work they perform at home in raising their children.

Demographics of working mothers

As of the early 2000s, more mothers in the United States are working than ever before. In the mid-1990s, 58 percent of mothers with children under the age of six, and nearly 75 percent of those with children between the ages of six and 18 were part of the paid labor force. The number of single mothers with full-time year-round jobs increased from 39 percent in 1996 to 49 percent in 2002. A growing percentage of married women living with their husbands work as well: 40 percent worked full time

in 1992, compared with 16 percent in 1970. The rapid influx of women into the labor force that began in the 1970s was marked by the confidence of many women in their ability to successfully pursue a career while meeting the needs of their children. Throughout the 1970s and 1980s the dominant ideal of the working mother was the "Supermom"; juggling meetings, reports, and presentations with birthday parties, science projects, and soccer games. With growing numbers of women confronting the competing pressures of work and home life, observers predicted that these women's needs would be accommodated by significant changes in how things were managed on both fronts: a domestic revolution in sex roles at home and a major shift toward enlightened attitudes and policies toward women in the workplace. Although there have been some changes, they have not been substantial enough to prevent many working mothers from feeling that the price for "having it all" is too high. In the early twenty-first century, some working mothers express disenchantment with the "Supermom" ideal and look for alternatives to help them create a better balance between work and **family**.

Social and economic factors affecting working mothers

It is important to recognize that mothers in the U.S. workforce are not a homogeneous group of people; there is no typical working mother. Their attitudes toward their jobs and their decisions about child care are shaped by a range of social and economic factors:

- Marital status and family structure: Statistics indicate that working mothers who are married to the fathers of their children have more stable families. Working mothers who are single or in nontraditional relationships have a more difficult time maintaining family stability even apart from the demands of their jobs. As of 2003, only 68 percent of children in the United States under the age of 18 are living with both biological parents.
- Type of work: Working mothers in business or the professions usually earn more than women with less education and often find their work psychologically satisfying. They are also often on call outside the office and may find it difficult to leave the demands of their work behind when they go home.
- Income level: Working mothers with well-paying jobs have more choices about housing, transportation, and child care arrangements than those with limited incomes.
- Number, ages, and special needs of children: All other factors being equal, women with fewer, widely spaced, and healthy children find it easier to juggle the demands of a job with those of child care than women with several children born close together or women whose children suffer from chronic illnesses or developmental difficulties.
- Age: Working mothers over 40 are more likely to develop job-related health problems than younger women. In addition, women in this age group are often coping with the care of aging parents as well as their own offspring.

Description

There are a number of different strategies that working mothers use to balance the demands of workplace and family.

The "Mommy track"

Working mothers in many fields experience conflicts between motherhood and professional advancement. Many report that once they have children their professional aspirations are not taken as seriously by colleagues or superiors. In particular, if they quit working for a time to stay home with their children, the gap in their resumes is regarded with suspicion. One study found that the earnings of women with MBAs who took even nine months off after their children were born were still 17 percent lower 10 years later than those of employees with similar qualifications but no comparable gap in their employment record. Some women feel too threatened by the repercussions of time off the job to even take a maternity leave; others report problems on reentering the workforce after such a leave. Women in highly competitive professions are especially reluctant to lighten their work loads or schedules for **fear** that such measures will signal a lower level of commitment or ability than that of their peers, and they will be automatically assigned to the infamous "Mommy track." Many women—both with and without children—in traditionally male professions still earn lower salaries and carry greater workloads than those of male colleagues with comparable credentials and work experience because of the perception that they are not the breadwinners in their families.

Household responsibilities

On the home front, married working mothers, even those whose husbands espouse an egalitarian philosophy, still find themselves saddled with most of the housework and child care responsibilities. In effect, they often have the equivalent of two jobs, a phenomenon expressed in the title of Arlie Hochschild's highly regarded study *The Second Shift*. The book reported that the husbands of

working mothers shoulder, on average, only one-third of the couple's household duties. Hochschild also noted that the tasks performed most often by men, such as repairs and home maintenance chores, can often be done at their convenience, as opposed to women's duties, such as cooking, which must be done on a daily basis and at specific times, giving women less control over their schedules. In 1990 a survey of 5,000 couples found that only 50 percent of husbands took out the garbage, 38 percent did laundry, and 14 percent ironed. Working mothers also received less help from their children, with one important exception—working single mothers, whose children helped out at home twice as much as children in other families. In addition, they often worked at tasks traditionally done by the opposite sex: boys cooked, cleaned, and babysat; girls helped with home repairs and yard work. A supplementary benefit of this development is that the daughters of single mothers have a greater than average likelihood of entering traditionally male professions offering higher pay and better opportunities for advancement.

There are signs that this "second shift" pattern may be changing. The U. S. Department of Commerce's Survey of Income and Program Participation (SIPP) reported in 1997 that one married father in four provided care for at least one child under the age of 15 while the child's mother was working. The study found that fathers who provided child care were more likely to be employed in lower-income occupations; more likely to work in service occupations (police, firefighting, maintenance, security); more likely to be military veterans; and more likely to live in the Northeast than in other parts of the United States.

Day care arrangements

More than 8 million school-age and 15 million preschool-age children in the United States are placed in the charge of substitute care givers during the hours their mothers are working. The major options for child care include staggered work hours that allow parents to meet all child care needs themselves; care by relatives or close friends; hiring a babysitter or housekeeper; and child care in a private home or at public facilities, including **day care** centers, nursery schools or preschools, and company-sponsored programs. In 1990, provisions for children under the age of five were split almost equally between in-home care by parents or other relatives and out-of-home care by nonrelatives. The percentage of child care provided by day care centers had increased from 6 percent in 1965 to 28 percent in 1990, partly because the influx of women into the workforce had narrowed the pool of female relatives and friends available to take care of other people's children. Between 1985 and 2005, employment by day care centers increased over 250 percent, representing a gain of almost 400,000 new jobs. Workplace child care facilities did not grow at the same rate: a 1995 survey found that only 10 percent of the nation's 681 major employers offered on-site care programs to their employees.

There are also a number of options for part-time child care as of the early 2000s:

- Parent babysitting cooperatives: A group of families share responsibilities for child care. Most cooperatives operate on a point basis rather than charging a monetary fee. Points are assigned to each family according to the number of its children and the number of hours of care they require.
- Sick child care: These programs send an adult caregiver to the home of a sick child on an as-needed basis. There are also day care programs run exclusively for chronically ill children.
- **Play** groups: Play groups are similar to cooperative babysitting in that several parents get together to provide opportunities for supervised play for a group of children. Most play groups meet once or twice a week for two or three hours.
- Drop-in care: Drop-in care is an option offered by some child care centers on an as-needed basis. Parents must pre-register and pay for this service, usually on an hourly basis. Drop-in care allows parents to bring their child in for three to four hours of supervised play on an occasional basis. Most child care centers that offer a drop-in option set an upper limit of 45–50 hours per child in any given month.

Alternative work arrangements

Given the failure of either home or workplace demands to ease significantly, working mothers routinely sacrifice time for themselves, and many report high levels of stress, **anxiety**, and fatigue. In addition, many still feel torn between the conflicting demands of family and career and guilt for not being able to spend more time with their children. Increasing numbers of working mothers also feel responsible for helping their own aging parents as they develop health problems and become less able to handle their own affairs. (And parents traditionally place greater demands on grown daughters than on sons.) In addition, working mothers are often expected to assume most of the responsibility in family emergencies, such as the illness of a child, which periodically disrupt their already overloaded schedules.

FLEX-TIME AND PART-TIME WORK Dissatisfied with the pressures and sacrifices of combining mothering with full-time work, many women have sought alternatives

that allow them to relax the hectic pace of their lives but still maintain jobs and careers. According to one study, the number of companies offering some type of employment flexibility to their workers rose from 51 percent in 1990 to 73 percent in 1995. Fifty-five percent offered flex-time, while 51 percent offered part-time work. In 2004, *Working Mother* magazine reported that 97 percent of the companies on their list of the 100 best companies for mothers in the workforce offered compressed workweeks or job sharing opportunities. Mothers who work part-time gain more flexibility and more time with their children, as well as time to devote to their own needs. They are able to be there when their children get home from school, attend school plays and other functions, and take their children to doctor appointments without facing conflicts at work. However, part-time work also has disadvantages, aside from the cut in pay. Many part-timers carry workloads disproportionate to the number of hours they put in, sometimes being required to be available by telephone to clients or colleagues during their hours at home. In most cases they lose health insurance coverage. They may also face the resentment of coworkers who are required to keep a nine-to-five schedule. In addition, part-time work, like time taken off the job, usually places women at a disadvantage in terms of professional advancement. Promotions come later, and the "fast-track" positions are often out of reach altogether.

SHIFT WORK Another employment pattern that works well for some couples is working different job shifts, so that the father can provide child care when the mother is at work and vice versa. Many fathers in service occupations are able to share childcare responsibilities because they can work evening or night shifts. Although shift work has the advantage of allowing both parents to work full-time jobs and increase the family's total income, its disadvantage is that it decreases the time available for all family members to share meals and other activities. One study of 4,400 dual wage-earner Canadian families with children below the age of 11 found that children whose parents worked nonstandard schedules were more likely to develop difficulties than children whose parents did not do shift work. The researchers found that this correlation held whether it was the father, the mother, or both parents who worked nonstandard hours.

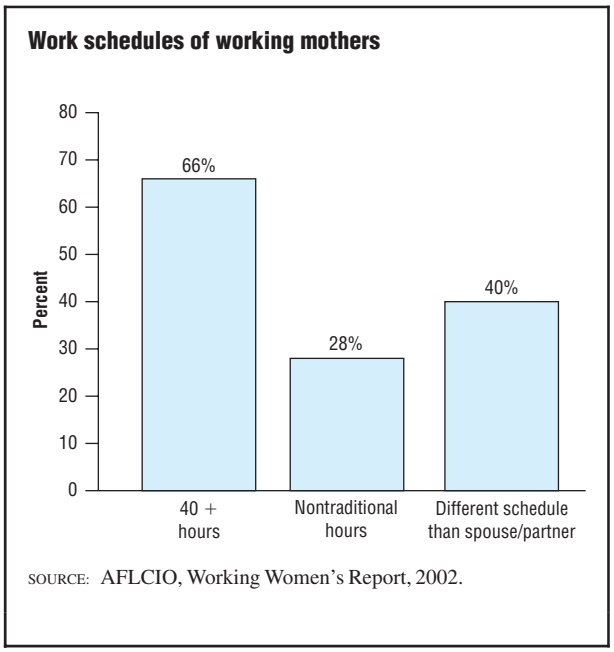
JOB SHARING An employment arrangement is job sharing, in which two people jointly fill one full-time position. They may alternate their hours in a variety of ways depending on what arrangement best suits the personal and professional needs of both people. For example, one pair of job sharers may work alternate days, while another arrangement may have each person working two days in a row and part of a third day. Job sharing opens up a wider arena of employment than that normally available to holders of traditional part-time jobs, and unlike most part-time employees, women who job share generally receive benefits prorated in accordance with the number of hours each works. For working mothers another advantage of job sharing is that people who job share often cover for each other when unusual family needs arise. In successful job sharing arrangements, the partners have a cooperative and supportive relationship, staying in close touch to maintain continuity on the job. Job sharing may be an option for a husband and wife in the same field as well as for two unrelated workers; some colleges and universities have allowed faculty couples to share a teaching position.

TELECOMMUTING The computer revolution makes possible yet another alternative work option for mothers seeking extra time and a more flexible schedule: telecommuting or working from home. According to reports in both the *Wall Street Journal* and the *New York Times*, telecommuting was the fastest-growing type of alternative work arrangement in the United States as of 2004. It can replace either all or part of one's hours at the workplace, and a telecommuter can work either part- or full-time. Telecommuters receive and send documents via their company's computer networks and can be available, if necessary, by e-mail, voice mail, and pager. Even when a telecommuting employee is expected to adhere to fixed work hours, the arrangement still provides a significant savings in time spent dressing for work, commuting, and socializing with other employees. Experts caution, however, that a woman who works at home should not expect to simultaneously take care of her children. Telecommuting mothers may want to arrange for child care during their working hours and may be interested in establishing boundaries between their work and their family life. Some employers may change the employment status of telecommuters to that of independent contractors, resulting in a loss of benefits for the workers.

ROLE REVERSAL A less common option is for the mother to become the sole family breadwinner while the father assumes the role of "househusband." Some men choose to become "stay-at-home dads" because their wives earn considerably higher salaries than they do; others simply want to spend more time with their children. One finding reported by the American Heart Association in April 2002, however, was that househusbands have a significantly higher risk of developing heart disease than men employed outside the home. The researchers who wrote the report theorized that the increased risk of heart disease is the result of stress caused by violating social expectations rather than the demands of child care.

SELF-EMPLOYMENT Some working mothers who want a challenging but flexible work schedule are drawn

to self-employment. While the number of entrepreneurs in the United States increased 56 percent overall in the 1980s, the number of female entrepreneurs grew 82 percent. Women were expected to start 2.5 million companies in the 1990s and own half of all American businesses by 2000. In the early 1990s home-based businesses started by women were the fastest-growing type of small business. The number of women employed in these ventures tripled between 1985 and 1991. Self-employment accommodated a wide range of skills and employment backgrounds, from cooking and crafts to consulting, writing, and practicing tax law. Self-employed women working at home may put in long hours and those leaving high-powered corporate jobs usually earn less money, at least initially, but they gain flexibility and control over their schedules. Like telecommuters, self-employed women may want to daycare arrangements and find strategies for separating their business and personal lives. Fortunately, start-up costs for home-based businesses are relatively low. For women requiring assistance, low-interest loans can be obtained through the Small Business Administration, which also runs a variety of training and networking programs for female entrepreneurs. A number of states also offer programs that aid women-owned businesses.



This graph of 2002 data on working mothers shows that well over half of all working mothers work over 40 hours a week. *(Graph by GGS Information Services.)*

Children of working mothers

The NICHD Study of Early Child Care, or SECC, was launched in 1991 with the enrollment of a diverse sample of 1364 children at ten different locations across the United States. Phase I of the SECC study followed these children from birth through three years of age and was completed in 1994. Phase II followed the 1226 children who remained in the study from age three through second grade between 1995 and 2000. Phase III follows the remaining 1100 participating children through 2005.

The SECC researchers reported in April 2001 that over 90 percent of the children enrolled in the study had spent some time in the care of people other than the mother, with 50 percent of the children spending 30 hours or more per week in the care of others. The report contained three major findings:

- A small minority of children (16%) who spent 30 hours or more per week in child care settings were reported to have higher levels of problem behaviors (such as fighting) than children who spent less time in care.
- The quality of nonmaternal child care makes a major difference. Children who were placed in high-quality childcare settings had better language skills and social/emotional development than those who were placed in centers with poorly trained adults or a high number of children per adult caregiver. (A good childcare center should have no more than five children per adult caregiver.) The study found that the type of care (relatives versus nonrelatives) was not significant.
- The most important single element in the children's development was their families of origin and the quality of their relationship with the mother when she was not at work.

Common problems

Common problems that working mothers confront can be summarized as follows:

- Logistical problems: These problems have to do with coordinating the details of the mother's working day, including use of the family car, arranging one's hours at work, dealing with a sick child, taking the child to the doctor for checkups, etc. Nursing mothers who return to work before an infant is weaned often have to make complicated arrangements for expressing and storing breast milk during the working day.
- Financial issues: These include the cost of child care arrangements, problems with continuity of health insurance coverage, and loss of income related to missed work. One report indicates that working mothers miss an average of 17 days of work per year due to children's healthcare needs.

- Professional development issues: Working mothers who cut back their employment to part-time work often lose opportunities for promotions as well as such benefits as health insurance. In addition, other employees often resent having to cover for working mothers who come in late, leave early, or miss work on short notice because of their child's needs.
- Health issues: Working mothers are often more vulnerable to stress-related illnesses than those who remain at home with their children. Some of the stress is related to ongoing social controversy about changing sex roles and family structures; many working mothers are made to feel guilty about their decision to continue working. In addition, working mothers often do not get enough **sleep**. Sleep deficits are known to make people more susceptible to infectious illnesses as well as automobile or workplace accidents.
- Interpersonal issues: Many working mothers, particularly those whose jobs give them little control over their work (such as food service, factory assembly-line work, retail sales work, etc.) come home at night feeling emotionally frustrated as well as physically tired. They are often concerned about the effects of job-related stress on other family members. If the family is coping with the care of elderly relatives as well as children, interpersonal stress is intensified. Parents may find themselves withdrawing emotionally from their children as well as quarreling more often with each other.

Parental concerns

Parental concerns about a mother's employment include several long-term as well as short-term issues:

- Children's future well-being: A major concern is the impact of the mother's work on her children's long-term academic success, mental health, and ability to form relationships. The SECC study appears to confirm earlier findings that the children of working mothers often benefit from her involvement in the outside world in terms of cultivating their own interests. In addition, good child care experiences often help children's social as well as emotional development.
- Stability of the marital relationship: Some couples worry about the impact of the mother's work on her relationship with her partner. In general, married couples appear to be less affected by this issue than cohabiting or lesbian couples. Many men feel less burdened by economic concerns when their wives are contributing to the family's income and report that having fewer anxieties about money actually improves their relationship with their spouse. On the other hand, some men whose wives earn higher incomes than they do may come to resent the demands of the wife's job.
- Household **safety** and security: Household safety is most likely to become an issue when the children of a working mother are too old for day care and must stay in an empty home for several hours after school before the parents return from work. Such children are sometimes called "latchkey children" because they are usually given a key to the house or apartment so that they can let themselves in when they get home. The American Academy of Adolescent and Child Psychiatry (AACAP) maintains that parents should limit as much as possible the time children must be at home alone because of the many risks involved. These risks range from physical dangers in the home (matches, knives, gas stoves, and household cleaners and other dangerous chemicals, etc.) to medical emergencies and strangers ringing the doorbell. AACAP recommends that older children should not be allowed access to "adult" channels on cable television or similar sites on the Internet. As a partial solution to parental concerns about latchkey children, some schools, churches, and synagogues offer after-school programs for children who would otherwise spend several hours at home alone.

See also Day care; Family; Parent-child relationships.

KEY TERMS

Cohabitation—Sexual partners living together outside of marriage.

Flex-time—A system that allows employees to set their own work schedules within guidelines or limits set by the employer.

Latchkey child—A child who must spend part of the after-school day at home without supervision while the parents are at work. The name comes from the fact that such children are given a house or apartment key so that they can let themselves in when they get home from school.

Telecommuting—A form of employment in which the employee works at home on a computer linked to the company's central office.

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American Academy of Pediatrics. 141 Northwest Point Boulevard, Elk Grove Village, IL 60007. Web site: <www.aap.org>.

Families and Work Institute (FWI). 267 Fifth Avenue, Floor 2, New York, NY 10016. Web site: <www.familiesandwork.org>.

Mothers' Home Business Network. PO Box 423, East Meadow, NY 11554. Web site: <www.homeworkingmom.com>.

National Institute of Child Health and Human Development (NICHD). PO Box 3006, Rockville, MD 20847. Web site: <www.nichd.nih.gov>.

United States Small Business Administration, Online Women's Business Center. Office of Women's Business Ownership, Small Business Administration, 409 Third Street SW, Fourth Floor, Washington, DC 20416. Web site: <www.onlinewbc.gov>.

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Rebecca Frey, PhD

Wounds

Definition

A wound occurs when the integrity of any tissue is compromised (e.g. skin breaks, muscle tears, **burns**, or bone **fractures**). A wound may be caused by an act (such as a gunshot, a fall, or a surgical procedure), by an infectious disease, or by an underlying condition.

Description

Types and causes of wounds are wide ranging, and healthcare professionals have several different ways of classifying them. They may be chronic, such as the skin ulcers caused by **diabetes mellitus**; or acute, such as a gunshot wound or animal bite. Wounds may also be

referred to as open, in which the skin has been compromised and underlying tissues are exposed, or closed, in which the skin has not been compromised, but trauma to underlying structures has occurred (e.g. a bruised rib or cerebral contusion). Emergency personnel and first-aid workers generally place acute wounds in one of eight categories:

- Abrasions. Also called scrapes, they occur when the skin is rubbed away by friction against another rough surface (e.g. rope burns and skinned knees).
- Avulsions. These occur when an entire structure or part of it is forcibly pulled away, such as the loss of a permanent tooth or an ear lobe. Explosions, gunshots, and animal **bites** may cause avulsions.
- Contusions. Also called **bruises**, these are the result of a forceful trauma that injures an internal structure without breaking the skin. Blows to the chest, abdomen, or head with a blunt instrument (e.g. a football or a fist) can cause contusions.
- Crush wounds. These occur when a heavy object falls onto a person, splitting the skin and shattering or tearing underlying structures.
- Cuts. These slicing wounds are made with a sharp instrument, leaving even edges. They may be as minimal as a paper cut or as significant as a surgical incision.
- Lacerations. Also called tears, these are separating wounds that produce ragged edges. They are produced by a tremendous force against the body, either from an internal source as in **childbirth**, or from an external source like a punch.
- Missile wounds. Also called velocity wounds, they are caused by an object entering the body at a high speed, typically a bullet.
- Punctures. These deep, narrow wounds are produced by sharp objects such as nails, knives, and broken glass.

Demographics

Wounds are very common. Nearly everyone has had a wound of one type or another. Minor wounds are especially common in childhood because children engage in so much rough-and-tumble **play**.

Causes and symptoms

Acute wounds have a wide range of causes. Often they are the unintentional results of motor vehicle accidents, falls, mishandling of sharp objects, or sports-related injury. Wounds may also be an intentional result of violence involving assault with weapons, including fists, knives, or guns.

The general symptoms of a wound are localized **pain** and bleeding. Specific symptoms include the following:

- An abrasion usually appears as lines of scraped skin with tiny spots of bleeding.
- An avulsion has heavy, rapid bleeding and a noticeable absence of tissue.
- A contusion may appear as a bruise beneath the skin or may appear only on imaging tests. An internal wound may also generate symptoms such as weakness, perspiration, and pain.
- A crush wound may have irregular margins like a laceration; however, the wound will be deeper and trauma to muscle and bone may be apparent.
- A cut may have little or profuse bleeding depending on its depth and length. Its even edges readily line up.
- A laceration too may have little or profuse bleeding, the tissue damage is generally greater, and the wound's ragged edges do not readily line up.
- A missile entry wound may be accompanied by an exit wound, and bleeding may be profuse, depending on the nature of the injury.
- A puncture wound's depth will be greater than its length; therefore, there is usually little bleeding around the outside of the wound and more bleeding inside, causing discoloration.

When to call the doctor

A child who has become impaled on a fixed object, such as a fence post or a stake in the ground, should only be moved by emergency medical personnel. **Foreign objects** embedded in the eye should only be removed by a doctor. Larger penetrating objects, such as a fishhook or an arrow, should only be removed by a doctor to prevent further damage as they exit.

Many times wounds can be treated at home; however, additional medical attention is necessary in several instances. Wounds which penetrate the muscle beneath the skin should be cleaned and treated by a doctor. Such a wound may require stitches to keep it closed during healing. Some deep wounds that do not extend to the underlying muscle may only require butterfly bandages to keep them closed during healing. Wounds to the face and neck, even small ones, should always be examined and treated by a doctor to preserve sensory function and minimize scarring. Deep wounds to the hands and wrists should be examined for nerve and tendon damage. Puncture wounds may require a **tetanus** shot to prevent

serious infection. Animal bites should always be examined and the possibility of **rabies** infection determined.

Infection

Wounds that develop signs of infection should also be brought to a doctor's attention. Signs of infection are swelling, redness, tenderness, throbbing pain, localized warmth, **fever**, swollen lymph glands, the presence of pus either in the wound or draining from it, and red streaks spreading away from the wound.

Emergency treatment

Even with the loss of less than one quart of blood, a child may lose consciousness and go into traumatic shock. Because this condition is life-threatening, emergency medical assistance should be called immediately. If the child stops breathing, artificial respiration (also called mouth-to-mouth resuscitation or rescue breathing) should be administered. In the absence of a pulse, **cardiopulmonary resuscitation** (CPR) must be performed. Once the child is breathing unassisted, the bleeding may be attended to.

In cases of severe blood loss, medical treatment may include the intravenous replacement of body fluids. This treatment may be infusion with saline or plasma or a transfusion of whole blood.

Diagnosis

A diagnosis is made by visual examination and may be confirmed by a report of the causal events. Medical personnel will also assess the extent of the wound and what effect it has had on the patient's well being.

Treatment of wounds involves stopping any bleeding then cleaning and dressing the wound to prevent infection. Additional medical attention may be required if the effects of the wound have compromised the body's ability to function effectively.

Treatment

Stopping the bleeding

Most bleeding may be stopped by direct pressure. Direct pressure is applied by placing a clean cloth or dressing over the wound and pressing the palm of the hand over the entire area. This pressure limits local bleeding without disrupting a significant portion of the circulation. The cloth absorbs blood and allows clot formation. The clot should not be disturbed, so if blood soaks through the cloth, another cloth should be placed directly on top rather than replacing the original cloth.

If the wound is on an arm or leg that does not appear to have a broken bone, the wound should be elevated to a height above the child's heart while direct pressure is applied. Elevating the wound allows gravity to slow down the flow of blood to that area.

If severe bleeding cannot be stopped by direct pressure or with elevation, the next step is to apply pressure to the major artery supplying blood to the area of the wound. In the arm, pressure would be applied to the brachial artery by pressing the inside of the upper arm against the bone. In the leg, pressure would be applied to the femoral artery by pressing on the inner crease of the groin against the pelvic bone.

If the bleeding from an arm or leg is so extreme as to be life-threatening and if it cannot be stopped by any other means, a tourniquet may be required. However, in the process of limiting further blood loss, the tourniquet also drastically deprives the limb tissues of oxygen. As a result, the patient may live but the limb may die.

Dressing the wound

Once the bleeding has been stopped, cleaning and dressing the wound is important for preventing infection. Although the flowing blood flushes debris from the wound, running water should also be used to rinse away dirt. Embedded particles such as wood slivers and glass splinters, if not too deep, may be removed with a needle or pair of tweezers that has been sterilized in rubbing alcohol or in the heat of a flame. Once the wound has been cleared of foreign material and washed, it should be gently blotted dry, with care not to disturb the blood clot. An antibiotic ointment may be applied. The wound should then be covered with a clean dressing and bandaged to hold the dressing in place.

Alternative treatment

In addition to the conventional treatments described above, there are alternative therapies that may help support the injured person. Homeopathy can be very effective in acute wound situations. Ledum (*Ledum palustre*) is recommended for puncture wounds (taken internally). Calendula (*Calendula officinalis*) is the primary homeopathic remedy for wounds. An antiseptic, it is used topically as a succus (juice), tea, or salve. Another naturally occurring antiseptic is tea tree oil (*Melaleuca* spp.), which can be mixed with water for cleaning wounds. Aloe (*Aloe barbadensis*) can be applied topically to soothe skin during healing. When wounds affect the nerves, especially in the arms and legs, St. John's wort (*Hypericum perforatum*) can be helpful when taken internally or applied topically. Acupuncture can help support the healing process by restoring the energy flow in the meridians that have been affected by

KEY TERMS

Abrasion—Also called a scrape. The rubbing away of the skin surface by friction against another rough surface.

Avulsion—The forcible separation of a piece from the entire structure.

Butterfly bandage—A narrow strip of adhesive with wider flaring ends (shaped like butterfly wings) used to hold the edges of a wound together while it heals.

Cut—A slicing wound made with a sharp instrument, leaving even edges.

Laceration—A cut or separation of skin or other tissue by a tremendous force, producing irregular edges. Also called a tear.

Plasma—A watery fluid containing proteins, salts, and other substances that carries red blood cells, white blood cells, and platelets throughout the body. Plasma makes up 50% of human blood.

Puncture—An injury caused by a sharp, narrow object deeply penetrating the skin.

Tourniquet—Any device that is used to compress a blood vessel to stop bleeding or as part of collecting a blood sample. Phlebotomists usually use an elastic band as a tourniquet.

Traumatic shock—A condition of depressed body functions as a reaction to injury with loss of body fluids or lack of oxygen. Signs of traumatic shock include weak and rapid pulse, shallow and rapid breathing, and pale, cool, clammy skin.

Whole blood—Blood which contains red blood cells, white blood cells, and platelets in plasma.

the wound. In some cases, vitamin E taken orally or applied topically can speed healing and prevent scarring.

Prognosis

Without the complication of infection, most wounds heal well with time. Depending on the depth and size of the wound, it may or may not leave a visible scar. Individuals with certain underlying diseases such as diabetes mellitus may have more difficulty healing.

Prevention

Most actions that result in wounds are preventable. Injuries from motor vehicle accidents may be reduced by wearing seat belts and placing children in size-appropriate car seats in the back seat. Sharp, jagged, or pointed objects or machinery parts should be used according to the manufacturer's instructions and only for their intended purpose. Firearms and explosives should be used only by adults with explicit training; they should also be kept locked and away from children. Children engaging in **sports**, games, and recreational activities should wear proper protective equipment and follow **safety** rules.

Parental concerns

Children need to be instructed not to pick at scabs, because it slows the healing process and increases the risk of infection. Wounds tend to occur often during childhood, but most of them are minor and can successfully be treated at home.

Resources

BOOKS

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Wryneck

Definition

Wryneck, also called twisted neck or torticollis, is a deformity in which the neck is twisted and held at an angle to one side. A congenital (present at birth) form called congenital torticollis is the most common type of wryneck seen in children.

Description

The sternocleidomastoid (SCM) muscle runs down either side of the neck. One end is attached to the occipital bone of the skull. The other end splits, with one end attaching to the clavicle (collarbone) and the other to the top of the sternum (breastbone). This muscle is involved

in the complex movements of flexing the neck bones (cervical spine) and rotating the head up and down and sideways. Wryneck affects the SCM muscle, usually on only one side of the neck, causing the neck to spasm painfully and twist.

There are several different types of wryneck. Acute wryneck is the most common type. It develops suddenly, often for no apparent reason, and causes painful spasms that make the individual tilt the neck at an angle. The condition lasts one to two weeks, then symptoms disappear on their own without medical intervention. This type of wryneck is seen most often in older children and adults.

Adults can also develop spasmodic torticollis with head tilt and jerky head movements. This condition can develop from injury to the bones of the neck or because of infection, inflammation, or tumors of the soft tissue of the head and neck. Most often adult torticollis develops between the ages of 30 and 60. Adult onset torticollis is not be discussed here.

Congenital muscular torticollis is a neck deformity that affects newborns. It limits the range of neck motion and causes infants to tilt their head. It is the most common type of wryneck seen in young children and is different from acute wryneck, because it does not cause **pain** and does not resolve on its own. It arises from different causes than adult-onset torticollis.

Infants who have congenital muscular torticollis appear normal when they are born. However, within about a month, they often develop a non-tender lump on the side of the neck. Although this lump disappears by itself after about three months, the SCM muscle becomes tight, contracted, and fibrous. It does not stretch. The child then begins to tilt his head toward the affected side and point his chin toward the opposite shoulder. About three-quarters of the time, the right side is affected, causing the head to tilt to the right.

Demographics

Congenital torticollis is a rare disorder. It affects fewer than 0.4 percent of newborns. It is more common in first children than in later children and appears to be more common in babies born with a breech presentation (feet first). For reasons that are not understood, about 20 percent of children with congenital muscular torticollis also have **congenital hip dysplasia**. Hip dysplasia is a deformity in which the ball and socket of the hip joint do not mate properly.

Causes and symptoms

Congenital torticollis is thought to be caused by trauma around the time of birth. There are two theories about how this trauma occurs. One theory suggests that damage occurs during the birth causing a blood clot to form in the SCM muscle. This blood clot eventually leads to scarring in the muscle. The scar tissue does not stretch and causes the muscle to shorten and pull the neck out of position. Support for this theory comes from the observation that children with congenital torticollis are often breech or difficult forceps deliveries.

A second theory is that the trauma occurs before birth. It is believed that either pressure on the SCM muscle due to position of the head in the uterus causes the muscle to become fibrous and shorten or that the blood supply to the muscle is disturbed and the muscle becomes scarred. In either case, the result is scarring and shortening of the SCM muscle.

In rare cases, congenital torticollis can also be a symptom of other congenital disorders including abnormalities of the neck vertebra such as **spina bifida** or Arnold-Chiari syndrome. Torticollis can also be caused at an older age by fracture or dislocation of the neck vertebra or juvenile rheumatoid arthritis.

The causes of acute wryneck in older children and adults are not usually clear but seem to be related to wrenching the neck muscles, sleeping with the neck in an odd position or similar causes. Acute wryneck is briefly uncomfortable but not serious.

Symptoms of congenital torticollis are a painless mass on the neck appearing during the first two months of life and a persistent tilt of the head to one side for no other apparent reason. The child has limited ability to turn his head or move his neck (limited range of motion).

Symptoms of acute wryneck are sudden development (often overnight) of pain and stiffness in the neck sometimes accompanied by **muscle spasms** that cause an individual to hold the neck at an angle to try to relieve the pain.

When to call the doctor

Parents of newborns should call the doctor if they notice a lump on their child's neck or any time that their child persists in holding the head at an angle.

Diagnosis

Normally x rays of the neck are done to check for **fractures**. A **computed tomography** (CT) scan and/or a **magnetic resonance imaging** (MRI) scan is done to

check for abnormalities in the soft tissue, such as tumors. Electromyography (EMG) is a technique that records the electrical activity of skeletal muscles. This exam can be useful in determining the extent of muscle and nerve involvement. Electromyography is usually done before surgery.

Treatment

Treatment should begin immediately for infants with torticollis. Delayed treatment increases the chance that the head tilt will not be reversed by non-surgical means. In addition, as the child grows, the face on the tilted side may become flattened. This flattening can be reversed while the bones are young and soft but after one year of age is likely to be permanent. Another reason to begin treatment early is that children with head tilt have more difficulty learning to walk and fall more often because their balance is affected by the way their head is twisted to one side of the body.

Conservative treatment

Conservative treatment for congenital torticollis should begin as soon as the condition is diagnosed. Physical therapy is begun with turning and bending the child's head four to six times per day for 15 to 20 minutes at a time. The goal of the physical therapy is to stretch and loosen the muscle and improve the range of motion. Physical therapy is continued until the child is at least one year old before surgery is considered.

Parents can supplement physical therapy by placing **toys** in positions such that the child must turn his head to see the object. This encourages use of the affected muscle. If there is improvement in the angle of head tilt and range of motion, therapy is continued. If there is no improvement after at least one year, surgery is considered.

Another conservative treatment to supplement stretching exercises is a tubular orthosis for torticollis (TOT) collar. The TOT collar is fitted by a physical therapist on infants who are at least four months old. It is made of soft plastic tubing with hard wedges of plastic inserted on the tilt side. When the head tilts into the hard plastic, it is uncomfortable, so the child tries to straighten his neck, thus exercising and stretching the affected muscles. Children wear the collar while awake and directly supervised from the age of about four months until they begin to walk.

When congenital torticollis is caused by deformities of the neck bones (vertebrae), conservative treatment involves the use of neck braces or body jackets.

Acute wryneck is treated with heat and over-the-counter non-aspirin pain medication (**acetaminophen**, ibuprofen).

Surgical treatment

The goal of surgery in congenital muscular torticollis is to cut and then reattach the SCM muscle in a way that will remove the constricting bands of fibrous tissue, improve range of motion, and allow the head to be held vertically. Several different surgical techniques can be used. A uniploar SCM release, sometimes called an inferior open tenotomy of the SCM, cuts and then reattaches the SCM muscle where it meets both the breastbone and collarbone. This operation requires only one incision. A bipolar SCM release, also called a bipolar z-plasty, releases muscle where it is attached to the skull and at the collarbone. It requires two incisions. The SCM muscle is cut apart in a Z and then reattached. An endoscopic technique has been pioneered by plastic surgeons. This surgery involves making a small incision behind the ear and with the help of an endoscope clipping the muscle. Other surgeries are done when the cause of torticollis is a bone deformity.

After surgery children are fitted with a soft cervical collar that is worn continuously except during physical therapy and bathing or a stiff orthopedic collar that is worn during waking hours except for physical therapy. These collars are worn for about 10 weeks while the muscles heal and strengthen. Physical therapy begins about one week after surgery. The therapy involves stretching and strengthening exercises for the neck. Surgery does not instantly allow the head to be held vertically, so physical therapy and home exercises continue at least until the head tilt disappears.

Alternative treatment

Massage is said to be helpful both in stretching and releasing the muscles as a supplement, but not a replacement for, physical therapy.

Prognosis

When discovered during the first few months of life and treated promptly and consistently with stretching exercises, about 80 to 90 percent of children recover from uncomplicated congenital muscular torticollis with conservative treatment alone. Surgery is highly successful on children who do not respond to conservative treatment, so long as their torticollis is caused by restriction of the SCM muscle. In cases where torticollis is caused by or complicated by bone deformities or other congenital defects, the outcome is less likely to be successful.

KEY TERMS

Arnold-Chiari syndrome—A congenital malformation of the base of the brain.

Cervical spine—The seven bones of the neck that form the uppermost part of the spinal column.

Endoscope—A medical instrument that can be passed into an area of the body (the bladder or intestine, for example) to allow visual examination of that area. The endoscope usually has a fiberoptic camera that allows a greatly magnified image to be shown on a television screen viewed by the operator. Many endoscopes also allow the operator to retrieve a small sample (biopsy) of the area being examined, to more closely view the tissue under a microscope.

Spina bifida—A birth defect (a congenital malformation) in which part of the vertebrae fail to develop completely so that a portion of the spinal cord, which is normally protected within the vertebral column, is exposed. People with spina bifida can suffer from bladder and bowel incontinence, cognitive (learning) problems, and limited mobility.

Torticollis is unlikely to recur if stretching and flexibility exercises for the neck are continued.

Prevention

There is no sure way to prevent wryneck and congenital torticollis; however, care should be taken to avoid as much trauma to the child as possible during delivery.

Parental concerns

Parents' concerns often are focused on the psychological impact of torticollis in children who do to respond completely to treatment. Holding the head at an angle is an obvious deformity that can cause a child to retreat from social situations. The parents of a young child who has head tilt and is just learning to walk may also be concerned about the frequency with which their child loses his balance, increasing the risk of injury.

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X

X rays

Definition

X rays are electromagnetic radiation that differentially penetrates structures within the body and creates images of these structures on photographic film or a fluorescent screen. These images are called diagnostic x rays.

Purpose

Diagnostic x rays are useful in detecting abnormalities within the body. They are a painless, non-invasive way to help diagnose problems such as broken bones, tumors, dental decay, and the presence of foreign bodies.

Description

X rays are a form of radiation similar to light rays, except that they are more energetic than light rays and are invisible to the human eye. They are created when an electric current is passed through a vacuum tube. X rays were accidentally discovered in 1895 by German physicist Wilhem Roentgen (1845-1923), who was later awarded the first Nobel Prize in physics for his discovery. Roentgen was also a photographer and almost immediately realized that the shadows created when x rays passed through the body could be permanently recorded on photographic plates. His first x-ray picture was of his wife's hand. Within a few years, x rays became a valued diagnostic tool of physicians world-wide.

How x rays work

X rays pass easily through air and soft tissue of the body. When they encounter more dense material, such as a tumor, bone, or a metal fragment, they are stopped. Diagnostic x rays are performed by positioning the part of the body to be examined between a focused beam of x rays and a plate containing film. This process is painless. The greater the density of the material that the x rays pass through, the more rays are absorbed. Thus bone absorbs more x rays than muscle or fat, and tumors may absorb more x rays than surrounding tissue. The x rays that pass through the body strike the photographic plate and interact with silver molecules on the surface of the film.

Once the film plates have been processed, dense material such as bone shows up as white, while softer tissue shows up as shades of gray, and airspaces look black. A radiologist, who is a physician trained to interpret diagnostic x rays, examines the pictures and reports to the doctor who ordered the tests. Plain film x rays normally take only a few minutes to perform and can be done in a hospital, radiological center, clinic, doctor's or dentist's office, or at bedside with a portable x-ray machine.

Special types of x-ray procedures

Mammograms are fixed plate x rays that are designed to locate tumors within the breasts. Dental x rays are designed to locate decay within the tooth. Sometimes a liquid called contrast material (for example, barium) is used to help outline internal organs such as the intestines. The contrast material absorbs x rays, helping to make soft tissue more easily visible on the x-ray films. Contrast material is commonly used in making x rays of the digestive system. The contrast liquid can be swallowed or injected, depending on the part of the body being x rayed. This may cause some minor discomfort.

Fluoroscopy is a special x-ray technique that produces real-time images on a television monitor. With fluoroscopy, contrast material is injected into a blood vessel. The physician can then watch the real-time movement of the contrast material to determine if there are blockages in circulation. Fluoroscopy is also used to help guide catheters into place in the heart during cardiac catheterization or to guide an endoscope during endoscopic surgery.

Computed tomography or CT scan works on the same principles as fixed plate x rays, only with a CT scan, an x ray tube rotates around the individual, taking hundreds of images that are then compiled by a computer to produce a two-dimensional cross section of the body. Although many images are taken to produce a CT scan, the total dose of radiation the individual is exposed to is low. Other common imaging techniques such as **magnetic resonance imaging** (MRI) and ultrasound do not use x rays.

How x rays are performed

Fixed plate x rays are extremely common diagnostic tests. A trained x-ray technologist takes the x ray. The individual is first asked to remove clothing and jewelry and to wear a hospital gown. The x ray technologist positions the patient appropriately, so that the part of the body to be x rayed will be between the x-ray beam and the film plate. Usually the individual either lies on an adjustable table or stands. Parts of the body that are especially sensitive to damage by x rays (for example, the reproductive organs, the thyroid) are shielded with a lead apron. Lead is very dense and effectively protects the body by stopping all x rays.

It is essential to remain motionless during the x ray, since movement causes the resulting picture to be blurry. Sometimes patients are asked to hold their breath briefly during the procedure. Children who are not old enough follow directions or who cannot stay still may need to be restrained or given medication to sedate them in order to keep them still enough to obtain useful results. Sometimes parents can stay with children during an x ray, unless the mother is pregnant, in which case she must protect the fetus from x-ray exposure.

If a contrast material is to be used, the individual will be given special instructions to prepare for the procedure and may be asked to remain afterwards until recovery is complete. (See Preparation and Aftercare below.)

Precautions

Although unnecessary exposure to radiation should be avoided, the low levels of radiation one is exposed to during an x ray does not cause harm with a few exceptions. Pregnant women should not have x rays unless in emergencies the benefits highly outweigh the risks. Exposure of the fetus to x rays, especially during early pregnancy can increase the risk of the child later developing leukemia. Body parts not being x rayed should be shielded with a lead apron, especially the testes, ovaries, and thyroid.

KEY TERMS

Contrast agent—Also called a contrast medium, this is usually a barium or iodine dye that is injected into the area under investigation. The dye makes the interior body parts more visible on an x-ray film.

Electromagnetic radiation—Packets of energy that develop when an electric current passes through a vacuum tube.

Endoscope—A medical instrument that can be passed into an area of the body (the bladder or intestine, for example) to allow visual examination of that area. The endoscope usually has a fiber-optic camera that allows a greatly magnified image to be shown on a television screen viewed by the operator. Many endoscopes also allow the operator to retrieve a small sample (biopsy) of the area being examined, to more closely view the tissue under a microscope.

Preparation

No special preparation is needed for fixed plate x rays unless contrast material is used. When x rays are scheduled that involve the use of contrast material, the physician will give specific instructions for preparation. For example, in a lower GI series, the individual may have to fast and use special **laxatives** to cleanse the bowel before swallowing the contrast material. Parents can prepare children for x rays be explaining what will happen and that these tests are short and painless.

Aftercare

Little aftercare is needed following an x ray. In complicated x rays where contrast material is injected into a blood vessel, the individual may need to remain under medical care for a short while to assure that there is no allergic reaction to the contrast material and recovery is complete.

Risks

Low dose exposure to x rays creates minimal cell damage and minimal risk when x rays are performed in an accredited facility. There is an increased risk that a developing fetus will develop leukemia during childhood if exposed to x-ray radiation; pregnant or potentially pregnant women should avoid x rays. There is also a

slight risk of an allergic reaction to the contrast material or dye used in certain x rays.

Parental concerns

Some parents are concerned about health consequences of their child's exposure to x-ray radiation. However, doses of radiation received in most x rays are quite similar to the environmental (background) radiation one is exposed to simply by living on Earth. Although unnecessary x rays should be avoided, in most cases, the benefits greatly outweigh the potentially small increased risk of exposure.

See also Computed tomography.

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XLA *see* **Bruton's agammaglobulinemia**

X-linked agammaglobulinemia *see* **Bruton's agammaglobulinemia**

XXY syndrome *see* **Klinefelter syndrome**

Y

Yoga

Definition

The term "yoga" comes from a Sanskrit word meaning "union." Yoga combines physical exercises, mental meditation, and breathing techniques to strengthen the muscles and relieve stress.

Purpose

Yoga has been practiced for thousands of years as a life philosophy to join the individual self with what practitioners call the Divine, Universal Spirit, or Cosmic Consciousness. However, very few individuals in the United States as of 2004 practiced yoga in this way; rather, yoga is performed as part of an **exercise** program to increase general health, reduce stress, improve flexibility and muscle strength, and alleviate certain physical symptoms, such as chronic **pain**. Because yoga is a low-impact activity and can include gentle movements, it is commonly used as part of physical therapy and rehabilitation of injuries.

Clinical and psychological studies have demonstrated that performing yoga has the following benefits:

- Physical postures strengthen and tone muscles, and when performed in rapid succession, can provide cardiovascular conditioning.
- Meditation and deep breathing can reduce stress, thereby lowering blood pressure and inducing relaxation.
- Mind/body awareness can influence mood and **self-esteem** to improve quality of life.

In addition to exercise and stress reduction, yoga is also used therapeutically to help children and adolescents with medical conditions. Yoga instructors experienced in adapting yoga postures for individuals with special needs teach yoga to children and adolescents with **Down syndrome**, **cerebral palsy**, seizure disorders, **spinal cord injury**, multiple sclerosis, **cancer**, **autism**, Asperger's syndrome, attention deficit hyperactivity disorder (ADHD), psychiatric disorders, learning disabilities, and other disabilities to help improve physical and mental functioning. Many physicians may recommend yoga for patients with **hypertension**, **asthma**, stress-related disorders, and depression. Growing interest in alternative and complementary medicine has increased the popularity of yoga in the United States and spurred research into its medical benefits. Many hospitals offer alternative or integrative medicine centers that include yoga classes.

Some yoga instructors have even pioneered yoga for infants and toddlers, practiced with one or both parents. Yoga for infants and toddlers can improve **sleep**, ease digestive problems, facilitate neuromuscular development, strengthen the immune system, deepen parent-child bonds, serve as an outlet for creative **play** and self-expression, and reduce stress and **anxiety** for both parents and children.

Description

Yoga originated in ancient India and is considered one of the longest surviving philosophical systems in the world. Some scholars have estimated that yoga is as old as 5,000 years; artifacts detailing yoga postures have been found in India from over 3000 B.C. A recent poll conducted by *Yoga Journal* found that 11 million Americans do yoga at least occasionally and 6 million perform it regularly.

Hatha yoga is the most commonly practiced branch of yoga in the United States, and it is a highly developed system of nearly 200 physical postures, movements, and breathing techniques. The yoga philosophy maintains that the breath is the most important facet of health, as the breath is the largest source of "prana," or life force, and hatha yoga uses "pranayama," which literally means the science or control of breathing.

A typical hatha yoga routine consists of a sequence of physical poses, called asanas, and the sequence is designed to work all parts of the body, with particular

emphasis on making the spine supple and increasing circulation. Each asana is named for a common thing it resembles, like the sun salutation, cobra, locust, plough, bow, eagle, tree, and the head to knee pose, to name a few. Poses named after animals are especially appealing to children, and children's yoga programs focus on those poses that mimic animals and trees. Each pose has steps for entering and exiting it, and each posture requires proper form and alignment. A pose is held for some time, depending on its level of difficulty and one's strength and stamina, and the instructor cues participants when to inhale and exhale at certain points in each posture, as breathing properly is a fundamental aspect of yoga postures. Breathing should be deep and through the nose. Mental concentration in each position is also very important, which improves awareness, poise, and posture. During a yoga routine there is often a position in which to perform meditation, called dyana, if deep relaxation is one of the goals of the sequence.

Yoga routines can take anywhere from 20 minutes to two or more hours, with one hour being a good time investment to perform a sequence of postures and a meditation. For children, 30 minutes may be the maximum span of attention for practicing yoga. Some yoga routines, depending on the teacher and school, can be as strenuous as the most difficult workout, especially those called ashtanga, or power, yoga. Other routines merely stretch and align the body while the breath and heart rate are kept slow and steady. Power yoga is only appropriate for children and adolescents who have practiced yoga for some time, or who are engaged in advanced athletic activities. Yoga achieves its best results when it is practiced as a daily discipline, and yoga can be a life-long exercise routine, offering deeper and more challenging positions as a practitioner becomes more adept. The basic positions can increase a person's strength, flexibility, and sense of well-being almost immediately, but it can take years to perfect and deepen them, which is an appealing and stimulating aspect of yoga for many.

Precautions

Children and adolescents with injuries, medical conditions, or spinal problems should consult a physician before beginning yoga. For children with special needs, parents should find a yoga teacher who is properly trained and experienced and can give children individual attention. Certain yoga positions should not be performed by a person who has a **fever** or is menstruating.

Children and adolescents who are beginners at yoga should always be properly supervised, since injuries are possible, and some advanced yoga postures, like the headstand and full lotus position, can be difficult and require strength, flexibility, and gradual preparation. Proper form and alignment should always be maintained during a stretch or posture, and the stretch or posture should be stopped if pain, **dizziness**, or excessive fatigue occurs.

KEY TERMS

Asana—A position or stance in yoga.

Dyana—The yoga term for meditation.

Hatha yoga—A form of yoga using postures, breathing methods, and meditation.

Meditation—A practice of concentrated focus upon a sound, object, visualization, the breath, movement, or attention itself in order to increase awareness of the present moment, reduce stress, promote relaxation, and enhance personal and spiritual growth.

Pranayama—The yoga practice of breathing cirrectly and deeply.

Yogi (female, yogini)— A trained yoga expert.

While yoga can be used therapeutically to help alleviate certain symptoms in children with various medical conditions, it is not a cure. A physician should be consulted for standard medical treatment.

Risks

Injuries have been reported when yoga postures were performed without proper form or concentration, or by attempting difficult positions without working up to them gradually or having appropriate supervision. Beginners sometimes report muscle soreness and fatigue after performing yoga, but these side effects diminish with practice.

Parental concerns

Parents should make sure that the yoga instructor is qualified to teach yoga to children. Yoga instructors experienced in teaching adults may not understand that teaching children requires different skills and methods. Yoga certifications and/or training in teaching children are available.

Yoga classes for children, adolescents, and teens are held at local schools, community centers, fitness clubs, and YMCAs. In addition, yoga videos for children are available online at <www.collagevideo.com>. For children

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GLOSSARY

A

ABANDONMENT. Legally, the refusal to provide adequate financial support for one's dependent child; the failure to maintain a parental relationship with one's dependent child.

ABDUCTION. Turning away from the body.

ABLATION. To remove or destroy tissue or a body part, such as by burning or cutting.

ABO INCOMPATABILITY. The reaction that occurs with blood groups that are of a different type.

ABRASION. Also called a scrape. The rubbing away of the skin surface by friction against another rough surface.

ABRUPTIO PLACENTAE. Premature separation of the placenta from the uterine wall. It occurs late in pregnancy and results in bleeding that may or may not establish an obstetrical emergency.

ABSCESS. A localized collection of pus in the skin or other body tissue caused by infection.

ABSENCE SEIZURE. A brief seizure with an accompanying loss of awareness or alertness. Also known as a petit mal seizure.

ACCOMMODATION. The ability of the lens to change its focus from distant to near objects and vice versa. It is achieved through the action of the ciliary muscles that change the shape of the lens.

ACELLUAR. Without whole cells. An acelluar vaccine contains on parts of the cells which can produce immunity in a person receiving the vaccine.

ACETABULUM. The large cup-shaped cavity at the junction of pelvis and femur (thigh bone).

ACETAMINOPHEN. A drug used for pain relief as well as to decrease fever. A common trade name for the drug is Tylenol.

ACETYLCHOLINE. A chemical called a neurotransmitter that functions primarily to mediate activity of the nervous system and skeletal muscles.

ACETYLSALICYLIC ACID. Aspirin; an analgesic, antipyretic, and antirheumatic drug prescribed to reduce fever and to relieve pain and inflammation.

ACHONDROPLASIA. A congenital disturbance of growth plate development in long bones that results in a person having shortened limbs and a normal trunk.

ACIDOSIS. A disturbance of the balance of acid to base in the body causing an accumulation of acid or loss of alkali (base). Blood plasma normally has a pH of 7.35-7.45. Alkaline blood has a pH value greater than pH 7.45. When the blood pH value is less than 7.35, the patient is in acidosis.

ACNE. A chronic inflammation of the sebaceous glands that manifests as blackheads, whiteheads, and/or pustules on the face or trunk.

ACOUSTIC NEUROMA. A benign tumor that grows on the nerve leading from the inner ear to the brain. As the tumor grows, it exerts pressure on the inner ear and causes severe vertigo.

ACQUIRED IMMUNODEFICIENCY SYNDROME (AIDS). An infectious disease caused by the human immunodeficiency virus (HIV). A person infected with HIV gradually loses immune function, becoming less able to resist other infections and certain cancers.

ACROCYANOSIS. A condition characterized by blueness, coldness, and sweating of the extremities. A slight cyanosis, or blueness, of the hands and feet of the newborn is considered normal.

ACROMEGALY. A rare disease resulting from excessive growth hormone caused by a benign tumor. If such a tumor develops within the first ten years of life, the result is gigantism (in which growth is accelerated) and not acromegaly. Symptoms include coarsening of the facial

features, enlargement of the hands, feet, ears, and nose, jutting of the jaw, and a long face.

ACTIVE IMMUNITY. Produced by the body when the immune system is triggered to produce antibodies, either by immunization or a disease.

ACTIVE IMMUNIZATION. Treatment that provides immunity by challenging an individual's own immune system to produce antibody against a particular organism.

ACTIVE MOTION. Spontaneous; produced by active efforts. Active range of motion exercises are those that are performed by the patient without assistance.

ACTIVITIES OF DAILY LIVING (ADL). The activities performed during the course of a normal day, for example, eating, bathing, dressing, toileting, etc.

ACUPRESSURE. A traditional Chinese medical technique based on theory of *qi* (life energy) flowing in energy meridians or channels in the body. Applying pressure with the thumb and fingers to acupressure points can relieve specific conditions and promote overall balance and health.

ACUPUNCTURE. Based on the same traditional Chinese medical foundation as acupressure, acupuncture uses sterile needles inserted at specific points to treat certain conditions or relieve pain.

ACUTE. Refers to a disease or symptom that has a sudden onset and lasts a relatively short period of time.

ACUTE OTITIS MEDIA. Inflammation of the middle ear with signs of infection lasting less than three months.

ACUTE PAIN. Pain in response to injury or another stimulus that resolves when the injury heals or the stimulus is removed.

ACUTE PHASE REACTANTS. Blood proteins whose concentrations increase or decrease in reaction to the inflammation process.

ACUTE SPLENIC SEQUESTRATION. Retention of blood in the spleen.

ACYCLOVIR. An antiviral drug, available under the trade name Zovirax, used for combating chickenpox and other herpes viruses.

ADAPTIVE BEHAVIOR. The ability to do things on one's own without getting into trouble and to adapt to and manage one's surroundings.

ADDICTION. The state of being both physically and psychologically dependent on a substance or activity.

ADDUCTION. Movement toward the body.

ADENOIDS. Common name for the pharyngeal tonsils, which are lymph masses in the wall of the air passageway (pharynx) just behind the nose.

ADENOMA. A type of noncancerous (benign) tumor that often involves the overgrowth of certain cells found in glands. These tumors can secrete hormones or cause changes in hormone production in nearby glands.

ADENOMYOSIS. Uterine thickening caused when endometrial tissue, which normally lines the uterus, extends outward into the fibrous and muscular tissue of the uterus.

ADENOSINE DEAMINASE (ADA). An enzyme that is lacking in a specific type of severe combined immunodeficiency disease (SCID). Children with an ADA deficiency have low levels of both B and T cells.

ADENOVIRUS. A type of virus that can cause upper respiratory tract infections.

ADIPOSE TISSUE. Fat tissue.

ADJUVANT THERAPY. A treatment that is intended to aid primary treatment.

ADOLESCENCE. A period of life in which the biological and psychosocial transition from childhood to adulthood occurs.

ADOPTEE. A person who has been adopted.

ADOPTION. The legal process that creates a parent and child relationship between two individuals who are not biologically related at birth.

ADOPTION SUBSIDY. A short-term or long-term financial payment, either in the form of cash or services, to help an adoptive family provide for the on-going care of an adopted child. A subsidy can be medical insurance for the child, counseling services for the family, respite care for the adoptive parents; or a monthly cash allowance to help cover other extraordinary expenses and services associated with the adoption.

ADRENAL GLAND. A small gland located above the kidney (one on each side) that secretes various hormones.

ADRENALINE. Another name for epinephrine, the hormone released by the adrenal glands in response to stress. It is the principal blood-pressure raising hormone and a bronchial and intestinal smooth muscles relaxant.

ADRENOCORTICOTROPIC HORMONE (ACTH). Also called adrenocorticotropin or corticotropin, this hormone is produced by the pituitary gland to stimulate the adrenal cortex to release various corticosteroid hormones.

ADVERSE EFFECT. A negative side effect of a vaccine, drug, or other treatment.

AEROBIC. An organism that grows and thrives only in environments containing oxygen.

AFFECTIVE DISORDER. An emotional disorder involving abnormal highs and/or lows in mood. Now termed mood disorder.

AGAMMAGLOBULINEMIA. The lack of gamma globulins in the blood associated with an increased susceptibility to infection.

AGAR. A gel made from red algae that is used to culture certain disease agents in the laboratory.

AGE OUT. Becoming a legal adult at age 18 and moving out of foster care.

AGGRAVATED SEXUAL ABUSE. When an individual is forced to submit to sexual acts by use of physical force; threats of death, injury, or kidnapping; or substances that render that individual unconscious or impaired.

AGORAPHOBIA. Abnormal anxiety regarding public places or situations from which the person may wish to flee or in which he or she would be helpless in the event of a panic attack.

AILUROPHOBIA. Fear of cats.

AIRWAY OBSTRUCTION INJURY. An injury that obstructs the airway and prevents proper breathing, either through strangulation, suffocation, or choking.

ALACTASIA. A rare inherited condition causing the lack of the enzyme needed to digest milk sugar.

ALBINISM. An inherited condition that causes a lack of pigment. People with albinism typically have light skin, white or pale yellow hair, and light blue or gray eyes.

ALBUMIN. A blood protein that is made in the liver and helps to regulate water movement in the body.

ALCOHOL USE DISORDER (AUD). The repetitive, long-term ingestion of alcohol in ways that impair psychosocial functioning and health, leading to problems with personal relationships, school, or work. Alcohol use disorders include alcohol dependence, alcohol abuse, alcohol intoxication, and alcohol withdrawal.

ALCOHOL USE DISORDERS INVENTORY TEST (AUDIT). A test for alcohol use developed by the World Health Organization (WHO). Its ten questions address three specific areas of drinking over a 12-month period: the amount and frequency of drinking, dependence upon alcohol, and problems that have been encountered due to drinking alcohol.

ALDOLASE B. Also called fructose 1-phosphate aldolase, this chemical is produced in the liver, kidneys, and brain. It is needed for the breakdown of fructose, a sugar found in fruits, vegetables, honey, and other sweeteners.

ALDOSTERONE. A hormone secreted by the adrenal glands that is important for maintaining salt and water balance in the body.

ALEXANDER'S DISEASE. A progressive, degenerative disorder of the central nervous system.

ALKALOID. A type of chemical commonly found in plants and often having medicinal properties.

ALKYLATING AGENT. A chemical that alters the composition of the genetic material of rapidly dividing cells, such as cancer cells, causing selective cell death; used as a chemotherapeutic agent.

ALLELE. One of two or more alternate forms of a gene.

ALLERGEN. A foreign substance that provokes an immune reaction or allergic response in some sensitive people but not in most others.

ALLERGIC CONJUNCTIVITIS. Inflammation of the membrane lining the eyelid and covering the eyeball; congestion of the conjunctiva, with mucus secretion.

ALLERGIC REACTION. An immune system reaction to a substance in the environment; symptoms include rash, inflammation, sneezing, itchy watery eyes, and runny nose.

ALLERGIC RHINITIS. Swelling and inflammation of the nasal membranes caused by sensitivity to airborne matter like pollen or cat hair.

ALLERGISTS. Doctors who specialize in treating allergies.

ALLERGY. A hypersensitivity reaction in response to exposure to a specific substance.

ALLERGY SHOTS. Injections given by an allergy specialist to desensitize an allergic person. Also known as immunotherapy treatment.

ALOPECIA. The loss of hair, or baldness.

ALPHA FETOPROTEIN (AFP). A substance produced by a fetus's liver that can be found in the amniotic fluid and in the mother's blood.

ALPHA FETOPROTEIN TEST. A screening blood test that can be done after the sixteenth week of pregnancy to evaluate the possibility of spina bifida and other neural tube defects in the fetus.

ALTERNATIVE SCHOOL. An educational setting designed to accommodate educational, behavioral, and/or medical needs of children and adolescents that cannot

be adequately addressed in a traditional school environment.

ALTRUISTIC. Thinking of others' welfare.

ALVEOLAR BONE. A set of ridges from the jawbones.

ALVEOLI. The tiny air sacs clustered at the ends of the bronchioles in the lungs in which oxygen-carbon dioxide exchange takes place.

AMALGAM. A mixture (alloy) of silver and several other metals used by dentists to make fillings for cavities.

AMBIDEXTROUS. Equally competent with either hand.

AMBIENT. Surrounding.

AMBLYOPIA. Decreased visual acuity, usually in one eye, in the absence of any structural abnormality in the eye.

AMENORRHEA. The absence or abnormal stoppage of menstrual periods.

AMINO ACID. An organic compound composed of both an amino group and an acidic carboxyl group. Amino acids are the basic building blocks of proteins. There are 20 types of amino acids (eight are "essential amino acids" which the body cannot make and must therefore be obtained from food).

AMNESIA. A general medical term for loss of memory that is not due to ordinary forgetfulness. Amnesia can be caused by head injuries, brain disease, or epilepsy, as well as by dissociation.

AMNIOCENTESIS. A procedure performed at 16-18 weeks of pregnancy in which a needle is inserted through a woman's abdomen into her uterus to draw out a small sample of the amniotic fluid from around the baby for analysis.

AMNIOINFUSION. A procedure whereby a physiologic solution such as normal saline or lactated ringer's solution is infused through a lumen in an intrauterine pressure catheter into the uterus to alleviate cord compression and to help dilute meconium staining.

AMNION. Thin, tough, innermost layer of the amniotic sac.

AMNIOTIC FLUID. The liquid in the amniotic sac that cushions the fetus and regulates temperature in the placental environment. Amniotic fluid also contains fetal cells.

AMNIOTIC MEMBRANE. The thin tissue that creates the walls of the amniotic sac.

AMNIOTIC SAC. The membranous sac that contains the fetus and the amniotic fluid during pregnancy.

AMNIOTOMY. Rupturing or breaking the amniotic sac (bag of waters) to permit the release of fluid.

AMOK. A culture-specific psychiatric syndrome first described among the Malays, in which adolescent or adult males are overcome by a sudden fit of murderous fury provoked by a perceived insult or slight. Some researchers consider amok to be a variant of intermittent explosive disorder.

AMPLIFICATION. A process by which something is made larger. In clotting, only a very few chemicals are released by the initial injury; they trigger a cascade of chemical reactions which produces increasingly larger quantities of different chemicals, resulting in an appropriately sized, strong fibrin clot.

AMPUTATION. Surgical removal of any portion of the body.

AMYGDALA. An almond-shaped brain structure in the limbic system that is activated in stressful situations to trigger the emotion of fear. It is thought that the emotional overreactions in Alzheimer's patients are related to the destruction of neurons in the amygdala.

AMYLOID. A waxy, translucent, starch-like protein that is deposited in tissues during the course of certain chronic diseases such as rheumatoid arthritis and Alzheimer's disease.

AMYLOIDOSIS. The accumulation of amyloid deposits in various organs and tissues in the body so that normal functioning is compromised. Primary amyloidosis usually occurs as a complication of multiple myeloma. Secondary amyloidosis occurs in patients suffering from chronic infections or inflammatory diseases such as tuberculosis, rheumatoid arthritis, and Crohn's disease.

AMYLOPHAGIA. The compulsive eating of purified starch, typically cornstarch or laundry starch.

ANABOLIC. Refers to metabolic processes characterized by the conversion of simple substances into more complex compounds.

ANAEROBIC. An organism that grows and thrives in an oxygen-free environment.

ANALGESICS. A class of pain-relieving medicines, including aspirin and acetaminophen (Tylenol).

ANAPHYLACTOID. A non-allergic sensitivity response resembling anaphylaxis.

ANAPHYLAXIS. Also called anaphylactic shock; a severe allergic reaction characterized by airway constriction, tissue swelling, and lowered blood pressure.

ANASTOMOSIS. Surgical reconnection of two ducts, blood vessels, or bowel segments to allow flow between the two.

ANDROGENS. Hormones (specifically testosterone) responsible for male sex characteristics.

ANEMIA. A condition in which there is an abnormally low number of red blood cells in the bloodstream. It may be due to loss of blood, an increase in red blood cell destruction, or a decrease in red blood cell production. Major symptoms are paleness, shortness of breath, unusually fast or strong heart beats, and tiredness.

ANENCEPHALY. A genetic defect resulting in the partial to complete absence of the brain and malformation of the brainstem.

ANESTHESIA. Treatment with medicine that causes a loss of feeling, especially pain. Local anesthesia numbs only part of the body; general anesthesia causes loss of consciousness.

ANESTHESIOLOGIST. A medical specialist who has special training and expertise in the delivery of anesthetics.

ANEURYSM. A weakened area in the wall of a blood vessel which causes an outpouching or bulge. Aneurysms may be fatal if these weak areas burst, resulting in uncontrollable bleeding.

ANGIOEDEMA. Patches of circumscribed swelling involving the skin and its subcutaneous layers, the mucous membranes, and sometimes the organs frequently caused by an allergic reaction to drugs or food. Also called angioneurotic edema, giant urticaria, Quincke's disease, or Quincke's edema.

ANGIOGRAPHY. Radiographic examination of blood vessels after injection with a radiopaque contrast substance or dye.

ANGIOMA. A tumor (such as a hemangioma or lymphangioma) that mainly consists of blood vessels or lymphatic vessels.

ANGIOPLASTY. A medical procedure in which a catheter, or thin tube, is threaded through blood vessels. The catheter is used to place a balloon or stent (a small metal rod) at a narrowed or blocked area and expand it mechanically.

ANISOMETROPIA. An eye condition in which there is an inequality of vision between the two eyes. There may be unequal amounts of nearsightedness, farsightedness, or astigmatism, so that one eye will be in focus while the other will not.

ANODYNE. A medicinal herb or other drug that relieves or soothes pain.

ANOMALY. Something that is different from what is normal or expected. Also an unusual or irregular structure.

ANOREXIA NERVOSA. An eating disorder marked by an unrealistic fear of weight gain, self-starvation, and distortion of body image. It most commonly occurs in adolescent females.

ANOXIA. Lack of oxygen.

ANTEPARTUM. The time period of the woman's pregnancy from conception and onset of labor.

ANTHRAX. A bacterial infection, primarily of livestock, that can be spread to humans. In humans it affects the skin, intestines, or lungs.

ANTHROPOMORPHIC. Taking on human characteristics or looking like humans.

ANTIANDROGEN. A substance that blocks the action of androgens, the hormones responsible for male characteristics.

ANTIBIOTICS. Drugs that are designed to kill or inhibit the growth of the bacteria that cause infections.

ANTIBODY. A special protein made by the body's immune system as a defense against foreign material (bacteria, viruses, etc.) that enters the body. It is uniquely designed to attack and neutralize the specific antigen that triggered the immune response.

ANTICHOLINERGIC DRUG. Drugs that block the action of the neurotransmitter acetylcholine. They are used to lessen muscle spasms in the intestines, lungs, bladder, and eye muscles.

ANTICOAGULANT DRUG. A drug used to prevent clot formation or to prevent a clot that has formed from enlarging. Anticoagulant drugs inhibit clot formation by blocking the action of clotting factors or platelets. They fall into three groups: inhibitors of clotting factor synthesis, inhibitors of thrombin, and antiplatelet drugs.

ANTICONVULSANT. Drugs used to prevent convulsions or seizures. They often are prescribed in the treatment of epilepsy.

ANTIDEPRESSANT DRUG. A medication prescribed to relieve major depression. Classes of antidepressants include selective serotonin reuptake inhibitors (fluoxetine/Prozac, sertraline/Zoloft), tricyclics (amitriptyline/Elavil), MAOIs (phenelzine/Nardil), and heterocyclics (bupropion/Wellbutrin, trazodone/Desyrel).

ANTIDIURETIC HORMONE (ADH). Also called vasopressin, a hormone that acts on the kidneys to regulate water balance.

ANTIDOTE. A remedy to counteract a poison or injury. Also refers to a substance which cancels the effect of homeopathic remedies

ANTIEMETIC DRUG. A medication that helps control nausea; also called an antinausea drug.

ANTIGEN. A substance (usually a protein) identified as foreign by the body's immune system, triggering the release of antibodies as part of the body's immune response.

ANTIHISTAMINE. A drug used to treat allergic conditions that blocks the effects of histamine, a substance in the body that causes itching, vascular changes, and mucus secretion when released by cells.

ANTI-INFLAMMATORY DRUGS. A class of drugs, including nonsteroidal anti-inflammatory drugs (NSAIDs) and corticosteroids, used to relieve swelling, pain, and other symptoms of inflammation.

ANTIMETABOLITE. A drug or other substance that interferes with a cell's growth or ability to multiply.

ANTIMOTILITY DRUG. A medication, such as loperamide (Imodium), dephenoxylate (Lomotil), or medications containing codeine or narcotics that decrease the ability of the intestine to contract.

ANTIOXIDANT. Any substance that reduces the damage caused by oxidation, such as the harm caused by free radicals.

ANTIPHOSPHOLIPID ANTIBODY SYNDROME. An immune disorder that occurs when the body recognizes phospholipids (part of a cell's membrane) as foreign and produces abnormal antibodies against them. This syndrome is associated with abnormal blood clotting, low blood platelet counts, and migraine headaches.

ANTIPLATELET DRUG. Drugs that inhibit platelets from aggregating to form a plug. They are used to prevent clotting and alter the natural course of atherosclerosis.

ANTIPSYCHOTIC DRUG. A class of drugs used to control psychotic symptoms in patients with psychotic disorders such as schizophrenia and delusional disorder. Antipsychotics include risperidone (Risperdal), haloperidol (Haldol), and chlorpromazine (Thorazine).

ANTIPYRETIC DRUG. Medications, like aspirin or acetaminophen, that lower fever.

ANTIRETROVIRAL DRUGS. Several classes of drugs that are used to treat HIV.

ANTISERUM. Human or animal blood serum containing specific antibodies.

ANTISOCIAL. Actions described as impulsively aggressive, sometimes violent, that do not comply with established social and ethical codes.

ANTISOCIAL BEHAVIOR. Behavior characterized by high levels of anger, aggression, manipulation, or violence.

ANTISOCIAL PERSONALITY DISORDER. A disorder characterized by a behavior pattern that disregards for the rights of others. People with this disorder often deceive and manipulate, or their behavior might include aggression to people or animals or property destruction, for example. This disorder has also been called sociopathy or psychopathy.

ANTITOXIN. An antibody against an exotoxin, usually derived from horse serum.

ANTITUSSIVE DRUG. A drug used to suppress coughing.

ANTIVIRAL DRUG. A medication that can destroy viruses and help treat illnesses caused by them.

ANUS. The opening at the end of the intestine through which solid waste (stool) passes as it leaves the body.

ANXIETY. Worry or tension in response to real or imagined stress, danger, or dreaded situations. Physical reactions, such as fast pulse, sweating, trembling, fatigue, and weakness, may accompany anxiety.

ANXIETY DISORDER. A mental disorder characterized by prolonged, excessive worry about circumstances in one's life. Anxiety disorders include agoraphobia and other phobias, obsessive-compulsive disorder, post-traumatic stress disorder, and panic disorder.

AORTA. The main artery located above the heart that pumps oxygenated blood out into the body. The aorta is the largest artery in the body.

AORTIC VALVE. The valve between the heart's left ventricle and ascending aorta that prevents regurgitation of blood back into the left ventricle.

AORTIC VALVE STENOSIS. Narrowing of the aortic valve.

APERT SYNDROME. A craniofacial abnormality characterized by abnormal head shape, small upper jaw, and fusion of fingers and toes.

APGAR SCORE. The results of an evaluation of a newborn's physical status, including heart rate, respiratory effort, muscle tone, response to stimulation, and color of skin.

APHASIA. The loss of the ability to speak, or to understand written or spoken language. A person who

cannot speak or understand language is said to be aphasic.

APHTHOUS STOMATITIS. A specific type of stomatitis presenting with shallow, painful ulcers. Also known as canker sores.

APLASTIC ANEMIA. A disorder in which the bone marrow greatly decreases or stops production of blood cells.

APNEA. The temporary absence of breathing. Sleep apnea consists of repeated episodes of temporary suspension of breathing during sleep.

APPENDECTOMY. Surgical removal of the appendix.

APPENDIX. The worm-shaped pouch attached to the cecum, the beginning of the large intestine.

APPERCEPTION. The process of understanding through linkage with previous experience.

APPETITE SUPPRESSANT. Drug that decreases feelings of hunger. Most work by increasing levels of serotonin or catecholamine, chemicals in the brain that control appetite.

APRAXIA. Impairment of the ability to make purposeful movements, but not paralysis or loss of sensation.

ARACHNID. A large class of arthropods that includes spiders, scorpions, mites, and ticks.

ARACHNIDISM. Poisoning resulting from the bite or sting of an arachnid.

ARACHNODACTYLY. A condition characterized by abnormally long and slender fingers and toes.

ARNOLD-CHIARI SYNDROME. A congenital malformation of the base of the brain.

ARRHYTHMIA. Any deviation from a normal heart beat.

ARTERIOLE. The smallest type of artery.

ARTERIOSCLEROSIS. A chronic condition characterized by thickening, loss of elasticity, and hardening of the arteries and the build-up of plaque on the arterial walls. Arteriosclerosis can slow or impair blood circulation. It includes atherosclerosis, but the two terms are often used synonymously.

ARTERITIS. Inflammation of an artery.

ARTERY. A blood vessel that carries blood away from the heart to the cells, tissues, and organs of the body.

ARTHRITIS. A painful condition that involves inflammation of one or more joints.

ARTHROCHALASIA. Excessive looseness of the joints.

ARTICULAR BONES. Two or more bones that are connected to each other via a joint.

ASANA. A position or stance in yoga.

ASCITES. An abnormal accumulation of fluid within the abdominal cavity.

ASEPTIC. Sterile; containing no microorganisms, especially no bacteria.

ASPERGER SYNDROME. A developmental disorder of childhood characterized by autistic behavior but without the same difficulties acquiring language that children with autism have.

ASPHYXIA. Lack of oxygen.

ASPHYXIA NEONATORUM. Respiratory failure in a newborn.

ASPHYXIATION. Oxygen starvation of tissues. Chemicals such as carbon monoxide prevent the blood from carrying sufficient oxygen to the brain and other organs. As a result, the person may lose consciousness, stop breathing, and die without artificial respiration (assisted breathing) and other means of elevating the blood oxygen level.

ASPIRATION. The process of removing fluids or gases from the body by suction. Also refers to the inhalation of food or liquids into the lungs.

ASSESSMENT. In the context of psychological assessment (a structured interview), assessment is information-gathering to diagnose a mental disorder.

ASSIMILATION. The process of taking in new information by incorporating it into an existing schema.

ASSOCIATIVE PLAY. A type of play in which preschoolers engage. They participate in a similar activity but with little organization or responsibility.

ASTHMA. A disease in which the air passages of the lungs become inflamed and narrowed, causing wheezing, coughing, and shortness of breath.

ASTIGMATISM. An eye condition in which the cornea doesn't focus light properly on the retina, resulting in a blurred image.

ASYMMETRIC. Not occurring equally on both sides of the body.

ASYMPTOMATIC. Persons who carry a disease and are usually capable of transmitting the disease but who do not exhibit symptoms of the disease are said to be asymptomatic.

ATAXIA. A condition marked by impaired muscular coordination, most frequently resulting from disorders in the brain or spinal cord.

ATHEROSCLEROSIS. A disease process whereby plaques of fatty substances are deposited inside arteries, reducing the inside diameter of the vessels and eventually causing damage to the tissues located beyond the site of the blockage.

ATHETOSIS. A condition marked by slow, writhing, involuntary muscle movements.

ATOPIC DERMATITIS. An intensely itchy inflammation often found on the face, in the bend of the elbow, and behind the knees of people prone to allergies. In infants and young children, this condition is called infantile eczema.

ATOPY. A state that makes persons more likely to develop allergic reactions of any type, including the inflammation and airway narrowing typical of asthma.

ATRESIA. The congenital absence of a normal body opening or duct.

ATRIAL. Referring to the upper chambers of the heart.

ATRIAL FIBRILLATION. A type of heart arrhythmia in which the upper chamber of the heart quivers instead of pumping in an organized way. In this condition, the upper chambers (atria) of the heart do not completely empty when the heart beats, which can allow blood clots to form.

ATRIAL SEPTAL DEFECT. An opening between the right and left atria (upper chambers) of the heart.

ATROPHY. The progressive wasting and loss of function of any part of the body.

ATTACHMENT. A bond between an infant and a caregiver, usually its mother. Attachment is generally formed within the context of a family, providing the child with the necessary feelings of safety and nurturing at a time when the infant is growing and developing. This relationship between the infant and his caregiver serves as a model for all future relationships.

ATTACHMENT BEHAVIOR. Any behavior that an infant uses to seek and maintain contact with and elicit a response from the caregiver. These behaviors include crying, searching, grasping, following, smiling, reaching, and vocalizing.

ATTENDING PHYSICIAN. The doctor who is in charge of the patient's overall care and treatment in the hospital. This doctor may or may not be the child's primary physician.

ATTENTION DEFICIT HYPERACTIVITY DISORDER (ADHD). A condition in which a person (usually a child) has an unusually high activity level and a short attention span. People with the disorder may act impulsively and may have learning and behavioral problems. Sometimes called attention deficit disorder (ADD).

ATTENUATED. A live but weakened microorganism that can no longer produce disease.

AUDIOGRAM. A chart or graph of the results of a hearing test conducted with audiographic equipment. The chart reflects the softest (lowest volume) sounds that can be heard at various frequencies or pitches.

AUDIOLOGIST. A person with a degree and/or certification in the areas of identification and measurement of hearing impairments and rehabilitation of those with hearing problems.

AUDIOMETRY. The measurement of hearing ability, usually with the an audiometer.

AUDITORY BRAINSTEM RESPONSE (ABR). Brainstem auditory evoked response (BAER), brainstem evoked response (BSER), auditory evoked response (AER); a hearing test that records electrical activity in the brain in response to sound via electrodes on the scalp; used for newborns, infants, and young children.

AUDITORY DISCRIMINATION. The ability to detect small similarities and differences between sounds.

AUDITORY EVOKED POTENTIAL (AEP). A change in the neural-electrical activity in the brain in response to auditory signals.

AUDITORY PERCEPTION. The ability to comprehend and interpret auditory signals.

AURA. A subjective sensation or motor phenomenon that precedes and indicates the onset of a neurological episode, such as a migraine or an epileptic seizure. This term also is used to refer to the emanation of light from living things (plants and animals) that can be recorded by Kirlian photography.

AUTHENTIC TASK ASSESSMENT. Evaluation of a task performed by a student that is similar to tasks performed in the outside world.

AUTISM. A developmental disability that appears early in life, in which normal brain development is disrupted and social and communication skills are retarded, sometimes severely.

AUTOGENIC TRAINING. A form of self-hypnosis developed in Germany that appears to be beneficial to migraine sufferers.

AUTOIMMUNE. Pertaining to an immune response by the body against its own tissues or types of cells.

AUTONOMIC NERVOUS SYSTEM. The part of the nervous system that controls so-called involuntary functions, such as heart rate, salivary gland secretion, respiratory function, and pupil dilation.

AUTOSOMAL. Relating to any chromosome besides the X and Y sex chromosomes. Human cells contain 22 pairs of autosomes and one pair of sex chromosomes.

AUTOSOMAL DOMINANT. A pattern of inheritance in which only one of the two copies of an autosomal gene must be abnormal for a genetic condition or disease to occur. An autosomal gene is a gene that is located on one of the autosomes or non-sex chromosomes. A person with an autosomal dominant disorder has a 50% chance of passing it to each of their offspring.

AUTOSOMAL INHERITANCE. Inheritance involving any of the autosomes (22 pairs) and not involving sex-linked chromosomes X and Y.

AUTOSOMAL RECESSIVE. A pattern of inheritance in which both copies of an autosomal gene must be abnormal for a genetic condition or disease to occur. An autosomal gene is a gene that is located on one of the autosomes or non-sex chromosomes. When both parents have one abnormal copy of the same gene, they have a 25% chance with each pregnancy that their offspring will have the disorder.

AUTOSOME. A chromosome not involved in sex determination.

AVOIDANT PERSONALITY DISORDER. Chronic and longstanding fear of negative evaluation and tendency to avoid interpersonal situations without a guarantee of acceptance and support, accompanied by significant fears of embarrassment and shame in social interaction.

AVULSION. The forcible separation of a piece from the entire structure.

AVULSION FRACTURE. A fracture caused by the tearing away of a fragment of bone where a strong ligament or tendon attachment forcibly pulls the fragment away from the bone tissue.

AXILLARY. Located in or near the armpit.

AXIS. A line that passes through the center of the body or body part.

AXON. A long, threadlike projection that is part of a neuron (nerve cell).

B

BABESIOSIS. A infection transmitted by the bite of a tick and characterized by fever, headache, nausea, and muscle pain.

BABINSKI SIGN. Downward bending of the big toe on stimulating the sole of the foot.

BACILLUS. A rod-shaped bacterium, such as the diphtheria bacterium.

BACTEREMIA. Bacterial infection of the blood.

BACTERIA. Singular, bacterium; tiny, one-celled forms of life that cause many diseases and infections.

BACTERIAL ENDOCARDITIS. An infection caused by bacteria that enter the bloodstream and settle in the heart lining, a heart valve, or a blood vessel. People with congenital cardiovascular defects have an increased risk of developing bacterial endocarditis, so preventive antibiotics are prescribed before surgery, invasive tests or procedures, and dental work to reduce this risk.

BACTERIAL MENINGITIS. Meningitis caused by bacteria. Depending on the type of bacteria responsible for the infection, bacterial meningitis is either classified as monococcal or pneumococcal.

BACTERIAL SPECTRUM. The number of bacteria an antibiotic is effective against. Broad-spectrum antibiotics treat many different kinds of bacteria. Narrow-spectrum antibiotics treat fewer kinds.

BACTERIOSTATIC. An agent that prevents the growth of bacteria.

BACTERIURIA. The presence of bacteria in the urine.

BALNEOTHERAPY. The medical term for the use of baths to treat disease.

BARIATRICS. The branch of medicine that deals with the prevention and treatment of obesity and related disorders.

BARIUM ENEMA. An x ray of the bowel using a liquid called barium to enhance the image of the bowel. This test is also called a lower GI (gastrointestinal) series.

BASAL GANGLIA. Brain structure at the base of the cerebral hemispheres involved in controlling movement.

BATIK. A method of hand-printing a fabric by covering with removable wax the parts that will not be dyed.

B-CELL (B LYMPHOCYTE). A small white blood cell from bone marrow responsible for producing antibody and serving as a precursor for plasma cells.

B-CELL LYMPHOMAS. Non-Hodgkin's lymphomas that arise from B cells.

BECKER MUSCULAR DYSTROPHY (BMD). A type of muscular dystrophy that affects older boys and men and usually follows a milder course than Duchenne muscular dystrophy.

BEERY-BUKTENICA TEST. A test that identifies problems with visual perception, fine motor skills (especially hand control), and hand-eye coordination.

BEHAVIOR. A stereotyped motor response to an internal or external stimulus.

BEHAVIOR MODIFICATION. A form of therapy that uses rewards to reinforce desired behavior. An example would be to give a child a piece of chocolate for grooming appropriately.

BELL'S PALSY. Facial paralysis or weakness with a sudden onset, caused by swelling or inflammation of the seventh cranial nerve, which controls the facial muscles. Disseminated Lyme disease sometimes causes Bell's palsy.

BENIGN. In medical usage, benign is the opposite of malignant. It describes an abnormal growth that is stable, treatable, and generally not life-threatening.

BENIGN TUMOR. An abnormal proliferation of cells that does not spread to other parts of the body.

BENZODIAZEPINE. One of a class of drugs that have a hypnotic and sedative action, used mainly as tranquilizers to control symptoms of anxiety. Diazepam (Valium), alprazolam (Xanax), and chlordiazepoxide (Librium) are all benzodiazepines.

BEREAVEMENT. The emotional experience of loss after the death of a friend or relative.

BETA BLOCKERS. The popular name for a group of drugs that are usually prescribed to treat heart conditions, but that also are used to reduce the physical symptoms of anxiety and phobias, such as sweating and palpitations. These drugs, including nadolol (Corgard) and digoxin (Lanoxin), block the action of beta receptors that control the speed and strength of hear muscle contractions and blood vessel dilation. Beta blockers are also called beta-adrenergic blocking agents and antiadrenergics.

BETA-LACTAMASE. An enzyme produced by some bacteria that destroys penicillins.

BICEPS. The muscle in the front of the upper arm.

BICUSPID. Premolar; the two-cupped tooth between the first molar and the cuspid.

BICUSPID AORTIC VALVE. A condition in which the major blood vessel from the heart has only two rather than three components to the valve regulating blood flow.

BILATERAL. Occurring on two sides. For example, a patient with bilateral retinoblastoma has this retinal tumor in both eyes.

BILATERAL CLEFT LIP. A cleft that occurs on both sides of the lip.

BILE. A bitter yellow-green substance produced by the liver. Bile breaks down fats in the small intestine so that they can be used by the body. It is stored in the gallbladder and passes from the gallbladder through the common bile duct to the top of the small intestine (duodenum) as needed to digest fat.

BILE DUCTS. Tubes that carry bile, a thick yellow-green fluid that is made by the liver, stored in the gallbladder, and helps the body digest fats.

BILIARY ATRESIA. An obstruction or inflammation of a bile duct that causes bilirubin to back up into the liver.

BILIRUBIN. A reddish yellow pigment formed from the breakdown of red blood cells, and metabolized by the liver. When levels are abnormally high, it causes the yellowish tint to eyes and skin known as jaundice. Levels of bilirubin in the blood increase in patients with liver disease, blockage of the bile ducts, and other conditions.

BINGE DRINKING. Consumption of five or more alcoholic drinks in a row on a single occasion.

BINGE EATING. A pattern of eating marked by episodes of rapid consumption of large amounts of food; usually food that is high in calories.

BINOCULAR. Affecting or having to do with both eyes.

BINOCULAR VISION. Using both eyes at the same time to see an image.

BIOCHEMICAL TESTING. Measuring the amount or activity of a particular enzyme or protein in a sample of blood or urine or other tissue from the body.

BIOFEEDBACK. A training technique that enables an individual to gain some element of control over involuntary or automatic body functions.

BIOLOGICAL CLOCK. A synonym for the body's circadian rhythm, the natural biological variations that occur over the course of a day.

BIOPSY. The surgical removal and microscopic examination of living tissue for diagnostic purposes or to follow the course of a disease. Most commonly the term refers to the collection and analysis of tissue from a suspected tumor to establish malignancy.

BIOSYNTHESIS. The manufacture of materials in a biological system.

BIPOLAR DISORDER. A severe mental illness, also known as manic depression, in which a person has extreme mood swings, ranging from a highly excited state, sometimes with a false sense of well being, to depression.

BIRTH MULTIPLES. Children born in multiple births; e.g. twins, triplets, quads, etc.

BIRTH PARENTS. The biological parents of a child.

BLADDER. The muscular sac which receives urine from the kidneys, stores it, and ultimately works to remove it from the body during urination.

BLASTEMAL. An immature material from which cells and tissues develop.

BLENDED FAMILY. A family formed by the remarriage of a divorced or widowed parent. It includes the new husband and wife, plus some or all of their children from previous marriages.

BLOOD CELL COUNT. The number of red blood cells, white blood cells, and platelets in a sample of blood. Also called a complete blood count (CBC).

BLOOD CLOTTING. Also called coagulation. A natural process in which blood cells and fibrin strands clump together to stop bleeding after a blood vessel has been injured.

BLOOD CULTURE. A procedure where blood is collected from a vein and is placed in a small bottle that contains a special liquid; the liquid will make any organisms that are present in the blood sample grow. These organisms can then be grown and identified in the laboratory so that the proper antibiotic can be given to the patient.

BLOOD SERUM. A component of blood.

BLOOD VESSELS. General term for arteries, veins, and capillaries that transport blood throughout the body.

BLOOD-BRAIN BARRIER. An arrangement of cells within the blood vessels of the brain that prevents the passage of toxic substances, including infectious agents, from the blood and into the brain. It also makes it difficult for certain medications to pass into brain tissue.

BODY DYSMORPHIC DISORDER. A psychiatric disorder marked by preoccupation with an imagined physical defect.

BODY LANGUAGE. Communication without words, also sometimes referred to as ‘‘non-verbal communication’’; conscious or unconscious bodily movements and gestures that communicate to others a person’s attitudes and feelings.

BONE MARROW. The spongy tissue inside the large bones in the body that is responsible for making the red blood cells, most white blood cells, and platelets.

BONE MARROW TRANSPLANTATION. A medical procedure in which a quantity of bone marrow is extracted through a needle from a donor, and then passed into a patient to replace the patient’s diseased or absent bone marrow.

BOOSTER IMMUNIZATION. An additional dose of a vaccine to maintain immunity to the disease.

BORDERLINE PERSONALITY DISORDER (BPD). A pattern of behavior characterized by impulsive acts, intense but chaotic relationships with others, identity problems, and emotional instability.

BOTULINUM TOXIN. A potent bacterial toxin or poison made by *Clostridium botulinum*; causes paralysis in high doses, but is used medically in small, localized doses to treat disorders associated with involuntary muscle contraction and spasms, in addition to strabismus. Commonly known as Botox.

BOWEL. The intestine; a tube-like structure that extends from the stomach to the anus. Some digestive processes are carried out in the bowel before food passes out of the body as waste.

BRACES. An orthodontic appliance consisting of brackets cemented to the surface of each tooth and wires of stainless steel or nickel titanium alloy. Braces are used to treat malocclusion by changing the position of the teeth.

BRACHIAL PLEXUS. A group of lower neck and upper back spinal nerves supplying the arm, forearm, and hand.

BRACHYCEPHALY. An abnormal thickening and widening of the skull.

BRACING. Using orthopedic devices to hold joints or limbs in place.

BRADYCARDIA. A slow heart rate, usually under 60 beats per minute.

BRADYKINESIA. Extremely slow movement.

BRAIN LATERALIZATION. A function that is dominated by either the left or the right hemisphere of the brain.

BRAINSTEM. The stalk of the brain which connects the two cerebral hemispheres with the spinal cord. It is involved in controlling vital functions, movement, sensation, and nerves supplying the head and neck.

BREAKTHROUGH INFECTION. A disease that is contracted despite a successful vaccination against it.

BREECH BIRTH. Birth of a baby bottom-first, instead of the usual head first delivery. This can add to labor and delivery problems because the baby's bottom doesn't mold a passage through the birth canal as well as does the head.

BREECH PRESENTATION. The condition in which the baby enters the birth canal with its buttocks or feet first.

BROAD SPECTRUM. A term applied to antibiotics to indicate that they are effective against many different types of bacteria.

BROMOCRIPTINE. Also known as Parlodel, it is a dopamine receptor agonist used to treat galactorrhea by reducing levels of the hormone prolactin and is also used to treat Parkinson's disease.

BRONCHI. Singular, bronchus; the large tubular passages that carry air to the lung and allow air to be expelled from the lungs.

BRONCHIAL TUBES. The major airways to the lungs and their main branches.

BRONCHIECTASIS. A disorder of the bronchial tubes marked by abnormal stretching, enlargement, or destruction of the walls. Bronchiectasis is usually caused by recurrent inflammation of the airway.

BRONCHIOLE. Tubes in the lungs that carry air from the bronchi to lung tissues.

BRONCHITIS. Inflammation of the air passages of the lungs.

BRONCHODILATOR. A drug that when inhaled helps to expand the airways.

BRONCHOSCOPE. A lighted instrument that is inserted into the windpipe to view the bronchi and bronchioles, to remove obstructions, or to withdraw specimens for testing.

BRONCHOSCOPY. A procedure in which a hollow tube (bronchoscope) is inserted into the airway to allow visual examination of the larynx, trachea, bronchi, and bronchioles. It is also used to collect specimens for biopsy or culturing, and to remove airway obstructions.

BRONCHOSPASM. The tightening of the muscle bands that surround the airways, causing the airways to narrow.

BRUTON'S AGAMMAGLOBULINEMIA TYROSINE KINASE (BTK). An enzyme vital for the maturation of B cells.

BRUXISM. Habitual clenching and grinding of the teeth, especially during sleep.

BULB. The hair bulb is the expanded portion on the lower end of the hair root.

BULIMIA NERVOSA. An eating disorder characterized by binge eating and inappropriate compensatory behavior, such as vomiting, misusing laxatives, or excessive exercise.

BURSITIS. Inflammation of a bursa, a fluid-filled cavity or sac. In the body, bursae are located at places where friction might otherwise develop.

BUTTERFLY BANDAGE. A narrow strip of adhesive with wider flaring ends (shaped like butterfly wings) used to hold the edges of a wound together while it heals.

C

CAGE. A four-question assessment for the presence of alcoholism in both adults and children.

CALCIFICATION. A process in which tissue becomes hardened due to calcium deposits.

CALCINOSIS. A condition in which calcium salts are deposited in various body tissues. In juvenile dermatomyositis, calcinosis usually takes the form of small lumps of calcium compounds deposited in muscles or under the skin.

CALCULUS. Plural, calculi. Any type of hard concretion (stone) in the body, but usually found in the gallbladder, pancreas, and kidneys. They are formed by the accumulation of excess mineral salts and other organic material such as blood or mucous. Calculi (pl.) can cause problems by lodging in and obstructing the proper flow of fluids, such as bile to the intestines or urine to the bladder. In dentistry, calculus refers to a hardened yellow or brown mineral deposit from unremoved plaque, also called tartar.

CALISTHENICS. Exercise involving free movement without the aid of equipment.

CANAVAN DISEASE. A serious genetic disease more common in the Eastern European Jewish population that causes mental retardation and early death. Canavan disease is caused by the lack of an enzyme called aspartoacylase.

CANCER. A disease caused by uncontrolled growth of the body's cells.

CANINES. The two sharp teeth located next to the front incisor teeth in mammals that are used to grip and tear. Also called cuspids.

CAPILLARIES. The tiniest blood vessels with the smallest diameter. These vessels receive blood from the arterioles and deliver blood to the venules. In the lungs, capillaries are located next to the alveoli so that they can pick up oxygen from inhaled air.

CARBOHYDRATES. Compounds, such as cellulose, sugar, and starch, that contain only carbon, hydrogen, and oxygen, and are a major part of the diets of people and other animals.

CARBON DIOXIDE. A heavy, colorless gas that dissolves in water.

CARBOXYHEMOGLOBIN. Hemoglobin that is bound to carbon monoxide instead of oxygen.

CARCINOGENIC. A substance that can cause cancer to develop.

CARDIAC ARREST. Temporary or permanent cessation of the heartbeat.

CARDIAC CATHETERIZATION. A procedure to passes a catheter through a large vein into the heart and its vessels for the purpose of diagnosing coronary artery disease, assessing injury or disease of the aorta, or evaluating cardiac function.

CARDIOLOGIST. A physician who specializes in diagnosing and treating heart diseases.

CARDIOMEGALY. An enlarged heart.

CARDIOMYOPATHY. A disease of the heart muscle.

CARDIOPULMONARY. Relating to the heart and lungs.

CARDIOPULMONARY RESUSCITATION (CPR). An emergency procedure designed to stimulate breathing and blood flow through a combination of chest compressions and rescue breathing. It is used to restore circulation and prevent brain death to a person who has collapsed, is unconscious, is not breathing, and has no pulse.

CARDIOVASCULAR. Relating to the heart and blood vessels.

CARIES. The medical term for tooth decay.

CARING. The demonstration of an awareness of and a concern for the good of others.

CARNASSIALS. The last upper premolar teeth in the mouths of cats and other carnivores, adapted to shear or puncture food. Carnassial teeth often cause puncture wounds when a cat bites a human.

CAROTID ARTERY. One of the major arteries supplying blood to the head and neck.

CARRIER. A person who possesses a gene for an abnormal trait without showing signs of the disorder. The person may pass the abnormal gene on to offspring. Also refers to a person who has a particular disease agent present within his/her body, and can pass this agent on to others, but who displays no symptoms of infection.

CARRIER STATE. The continued presence of an organism (bacteria, virus, or parasite) in the body that does not cause symptoms, but is able to be transmitted and infect other persons.

CARTILAGE. A tough, elastic connective tissue found in the joints, outer ear, nose, larynx, and other parts of the body.

CASE MANAGER. A professional who designs and monitors implementation of comprehensive care plans (i.e., services addressing medical, financial, housing, psychiatric, vocational, social needs) for individuals seeking mental health or social services.

CATABOLISM. A process of metabolism that breaks down complex substances into simple ones.

CATAPLEXY. A symptom of narcolepsy in which there is a sudden episode of muscle weakness triggered by emotions. The muscle weakness may cause the person's knees to buckle, or the head to drop. In severe cases, the patient may become paralyzed for a few seconds to minutes.

CATARACT. A condition in which the lens of the eye turns cloudy and interferes with vision.

CATHARTIC COLON. A poorly functioning colon, resulting from the chronic abuse of stimulant cathartics.

CATHETER. A thin, hollow tube inserted into the body at specific points in order to inject or withdraw fluids from the body.

CAVITY. A hole or weak spot in the tooth surface caused by decay.

CD4+ CELLS. Called helper T-cells, these cells work in cell-mediated immunity by causing a form of inflammation to wall off and destroy foreign material as with a bacterial infection.

CELIAC DISEASE. A disease, occurring in both children and adults, which is caused by a sensitivity to gluten, a protein found in grains. It results in chronic inflammation and shrinkage of the lining of the small intestine. Also called gluten enteropathy or nontropical sprue.

CELLULAR RESPIRATION. The process by which food molecules are converted into high-energy molecules used as a source of energy.

CELLULOSE. The primary substance composing the cell walls or fibers of all plant tissues.

CEMENTUM. A bony substance that covers the root of the tooth.

CENTER-BASED CARE. Also called childcare centers or daycare centers, these facilities care for children in groups.

CENTRAL AUDITORY PROCESSING SKILLS. The skills needed for auditory perception, including auditory discrimination, auditory memory, auditory blending, and auditory comprehension.

CENTRAL NERVOUS SYSTEM. Part of the nervous system consisting of the brain, cranial nerves, and spinal cord.

CEPHALOCAUDAL DEVELOPMENT. Motor development which occurs in the first two years of life: head before arms and trunk, arms and trunk before legs.

CEPHALOHEMATOMA. A benign swelling of the scalp in a newborn due to an effusion of blood beneath the connective tissue that surrounds the skull, often resulting from birth trauma.

CEPHALOPELVIC DISPROPORTION. The condition in which the baby's head is too large to fit through the mother's pelvis.

CEREBELLUM. The part of the brain involved in the coordination of movement, walking, and balance.

CEREBRAL. Pertaining to the brain.

CEREBRAL EDEMA. The collection of fluid in the brain, causing tissue to swell.

CEREBRAL PALSY. A nonprogressive movement disability caused by abnormal development of or damage to motor control centers of the brain.

CEREBROSPINAL FLUID. The clear, normally colorless fluid that fills the brain cavities (ventricles), the subarachnoid space around the brain, and the spinal cord and acts as a shock absorber.

CEREBROSPINAL FLUID ANALYSIS. A laboratory test, important in diagnosing diseases of the central nervous system, that examines a sample of the fluid surrounding the brain and spinal cord. The fluid is withdrawn through a needle in a procedure called a lumbar puncture.

CEREBRUM. The largest section of the brain, which is responsible for such higher functions as speech, thought, vision, and memory.

CERUMEN. The medical term for earwax.

CERVICAL CERCLAGE. A procedure in which the cervix of the uterus is sewn closed, it is used in cases when the cervix starts to dilate too early in a pregnancy to allow the birth of a healthy baby.

CERVICAL NERVES. The eight pairs of nerves (C1C8) originating in the cervical (neck) region of the spinal cord.

CERVICAL SPINE. The seven bones of the neck that form the uppermost part of the spinal column.

CERVIX. A small, cylindrical structure about an inch or so long and less than an inch around that makes up the lower part and neck of the uterus. The cervix separates the body and cavity of the uterus from the vagina.

CESAREAN SECTION. Delivery of a baby through an incision in the mother's abdomen instead of through the vagina; also called a C-section, cesarean birth, or cesarean delivery.

CGG OR CGG SEQUENCE. Shorthand for the DNA sequence: cytosine-guanine-guanine. Cytosine and guanine are two of the four molecules, called nucleic acids, that make up DNA.

CHALAZION. A condition in which clogging of the meibomiam gland causes a cyst inside the eyelid.

CHARACTER. An individual's set of emotional, cognitive, and behavioral patterns learned and accumulated over time.

CHARITY. Giving money or providing help to the poor and needy. To make a donation of money to a religious organization.

CHELATION. The process by which a molecule encircles and binds to a metal and removes it from tissue.

CHELATION THERAPY. A treatment using chelating agents, compounds that surround and bind to target substances allowing them to be excreted from the body.

CHEMOTHERAPY. Any treatment of an illness with chemical agents. The term is usually used to describe the treatment of cancer with drugs that inhibit cancer growth or destroy cancer cells.

CHEST X RAY. Brief exposure of the chest to radiation to produce an image of the chest and its internal structures.

CHIARI II ANOMALY. A structural abnormality of the lower portion of the brain (cerebellum and brainstem) associated with spina bifida. The lower structures of the brain are crowded and may be forced into the foramen magnum, the opening through which the brain and spinal cord are connected.

CHILD DEVELOPMENT SPECIALIST. A professional who is trained in infant and toddler development and in the tools used to identify developmental delays and disabilities.

CHILD PROTECTIVE SERVICES (CPS). The designated social services agency (in most states) to receive reports, investigate, and provide intervention and treatment services to children and families in which child maltreatment has occurred. Frequently this agency is located within larger public social service agencies, such as Departments of Social Services.

CHIROPRACTIC. A method of treatment based on the interactions of the spine and the nervous system. Chiropractors adjust or manipulate segments of the patient's spinal column in order to relieve pain.

CHLAMYDIA. The most common bacterial sexually transmitted disease in the United States. It often accompanies gonorrhea and is known for its lack of evident symptoms in the majority of women.

CHOLANGIOPANCREATOGRAPHY. An examination of the bile ducts and pancreas.

CHOLERA. An infection of the small intestine caused by a type of bacterium. The disease is spread by drinking water or eating foods that have been contaminated with the feces of infected people. It occurs in parts of Asia, Africa, Latin America, India, and the Middle East. Symptoms include watery diarrhea and exhaustion.

CHOLESTEROL. A steroid fat found in animal foods that is also produced in the human body from saturated fat. Cholesterol is used to form cell membranes and process hormones and vitamin D. High cholesterol levels contribute to the development of atherosclerosis.

CHORDEE. An abnormal curvature of the penis.

CHOREOATHETOSIS. Involuntary rapid, irregular, jerky movements or slow, writhing movements that flow into one another.

CHORES. A small or minor job; a routine duty of a household or farm.

CHORION. The outer membrane of the amniotic sac. Chorionic villi develop from its outer surface early in pregnancy. The villi establish a physical connection with the wall of the uterus and eventually develop into the placenta.

CHORIONIC VILLUS SAMPLING. A procedure used for prenatal diagnosis at 10–12 weeks gestation. Under ultrasound guidance a needle is inserted either through the mother's vagina or abdominal wall and a sample of the chorionic membrane. These cells are then tested for chromosome abnormalities or other genetic diseases.

CHOROID PLEXUS. Specialized cells located in the ventricles of the brain that produce cerebrospinal fluid.

CHOROIDAL HEMANGIOMA. A nonmalignant blood vessel tumor in the eye.

CHROMOSOME. A microscopic thread-like structure found within each cell of the human body and consisting of a complex of proteins and DNA. Humans have 46 chromosomes arranged into 23 pairs. Chromosomes contain the genetic information necessary to direct the development and functioning of all cells and systems in the body. They pass on hereditary traits from parents to child (like eye color) and determine whether the child will be male or female.

CHRONIC. Refers to a disease or condition that progresses slowly but persists or recurs over time.

CHRONIC BRONCHITIS. A smoking-related respiratory illness in which the membranes that line the bronchi, or the lung's air passages, narrow over time. Symptoms include a morning cough that brings up phlegm, breathlessness, and wheezing.

CHRONIC OTITIS MEDIA. Inflammation of the middle ear with signs of infection lasting three months or longer.

CHRONIC PAIN. Pain that lasts over a prolonged period and threatens to disrupt daily life.

CILIA. Tiny hairlike projections on certain cells within the body. Cilia produce lashing or whipping movements to direct or cause motion of substances or fluids within the body. Within the respiratory tract, the cilia act to move mucus along, in an effort to continually flush out and clean the respiratory tract.

CIRCADIAN RHYTHM. Any body rhythm that recurs in 24-hour cycles. The sleep-wake cycle is an example of a circadian rhythm.

CIRCUMCISION. A surgical procedure, usually with religious or cultural significance, where the prepuce or skin covering the tip of the penis on a boy, or the clitoris on a girl, is cut away.

CIRCUMVALLATE PLACENTA. The existence of a thick, round, white, opaque ring around the periphery of the placenta that limits the expansion of the fetal vessels.

CIRRHOSIS. A chronic degenerative disease of the liver, in which normal cells are replaced by fibrous tissue and normal liver function is disrupted. The most common symptoms are mild jaundice, fluid collection in the tissues, mental confusion, and vomiting of blood.

CLAUDICATION. Cramping or pain in a leg caused by poor blood circulation. This condition is frequently caused by hardening of the arteries (atherosclerosis). Intermittent claudication occurs only at certain times, usually after exercise, and is relieved by rest.

CLAUSTROPHOBIA. Fear of small, enclosed spaces.

CLEFT. An elongated opening or slit in an organ.

CLEFT PALATE. A congenital malformation in which there is an abnormal opening in the roof of the mouth that allows the nasal passages and the mouth to be improperly connected.

CLINICAL NURSE SPECIALIST. A nurse with advanced training as well as a master's degree.

CLIQUE. A close group of friends having similar interests and goals and whom outsiders regard as excluding them.

CLITORIS. The most sensitive area of the external female genitals. Stimulation of the clitoris causes most women to reach orgasm.

CLONIC. Referring to clonus, a series of muscle contractions and partial relaxations that alternate in some nervous diseases in the form of convulsive spasms.

CLOSED-FIST INJURY. A hand wound caused when the skin of the fist is torn open by contact with teeth.

CLOT. A soft, semi-solid mass that forms when blood coagulates.

CLOT BUSTERS. Also called thrombolytics. Medications used to break up a blood clot.

CLOTTING FACTORS. Substances in the blood, also known as coagulation factors, that act in sequence to stop bleeding by triggering the formation of a clot. Each clotting factor is designated with a Roman numeral I through XIII.

COAGULATE. To clot or cause hemostasis; in electrosurgery, to cause tissue dehydration without cutting.

COAGULOPATHY. A disorder in which blood is either too slow or too quick to coagulate (clot).

COARCTATION OF THE AORTA. A congenital defect in which severe narrowing or constriction of the aorta obstructs the flow of blood.

COBB ANGLE. A measure of the curvature of scoliosis, determined by measurements made on x rays.

COCHLEA. The hearing part of the inner ear. This snail-shaped structure contains fluid and thousands of microscopic hair cells tuned to various frequencies, in addition to the organ of Corti (the receptor for hearing).

COCHLEAR IMPLANTATION. A surgical procedure in which a small electronic device is placed under the skin behind the ear and is attached to a wire that stimulates the inner ear, allowing people who have hearing loss to hear useful sounds.

COERCIVE BEHAVIOR. Maladaptive behaviors engaged in as a means of avoiding or escaping aversive events. Coercive behavior may include whining, noncompliance, and lying.

COGNITION. The act or process of knowing or perceiving.

COGNITIVE. The ability (or lack of) to think, learn, and memorize.

COGNITIVE ABILITY. Relating to the process of acquiring knowledge by using reasoning, intuition, or perception.

COGNITIVE PROCESSES. Thought processes (i.e., reasoning, perception, judgment, memory).

COGNITIVE SKILLS. Skills required to perform higher cognitive processes, such as knowing, learning, thinking, and judging.

COGNITIVE THERAPY. Psychological treatment aimed at changing a person's way of thinking in order to change his or her behavior and emotional state.

COGNITIVE-BEHAVIORAL THERAPY. A type of psychotherapy in which people learn to recognize and change negative and self-defeating patterns of thinking and behavior.

COHABITATION. Sexual partners living together outside of marriage.

CO-INFECTION. Concurrent infection of a cell or organism with two microorganisms (pneumonia caused by coinfection with a cytomegalovirus and streptococcus).

COLCHICINE. A drug used to treat painful flare-ups of gout. It is also effective in reducing the frequency and severity of attacks in familial Mediterranean fever.

COLITIS. Inflammation of the colon (large intestine).

COLLAGEN. The main supportive protein of cartilage, connective tissue, tendon, skin, and bone.

COLOBOMA. A birth defect in which part of the eye does not form completely.

COLON. The part of the large intestine that extends from the cecum to the rectum. The sigmoid colon is the area of the intestine just above the rectum; linking the descending colon with the rectum. It is shaped like the letter S.

COLONIZATION. The presence of bacteria on a body surface (like on the skin, mouth, intestines or airway) without causing disease in the person.

COLONOSCOPY. An examination of the lining of the colon performed with a colonoscope.

COLOSTOMY. A surgical procedure in which an opening is made in the wall of the abdomen to allow a part of the large intestine (the colon) to empty outside the body. Colostomies are usually required because portions of the intestine have been removed or an intestinal obstruction exists.

COLOSTRUM. Milk secreted for a few days after birth and characterized by high protein and antibody content.

COMA. A condition of deep unconsciousness from which the person cannot be aroused

COMEDO. A hard plug composed of sebum and dead skin cells, also called a blackhead. The mildest type of acne.

COMEDOLYTIC DRUGS. Medications that break up comedones and open clogged pores.

COMING OUT. The process by which gays and bisexuals become public or tell others about their sexual orientation.

COMMINUTED FRACTURE. A fracture where there are several breaks in a bone creating numerous fragments.

COMMON COLD. A mild illness caused by upper respiratory viruses. Usual symptoms include nasal congestion, coughing, sneezing, throat irritation, and a low-grade fever.

COMMUNICATION. The act of transmitting and receiving information.

COMORBIDITY. A disease or condition that coexists with the disease or condition for which the patient is being primarily treated.

COMPARTMENT SYNDROME. A condition in which the blood supply to a muscle is cut off because the muscle swells but is constricted by the connective tissue around it.

COMPLEMENT. One of several proteins in the blood that acts with other proteins to assist in killing bacteria.

COMPLETE BLOOD COUNT (CBC). A routine analysis performed on a sample of blood taken from the patient's vein with a needle and vacuum tube. The measurements taken in a CBC include a white blood cell count, a red blood cell count, the red cell distribution width, the hematocrit (ratio of the volume of the red blood cells to the blood volume), and the amount of hemoglobin (the blood protein that carries oxygen).

COMPLETE BREECH. A breech position in which the baby is "sitting" bottom first on the cervix with legs crossed.

COMPLETE CLEFT. A cleft that extends through the entire affected mouth structure.

COMPULSION. A repetitive or ritualistic behavior that a person performs to reduce anxiety. Compulsions often develop as a way of controlling or "undoing" obsessive thoughts.

COMPUTED TOMOGRAPHY (CT). An imaging technique in which cross-sectional x rays of the body are compiled to create a three-dimensional image of the body's internal structures; also called computed axial tomography.

COMVAX. Hib-HepB, a combination vaccine that protects against the *Haemophilus influenzae* type B bacterium and the hepatitis B virus.

CONCEPTION. The union of egg and sperm to eventually form a fetus.

CONCUSSION. An injury to the brain, often resulting from a blow to the head, that can cause temporary disorientation, memory loss, or unconsciousness.

CONDUCT DISORDER. A behavioral and emotional disorder of childhood and adolescence. Children with a conduct disorder act inappropriately, infringe on the rights of others, and violate societal norms.

CONDUCTING MATERIALS. Materials that conduct electricity, materials through which electric current travels easily. Examples are metals and water.

CONDUCTIVE HEARING IMPAIRMENT. Hearing impairment associated with the outer or middle ear, often caused by infection.

CONDYLOMA ACUMINATA. Another term for a genital wart.

CONES. Receptor cells, located in the retina of the eye, that allow the perception of colors.

CONFUSIONAL AROUSAL. A partial arousal state occurring during the fourth stage of deepest sleep. Childhood night terrors are a form of confusional arousal.

CONGENITAL. Present at birth.

CONGENITAL CYSTIC ADENOMATOID MALFORMATION (CCAM). A condition in which one or more lobes of the fetal lungs develop into fluid-filled sacs called cysts.

CONGENITAL DIAPHRAGMATIC HERNIA (CDH). A condition in which the fetal diaphragm (the muscle dividing the chest and abdominal cavity) does not close completely.

CONGENITAL MALFORMATION. A deformity present at birth.

CONGENITAL RUBELLA SYNDROME (CRS). Viral illness caused by a togavirus of the genus *Rubivirus*. When rubella infection occurs during pregnancy, fetal infection is likely and often causes congenital rubella syndrome (CRS), resulting in miscarriages, stillbirths, and severe birth defects.

CONJUNCTIVA. Plural, conjunctivae. The mucous membrane that covers the white part of the eyes (sclera) and lines the eyelids.

CONJUNCTIVITIS. Inflammation of the conjunctiva, the mucous membrane covering the white part of the eye (sclera) and lining the inside of the eyelids also called pinkeye.

CONNECTIVE TISSUE. A group of tissues responsible for support throughout the body; includes cartilage, bone, fat, tissue underlying skin, and tissues that support organs, blood vessels, and nerves throughout the body.

CONSOLIDATION. A condition in which lung tissue becomes firm and solid rather than elastic and air-filled, arising because of accumulated fluids and tissue debris.

CONSTIPATION. Difficult bowel movements caused by the infrequent production of hard stools.

CONSTRICTED. Made smaller or narrower.

CONTACT DERMATITIS. Skin inflammation as a result of contact with a foreign substance.

CONTAGIOUS. The movement of disease between people. All contagious disease is infectious, but not all infections are contagious.

CONTINGENCIES. Naturally occurring or artificially designated reinforcers or punishers that follow a behavior.

CONTRACEPTIVE. A device or medication designed to prevent pregnancy by either suppressing ovulation, preventing sperm from passing through the cervix to fertilize an egg, or preventing implantation of a fertilized egg.

CONTRACTION. A tightening of the uterus during pregnancy. Contractions may or may not be painful and may or may not indicate labor.

CONTRACTURE. A tightening or shortening of muscles that prevents normal movement of the associated limb or other body part.

CONTRAST AGENT. Also called a contrast medium, this is usually a barium or iodine dye that is injected into the area under investigation. The dye makes the interior body parts more visible on an x-ray film.

CONTRAST HYDROTHERAPY. A series of hot and cold water applications. A hot compress (as hot as an individual can tolerate) is applied for three minutes followed by an ice-cold compress for 30 seconds. These applications are repeated three times each and ending with the cold compress.

CONVERGENCE. The natural movement of the eyes inward to view objects close-up.

CONVERGENT THINKING. The ability to come up with a single correct answer.

CONVULSIONS. Also termed seizures; a sudden violent contraction of a group of muscles.

COOPERATIVE PLAY. A type of play in which school-age children participate in activities with an organized structure or compete for goal or outcome.

COORDINATION. The ability to perform activities with precision and proficiency.

COPING. In psychology, a term that refers to a person's patterns of response to stress.

COPROLALIA. The involuntary use of obscene language.

COPROPRAXIA. The involuntary display of unacceptable/obscene gestures.

CORD BLOOD. The blood that remains in the umbilical cord and placenta after birth. Stem cells from cord blood can be used in place of bone marrow for treating primary immunodeficiency disorders.

CORE GENDER IDENTITY. The deep inner feeling a child has about whether he or she is a male or female.

CORNEA. The clear, dome-shaped outer covering of the eye that lies in front of the iris and pupil. The cornea lets light into the eye.

CORNEAL ABRASION. A scratch on the surface of the cornea.

CORONAL SUTURE. Skull suture that lies behind the forehead area, across the head from left side to the right side.

CORONAVIRUS. A genus of viruses that cause respiratory diseases and gastroenteritis.

CORTICOSPINAL TRACT. A tract of nerve cells that carries motor commands from the brain to the spinal cord.

CORTICOSTEROIDS. A group of hormones produced naturally by the adrenal gland or manufactured synthetically. They are often used to treat inflammation. Examples include cortisone and prednisone.

CORTISOL. A steroid hormone secreted by the adrenal cortex that is important for maintenance of body fluids, electrolytes, and blood sugar levels. Also called hydrocortisone.

CORTISONE. Glucocorticoid produced by the adrenal cortex in response to stress. Cortisone is a steroid with anti-inflammatory and immunosuppressive properties.

CO-SLEEPING. Having an infant sleep with the mother in her bed.

COUGH SUPPRESSANT. A medication that stops or prevents coughing.

COXSACKIE VIRUS. A type of enterovirus that may produce a variety of illnesses, including upper respiratory infections, myocarditis, and pericarditis. Coxsackie viruses resemble the virus that causes polio.

CRABS. An informal or slang term for pubic lice.

CRANIAL NERVES. The set of 12 nerves found on each side of the head and neck that control the sensory and muscle functions of the eyes, nose, tongue, face, and throat.

CRANIOPHARYNGIOMA. A tumor near the pituitary gland in the craniopharyngeal canal that often results in intracranial pressure.

CRANIOSYNOSTOSIS. A premature closure of one or more of the joints (fissures) between the bones of the skull, which causes an abnormally shaped skull.

CREEPING. A form of locomotion in infants, in which the baby pulls the body forward with the arms while the belly and legs drag behind.

CREEPING ERUPTION. Itchy, irregular, wandering red lines on the foot made by burrowing larvae of the hookworm family and some roundworms.

CREPITUS. A crackling sound.

CRETINISM. Severe hypothyroidism that is present at birth and characterized by severe mental retardation.

CRITERION-REFERENCED TEST. An assessment that measures the achievement of specific information or skills against a standard as opposed to being measured against how others perform.

CROHN'S DISEASE. A chronic, inflammatory disease, primarily involving the small and large intestine, but which can affect other parts of the digestive system as well.

CROSSBITE. The condition in which the upper teeth bite inside the lower teeth.

CROSS-REACTION. A reaction that occurs in blood testing when a disease agent reacts to the specific antibody for another disease agent.

CROUZON SYNDROME. A disorder characterized by malformations of the skull and face.

CROWN. The natural part of the tooth covered by enamel. A restorative crown is a protective shell that fits over a tooth.

CRYOTHERAPY. The use of a very low-temperature probe to freeze and thereby destroy tissue. Cryotherapy is used in the treatment skin lesions, Parkinson's disease, some cancers, retinal detachment, and cataracts. Also called cryosurgery.

CRYPTORCHIDISM. Undescended testes, a condition in which a boy is born with one or both testicles in the lower abdomen rather than the scrotum.

CULTURE. A test in which a sample of body fluid is placed on materials specially formulated to grow microorganisms. A culture is used to learn what type of bacterium is causing infection.

CURETTE. Also spelled curet; a small loop or scoop-shaped surgical instrument with sharpened edges that can be used to remove tissue, growths, or debris.

CUSTODIAL PARENT. A parent who has legal custody of their child or children.

CUSTODY. The care, control, and maintenance of a child, which in abuse and neglect cases can be awarded by the court to an agency or in divorce to parents. Foster parents do not have legal custody of the children who are in their care.

CUT. A slicing wound made with a sharp instrument, leaving even edges.

CUTANEOUS. Pertaining to the skin

CUTANEOUS ANGIOLIPOMAS. Benign growths consisting of fat cells and blood vessels just underneath the skin.

CYANOSIS. A bluish tinge to the skin that can occur when the blood oxygen level drops too low.

CYANOTIC. Marked by a bluish tinge to the skin that occurs when the blood oxygen level drops too low. It is one of the types of congenital heart disease.

CYCLIC VOMITING. Uncontrolled vomiting that occurs repeatedly over a certain period of time.

CYCLOOXYGENASE 2 (COX 2). The cyclooxygenase that helps mediate inflammation and that helps the brain feel pain and regulate fever.

CYST. An abnormal sac or enclosed cavity in the body filled with liquid or partially solid material. Also refers to a protective, walled-off capsule in which an organism lies dormant.

CYSTIC FIBROSIS TRANSMEMBRANE CONDUCTANCE REGULATOR (CFTR). The protein responsible for regulating chloride movement across cells in some tissues. Cystic fibrosis results when a person has two defective copies of the CFTR gene.

CYSTOSCOPY. A diagnostic procedure in which a hollow lighted tube (cystoscope) is used to look inside the bladder and the urethra.

CYTOKINES. Chemicals made by the cells that act on other cells to stimulate or inhibit their function. They are important controllers of immune functions.

CYTOMEGALOVIRUS (CMV). A common human virus causing mild or no symptoms in healthy people, but permanent damage or death to an infected fetus, a transplant patient, or a person with HIV.

CYTOTOXIC. The characteristic of being destructive to cells.

D

DACRON. A synthetic polyester fiber used to surgically repair damaged sections of heart muscle and blood vessel walls.

DACTYLITIS. Inflammation of the hands or feet.

DANDER. Loose scales shed from the fur or feathers of household pets and other animals. Dander can cause allergic reactions in susceptible people.

DARWINIAN REFLEX. An unconscious action in infants in which if a palm is touched, the infant makes a very tight fist. This instinct disappears within two to three months.

DEADBEAT PARENT. A mother or father who has abandoned his or her child or children and does not pay child custody as required by a court.

DEBRIDEMENT. The surgical removal of dead tissue and/or foreign bodies from a wound or cut.

DECELERATION. A decrease in the fetal heart rate that can indicate inadequate blood flow through the placenta.

DECIBEL. A unit of the intensity of sound or a measure of loudness. Normal speech is typically spoken in the range of about 20-50 decibels.

DECOMPRESSION. A decrease in pressure from the surrounding water that occurs with decreasing diving depth.

DECONGESTANTS. A group of medications, such as pseudoephedrine, phenylephrine, and phenylpropanolamine, that shrink blood vessels and consequently mucus membranes.

DECREASED PENETRANCE. Individuals who inherit a changed disease gene but do not develop symptoms.

DECUBITUS ULCER. A pressure sore resulting from ulceration of the skin occurring in persons confined to bed for long periods of time

DEEP BITE. A closed bite; a deep or excessive overbite in which the lower incisors bite too closely to or into the gum tissue or palate behind the upper teeth.

DEEP BREATHING. Deep breathing helps expand the lungs and forces better distribution of the air into all sections of the lung. The patient either sits in a chair or sits upright in bed and inhales, pushing the abdomen out to force maximum amounts of air into the lung. The abdomen is then contracted, and the patient exhales.

DEFECATION. The act of having a bowel movement, or the passage of feces through the anus.

DEFENSE MECHANISMS. Indirect strategies used to reduce anxiety rather than directly facing the issues causing the anxiety.

DEFIBRILLATION. A procedure to stop the type of irregular heart beat called ventricular fibrillation, usually by using electric shock.

DEFICIT. A shortfall or slowdown in development, possibly related to a disorder that slows or interrupts normal childhood development.

DEFORMATIONAL PLAGIOCEPHALY (POSITIONAL MOLDING). A form of craniosynostosis in which the head is misshapen, the result of constant pressure to the same area of the head.

DEHYDRATION. An excessive loss of water from the body. It may follow vomiting, prolonged diarrhea, or excessive sweating.

DELETION. The absence of genetic material that is normally found in a chromosome. Often, the genetic material is missing due to an error in replication of an egg or sperm cell.

DELINQUENT. A term applied to young people who behave in a manner in defiance of established social and ethical codes.

DELIRIUM. Sudden confusion with a decreased or fluctuating level of consciousness.

DELIRIUM TREMENS. A complication that may accompany alcohol withdrawal. The symptoms include body shaking (tremulousness), insomnia, agitation, confusion, hearing voices or seeing images that are not really there (hallucinations), seizures, rapid heart beat, profuse sweating, high blood pressure, and fever.

DELUSION. A belief that is resistant to reason or contrary to actual fact. Common delusions include delusions of persecution, delusions about one's importance (sometimes called delusions of grandeur), or delusions of being controlled by others.

DEMYELINATION. Disruption or destruction of the myelin sheath, leaving a bare nerve. It results in a slowing or stopping of the impulses that travel along that nerve.

DENDRITE. A threadlike extension of the cytoplasm of a neuron that conducts electrical impulses toward the cell body of the neuron. Usually it spreads out into many branches.

DENDRITIC. Branched like a tree.

DENTAL CARIES. A disease of the teeth in which microorganisms convert sugar in the mouth to an acid that erodes the tooth. Commonly called a cavity.

DENTIN. The middle layer of a tooth, which makes up most of the tooth's mass.

DEOXYGENATED BLOOD. Blood that does not contain oxygen.

DEPENDENCE. A state in which a person requires a steady concentration of a particular substance to avoid experiencing withdrawal symptoms.

DEPIGMENTED. Characterized by a loss of normal color; discolored.

DEPRESSION. A mental condition in which a person feels extremely sad and loses interest in life. A person with depression may also have sleep problems and loss of appetite and may have trouble concentrating and carrying out everyday activities.

DEPRIVATIONAL DWARFISM. A condition where emotional disturbances are associated with growth failure and abnormalities of pituitary function.

DERMATITIS. Inflammation of the skin.

DERMATITIS HERPETIFORMIS. A chronic, very itchy skin disease with groups of red lesions that leave spots behind when they heal.

DERMATOLOGIST. A physician that specializes in diseases and disorders of the skin.

DERMATOLOGY. The branch of medicine that studies and treats disorders of the skin.

DERMATOPHYTE. A type of fungus that causes diseases of the skin, including tinea or ringworm.

DERMATOSPARAXIS. Skin fragility caused by abnormal collagen.

DERMIS. The basal layer of skin; it contains blood and lymphatic vessels, nerves, glands, and hair follicles.

DESENSITIZATION. A treatment for phobias which involves exposing the phobic person to the feared situation. It is often used in conjunction with relaxation techniques. Also used to describe a technique of pain reduction in which the painful area is stimulated with whatever is causing the pain.

DESFEROXAMINE. The primary drug used in iron chelation therapy. It aids in counteracting the life-threatening buildup of iron in the body associated with long-term blood transfusions.

DESMOPRESSIN ACETATE (DDAVP). A drug used to regulate urine production.

DETOXIFICATION. The process of physically eliminating drugs and/or alcohol from the system of a substance-dependent individual.

DEVELOPMENT, EMBRYONIC. The process whereby undifferentiated embryotic cells replicate and differentiate into limbs, organ systems, and other body components of the fetus.

DEVELOPMENTAL. Referring to the growth process, particularly the growth patterns and associated skills acquired in childhood.

DEVELOPMENTAL ASSESSMENT. The ongoing process of testing, observing, and analyzing a child's skills.

DEVELOPMENTAL COORDINATION DISORDER. A disorder of motor skills.

DEVELOPMENTAL DELAY. The failure of a child to meet certain developmental milestones, such as sitting, walking, and talking, at the average age. Developmental delay may indicate a problem in development of the central nervous system.

DEVELOPMENTAL DOMAINS. Areas of a child's development.

DEVELOPMENTAL MILESTONE. The age at which an infant or toddler normally develops a particular skill. For example, by nine months, a child should be able to grasp and toss a bottle.

DEVIATED SEPTUM. A shift in the position of the nasal septum, the partition that divides the two nasal cavities.

DEXTROSE. A sugar solution used in intravenous drips.

DIABETES MELLITUS. The clinical name for common diabetes. It is a chronic disease characterized by the inability of the body to produce or respond properly to insulin, a hormone required by the body to convert glucose to energy.

DIABETIC COMA. A life-threatening, reduced level of consciousness that occurs in persons with uncontrolled diabetes mellitus.

DIABETIC RETINOPATHY. A condition seen most frequently in individuals with poorly controlled diabetes mellitus where the tiny blood vessels to the retina, the tissues that sense light at the back of the eye, are damaged. This damage causes blurred vision, sudden blindness, or black spots, lines, or flashing light in the field of vision.

DIAGNOSIS. The art or act of identifying a disease from its signs and symptoms.

DIAGNOSTIC AND STATISTICAL MANUAL OF MENTAL DISORDERS, FOURTH EDITION (DSM-IV). This reference book, published by the American Psychiatric Association, is the diagnostic standard for most mental health professionals in the United States.

DIAGNOSTIC TESTING. Testing performed to determine if a person has a particular disease.

DIALYSIS. A process of filtering and removing waste products from the bloodstream, it is used as a treatment for patients whose kidneys do not function properly. Two main types are hemodialysis and peritoneal dialysis.

DIAPER DERMATITIS (DIAPER RASH). An inflammatory reaction to irritants in the diaper area.

DIAPHRAGM. The thin layer of muscle that separates the chest cavity containing the lungs and heart from the abdominal cavity containing the intestines and digestive organs. This term is also used for a dome-shaped device used to cover the back of a woman's vagina during intercourse in order to prevent pregnancy.

DIARRHEA. A loose, watery stool.

DIASTOLIC BLOOD PRESSURE. Diastole is the period in which the left ventricle relaxes so it can refill with blood; diastolic pressure is therefore measured during diastole.

DIDASKALEINOPHOBIA. Fear of going to school.

DIETARY FIBER. Mostly indigestible material in food that stimulates the intestine to peristalsis.

DIFFERENTIATION. The ability to retain one's identity within a family system while maintaining emotional connections with the other members.

DIGESTION. The mechanical, chemical, and enzymatic process in which food is converted into the substances suitable for use by the body.

DILATE. To expand in diameter and size.

DIMERCAPROL. A chemical agent used to remove excess lead from the body.

DIOPTER (D). A unit of measure for describing the refractive power of a lens.

DIPHTHERIA. A serious, frequently fatal, bacterial infection that affects the respiratory tract. Vaccinations given in childhood have made diphtheria very rare in the United States.

DIPHTHERIA-TETANUS-PERTUSSIS (DTP) VACCINE. The standard vaccine used to immunize children against diphtheria, tetanus, and whooping cough. A so-called "acellular pertussis" vaccine (aP) is usually used since its release in the mid-1990s.

DIPLEGIA. Paralysis affecting like parts on both sides the body, such as both arms or both legs.

DISABILITY. An inability to do something others can do; sometimes referred to as handicap or impairment.

DISCHARGE PLANNER. A health care professional who helps parents arrange for health and home care needs after their child goes home from the hospital.

DISCIPLINE. In health care, a specific area of preparation or training, i.e., social work, nursing, or nutrition.

DISCLOSURE. Release of information.

DISEASE-MODIFYING ANTI-RHEUMATIC DRUGS (DMARDS). A group of medications given to treat severe cases of arthritis, JDMS, and other diseases that affect the joints. All DMARDs work by modifying the immune system.

DISFLUENCY. An interruption in speech flow.

DISLOCATION. The displacement of bones at a joint or the displacement of any part of the body from its normal position.

DISSEMINATED. Spread to other tissues.

DISSOCIATIVE DISORDERS. A group of mental disorders in which dissociation is a prominent symptom. Patients with dissociative disorders have a high rate of self-mutilation.

DISTAL MUSCULAR DYSTROPHY (DD). A form of muscular dystrophy that usually begins in middle age or later, causing weakness in the muscles of the feet and hands.

DIURETICS. A group of drugs that helps remove excess water from the body by increasing the amount lost by urination.

DIVERGENT THINKING. The ability to come up with new and unusual answers.

DIVERTICULITIS. Inflammation of the diverticula (small outpouchings) along the wall of the colon, the large intestine.

DIVERTICULUM. Plural, diverticula; an outpouching in a tubular organ caused when the inner, lining layer bulges out (herniates) through the outer, muscular layer. Diverticula are present most often in the colon (large intestine), but are also found in the stomach and the small intestine.

DIZYGOTIC. From two zygotes, as in non-identical, or fraternal twins. The zygote is the first cell formed by the union of sperm and egg.

DNA. Deoxyribonucleic acid; the genetic material in cells that holds the inherited instructions for growth, development, and cellular functioning.

DNA TESTING. Analysis of DNA (the genetic component of cells) in order to determine changes in genes that may indicate a specific disorder.

DOPA. The common name for a natural chemical (3,4-dihydroxyphenylalanine) made by the body during the process of making melanin.

DOPAMINE. A neurotransmitter made in the brain that is involved in many brain activities, including movement and emotion.

DORMANT. The biological state of being relatively inactive or in a resting state in which certain processes are slowed down or suspended.

DORSAL RHIZOTOMY. A surgical procedure that cuts nerve roots to reduce spasticity in affected muscles.

DORSUM. The medical term for the bridge of the nose.

DOULA. A doula is someone who undergoes special training to enable them to support women during childbirth and into the postpartum period.

DOWN SYNDROME. A chromosomal disorder caused by an extra copy or a rearrangement of chromosome 21. Children with Down syndrome have varying degrees of mental retardation and may have heart defects.

DUBOWITZ EXAM. Standardized test that scores responses to 33 specific neurological stimuli to estimate an infant's neural development and, hence, gestational age.

DUCHENNE MUSCULAR DYSTROPHY (DMD). The most severe form of muscular dystrophy, DMD usually affects young boys and causes progressive muscle weakness, usually beginning in the legs.

DUCTUS ARTERIOSUS. The blood vessel that joins the pulmonary artery and the aorta. When the ductus does not close at birth, it causes a type of congenital heart disease called patent ductus arteriosus.

DUODENUM. The first of the three segments of the small intestine. The duodenum is about 10 in (25 cm) long and connects the stomach and the jejunum.

DURA MATER. The strongest and outermost of three membranes that protect the brain, spinal cord, and nerves of the cauda equina.

DUST MITES. Tiny insects, unable to be seen without a microscope, that are present in carpet, stuffed animals, upholstered furniture, and bedding, including pillows, mattresses, quilts, and other bed covers. Dust mites are one of the most common asthma triggers. They grow best in areas with high humidity.

DWARFISM, PITUITARY. Short stature. When caused by hGH deficiency, as opposed to late growth spurt or genetics, abnormally slow growth and short stature with normal proportions may be seen.

DYANA. The yoga term for meditation.

DYSENTERY. A disease marked by frequent watery bowel movements, often with blood and mucus, and characterized by pain, urgency to have a bowel movement, fever, and dehydration.

DYSKINESIA. Impaired ability to make voluntary movements.

DYSLEXIA. A type of reading disorder often characterized by reversal of letters or words.

DYSMENORRHEA. Painful menstruation.

DYSMOTILITY. Abnormally slow or fast rhythmic movement of the stomach or intestine.

DYSPHAGIA. Difficulty in swallowing.

DYSPHORIA. Feelings of anxiety, restlessness, and dissatisfaction.

DYSPLASIA. Abnormal changes in cells.

DYSPNEA. Difficulty in breathing, usually associated with heart or lung diseases.

DYSSOMNIA. A primary sleep disorder in which the patient suffers from changes in the quantity, quality, or timing of sleep.

DYSTOCIA. Failure to progress in labor, either because the cervix will not dilate (expand) further or because the head does not descend through the mother's pelvis after full dilation of the cervix.

DYSTONIA. Painful involuntary muscle cramps or spasms.

DYSTROPHIN. A protein that helps muscle tissue repair itself. Both Duchenne muscular dystrophy and Becker muscular dystrophy are caused by flaws in the gene that tells the body how to make this protein.

DYSURIA. Painful or difficult urination.

E

EAR CANDLING. An alternative method for removing impacted cerumen with a lighted hollow cone of paraffin or beeswax. It does not work and is not considered an acceptable treatment for any ear problem or disorder.

EAR SPECULUM. A cone- or funnel-shaped attachment for an otoscope that is inserted into the ear canal to examine the eardrum.

EARDRUM. A paper-thin covering stretching across the ear canal that separates the middle and outer ears.

ECCENTRIC. Deviating from the center; conduct and behavior departing from accepted norms and conventions.

ECCHYMOSIS. The medical term for a bruise, or skin discoloration caused by blood seeping from broken capillaries under the skin.

ECHOCARDIOGRAM. A record of the internal structures of the heart obtained from beams of ultrasonic waves directed through the wall of the chest.

ECHOCARDIOGRAPHY. A non-invasive technique, using ultrasound waves, used to look at the various structures and functions of the heart.

ECHOLALIA. Involuntary echoing of the last word, phrase, or sentence spoken by someone else.

ECHOPRAXIA. The imitation of the movement of another individual.

ECLAMPSIA. Coma and convulsions during or immediately after pregnancy, characterized by edema, hypertension, and proteinuria.

ECTOPIA LENTIS. Dislocation of the lens of the eye. It is one of the most important single indicators in diagnosing Marfan syndrome.

ECTOPIC. Out of place or located away from the normal position.

ECTOPIC PREGNANCY. A pregnancy that develops outside of the mother's uterus, such as in the fallopian tube. Ectopic pregnancies often cause severe pain in the lower abdomen and are potentially life-threatening because of the massive blood loss that may occur as the developing embryo/fetus ruptures and damages the tissues in which it has implanted.

ECZEMA. A superficial type of inflammation of the skin that may be very itchy and weeping in the early stages; later, the affected skin becomes crusted, scaly, and thick.

EDEMA. The presence of abnormally large amounts of fluid in the intercellular tissue spaces of the body.

EDETATE CALCIUM DISODIUM. A chemical chelating agent used to remove excess lead from the body.

EDTA. A colorless compound used to keep blood samples from clotting before tests are run.

EFFERENT NERVES. Peripheral nerves that carry signals away from the brain and spinal cord.

EFFICACY. The effectiveness of a drug in treating a disease or condition.

EFFUSION. The escape of fluid from blood vessels or the lymphatic system and its collection in a cavity.

EGOCENTRIC. Limited in outlook to things mainly relating to oneself or confined to one's own affairs or activities.

EISENMENGER'S SYNDROME. A condition in which high pressures in the pulmonary arteries cause them to thicken. To compensate, the right side of the heart works harder, causing it to stretch and weaken. Eisenmenger's syndrome is a serious condition that leads to heart failure and can result in death by age 40 if left untreated.

EJACULATION. The process by which semen (made up in part of prostatic fluid) is ejected by the erect penis.

ELBOW. Hinged joint between the forearm and upper arm.

ELECTRIC CURRENT. The rate of flow of electric charge, measured in amperes. Electric current can also be described as the flow of microscopic particles called electrons flowing through wires and electronic components and appliances.

ELECTRICAL RESISTANCE. Resistance to the flow of electrical current.

ELECTROCARDIAGRAM (ECG, EKG). A record of the electrical activity of the heart, with each wave being labeled as P, Q, R, S, and T waves. It is often used in the diagnosis of cases of abnormal cardiac rhythm and myocardial damage.

ELECTROCONVULSIVE THERAPY (ECT). A psychological treatment in which a series of controlled electrical impulses are delivered to the brain in order to induce a seizure within the brain. This type of therapy is used to treat major depression and severe mental illness that does not respond to medications.

ELECTRODE. A medium for conducting an electrical current.

ELECTROENCEPHALOGRAM (EEG). A record of the tiny electrical impulses produced by the brain's activity picked up by electrodes placed on the scalp. By measur-

ing characteristic wave patterns, the EEG can help diagnose certain conditions of the brain.

ELECTROENCEPHALOGRAPHY. The recording of electrical impulses produced by the brain's activity via electrodes attached to a patient's scalp.

ELECTROLYTES. Salts and minerals that produce electrically charged particles (ions) in body fluids. Common human electrolytes are sodium chloride, potassium, calcium, and sodium bicarbonate. Electrolytes control the fluid balance of the body and are important in muscle contraction, energy generation, and almost all major biochemical reactions in the body.

ELECTROMAGNETIC RADIATION. Packets of energy that develop when an electric current passes through a vacuum tube,

ELECTROMYOGRAPHY (EMG). A diagnostic test that records the electrical activity of muscles. In the test, small electrodes are placed on or in the skin; the patterns of electrical activity are projected on a screen or over a loudspeaker. This procedure is used to test for muscle disorders, including muscular dystrophy.

ELECTRONYSTAGMOGRAPHY. A method for measuring the electricity generated by eye movements. Electrodes are placed on the skin around the eye and the individual is subjected to a variety of stimuli so that the quality of eye movements can be assessed.

ELECTROOCULOGRAPHY (EOG). A diagnostic test that records the electrical activity of the muscles that control eye movement.

EMBOLISM. A blood clot, air bubble, or mass of foreign material that travels and blocks the flow of blood in an artery. When blood supply to a tissue or organ is blocked by an embolism, infarction, or death of the tissue the artery feeds, occurs. Without immediate and appropriate treatment, an embolism can be fatal.

EMBOLUS. Plural, emboli. An embolus is something that blocks the blood flow in a blood vessel. It may be a gas bubble, a blood clot, a fat globule, a mass of bacteria, or other foreign body that forms somewhere else and travels through the circulatory system until it gets stuck.

EMBRYO. In humans, the developing individual from the time of implantation to about the end of the second month after conception. From the third month to the point of delivery, the individual is called a fetus.

EMESIS. An act or episode of vomiting.

EMETIC. A medication intended to cause vomiting. Emetics are sometimes used in aversion therapy in place of electric shock. Their most common use in mainstream medicine is in treating accidental poisoning.

EMMENAGOGUE. A type of medication that brings on or increases a woman's menstrual flow.

EMMETROPIA. Normal vision.

EMOTIONAL INTELLIGENCE. The ability to perceive and interpret the emotions of others.

EMPATHY. A quality of the client-centered therapist, characterized by the therapist's conveying appreciation and understanding of the client's point of view.

EMPHYSEMA. A chronic respiratory disease that involves the destruction of air sac walls to form abnormally large air sacs that have reduced gas exchange ability and that tend to retain air within the lungs. Symptoms include labored breathing, the inability to forcefully blow air out of the lungs, and an increased susceptibility to respiratory tract infections. Emphysema is usually caused by smoking.

EMPIRICAL TREATMENT. Medical treatment that is given on the basis of the doctor's observations and experience.

ENAMEL. The hard, outermost surface of a tooth.

ENCEPHALITIS. Inflammation of the brain, usually caused by a virus. The inflammation may interfere with normal brain function and may cause seizures, sleepiness, confusion, personality changes, weakness in one or more parts of the body, and even coma.

ENCEPHALOMYELITIS. Encephalitis or another acute inflammation of the brain and spinal cord that can be caused by the rabies virus.

ENCEPHALOPATHY. Any abnormality in the structure or function of brain tissues.

ENCOPRESIS. Fecal incontinence that can occur as a result of stress or fear.

ENDEMIC. Natural to or characteristic of a particular place, population, or climate.

ENDEMIC DISEASE. An infectious disease that occurs frequently in a specific geographical locale. The disease often occurs in cycles.

ENDOCARDITIS. Inflammation of the inner membrane lining heart and/or of the heart valves caused by infection.

ENDOCHONDRAL OSSIFICATION. The process by which cartilage is converted into bone.

ENDOCRINE SYSTEM. A group of ductless glands and parts of glands that secrete hormones directly into the bloodstream or lymphatic system to control metabolic activity. Pituitary, thyroid, adrenals, ovaries, and testes are all part of the endocrine system.

ENDOCRINOLOGIST. A physician who specializes in treating patients who have diseases of the thyroid, parathyroid, adrenal glands, and/or the pancreas.

ENDODONTIST. A dentist who specializes in diagnosing and treating diseases of the pulp and other inner structures of the tooth.

ENDOMETRIOSIS. A condition in which the tissue that normally lines the uterus (endometrium) grows in other areas of the body, causing pain, irregular bleeding, and frequently, infertility.

ENDOMETRITIS. Inflammation of the endometrium or mucous membrane of the uterus.

ENDOMETRIUM. The mucosal layer lining the inner cavity of the uterus. The endometrium's structure changes with age and with the menstrual cycle.

ENDOSCOPE. A medical instrument that can be passed into an area of the body (the bladder or intestine, for example) to allow visual examination of that area. The endoscope usually has a fiber-optic camera that allows a greatly magnified image to be shown on a television screen viewed by the operator. Many endoscopes also allow the operator to retrieve a small sample (biopsy) of the area being examined, to more closely view the tissue under a microscope.

ENDOSCOPY. Visual examination of an organ or body cavity using an endoscope, a thin, tubular instrument containing a camera and light source. Many endoscopes also allow the retrieval of a small sample (biopsy) of the area being examined, in order to more closely view the tissue under a microscope.

ENDOSTEAL RESORPTION. The process by which bones are thinned from the inside.

ENDOTHELIAL CELLS. The cells lining the inner walls of a body cavity or the cardiovascular system. Also known as endothelium.

ENDOTRACHEAL TUBE. A hollow tube that is inserted into the trachea (windpipe) through the nose or mouth. It is used to administer anesthesia, to deliver oxygen under pressure, or to deliver medications (e.g. surfactants).

ENGLISH AS A SECOND LANGUAGE (ESL). English language instruction for English language learners (ELLs) that includes little or no use of a child's native language; a component of all bilingual education programs.

ENGLISH LANGUAGE LEARNER (ELL). A student who is learning English as a second language; also called limited English proficient (LEP).

ENTERAL NUTRITION. Liquid nutrition provided through tubes that enter the gastrointestinal tract, usually through the mouth or nose.

ENTERIC COATING. A coating or shell placed on a tablet that breaks up and releases the medicine into the intestine rather than the stomach.

ENTEROCOLITIS. Severe inflammation of the intestines that affects the intestinal lining, muscle, nerves and blood vessels.

ENTEROPATHY. A disease of the intestinal tract.

ENTEROVIRUS. Any of a group of viruses that primarily affect the gastrointestinal tract. The coxsackievirus and the poliovirus are both enteroviruses.

ENTERTAINMENT SOFTWARE RATING BOARD (ESRB). The industry board that rates video games.

ENTOMOPHOBIA. Fear of insects.

ENUCLEATION. Surgical removal of the eyeball.

ENZYME. A protein that catalyzes a biochemical reaction without changing its own structure or function.

EOSINOPHIL. A type of white blood cell containing granules that can be stained by eosin (a chemical that produces a red stain). Eosinophils increase in response to parasitic infections and allergic reactions.

EOSINOPHILIA. An abnormal increase in the number of eosinophils, a type of white blood cell.

EPIDEMIC. Refers to a situation in which a particular disease rapidly spreads among many people in the same geographical region in a relatively short period of time.

EPIDEMIC PAROTITIS. The medical name for mumps.

EPIDERMIS. The outermost layer of the human skin.

EPIGLOTTIS. A leaf-like piece of cartilage extending upwards from the larynx, which can close like a lid over the trachea to prevent the airway from receiving any food or liquid being swallowed.

EPIGLOTTITIS. Inflammation of the epiglottis, most often caused by a bacterial infection. The epiglottis is a piece of cartilage behind the tongue that closes the opening to the windpipe when a person swallows. An inflamed epiglottis can swell and close off the windpipe, thus causing the patient to suffocate. Also called supraglottitis.

EPILEPSY. A neurological disorder characterized by recurrent seizures with or without a loss of consciousness.

EPILEPTOLOGIST. A physician who specializes in the treatment of epilepsy.

EPINEPHRINE. A hormone produced by the adrenal medulla. It is important in the response to stress and partially regulates heart rate and metabolism. It is also called adrenaline.

EPISIOTOMY. An incision made in the perineum (the area between the vulva and the anus) during labor to assist in delivery and to avoid abnormal tearing of the perineum.

EPISODIC. Occurring once in a while, without a regular pattern.

EPISTAXIS. The medical term used to describe a bleeding from the nose.

EPITHELIUM. The layer of cells that covers body surfaces, lines body cavities, and forms glands.

EQUATOR. Imaginary line encircling the eyeball and dividing the eye into a front and back half.

ERB'S PALSY OR PARALYSIS. A condition caused by an injury to the upper brachial plexus, involving the cervical nerves C5, C6, and sometimes C7, affecting the upper arm and the rotation of the lower arm.

ERGOTAMINE. A drug used to prevent or treat migraine headaches. It can cause vomiting, diarrhea, and convulsions in infants and should not be taken by women who are nursing.

ERRATIC. Having no fixed course; behavior that deviates from common and accepted opinions.

ERUPTION. The process of a tooth breaking through the gum tissue to grow into place in the mouth.

ERYTHEMA. A diffuse red and inflamed area of the skin.

ERYTHEMA MIGRANS. A red skin rash that is one of the first signs of Lyme disease in about 75% of patients.

ERYTHROPOIESIS. The process through which new red blood cells are created; it begins in the bone marrow.

ERYTHROPOIETIC. Referring to the creation of new red blood cells.

ESCHERICHIA COLI. A type of enterobacterium that is responsible for most cases of severe bacterial diarrhea in the United States.

ESOPHAGEAL ATRESIA. Blockage or closure of the esophagus, the tube leading from the mouth to the stomach.

ESOPHAGOGASTRODUODENOSCOPY (EGD). An imaging test that involves visually examining the lining of the esophagus, stomach, and upper duodenum with a flexible fiberoptic endoscope.

ESOPHAGUS. The muscular tube that leads from the back of the throat to the entrance of the stomach. It is coated with mucus and surrounded by muscles, and pushes food to the stomach by sequential waves of contraction. It functions to transport food from the throat to the stomach and to keep the contents of the stomach in the stomach.

ESSENTIAL FATTY ACID (EFA). A fatty acid that the body requires but cannot make. It must be obtained from the diet. EFAs include omega-6 fatty acids found in primrose and safflower oils, and omega-3 fatty acids oils found in fatty fish and flaxseed, canola, soybean, and walnuts.

ESSENTIAL TREMOR. An uncontrollable (involuntary) shaking of the hands, head, and face. Also called familial tremor because it is sometimes inherited, it can begin in the teens or in middle age. The exact cause is not known.

ESTROGEN. Female hormone produced mainly by the ovaries and released by the follicles as they mature. Responsible for female sexual characteristics, estrogen stimulates and triggers a response from at least 300 tissues. After menopause, the production of the hormone gradually stops.

ESTRUS. A regular period of sexual excitement in females.

EUPHORIA. A feeling or state of well-being or elation.

EUROCENTRIC. Centered or focused on Europe or European peoples, especially in relation to historical or cultural influence.

EUSTACHIAN TUBE. A thin tube between the middle ear and the pharnyx. Its purpose is to equalize pressure on either side of the ear drum.

EX UTERO INTRAPARTUM TREATMENT (EXIT). A cesarean section in which the infant is removed from the uterus but the umbilical cord is not cut until after surgery for a congenital defect that blocks the air passage.

EXANTHEM. A skin eruption regarded as a characteristic sign of such diseases as measles, German measles, and scarlet fever.

EXERCISE-INDUCED BRONCHOSPASM. A sudden contraction in the lower airway that causes breathing problems and is brought about by heavy exercise.

EXFOLIATE. To shed skin. In skin care, the term exfoliate describes the process of removing dead skin cells.

EXOTOXIN. A poisonous secretion produced by bacilli that is carried in the bloodstream to other parts of the body.

EXPECTORANT. A drug that promotes the discharge of mucus from respiratory system.

EXPERIMENTAL PLAY THERAPY. Play therapy based on the belief that a child has the ability to solve his or her own problems within the context of a warm and caring therapeutic environment.

EXPRESSIVE APHASIA. A developmental disorder in which a child has lower-than-normal proficiency in vocabulary, production of complex sentences, and word recall, although language comprehension is normal.

EXPRESSIVE LANGUAGE. Communicating with language.

EXPRESSIVE LANGUAGE DEVELOPMENT. A style of language development in which a child's babble mimics the cadence and rhythm of adult speech.

EXSTROPHY. A congenital condition in which a hollow organ, such as the bladder, is turned inside out, establishing contact between the organ and the outside of the body.

EXTENDED FAMILY. Traditionally defined as the biological relatives of a nuclear family (the parents, sisters, and brothers of both members of a married couple); sometimes used to refer to the people living in the household as partners and parents with children.

EXTENDED FAMILY FIELD. A person's family of origin plus grandparents, in-laws, and other relatives.

EXTERNAL CEPHALIC VERSION. Manual manipulation of the abdomen in order to turn a breech baby; also known as version.

EXTRACTION. The removal of a tooth from its socket in the bone.

EXTRAOCULAR RETINOBLASTOMA. Cancer that has spread from the eye to other parts of the body.

EXTRAUTERINE. Occurring outside the uterus.

EXTRAVASATION. To pass from a blood vessel into the surrounding tissue.

EXTROVERSION. A personal preference for socially engaging activities and settings.

EXTROVERT. A person who is outgoing and performs well socially.

EXTUBATION. The removal of a breathing tube.

EXUDATE. Cells, protein, fluid, or other materials that pass through cell or blood vessel walls. Exudates may accumulate in the surrounding tissue or may be discharged outside the body.

EXUDATION. Leakage of cells, proteins, and fluids through the blood vessel wall into the surrounding tissue.

F

FACIOSCAPULOHUMERAL MUSCULAR DYSTROPHY (FSH). This form of muscular dystrophy, also known as Landouzy-Dejerine disease, begins in late childhood to early adulthood and affects both men and women, causing weakness in the muscles of the face, shoulders, and upper arms.

FACTITIOUS DISORDER. A mental condition in which symptoms are deliberately manufactured by patients in order to gain attention and sympathy. Patients with factitious diseases do not fake symptoms for obvious financial gain or to evade the legal system.

FACTOR VIII. A protein involved in blood clotting that requires von Willebrand factor for stability and long-term survival in the bloodstream.

FALLOPIAN TUBES. The pair of narrow tubes leading from a woman's ovaries to the uterus. After an egg is released from the ovary during ovulation, fertilization (the union of sperm and egg) normally occurs in the fallopian tubes.

FAMILY. Two or more emotionally involved people living in close proximity and having reciprocal obligations with a sense of commonness, caring, and commitment.

FAMILY SYSTEMS THEORY. An approach to treatment that emphasizes the interdependency of family members rather than focusing on individuals in isolation from the family. This theory underlies the most influential forms of contemporary family therapy.

FAMILY THERAPY. A type of therapy in which the entire immediate family participates.

FANCONI'S SYNDROME. A group of disorders involving kidney tubule malfunction and glucose, phosphate, and bicarbonate in the urine. Two forms of this syndrome have been identified: an inherited form and an acquired form caused by vitamin D deficiency or exposure to heavy metals.

FANTASY PLAY. Play activities in which children act out their fantasies.

FASCICULATIONS. Small involuntary muscle contractions visible under the skin.

FASCIITIS. Inflammation of the fascia (plural, fasciae), which refers to bands or sheaths of connective tissue that cover, support, or connect the muscles and internal organs. Human bites can lead to infection of the fasciae in the hand.

FAST-ACTING CARBOHYDRATE. A carbohydrate that causes blood sugar levels to rise quickly rather than

slowly and steadily. Also called simple sugars. Examples include glucose tablets, honey, fructose, hard candy, and cake frosting.

FAT-SOLUBLE VITAMIN. A vitamin that dissolves easily in fat or oil, but not in water. The fat-soluble vitamins are vitamins D, E, A, and K.

FATTY ACID. The primary component of lipids (fats) in the body. The body requires some, called essential fatty acids, to form membranes and synthesize important compounds.

FEBRILE SEIZURE. Convulsions brought on by fever.

FECES. The solid waste, also called stool, that is left after food is digested. Feces form in the intestines and pass out of the body through the anus.

FEMALE ATHLETE TRIAD. A combination of disorders frequently found in female athletes that includes disordered eating, osteoporosis, and oligo- or amenorrhea. The triad was first officially named in 1993.

FEMUR. The thigh bone.

FERTILIZATION. The joining of the sperm and the egg; conception.

FETAL PROTEINS. Proteins that are usually produced during fetal development but may persist at high blood levels in some conditions after birth.

FETAL TISSUE TRANSPLANTATION. A method of treating Parkinson's and other neurological diseases by grafting brain cells from human fetuses onto the basal ganglia.

FETOSCOPE. A fiber optic instrument for viewing the fetus inside the uterus.

FETUS. In humans, the developing organism from the end of the eighth week to the moment of birth. Until the end of the eighth week the developing organism is called an embryo.

FIBER. Carbohydrate material in food that cannot be digested.

FIBRILLIN. A protein that is an important part of the structure of the body's connective tissue. In Marfan's syndrome, the gene responsible for fibrillin has mutated, causing the body to produce a defective protein.

FIBRIN. The last step in the blood coagulation process. Fibrin forms strands that add bulk to a forming blood clot to hold it in place and help "plug" an injured blood vessel wall.

FIBROID TUMOR. A non-cancerous tumor of connective tissue made of elongated, threadlike structures, or fibers, which usually grow slowly and are contained within an irregular shape. Fibroids are firm in consistency but may become painful if they start to break down or apply pressure to areas within the body.

FIGHT BITE. Another name for closed-fist injury.

FINANCIAL COUNSELOR. Professional who can provide assistance with financial matters associated with the patient's hospital stay. The financial counselor can help families evaluate their insurance plan's hospitalization coverage, determine a payment plan for medical expenses that are not covered, and discuss possible sources of financial aid.

FINE MOTOR SKILL. The abilities required to control the smaller muscles of the body for writing, playing an instrument, artistic expression, and craft work. The muscles required to perform fine motor skills are generally found in the hands, feet, and head.

FISTULA. An abnormal channel that connects two organs or connects an organ to the skin.

FLACCID. Flabby, limp, weak, or floppy.

FLACCID PARALYSIS. Paralysis characterized by limp, unresponsive muscles.

FLARE. A sudden worsening or recurrence of a disease.

FLAT AFFECT. Showing no emotion.

FLAVONOID. A food chemical that helps to limit oxidative damage to the body's cells, and protects against heart disease and cancer.

FLEXION. The act of bending or condition of being bent.

FLEXOR MUSCLE. A muscle that serves to flex or bend a part of the body.

FLEX-TIME. A system that allows employees to set their own work schedules within guidelines or limits set by the employer.

FLUORAPATITE. Fluoride-substituted hydroxyapatite.

FLUORESCENCE IN SITU HYBRIDIZATION (FISH). A technique for diagnosing genetic disorders before birth by analyzing cells obtained by amniocentesis with DNA probes.

FLUORESCENT ANTIBODY TEST. A test in which a fluorescent dye is linked to an antibody for diagnostic purposes.

FLUORIDE. A chemical compound containing fluorine that is used to treat water or applied directly to teeth to prevent decay.

FLUOROQUINOLONES. A relatively new group of antibiotics used to treat infections with many gram-negative bacteria, such as *Shigella*. One drawback is that they

should not be used in children under 17 years of age, because of possible effect on bone or cartilage growth.

FLUOROSIS. Mottled discoloration of tooth enamel due to excessive systemic ingestion of fluoride during tooth development.

FMR-1 GENE. A gene found on the X chromosome. Its exact purpose is unknown, but it is suspected that the gene plays a role in brain development.

FOLLICLE-STIMULATING HORMONE (FSH). A pituitary hormone that in females stimulates the ovary to mature egg capsules (follicles) and in males stimulates sperm production.

FONTANELLE. One of several "soft spots" on the skull where the developing bones of the skull have yet to fuse.

FOOD-BORNE ILLNESS. A disease that is transmitted by eating or handling contaminated food.

FORAMEN MAGNUM. The opening at the base of the skull, through which the spinal cord and the brainstem pass.

FORAMEN OVALE. A fetal cardiac structure that allows the blood in both upper chambers (atria) of the heart to mix. After birth, the pressure rises in the left atrium pushing this opening closed, allowing the heart to function in a two-sided fashion: the right side carries the unoxygenated blood to the lungs, and the left side pumps the oxygenated blood out into the body.

FORCED EXHALATION. Blowing as much air out of the lungs as possible.

FORCIBLE SODOMY. Forced oral or anal intercourse.

FORENSIC. Pertaining to courtroom procedure or evidence used in courts of law.

FORESKIN. A covering fold of skin over the tip of the penis.

FRAGILE X SYNDROME. A genetic condition related to the X chromosome that affects mental, physical, and sensory development. It is the most common form of inherited mental retardation.

FRANK BREECH. A breech position where the baby is bottom first and his legs are extended upward so that his feet are near his head.

FREE RADICAL. An unstable molecule that causes oxidative damage by stealing electrons from surrounding molecules, thereby disrupting activity in the body's cells.

FREQUENCY. Sound, whether traveling through air or the human body, produces vibrations—molecules bouncing into each other—as the sound wave travels along. The frequency of a sound is the number of vibrations per second. Within the audible range, frequency means pitch—the higher the frequency, the higher a sound's pitch.

FUNDAL HEIGHT. Measured by a tape measure from the top of the symphysis pubis, over the arch of the growing uterus, to the top of the fundus.

FUNDOPLICATION. A surgical procedure that increases pressure on the lower esophageal sphincter by stretching and wrapping the upper part of the stomach around the sphincter.

FUNDUS. The inside of an organ. In the eye, fundus refers to the back area that can be seen with the ophthalmoscope.

FUNGAL. Caused by a fungus.

G

GAIT. Walking motions.

GALACTOSE. One of the two simple sugars (glucose is the other one) that makes up the protein, lactose, found in milk. Galactose can be toxic in high levels.

GANGLION. Plural, ganglia. A mass of nerve tissue or a group of neurons.

GANGLIOSIDE. A fatty (lipid) substance found within the brain and nerve cells.

GANGRENE. Decay or death of body tissue because the blood supply is cut off. Tissues that have died in this way must be surgically removed.

GASTRIC LAVAGE. Also called a stomach pump. For this procedure, a flexible tube is inserted through the nose, down the throat, and into the stomach. The contents of the stomach are then suctioned out. The inside of the stomach is rinsed with a saline (saltwater) solution.

GASTROENTERITIS. Inflammation of the stomach and intestines that usually causes nausea, vomiting, diarrhea, abdominal pain, and cramps.

GASTROENTEROLOGIST. A physician who specializes in diseases of the digestive system.

GASTROESOPHAGEAL REFLUX. The backflow of stomach contents into the esophagus.

GASTROESOPHAGEAL REFLUX DISEASE (GERD). A disorder of the lower end of the esophagus in which the lower esophageal sphincter does not open and close normally. As a result the acidic contents of the stomach can flow backward into the esophagus and irritate the tissues.

GASTROINTESTINAL. Pertaining to the digestive organs and structures, including the stomach and intestines.

GASTROINTESTINAL (GI) SYSTEM. The body system involved in digestion, the breaking down and use of food. It includes the stomach, small intestine, and large intestine. Also known as the gastrointestinal tract.

GASTROSTOMY TUBE. A tube that is inserted through a small incision in the abdominal wall and that extends through the stomach wall into the stomach for the purpose of introducing parenteral feedings. Also called a gastric tube, gastrointestinal tube, or stomach tube.

GAY BASHING. Physical or verbal violence directed against homosexuals.

GENDER IDENTITY DISORDER (GID). A strong and lasting cross-gender identification and persistent discomfort with one's biological gender (sex) role. This discomfort must cause a significant amount of distress or impairment in the functioning of the individual.

GENE. A building block of inheritance, which contains the instructions for the production of a particular protein, and is made up of a molecular sequence found on a section of DNA. Each gene is found on a precise location on a chromosome.

GENE THERAPY. An experimental treatment for certain genetic disorders in which a abnormal gene is replaced with the normal copy. Also called somatic-cell gene therapy.

GENERAL ANESTHESIA. Deep sleep induced by a combination of medicines that allows surgery to be performed.

GENETIC. Refers to genes, the basic units of biological heredity, which are contained on the chromosomes.

GENETIC DISEASE. A disease that is (partly or completely) the result of the abnormal function or expression of a gene; a disease caused by the inheritance and expression of a genetic mutation.

GENITAL. Refers to the sexual or reproductive organs that are visible outside the body.

GENITAL HERPES. A life-long, recurrent sexually transmitted infection caused by the herpes simplex virus (HSV).

GENOGRAM. A family tree diagram that represents the names, birth order, sex, and relationships of the members of a family. Therapists use genograms to detect recurrent patterns in the family history and to help the family members understand their problem(s).

GENU RECURVATUM. Hyperextension of the knee.

GEOPHAGIA. The compulsive eating of earth substances, including sand, soil, and clay.

GESTATION. The period from conception to birth, during which the developing fetus is carried in the uterus.

GESTATIONAL DIABETES. Diabetes of pregnancy leading to increased levels of blood sugar. Unlike diabetes mellitus, gestational diabetes is caused by pregnancy and goes away when pregnancy ends.

GHRELIN. A peptide hormone secreted by cells in the lining of the stomach. Ghrelin is important in appetite regulation and maintaining the body's energy balance.

GIGANTISM. Excessive growth, especially in height, resulting from overproduction of growth hormone during childhood or adolescence by a pituitary tumor. Untreated, the tumor eventually destroys the pituitary gland, resulting in death during early adulthood.

GINGIVA. The gum tissue surrounding the teeth.

GINGIVAL SULCUS. The space between the tooth and the gum that often traps food and bacteria, leading to periodontal disease.

GINGIVITIS. Inflammation of the gums in which the margins of the gums near the teeth are red, puffy, and bleeding. It is most often due to poor dental hygiene.

GLAND. A collection of cells whose function is to release certain chemicals (hormones) that are important to the functioning of other, sometimes distantly located, organs or body systems.

GLANS PENIS. The cone-shaped tip of the penis.

GLAUCOMA. A common eye disease characterized by increased fluid pressure in the eye that damages the optic nerve, which carries visual impulses to the brain. Glaucoma can be caused by another eye disorder, such as a tumor or congenital malformation, or can appear without obvious cause, but if untreated it generally leads to blindness.

GLOBIN. One of the component protein molecules found in hemoglobin. Normal adult hemoglobin has a pair each of alpha-globin and beta-globin molecules.

GLOMERULUS. Plural, glomeruli; a network of capillaries located in the nephron of the kidney where wastes are filtered from the blood.

GLOSSOPHOBIA. Fear of speaking.

GLOTTIS. The opening between the vocal cords at the upper part of the larynx.

GLUCAGON. A hormone produced in the pancreas that changes glycogen, a carbohydrate stored in muscles and the liver, into glucose. It can be used to relax mus-

cles for a procedure such as duodenography. An injectable form of glucagon is sometimes used to treat insulin shock.

GLUCOCORTICOIDS. A general class of adrenal cortical hormones that are mainly active in protecting against stress and in protein and carbohydrate metabolism. They are widely used in medicine anti-inflammatories and immunosuppresives.

GLUCOSE. A simple sugar that serves as the body's main source of energy.

GLUCOSE-6-PHOSPHATE DEHYDROGENASE (G6PD) DEFICIENCY. A sex-linked hereditary disorder in which the body lacks an enzyme that normally protects red blood cells from toxic chemicals. When people with this condition take certain drugs, their red blood cells break down, causing anemia.

GLUTEN. A protein found in wheat, rye, barley, and oats.

GLYCATED HEMOGLOBIN. A test that measures the amount of hemoglobin bound to glucose. It is a measure of how much glucose has been in the blood during a two to three month period beginning approximately one month prior to sample collection.

GLYCEMIC. The presence of glucose in the blood.

GOITER. Chronic enlargement of the thyroid gland.

GONADOTROPHIN. Hormones that stimulate the ovary and testicles.

GONADS. Organs that produce gametes (eggs or sperm), i.e. the ovaries and testes.

GONOCOCCAL. Refers to the bacterium *Neisseria gonorrheae*. This bacterium causes gonorrhea, a sexually transmitted infection of the genitals and urinary tract. The gonococcal organism may occasionally affect the eye, causing blindness if not treated.

GONORRHEA. A sexually transmitted disease that causes infection in the genital organs and may cause disease in other parts of the body.

GOODNESS OF FIT. A term first used by Thomas and Chess to describe the importance of children's interactions with their environment as well as their basic temperament in understanding their later growth and development.

GOUT. A metabolic disorder characterized by sudden recurring attacks of arthritis caused by deposits of crystals that build up in the joints due to abnormally high uric acid blood levels.

GRAFT. A transplanted organ or other tissue.

GRAM STAIN. A staining procedure used to visualize and classify bacteria. The Gram stain procedure allows the identification of purple (gram positive) organisms and red (gram negative) organisms. This identification aids in determining treatment.

GRAM-NEGATIVE. Refers to bacteria that have a cell wall composed of a thin layer of peptidoglycan surrounded by an outer membrane made of polysaccharides and proteins. They take on the red color of the counterstain used in the Gram stain procedure.

GRANULES. Small packets of reactive chemicals stored within cells.

GRANULOCYTOPENIA. A condition characterized by a deficiency of white blood cells.

GRANULOMA. An inflammatory swelling or growth composed of granulation tissue

GRAY MATTER. Areas of the brain and spinal cord that are comprised mostly of unmyelinated nerves.

GRIEF REACTION. The normal depression felt after a traumatic major life occurrence such as the loss of a loved one.

GROSS MOTOR SKILLS. The abilities required to control the large muscles of the body for walking, running, sitting, crawling, and other activities. The muscles required to perform gross motor skills are generally found in the arms, legs, back, abdomen and torso.

GROUP B STREPTOCOCCUS. A serotype of streptococcus, *Streptococcus agalactiae*, which is beta hemolytic and can cause neonatal sepsis, pneumonia, or meningitis if present in the birth canal at the time of delivery especially when the delivery is difficult.

GROWTH HORMONE. A hormone that eventually stimulates growth. Also called somatotropin.

GUILLAIN-BARRÉ SYNDROME. Progressive and usually reversible paralysis or weakness of multiple muscles usually starting in the lower extremities and often ascending to the muscles involved in respiration. The syndrome is due to inflammation and loss of the myelin covering of the nerve fibers, often associated with an acute infection. Also called acute idiopathic polyneuritis.

H

H2RAS. Medications used to treat some gastroesophageal reflux disease symptoms, for example, Tagamet, Pepcid, Axid.

***HAEMOPHILUS INFLUENZAE* TYPE B.** An anaerobic bacteria associated with human respiratory infections, conjunctivitis, and meningitis.

HAIR CELLS. Sensory receptors in the inner ear that transform sound vibrations into messages that travel to the brain.

HAIR FOLLICLE. The root of a hair (that portion of a hair below the skin surface) together with its epithelial and connective tissue coverings.

HAIRBULB. The root of a strand of hair from which growth and coloration of the hair develops.

HALO EFFECT. An observer bias in which the observer interprets a child's actions in a way that confirm the observer's preconceived ideas about the child.

HAND-EYE COORDINATION. The ability to grasp or touch an object while looking at it.

HATHA YOGA. A form of yoga using postures, breathing methods, and meditation.

HEAD START. A federal program started in 1965 that provides free education for young children in many low-income families across the United States.

HEART ATTACK. Damage that occurs to the heart when one of the coronary arteries becomes narrowed or blocked.

HEART FAILURE. A condition in which the heart is unable to pump enough blood to supply the needs of the body

HEARTBURN. A burning sensation in the chest that can extend to the neck, throat, and face. It is the primary symptom of gastroesophageal reflux (the movement of stomach acid into the esophagus).

HEAT EXHAUSTION. A condition of physical weakness or collapse often accompanied by nausea, muscle cramps, and dizziness, that is caused by exposure to intense heat.

HEAT STROKE. A serious condition that results from exposure to extreme heat. The body loses its ability to cool itself. Severe headache, high fever, and hot, dry skin may result. In severe cases, a person with heat stroke may collapse or go into a coma.

HEAVY METAL. One of 23 chemical elements that has a specific gravity (a measure of density) at least five times that of water.

HEIMLICH MANEUVER. An emergency procedure for removing a foreign object lodged in the airway that is preventing the person from breathing. To perform the Heimlich maneuver on a conscious adult, the rescuer stands behind the victim and encircles his waist. The rescuer makes a fist with one hand and places the other hand on top, positioned below the rib cage and above the waist. The rescuer then applies pressure by a series of upward and inward thrusts to force the foreign object back up the victim's trachea.

HEMANGIOMA. A benign skin tumor composed of abnormal blood vessels.

HEMATIN. A drug administered intravenously to halt an acute porphyria attack. It causes heme biosynthesis to decrease, preventing the further accumulation of heme precursors.

HEMATOCRIT. A measure of the percentage of red blood cells in the total volume of blood in the human body.

HEMATOMA. A localized collection of blood, often clotted, in body tissue or an organ, usually due to a break or tear in the wall of blood vessel.

HEMATURIA. The presence of blood in the urine.

HEME. The iron-containing molecule in hemoglobin that serves as the site for oxygen binding.

HEMIANOPSIA. Loss of half of the field of vision.

HEMIPLEGIA. Paralysis of one side of the body.

HEMOCHROMATOSIS. An inherited blood disorder that causes the body to retain excessive amounts of iron. This iron overload can lead to serious health consequences, including painful joints, diabetes, and liver damage, if the iron concentration is not lowered.

HEMOCYTOMETER. An instrument used to count platelets or other blood cells.

HEMOGLOBIN. An iron-containing pigment of red blood cells composed of four amino acid chains (alpha, beta, gamma, delta) that delivers oxygen from the lungs to the cells of the body and carries carbon dioxide from the cells to the lungs.

HEMOGLOBIN A. Normal adult hemoglobin that contains a heme molecule, two alpha-globin molecules, and two beta-globin molecules.

HEMOGLOBIN ELECTROPHORESIS. A laboratory test that separates molecules based on their size, shape, or electrical charge. It is used to identify abnormal hemoglobins in the blood.

HEMOGLOBINOPATHY. A disorder of hemoglobin, which can be either the presence of abnormal types of hemoglobin or abnormal levels of specific types of hemoglobin, i.e., sickle cell disease and thalessemia.

HEMOLYSIS. The process of breaking down of red blood cells. As the cells are destroyed, hemoglobin, the

component of red blood cells which carries the oxygen, is liberated.

HEMOLYTIC. Able to break down or dissolve red blood cells.

HEMOLYTIC ANEMIA. A form of anemia characterized by chronic premature destruction of red cells in the bloodstream. Hemolytic anemias are classified as either inherited or acquired.

HEMOLYTIC-UREMIC SYNDROME (HUS). A potentially fatal complication of *E. coli* infection characterized by kidney failure and destruction of red blood cells.

HEMOPHILIA. Any of several hereditary blood coagulation disorders occurring almost exclusively in males. Because blood does not clot properly, even minor injuries can cause significant blood loss that may require a blood transfusion, with its associated minor risk of infection.

HEMORRHAGE. Severe, massive bleeding that is difficult to control. The bleeding may be internal or external.

HENOCH-SCHÖNLEIN PURPURA. A syndrome sometimes classified as a hypersensitivity vasculitis, associated with a variety of digestive symptoms, pain in the joints, and kidney involvement. Purpura comes from the Latin word for ''purple'' and refers to the reddish-purple spots on the skin caused by leakage of blood from inflamed capillaries.

HEPARIN. An organic acid that occurs naturally in the body and prevents blood clots. Heparin is also made synthetically and can be given as an anticoagulant treatment.

HEPATIC. Refers to the liver.

HEPATITIS. An inflammation of the liver, with accompanying liver cell damage or cell death, caused most frequently by viral infection, but also by certain drugs, chemicals, or poisons. May be either acute (of limited duration) or chronic (continuing).

HEPATITIS A. Commonly called infectious hepatitis, caused by the hepatitis A virus (HAV). Most often spread by food and water contamination.

HEPATITIS B. An infection of the liver that is caused by a DNA virus, is transmitted by contaminated blood or blood derivatives in transfusions, by sexual contact with an infected person, or by the use of contaminated needles and instruments.

HEPATITIS B IMMUNE GLOBULIN. HBIG, a blood serum preparation containing anti-hepatitis-B antibodies (anti-HBs) that is administered along with HBV to children born to hepatitis-B-infected mothers.

HEPATITIS B VIRUS (HBV). Also called Hepadna virus, the pathogen responsible for hepatitis B infection.

HEREDITARY. Something which is inherited, that is passed down from parents to offspring. In biology and medicine, the word pertains to inherited genetic characteristics.

HEREDITARY ATAXIA. One of a group of hereditary degenerative diseases of the spinal cord or cerebellum. These diseases cause tremor, spasm, and wasting of muscle.

HERMANSKY-PUDLAK SYNDROME. A rare type of albinism, most common in the Puerto Rican community, which can cause pigment changes, lung disease, intestinal disorders, and blood disorders.

HERNIA. A rupture in the wall of a body cavity, through which an organ may protrude.

HERNIATION. Bulging of tissue through opening in a membrane, muscle, or bone.

HERNIORRHAPHY. Surgical repair of a hernia.

HERPES SIMPLEX VIRUS. A virus that can cause fever and blistering on the skin and mucous membranes. Herpes simplex 1 infections usually occur on the face (cold sores) and herpes simplex 2 infections usually occur in the genital region.

HERPES STOMATITIS. A form of stomatitis caused by the herpes 1 virus, usually seen in young children.

HERPES ZOSTER VIRUS. Acute inflammatory virus that attacks the nerve cells on the root of each spinal nerve with skin eruptions along a sensory nerve ending. It causes chickenpox and shingles. Also called varicella zoster virus.

HERPES VIRUS. A family of viruses including herpes simplex types 1 and 2, and herpes zoster (also called varicella zoster). Herpes viruses cause several infections, all characterized by blisters and ulcers, including chickenpox, shingles, genital herpes, and cold sores or fever blisters.

HERPETIC GINGIVOSTOMATITIS. A severe oral infection that affects children under five years of age; vesicles and ulcerations, edematous throat, enlarged painful cervical lymph nodes occur; chills, fever, malaise, bed breath, and drooling.

HETEROZYGOTE/HETEROZYGOUS. Having two different versions of the same gene.

HIATAL HERNIA. A condition in which part of the stomach protrudes through the diaphragm into the chest cavity.

HIB DISEASE. An infection caused by *Haemophilus influenza* type b (Hib). This disease mainly affects children under the age of five. In that age group, it is the leading cause of bacterial meningitis, pneumonia, joint and bone infections, and throat inflammations.

HIGH. The altered state of consciousness that a person seeks when abusing a substance.

HISTAMINE. A substance released by immune system cells in response to the presence of an allergen. It stimulates widening of blood vessels and increased porousness of blood vessel walls so that fluid and protein leak out from the blood into the surrounding tissue, causing localized inflammation of the tissue.

HISTOLOGY. The study of tissue structure.

HISTRIONIC PERSONALITY DISORDER. A mental disorder characterized by inappropriate attention-seeking behavior, rapid emotional shifts, and exaggerated expression of emotion.

HODGKIN'S DISEASE. One of two general types of lymphoma (cancers that arise in the the lymphatic system and can invade other organs), Hodgkin's disease is characterized by lymph node enlargement and the presence of a large polyploid cells called Reed-Sternberg cells.

HOME CARE. Health care services provided in the patient's home. If home health services will be needed after the patient is discharged, they can be arranged by the social worker or nursing staff.

HOMEOPATHY. A holistic system of treatment developed in the eighteenth century. It is based on the idea that substances that produce symptoms of sickness in healthy people will have a curative effect when given in very dilute quantities to sick people who exhibit those same symptoms. Homeopathic remedies are believed to stimulate the body's own healing processes.

HOMEOSTASIS. The balanced internal environment of the body and the automatic tendency of the body to maintain this internal "steady state." Also refers to the tendency of a family system to maintain internal stability and to resist change.

HOMOCYSTEINE. An sulfur-containing amino acid.

HOMOPHOBIA. An irrational hatred, disapproval, or fear of homosexuality and homosexuals.

HOMOZYGOTE/HOMOZYGOUS. Having two identical copies of a gene.

HONEYMOON PHASE. A period of time shortly following diagnosis of type 1 diabetes during which a child's need for insulin may decrease or disappear altogether. The honeymoon phase is transitional, and insulin requirements eventually increases again.

HORDEOLUM. The medical term for stye, an infection or small abscess formation in a hair follicle of the eyelids.

HORMONE. A chemical messenger secreted by a gland or organ and released into the bloodstream. It travels via the bloodstream to distant cells where it exerts an effect.

HORMONE THERAPY. Treating cancers by changing the hormone balance of the body, instead of by using cell-killing drugs.

HUMAN DIPLOID CELL VACCINE (HDCV). A rabies vaccine in which the virus is grown in cultures of human cells, concentrated, and inactivated for IM or ID injection.

HUMAN IMMUNODEFICIENCY VIRUS (HIV). A transmissible retrovirus that causes AIDS in humans. Two forms of HIV are now recognized: HIV-1, which causes most cases of AIDS in Europe, North and South America, and most parts of Africa; and HIV-2, which is chiefly found in West African patients. HIV-2, discovered in 1986, appears to be less virulent than HIV-1 and may also have a longer latency period.

HUMAN LEUCKOCYTE ANTIGEN (HLA). A group of protein molecules located on bone marrow cells that can provoke an immune response. A donor's and a recipient's HLA types should match as closely as possible to prevent the recipient's immune system from attacking the donor's marrow as a foreign material that does not belong in the body.

HUMAN PAPILLOMA VIRUS (HPV). A virus that causes common warts of the hands and feet, as well as lesions in the genital and vaginal area. More than 50 types of HPV have been identified, some of which are linked to cancerous and precancerous conditions, including cancer of the cervix.

HUNTINGTON'S DISEASE. A rare hereditary disease that causes progressive chorea (jerky muscle movements) and mental deterioration that ends in dementia. Huntington's symptoms usually appear in patients in their 40s. Also called Huntington's chorea.

HYALINE MEMBRANE. A fibrous layer that settles in the alveoli in respiratory distress syndrome and prevents oxygen from escaping from inhaled air to the bloodstream.

HYDROCELE. A collection of fluid between two layers of tissue surrounding the testicle; the most common cause of painless scrotal swelling.

HYDROCEPHALUS. An abnormal accumulation of cerebrospinal fluid within the brain. This accumulation can be harmful by pressing on brain structures, and damaging them.

HYDROGEN BREATH TEST. A test used to determine if a person is lactose intolerant or if abnormal bacteria are present in the colon.

HYDROPS FETALIS. A condition in which a fetus or newborn baby accumulates fluids, causing swollen arms and legs and impaired breathing.

HYDROTHERAPY. The use of water (hot, cold, steam, or ice) to relieve discomfort and promote physical well-being. Also called water therapy.

HYDROXYUREA. A drug that has been shown to induce production of fetal hemoglobin. Fetal hemoglobin has a pair of gamma-globin molecules in place of the typical beta-globins of adult hemoglobin. Higher-than-normal levels of fetal hemoglobin can ameliorate some of the symptoms of thalassemia.

HYPERACTIVE REFLEXES. Reflexes that persist too long and may be too strong. For example, a hyperactive grasp reflex may cause the hand to stay clenched in a tight fist.

HYPERALIMENTATION. A method of refeeding anorexics by infusing liquid nutrients and electrolytes directly into central veins through a catheter.

HYPERANDROGENISM. The excessive secretion of androgens.

HYPERBARIC OXYGEN THERAPY. Medical treatment in which oxygen is administered in specially designed chambers, under pressures greater than that of the atmosphere, in order to treat specific medical conditions, such as carbon monoxide poisoning, smoke inhalation, and certain bacterial infections.

HYPERBILIRUBINEMIA. A condition characterized by a high level of bilirubin in the blood. Bilirubin is a natural byproduct of the breakdown of red blood cells, however, a high level of bilirubin may indicate a problem with the liver.

HYPERCALCEMIA. A condition marked by abnormally high levels of calcium in the blood.

HYPERCOAGULABLE STATE. (Also called thromboembolic state or thrombophilia.) A condition characterized by excess blood clotting.

HYPEREXTENSIBILITY. The ability to extend a joint beyond the normal range.

HYPERGLYCEMIA. A condition characterized by excessively high levels of glucose in the blood. It occurs when the body does not have enough insulin or cannot use the insulin it does have to turn glucose into energy.

HYPERKALEMIA. An abnormally high level of potassium in the blood.

HYPERLIPIDEMIA. A condition characterized by abnormally high levels of lipids in blood plasma.

HYPERMOBILITY. Unusual flexibility of the joints, allowing them to be bent or moved beyond their normal range of motion.

HYPERNATREMIA. An abnormally high level of sodium in the blood.

HYPEROSMOTIC. Hypertonic, containing a higher concentration of salts or other dissolved materials than normal tissues.

HYPERPHAGIA. Over-eating.

HYPERPLASIA. A condition where cells, such as those making up the prostate gland, rapidly divide abnormally and cause the organ to become enlarged.

HYPERPLASTIC. Refers to an increase in the size of an organ or tissue due to an increase in the number of cells.

HYPERPLASTIC OBESITY. Excessive weight gain in childhood, characterized by an increase in the number of new fat cells.

HYPERPYREXIA. Fever greater than 105.8°F (41°C).

HYPERSENSITIVITY. A condition characterized by an excessive response by the body to a foreign substance. In hypersensitive individuals even a tiny amount of allergen can cause a severe allergic reaction.

HYPERSOMNIA. An abnormal increase of 25% or more in time spent sleeping. Individuals with hypersomnia usually have excessive daytime sleepiness.

HYPERTENSION. Abnormally high arterial blood pressure, which if left untreated can lead to heart disease and stroke.

HYPERTHERMIA. Body temperature that is much higher than normal (i.e. higher than 98.6°F [37°C]).

HYPERTHYROIDISM. A condition characterized by abnormal over-functioning of the thyroid glands. Patients are hypermetabolic, lose weight, are nervous, have muscular weakness and fatigue, sweat more, and have increased urination and bowel movements. Also called thyrotoxicosis.

HYPERTONIA. Having excessive muscular tone or strength.

HYPERTONIC SALINE SOLUTION. Fluid that contains salt in a concentration higher than that of healthy blood.

HYPERTROPHIC OBESITY. Excessive weight gain in adulthood, characterized by expansion of already existing fat cells.

HYPERTROPHY. An increase in the size of a tissue or organ brought about by the enlargement of its cells rather than cell multiplication.

HYPERVENTILATION. Rapid, deep breathing, possibly exceeding 40 breaths/minute. The most common cause is anxiety, although fever, aspirin overdose, serious infections, stroke, or other diseases of the brain or nervous system. Also refers to a respiratory therapy involving deeper and/or faster breathing to keep the carbon dioxide pressure in the blood below normal.

HYPNOGOGIC HALLUCINATION. A vivid, dreamlike hallucination, such as the sensation of falling, that occurs at the onset of sleep.

HYPNOSIS. The technique by which a trained professional induces a trance-like state of extreme relaxation and suggestibility in a patient. Hypnosis is used to treat amnesia and identity disturbances that occur in dissociative disorders.

HYPNOTICS. A class of drugs that are used as a sedatives and sleep aids.

HYPOCALCEMIA. A condition characterized by an abnormally low level of calcium in the blood.

HYPOCHROMIC. A descriptive term applied to a red blood cell with a decreased concentration of hemoglobin.

HYPOCRETINS. Chemicals secreted in the hypothalamus that regulate the sleep/wake cycle.

HYPOGLYCEMIA. A condition characterized by abnormally low levels of glucose in the blood.

HYPOGLYCEMIC UNAWARENESS. A condition in which normal warning signals of a blood sugar low, such as shakiness, sweating, or rapid heartbeat, are no longer felt.

HYPOKALEMIA. A condition characterized by a deficiency of potassium in the blood.

HYPOMANIA. A milder form of mania that is characteristic of bipolar II disorder.

HYPOPLASIA. An underdeveloped or incomplete tissue or organ usually due to a decrease in the number of cells.

HYPOPLASTIC. Refers to incomplete or underdeveloped tissues or organs. Hypoplastic left heart syndrome is the most serious type of congenital heart disease.

HYPOPNEA. Shallow or excessively slow breathing usually caused by partial closure of the upper airway during sleep, leading to disruption of sleep.

HYPOSPADIAS. A congenital abnormality of the penis in which the urethral opening is located on the underside of the penis rather than at its tip.

HYPOTENSION. Low blood pressure.

HYPOTHALAMUS. A part of the forebrain that controls heartbeat, body temperature, thirst, hunger, body temperature and pressure, blood sugar levels, and other functions.

HYPOTHERMIA. A serious condition in which body temperature falls below 95°F (35 °C). It is usually caused by prolonged exposure to the cold.

HYPOTHYROIDISM. A disorder in which the thyroid gland produces too little thyroid hormone causing a decrease in the rate of metabolism with associated effects on the reproductive system. Symptoms include fatigue, difficulty swallowing, mood swings, hoarse voice, sensitivity to cold, forgetfulness, and dry/coarse skin and hair.

HYPOTONIA. Having reduced or diminished muscle tone or strength.

HYPOXEMIA. A condition characterized by an abnormally low amount of oxygen in the arterial blood. It is the major consequence of respiratory failure, when the lungs no longer are able to perform their chief function of gas exchange.

I

IATROGENIC. A condition that is caused by the diagnostic procedures or treatments administered by medical professionals. Iatrogenic conditions may be caused by any number of things including contaminated medical instruments or devices, contaminated blood or implants, or contaminated air within the medical facility.

ICHTHYOSIS. A group of congenital skin disorders of keratinization characterized by dryness and scaling of the skin.

IDEAL WEIGHT. Weight corresponding to the lowest death rate for individuals of a specific height, gender, and age.

IDENTIFIED PATIENT (IP). The family member in whom the family's symptom has emerged or is most obvious.

IDENTITY. The condition of being the same with, or possessing, a character that is well described, asserted, or defined.

IDIOPATHIC. Refers to a disease or condition of unknown origin.

ILEUS. An obstruction of the intestines usually caused by the absence of peristalsis.

IMMERSION. A language education approach in which English is the only language used.

IMMUNE GLOBULIN. Preparation of antibodies that can be given before exposure for short-term protection against hepatitis A and for persons who have already been exposed to hepatitis A virus. Immune globulin must be given within two weeks after exposure to hepatitis A virus for maximum protection.

IMMUNE HYPERSENSITIVITY REACTION. An allergic reaction that is mediated by mast cells and occurs within minutes of allergen contact.

IMMUNE RESPONSE. A physiological response of the body controlled by the immune system that involves the production of antibodies to fight off specific foreign substances or agents (antigens).

IMMUNE SYSTEM. The system of specialized organs, lymph nodes, and blood cells throughout the body that work together to defend the body against foreign invaders (bacteria, viruses, fungi, etc.).

IMMUNITY. Ability to resist the effects of agents, such as bacteria and viruses, that cause disease.

IMMUNIZATION. A process or procedure that protects the body against an infectious disease by stimulating the production of antibodies. A vaccination is a type of immunization.

IMMUNOCOMPROMISED. A state in which the immune system is suppressed or not functioning properly.

IMMUNODEFICIENCY. A condition in which the body's immune response is damaged, weakened, or is not functioning properly.

IMMUNODEFICIENCY DISEASE. A disease characterized chiefly by an increased susceptibility to infection. It is caused by very low levels of immunoglobulins that result in an impaired immune system. Affected people develop repeated infections.

IMMUNOGLOBIN A. A sugar protein with a high molecular weight that acts like an antibody and is produced by white blood cells during an immune response.

IMMUNOGLOBULIN E (IGE). A type of protein in blood plasma that acts as an antibody to activate allergic reactions. About 50% of patients with allergic disorders have increased IgE levels in their blood serum.

IMMUNOGLOBULIN G (IGG). Immunoglobulin type gamma, the most common type found in the blood and tissue fluids.

IMMUNOSUPPRESSED. A state in which the immune system is suppressed by medications during the treatment of other disorders, like cancer, or following an organ transplantation.

IMMUNOTHERAPY. A mode of cancer treatment in which the immune system is stimulated to fight the cancer.

IMPACTED TOOTH. Any tooth that is prevented from reaching its normal position in the mouth by another tooth, bone, or soft tissue.

IMPACTION. A condition in which earwax has become tightly packed in the outer ear to the point that the external ear canal is blocked.

IMPETIGO. A bacterial infection of the skin characterized by skin blistering.

IMPLANTATION. The process in which the fertilized egg embeds itself in the wall of the uterus.

IMPRESSION. In dentistry, an imprint of the upper or lower teeth made in a pliable material that sets. When this material has hardened, it may be filled with plaster, plastic, or artificial stone to make an exact model of the teeth.

IMPRINTING. A process that silences a gene or group of genes. The genes are silenced depending on whether they are inherited through the egg or the sperm.

INBORN ERROR OF METABOLISM. One of a group of rare conditions characterized by an inherited defect in an enzyme or other protein. Inborn errors of metabolism can cause brain damage and mental retardation if left untreated. Phenylketonuria, Tay-Sachs disease, and galactosemia are inborn errors of metabolism.

INCARCERATED HERNIA. A hernia of the bowel that can not return to its normal place without manipulation or surgery.

INCEST. Unlawful sexual contact between persons who are biologically related. Many therapists, however, use the term to refer to inappropriate sexual contact between any members of a family, including stepparents and stepsiblings.

INCIDENCE. The rate of development of a disease in a given population over time.

INCISOR. One of the eight front teeth.

INCOMPLETE BREECH. Also called a footling breech, in this position the baby has one or both feet down towards the pelvis so that his leg(s) are poised to deliver first.

INCONTINENCE. A condition characterized by the inability to control urination or bowel functions.

INCUBATION PERIOD. The time period between exposure to an infectious agent, such as a virus or bacteria, and the appearance of symptoms of illness. Also called the latent period.

INDEX OF REFRACTION. A constant number for any material and any given color of light that is an indicator of the degree of bending of the light caused by that material.

INDIVIDUALIZED EDUCATIONAL PLAN (IEP). A detailed description of the educational goals, assessment methods, behavioral management plan, and educational performance of a student requiring special education services.

INFANTILE MASTURBATION. The masturbation by infants, also called gratification disorder.

INFARCT. An area of dead tissue caused by inadequate blood supply.

INFECTIOUS DISEASE. A disease caused by a virus or a bacterium. Examples of viruses causing an infectious disease are: HIV-1 virus, herpes simplex, cytomegalovirus, Epstein-Barr virus, leukemia virus. Examples of bacterial infectious diseases are: syphilis and tuberculosis.

INFLAMMATION. Pain, redness, swelling, and heat that develop in response to tissue irritation or injury. It usually is caused by the immune system's response to the body's contact with a foreign substance, such as an allergen or pathogen.

INFLAMMATORY. Pertaining to inflammation.

INFLECTION. Variations in the pitch or tone of a voice.

INFLUENZA. An infectious disease caused by a virus that affects the respiratory system, causing fever, congestion, muscle aches, and headaches.

INFLUENZA VIRUS TYPE. The nature of the proteins in the outer coat of an influenza virus. Depending on the proteins, influenza viruses may be classified as A, B, or C.

INFUSION. Introduction of a substance directly into a vein or tissue by gravity flow.

INHALANT. Medication that is breathed into the lungs.

INHIBITED. A type of child defined by Jerome Kagan and his colleagues as having a low level of responsiveness to strangers, a reluctance to initiate activities, and requiring a long time to relax in new situations. Children with inhibited temperaments appear to be more susceptible to anxiety disorders, depression, and certain personality disorders in their later years.

INNER EAR. The interior section of the ear, where sound vibrations and information about balance are translated into nerve impulses.

INORGANIC. Pertaining to chemical compounds that are not hydrocarbons or their derivatives.

INORGANIC CAUSES. Cases of failure to thrive brought on by a caregiver's actions.

INPATIENT SURGERY. Surgery that requires an overnight stay of one or more days in the hospital. The number of days spent in the hospital after surgery depends on the type of procedure performed.

INSECTICIDE. Any substance used to kill insects.

INSECURE ATTACHMENT. Develops when a primary caregiver does not consistently respond in ways that are warm, affectionate, and sensitive to a baby's needs.

INSOMNIA. A sleep disorder characterized by inability either to fall asleep or to stay asleep.

INSULIN. A hormone or chemical produced by the pancreas that is needed by cells of the body in order to use glucose (sugar), a major source of energy for the human body.

INTELLIGENCE QUOTIENT (IQ). A measure of somebody's intelligence, obtained through a series of aptitude tests concentrating on different aspects of intellectual functioning.

INTELLIGENCE TEST. A questionnaire or series of exercises designed to attempt to measure intelligence.

INTERCEPTIVE ORTHODONTICS. Preventative orthodontics; early, simpler orthodontic treatment.

INTERFERON. A potent immune-defense protein produced by virus-infected cells; used as an anti-cancer and anti-viral drug.

INTERNATIONAL UNIT (IU). A measurement of biological activity in which one IU is equal to one mg (milligram).

INTRACEREBRAL HEMORRHAGE. A cause of some strokes in which vessels within the brain begin bleeding.

INTRACUTANEOUS. Into the skin, in this case directly under the top layer of skin.

INTRADERMAL. An injection into a deep layer of skin.

INTRAMEMBRANOUS OSSIFICATION. The process by which bone tissue is formed within sheets of connective tissue.

INTRAMUSCULAR (IM) INJECTION. An injection into a muscle.

INTRAOCULAR RETINOBLASTOMA. Cancer of the retina that is limited to the eye and has not spread to other parts of the body.

INTRAPARTUM. Refers to the period of time that includes labor and delivery of a baby.

INTRAUTERINE. Situated or occurring in the uterus.

INTRAVENOUS. Into a vein; a needle is inserted into a vein in the back of the hand, inside the elbow, or some other location on the body. Fluids, nutrients, and drugs can be injected. Commonly called IV.

INTRAVENTRICULAR HEMORRHAGE (IVH). A condition in which fragile blood vessels within the brain burst and bleed into the hollow chambers (ventricles) of the brain and into the tissue surrounding them.

INTRINSIC BIAS. An assumed bias that favors one group over another; as in systems and hand implements that assume that all people are right-handed.

INTROVERSION. A personal preference for solitary, non-social activities and settings.

INTUBATION. A procedure in which a tube is inserted through the mouth and into the trachea to keep the airway open and to help a patient breathe.

INTUSSUSCEPTION. The slipping or telescoping of one part of the intestine into the section next to it.

INVOLUTION. The return of a large organ to normal size.

IONIZING RADIATION. Radiation that can damage living tissue by disrupting and destroying individual cells at the molecular level. All types of nuclear radiation—x rays, gamma rays, and beta rays—are potentially ionizing. Sound waves physically vibrate the material through which they pass, but do not ionize it.

IONTOPHORESIS. Application of a small electric current to the skin.

IQ. Intelligence quotient, a measure of intellectual functioning determined by performance on standardized intelligence tests. It is usually calculated by dividing an individual's mental age (determined by testing) by his/her chronological age and multiplying that result by 100.

IRON OVERLOAD. A side effect of frequent blood transfusions in which the body accumulates abnormally high levels of iron. Iron deposits can form in organs, particularly the heart, and cause life-threatening damage.

IRRIGATION. Cleansing a wound with large amounts of water and/or an antiseptic solution. Also refers to the technique of removing wax (cerumen) from the ear canal by flushing it with water.

ISCHEMIA. A decrease in the blood supply to an area of the body caused by obstruction or constriction of blood vessels.

ISOIMMUNIZATION. The development of antibodies in a species in response to antigens from the same species.

ISOTRETINOIN. A powerful vitamin A derivative used in the treatment of acne.

J

JAUNDICE. A condition in which the skin and whites of the eyes take on a yellowish color due to an increase of bilirubin (a compound produced by the liver) in the blood. Also called icterus.

JET LAG. A temporary disruption of the body's sleep-wake rhythm following high-speed air travel across several time zones. Jet lag is most severe in people who have crossed eight or more time zones in 24 hours.

JOINT. The connection point where two bones meet.

JOINT CONTRACTURES. Stiffness of the joints that prevents full extension.

JOINT DISLOCATION. The displacement of a bone from its socket or normal position.

JUVENILE ARTHRITIS. A chronic inflammatory disease characterized predominantly by arthritis with onset before the sixteenth birthday.

K

KALLMAN'S SYNDROME. A disorder of hypogonadotropic hypogonadism, delayed puberty, and anosmia. Kallman's syndrome is a birth defect in the brain that prevents release of hormones and appears as failure of male puberty.

KAPOSI'S SARCOMA. A cancer characterized by bluish-red nodules on the skin, usually on the lower extremities, that often occurs in people with AIDS.

KARYOTYPE. A standard arrangement of photographic or computer-generated images of chromosome pairs from a cell in ascending numerical order, from largest to smallest.

KARYOTYPING. A laboratory test used to study an individual's chromosome make-up. Chromosomes are separated from cells, stained, and arranged in order from largest to smallest so that their number and structure can be studied under a microscope.

KASABACH-MERRIT SYNDROME. A combination of rapidly enlarging hemangioma and thrombocytopenia; it is usually clinically evident during early infancy, but occasionally the onset is later. The hemangiomas are large and may increase in size rapidly and may cause severe anemia in infants.

KAUFMAN ASSESSMENT BATTERY FOR CHILDREN. An intelligence and achievement test for children ages 2.5 to 12.5 years.

KAWASAKI SYNDROME. A syndrome of unknown origin that affects the skin, mucous membranes, and the immune system of infants and young children. It is named for the Japanese pediatrician who first identified it in 1967.

KERATIN. A tough, nonwater-soluble protein found in the nails, hair, and the outermost layer of skin. Human hair is made up largely of keratin.

KERATOCONUS. An eye condition in which the central part of the cornea bulges outward, interfering with normal vision. Usually both eyes are affected.

KERNICTERUS. A potentially lethal disease of newborns caused by excessive accumulation of the bile pigment bilirubin in tissues of the central nervous system.

KETOACIDOSIS. Usually caused by uncontrolled type I diabetes, when the body isn't able to use glucose for energy. As an alternate source of energy, fat cells are broken down, producing ketones, toxic compounds that make the blood acidic. Symptoms of ketoacidosis include excessive thirst and urination, abdominal pain, vomiting, rapid breathing, extreme tiredness, and drowsiness.

KETONES. Poisonous acidic chemicals produced by the body when fat instead of glucose is burned for energy. Breakdown of fat occurs when not enough insulin is present to channel glucose into body cells.

KIESSELBACH'S PLEXUS. The mass of blood vessels on either side of the septum.

KILLER BEES. Hybrids of African bees accidentally introduced into the wild in South and North America in 1956 and first reported in Texas in 1990. They were first imported by Brazilian scientists attempting to create a new hybrid bee to improve honey production.

KINETIC ENERGY. The energy that the body has because of its motion.

KLEINE-LEVIN SYNDROME. A disorder that occurs primarily in young males, three or four times a year. The syndrome is marked by episodes of hypersomnia, hypersexual behavior, and excessive eating.

KLEPTOMANIA. An impulse control disorder in which one steals objects that are of little or no value.

KLUMPKE'S PALSY OR PARALYSIS. A condition caused by an injury to the lower brachial plexus, involving the cervical nerves C7 and C8, and sometimes the thoracic nerve T1, causing weakness or paralysis in the hands and fingers.

KOPLIK'S SPOTS. Tiny spots occurring inside the mouth, especially on the inside of the cheek. These spots consist of minuscule white dots (like grains of salt or sand) set onto a reddened bump and are characteristic of measles.

KYPHOSCOLIOSIS. Abnormal front-to-back and side-to-side curvature of the spine.

KYPHOSIS. An extreme, abnormal outward curvature of the spine, with a hump at the upper back.

L

LABYRINTH. The bony cavity of the inner ear.

LACERATION. A cut or separation of skin or other tissue by a tremendous force, producing irregular edges. Also called a tear.

LACTASE. The enzyme produced by cells that line the small intestine that allows the body to break down lactose.

LACTOBACILLUS ACIDOPHILUS. Commonly known as acidophilus, a bacteria found in yogurt that changes the balance of the bacteria in the intestine in a beneficial way.

LACTOBACILLUS BIFIDUS. A beneficial bacteria in breast milk that interferes with the growth of pathogenic bacteria in the gastrointestinal tracts of babies, reducing the incidence of diarrhea. *Lactobacillus bifidus* can be added to infant formulas to help control diarrhea.

LACTOGENESIS. The initiation of milk secretion.

LACTO-OVO VEGETARIAN. People who do not eat meat, but do include dairy products and eggs in their diets.

LACTOSE. A sugar found in milk and milk products.

LACTOSE INTOLERANCE. An inability to properly digest the lactose found in milk and dairy products.

LAMBDOIDAL SUTURE. The suture between the two parietal bones and the occipital bone in the skull.

LANUGO. A soft, downy body hair that covers a normal fetus beginning in the fifth month and usually shed by the ninth month. Also refers to the fine, soft hair that develops on the chest and arms of anorexic women. Also called vellus hair.

LAPAROSCOPIC SURGERY. Minimally invasive surgery in which a camera and surgical instruments are inserted through a small incision.

LAPAROSCOPY. A surgical procedure in which a small incision is made, usually in the navel, through which a viewing tube (laparoscope) is inserted. This allows the doctor to examine abdominal and pelvic organs. Other small incisions can be mad to insert instruments to perform procedures. Laparoscopy is done to diagnose conditions or to perform certain types of surgeries.

LARYNGOMALACIA. A birth defect that causes the tissues around the larynx to partially collapse and narrow the air passageway, causing noisy breathing.

LARYNGOSCOPE. An endoscope that is used to examine the interior of the larynx.

LARYNGOSPASM. Spasmodic closure of the larynx.

LARYNX. Also known as the voice box, the larynx is the part of the airway that lies between the pharynx and the trachea. It is composed of cartilage that contains the apparatus for voice production–the vocal cords and the muscles and ligaments that move the cords.

LATCHKEY CHILD. A child who must spend part of the after-school day at home without supervision while the parents are at work. The name comes from the fact that such children are given a house or apartment key so that they can let themselves in when they get home from school.

LATENT VIRUS. A nonactive virus that is in a dormant state within a cell. The herpes virus is latent in the nervous system.

LEARNING DISABILITIES. An impairment of the cognitive processes of understanding and using spoken and written language that results in difficulties with one or more academic skill sets (e.g., reading, writing, mathematics).

LEARNING DISORDERS. Academic difficulties experienced by children and adults of average to above-average intelligence that involve reading, writing, and/or mathematics, and which significantly interfere with academic achievement or daily living.

LEGUMES. A family of plants, including beans, peas, and lentils, that bear edible seeds in pods. These seeds are high in protein, fiber, and other nutrients.

LENNOX-GASTAUT SYNDROME. A severe form of epilepsy that is characterized by the onset in early childhood of frequent seizures of multiple types and by developmental delay.

LENS. The transparent, elastic, curved structure behind the iris (colored part of the eye) that helps focus light on the retina. Also refers to any device that bends light waves.

LEPTIN. A protein hormone that affects feeding behavior and hunger in humans. As of 2004 it was thought that obesity in humans may result in part from insensitivity to leptin.

LESION. A disruption of the normal structure and function of a tissue by an injury or disease process. Wounds, sores, rashes, and boils are all lesions.

LEUKEMIA. A cancer of the blood-forming organs (bone marrow and lymph system) characterized by an abnormal increase in the number of white blood cells in the tissues. There are many types of leukemias and they are classified according to the type of white blood cell involved.

LEUKOCYTE. A white blood cell that defends the body against invading viruses, bacteria, and cancer cells. There are five types of leukocytes—neutrophils, basophils, eosinophils, lymphocytes, and monocytes.

LEUKOCYTOSIS. An increased level of white cells in the blood. Leukocytosis is a common reaction to infections.

LEUKOTRIENE ANTAGONIST. An agent or class of drugs which exerts an action opposite to that of leukotrienes.

LEUKOTRIENES. Substances that are produced by white blood cells in response to antigens and contribute to inflammatory and asthmatic reactions.

LEVODOPA (L-DOPA). A substance used in the treatment of Parkinson's disease. Levodopa can cross the blood-brain barrier that protects the brain. Once in the brain, it is converted to dopamine and thus can replace the dopamine lost in Parkinson's disease.

LICHEN PLANUS. A noncancerous, chronic itchy skin disease that causes small, flat purple plaques on wrists, forearm, ankles.

LICHENIFICATION. Thickening of the outer layer of skin cells caused by prolonged scratching or rubbing and resulting in a leathery or bark-like appearance of the skin.

LIGAMENT. A type of tough, fibrous tissue that connects bones or cartilage and provides support and strength to joints.

LIGAND. Any type of small molecule that binds to a larger molecule. Hyper-IgM syndrome is caused by a lack of a ligand known as CD40 on the surfaces of the T cells in the child's blood.

LIMB-GIRDLE MUSCULAR DYSTROPHY (LGMD). A form of muscular dystrophy that begins in late childhood to early adulthood and affects both men and women. It causes weakness in the muscles around the hips and shoulders.

LIMBIC SYSTEM. A group of structures in the brain that includes the hypothalamus, amygdala, olfactory bulbs, and hippocampus. The limbic system plays an important part in regulation of human moods and emotions. Many psychiatric disorders are related to malfunctioning of the limbic system.

LIMITED ENGLISH PROFICIENT (LEP). Used to identify children who have insufficient English to succeed in English-only classrooms; also called English language learner (ELL).

LINCOLN-OSERETSKY MOTOR DEVELOPMENT SCALE. A test that assesses the development of motor skills.

LINDANE. A benzene compound that is used to kill body and pubic lice. Lindane is absorbed into the louse's central nervous system, causing seizures and death.

LIPIDS. Organic compounds not soluble in water, but soluble in fat solvents such as alcohol. Lipids are stored in the body as energy reserves and are also important components of cell membranes. Commonly known as fats.

LIPOSUCTION. A surgical technique for removing fat from under the skin by vacuum suctioning.

LISTERIA. An uncommon food-borne, life-threatening pathogen that can cause perinatal infection, which is associated with a high rate of fetal loss (including full-term stillbirths) and serious neonatal disease.

LITHIUM. A medication prescribed to treat the manic (excited) phases of bipolar disorder.

LIVER BIOPSY. A surgical procedure where a small piece of the liver is removed for examination. A needle or narrow tube may be inserted either directly through the skin and muscle or through a small incision and passed into the liver for collection of a sample of liver tissue.

LOCAL ANESTHESIA. Pain-relieving medication used to numb an area while the patient remains awake. Also see general anesthesia.

LOCOMOTION. The ability to move from one place to another.

LOEFFLER'S MEDIUM. A special substance used to grow diphtheria bacilli to confirm the diagnosis.

LOWER ESOPHAGEAL SPHINCTER (LES). A muscle located at the base of the esophagus which keeps the stomach contents from coming back into the esophagus.

LUMBAR PUNCTURE. A procedure in which the doctor inserts a small needle into the spinal cavity in the lower back to withdraw spinal fluid for testing. Also known as a spinal tap.

LUMEN. The inner cavity or canal of a tube-shaped organ, such as the bowel.

LUTEINIZING HORMONE. A hormone secreted by the pituitary gland that regulates the menstrual cycle and triggers ovulation in females. In males it stimulates the testes to produce testosterone.

LYME BORRELIOSIS. Another name for Lyme disease.

LYME DISEASE. An acute, recurrent, inflammatory disease involving one or a few joints, and transmitted by the bite of ticks carrying the spiral-shaped bacterium *Borrelia burgdorferi*. The condition was originally described in the community of Lyme, Connecticut, but has also been reported in other parts of the United States and other countries. Knees and other large joints are most commonly involved with local inflammation and swelling.

LYMPH FLUID. Clear, colorless fluid found in lymph vessels and nodes. The lymph nodes contain organisms that destroy bacteria and other disease causing organisms (also called pathogens).

LYMPH NODES. Small, bean-shaped collections of tissue located throughout the lymphatic system. They produce cells and proteins that fight infection and filter lymph. Nodes are sometimes called lymph glands.

LYMPHADENOPATHY. A disorder characterized by local or generalized enlargement of the lymph nodes or lymphatic vessels.

LYMPHANGIOMA. A benign skin tumor composed of abnormal lymph vessels.

LYMPHANGITIS. Inflammation of the lymphatic vessels. It often occurs together with lymphadenitis (inflammation of the lymph nodes).

LYMPHATIC VESSELS. Part of the lymphatic system, these vessels connect lymph capillaries with the lymph nodes. They carry lymph, a thin, watery fluid resembling blood plasma and containing white blood cells. Also called lymphatic channels.

LYMPHOCYTE. A type of white blood cell that participates in the immune response. The two main groups are the B cells that have antibody molecules on their surface and T cells that destroy antigens.

LYMPHOCYTIC LEUKEMIA. An acute form of childhood leukemia characterized by the development of abnormal cells in the bone marrow.

LYMPHOMA. A diverse group of cancers of the lymphatic system characterized by abnormal growth of lymphatic cells. Two general types are commonly recognized—Hodgkin's disease and non-Hodgkin's lymphoma.

LYSINE. A crystalline basic amino acid essential to nutrition.

LYSOSOME. A membrane-enclosed compartment in cells, containing many hydrolytic enzymes, where large molecules and cellular components are broken down.

M

MACROCYTIC. A descriptive term applied to a larger than normal red blood cell.

MACROPHAGE. A large white blood cell that engulfs and digests foreign invaders, such as bacteria and viruses, in an attempt to stop them from causing disease within the body.

MACULE. A flat, discolored area on the skin.

MAGNETIC RESONANCE IMAGING (MRI). An imaging technique that uses a large circular magnet and radio waves to generate signals from atoms in the body. These signals are used to construct detailed images of internal body structures and organs, including the brain.

MAJOR DEPRESSIVE DISORDER. A mood disorder characterized by profound feelings of sadness or despair.

MALABSORPTION. The inability of the digestive tract to absorb all the nutrients from food due to some malfunction or disability.

MALADAPTIVE BEHAVIOR. Undesirable and socially unacceptable behavior that interferes with the acquisition of desired skills or knowledge and with the performance of everyday activities.

MALAISE. The medical term for a general condition of unease, discomfort, or weakness.

MALATHION. An insecticide that can be used in 1% powdered form to disinfect the clothes of patients with body lice.

MALFORMATION. An irregular or abnormal formation or structure.

MALIGNANT. Cells that have been altered such that they have lost normal control mechanisms and are capable of local invasion and spread to other areas of the body. Often used to describe a cancer.

MALIGNANT HYPERTHERMIA. A type of reaction (probably with a genetic origin) that can occur during general anesthesia and in which the patient experiences a high fever, muscle rigidity, and irregular heart rate and blood pressure.

MALIGNANT MELANOMA. The most serious of the three types of skin cancer, malignant melanoma arises from the melanocytes, the skin cells that produce the pigment melanin.

MALIGNANT TUMOR. An abnormal proliferation of cells that can spread to other sites.

MALINGERING. Pretending to be sick in order to be relieved of an unwanted duty or obtain some other obvious benefit.

MALLEABILITY. A term that refers to the adaptability of human temperament; the extent to which it can be reshaped.

MALOCCLUSION. The misalignment of opposing teeth in the upper and lower jaws.

MAMMARY. Relating to the breast.

MANDIBLE. The lower jaw, a U-shaped bone attached to the skull at the temporomandibular joints.

MANIA. An elevated or euphoric mood or irritable state that is characteristic of bipolar I disorder. This state is characterized by mental and physical hyperactivity, disorganization of behavior, and inappropriate elevation of mood.

MANIC DEPRESSION. A psychiatric disorder characterized by extreme mood swings, ranging between episodes of acute euphoria (mania) and severe depression; also called bipolar depression.

MANIC EPISODE. A distinct period of abnormally and persistently elevated, expansive, or irritable mood, lasting at least one week, characterized by inflated sense of self-importance, decreased need for sleep, extreme

talkativeness, racing thoughts, and excessive participation in pleasure-seeking activities.

MANIPULATION. Moving muscles or connective tissue to enhance function, ease tension, and reduce pain in those tissues as well as other beneficial effects.

MANOMETRY. A technique for measuring changes in pressure.

MANTOUX TEST. A tuberculin skin test. Also called the PPD (purified protein derivative) test.

MASK. An expressionless look, caused by reduced movements of the face.

MAST CELLS. A type of immune system cell that is found in the lining of the nasal passages and eyelids. It displays a type of antibody called immunoglobulin type E (IgE) on its cell surface and participates in the allergic response by releasing histamine from intracellular granules.

MASTOID BONE. The prominent bone behind the ear that projects from the temporal bone of the skull.

MASTOIDITIS. An inflammation of the bone behind the ear (the mastoid bone) caused by an infection spreading from the middle ear to the cavity in the mastoid bone.

MATERNAL SERUM ANALYTE SCREENING. A medical procedure in which a pregnant woman's blood is drawn and analyzed for the levels of certain hormones and proteins. These levels can indicate whether there may be an abnormality in the unborn child. This test is not a definitive indicator of a problem and is followed by more specific testing such as amniocentesis or chorionic villus sampling.

MATERNAL UNIPARENTAL DISOMY. A chromosome abnormality in which both chromosomes in a pair are inherited from one's mother.

MATURATION. The process by which stem cells transform from immature cells without a specific function into a particular type of blood cell with defined functions.

MATURATION DELAY. Developmental language delay; a language delay caused by the slow maturation of speech centers in the brain; often causes late talking.

MATURITY. A state of full development or completed growth.

MAXILLA. The bone of the upper jaw which serves as a foundation of the face and supports the orbits.

MEAN CORPUSCULAR HEMOGLOBIN CONCENTRATION (MCHC). A measurement of the average concentration of hemoglobin in a red blood cell.

MEAN CORPUSCULAR VOLUME (MCV). A measurement of the average volume of a red blood cell.

MECKEL'S DIVERTICULUM. A congenital abnormality of the digestive tract consisting of a small pouch off the wall of the small bowel that was not reabsorbed before birth. A Meckel's diverticulum increases the risk that a foreign object in the digestive tract will get trapped or stuck in the small intestine and cause problems.

MECONIUM. A greenish fecal material that forms the first bowel movement of an infant.

MECONIUM ASPIRATION SYNDROME. Breathing in of meconium (a newborn's first stool) by a fetus or newborn, which can block air passages and interfere with lung expansion.

MEDICAID. A program jointly funded by state and federal governments that reimburses hospitals and physicians for the care of individuals who cannot pay for their own medical expenses. These individuals may be in low-income households or may have chronic disabilities.

MEDITATION. A practice of concentrated focus upon a sound, object, visualization, the breath, movement, or attention itself in order to increase awareness of the present moment, reduce stress, promote relaxation, and enhance personal and spiritual growth.

MEDULLARY THYROID CANCER. A slow-growing tumor associated with multiple endocrine neoplasia syndromes.

MEGACOLON. Abnormal dilation (enlargement) of the colon.

MEGALENCEPHALY. A condition in which the brain is abnormally large.

MEGALOBLAST. A large erythroblast (a red marrow cell that synthesizes hemoglobin).

MEGALOCEPHALY. An abnormally large head.

MELANIN. A pigment that creates hair, skin, and eye color. Melanin also protects the body by absorbing ultraviolet light.

MELANOMA. A tumor, usually of the skin.

MELENA. The passage of dark stools stained with blood pigments or with altered blood.

MEMBRANE OXYGENATOR. The artificial lung that adds oxygen and removes carbon dioxide.

MENARCHE. The first menstrual cycle in a girl's life.

MENINGES. The three-layer membranous covering of the brain and spinal cord, composed of the dura mater, arachnoid, and pia mater. It provides protection for the brain and spinal cord, as well as housing many blood

vessels and participating in the appropriate flow of cerebrospinal fluid.

MENINGITIS. An infection or inflammation of the membranes that cover the brain and spinal cord. It is usually caused by bacteria or a virus.

MENINGOENCEPHALITIS. Inflammation of the brain and its membranes; also called cerebromeningitis or encephalomeningitis.

MENKES DISEASE. A genetic disease caused by a mutation on the X chromosome and resulting in impaired transport of copper from the digestive tract. It was first identified in 1962.

MENSTRUATION. The periodic discharge from the vagina of blood and tissues from a nonpregnant uterus.

MENTAL RETARDATION. A condition where an individual has a lower-than-normal IQ, and thus is developmentally delayed.

METABOLIC. Refers to the chemical reactions in living organisms.

METABOLISM. The sum of all chemical reactions that occur in the body resulting in growth, transformation of foodstuffs into energy, waste elimination, and other bodily functions. These include processes that break down substances to yield energy and processes that build up other substances necessary for life.

METACOGNITION. Awareness of the process of cognition.

METALINGUISTIC SKILLS. The ability to analyze language and control internal language processing; important for reading development in children.

METASTASIS. A secondary tumor resulting from the spread of cancerous cells from the primary tumor to other parts of the body.

METHEMOGLOBIN. A compound formed from hemoglobin by oxidation of its iron component. Methemoglobin cannot carry oxygen.

METHIONINE. An amino acid that, when not metabolized properly, allows homocysteine to build up in the blood. Folic acid aids methionine metabolism.

METHOTREXATE. A drug that interferes with cell growth and is used to treat rheumatoid arthritis as well as various types of cancer. Side effects may include mouth sores, digestive upsets, skin rashes, and hair loss.

METHYLATION TESTING. DNA testing that detects if a gene is active or if it is imprinted.

METHYLPREDISOLONE. A steroid drug. Methylprednisolone administered within eight hours of acute spinal cord trauma is the first drug shown to improve recovery from spinal cord injury.

METOPIC SUTURE. Suture extending from the top of the head down the middle of the forehead to the nose.

MICROCEPHALY. An abnormally small head.

MICROCYTIC. A descriptive term applied to a smaller than normal red blood cell.

MICROFLORA. The bacterial population in the intestine.

MICRONUTRIENT. An organic compound such as vitamins or minerals essential in small amounts and necessary to the growth and health of humans and animals.

MICROORGANISM. An organism that is too small to be seen with the naked eye, such as a bacterium, virus, or fungus.

MIDDLE EAR. The cavity or space between the eardrum and the inner ear. It includes the eardrum, the three little bones (hammer, anvil, and stirrup) that transmit sound to the inner ear, and the eustachian tube, which connects the inner ear to the nasopharynx (the back of the nose).

MIDGET. An individual who is short statured but has normal body proportions. The term is considered to be offensive.

MIGRAINE. A throbbing headache that usually affects only one side of the head. Nausea, vomiting, increased sensitivity to light, and other symptoms often accompany a migraine.

MIND-BODY CONNECTION. Rather than relying on an understanding of the term ‘‘psychosomatic,‘‘ mind-body medicine acknowledges the influence of thinking and the cognitive process on the behavior of chemicals in the body, involving the mind in both creating the conditions for disease and helping to heal the effects of disease.

MINERALS. Inorganic chemical elements that are found in plants and animals and are essential for life. There are two types of minerals: major minerals, which the body requires in large amounts, and trace elements, which the body needs only in minute amounts.

MISBEHAVIOR. Behavior outside the norms of acceptance within the group.

MISCARRIAGE. Loss of the embryo or fetus and other products of pregnancy before the twentieth week. Often, early in a pregnancy, if the condition of the baby and/or the mother’s uterus are not compatible with sustaining life, the pregnancy stops, and the contents of the uterus

are expelled. For this reason, miscarriage is also referred to as spontaneous abortion.

MITE. An insect parasite belonging to the order Acarina. The organism that causes scabies is a mite.

MITOCHONDRIA. Spherical or rod-shaped structures of the cell. Mitochondria contain genetic material (DNA and RNA) and are responsible for converting food to energy.

MITOCHONDRIAL INHERITANCE. Inheritance associated with the mitochondrial genome which is inherited exclusively from the mother.

MITRAL VALVE PROLAPSE. A heart defect in which the mitral valve of the heart (which normally controls blood flow from the left atrium to the left ventricle) becomes floppy. Mitral valve prolapse may be detected as a heart murmur but there are usually no symptoms.

MITRAL VALVE STENOSIS. Narrowing of the mitral valve.

MIXED MANIA. A mental state in which symptoms of both depression and mania occur simultaneously. Also called mixed state.

MODELING. A type of teaching method used in social skills training. Therapists who use this method may offer positive and negative examples of the behaviors that make up a social skill.

MOLARS. The teeth behind the primary canines or the permanent premolars, with large crowns and broad chewing surfaces for grinding food.

MONOAMINE OXIDASE (MAO) INHIBITORS. A type of antidepressant that works by blocking the action of a chemical substance known as monoamine oxidase in the nervous system.

MONOCHORIONIC TWINS. Twins that share a single placenta.

MONONUCLEOSIS. An infection, caused by the Epstein-Barr virus, that causes swelling of lymph nodes, spleen, and liver, usually accompanied by extremely sore throat, fever, headache, and intense long-lasting fatigue. Also called infectious mononucleosis.

MONOZYGOTIC. From one zygote, as in identical twins. The zygote is the first cell formed by the union of sperm and egg.

MORBIDITY. A disease or abnormality. In statistics it also refers to the rate at which a disease or abnormality occurs.

MORO REFLEX. A startle response in a newborn, characterized by spreading the arms with the palms up and fingers flexed; the reflex usually disappears by two months of age.

MORPHINE. The principal alkaloid derived from the opium poppy for use as a pain reliever and sedative. In its purified form, it is a white, bitter-tasting crystalline powder.

MORPHOLOGY. Literally, the study of form. In medicine, morphology refers to size, shape, and structure rather than function.

MOSAIC. A term referring to a genetic situation in which an individual's cells do not have the exact same composition of chromosomes. In Down syndrome, this may mean that some of the individual's cells have a normal 46 chromosomes, while other cells have an abnormal 47 chromosomes.

MOSAICISM. A genetic condition resulting from a mutation, crossing over, or nondisjunction of chromosomes during cell division, causing a variation in the number of chromosomes in the cells.

MOTILITY. The movement or capacity for movement of an organism or body organ. Indigestion is sometimes caused by abnormal patterns in the motility of the stomach.

MOTOR COORDINATION (MC). Related to movement of parts of the body, particularly the use of the hands and coordination of eye-hand motion.

MOTOR NEURON. A nerve cell that specifically controls and stimulates voluntary muscles.

MOTOR SKILLS. Controlled movements of muscle groups. Fine motor skills involve tasks that require dexterity of small muscles, such as buttoning a shirt. Tasks such as walking or throwing a ball involve the use of gross motor skills.

MOTTLING. Fluorosis; spotting on the teeth due to excess fluoride as the tooth enamel is forming.

MOURN. To express grief or sorrow, usually for a death.

MUCIN. A protein in saliva that combines with sugars in the mouth to form plaque.

MUCOCILIARY ESCALATOR. The coordinated action of tiny projections on the surfaces of cells lining the respiratory tract, which moves mucus up and out of the lungs.

MUCOCUTANEOUS LYMPH NODE SYNDROME (MLNS). Another name for Kawasaki syndrome. The name comes from the key symptoms of the disease, which involve the mucous membranes of the mouth and

throat, the skin, and the lymph nodes. MLNS is a potentially fatal inflammatory disease of unknown cause.

MUCOLYTIC. An agent that dissolves or destroys mucin, the chief component of mucus.

MUCOPOLYSACCHARIDE. A complex molecule made of smaller sugar molecules strung together to form a chain. It is found in mucous secretions and intercellular spaces.

MUCOSAL. Refers to the mucous membrane.

MUCUS. The thick fluid produced by the mucous membranes that line many body cavities and structures. It contains mucin, white blood cells, water, inorganic salts, and shed cells, and it serve to lubricate body parts and to trap particles of dirt or other contaminants.

MULTICULTURAL EDUCATION. A social or educational theory that encourages interest in many cultures within a society rather than in only a mainstream culture.

MULTIFACTORIAL. Describes a disease that is the product of the interaction of multiple genetic and environmental factors.

MULTIFOCAL. Having many focal points. When referring to a disease, it means that damage caused by the disease occurs at multiple sites. When referring to a cancer, it means that more than one tumor is present.

MULTIPLE RETINAL HEMORRHAGES. Bleeding in the back of the eye.

MULTIPLE SCLEROSIS. A progressive, autoimmune disease of the central nervous system characterized by damage to the myelin sheath that covers nerves. The disease, which causes progressive paralysis, is marked by periods of exacerbation and remission.

MULTI-TASKING. Performing multiple duties or taking on multiple responsibilities and roles simultaneously.

MUNCHAUSEN SYNDROME. A factitious disorder in which a patient intentionally acts physically ill without obvious benefit.

MUSCLE SPASM. Localized muscle contraction that occurs when the brain signals the muscle to contract.

MUSCLE TONE. Also termed tonus; the normal state of balanced tension in the tissues of the body, especially the muscles.

MUSCLE WEAKNESS. Reduction in the strength of one or more muscles.

MUSCULAR DYSTROPHY. A group of inherited diseases characterized by progressive wasting of the muscles.

MUTATION. A permanent change in the genetic material that may alter a trait or characteristic of an individual, or manifest as disease. This change can be transmitted to offspring.

MYCTOPHOBIA. Fear of darkness.

MYELIN. A fatty sheath surrounding nerves throughout the body that helps them conduct impulses more quickly.

MYELOGRAM. An x-ray image of the spinal cord, spinal canal, and nerve roots taken with the aid of a contrast dye.

MYOCARDITIS. Inflammation of the heart muscle (myocardium).

MYOCLONUS. Involuntary contractions of a muscle or an interrelated group of muscles. Also known as myoclonic seizures.

MYOPATHY. Any abnormal condition or disese of muscle tissue, characterized by muscle weakness and wasting.

MYOSITIS. Inflammation of the muscle.

MYOTONIC DYSTROPHY. A form of muscular dystrophy, also known as Steinert's disease, that affects both men and women. It is characterized by delay in the ability to relax muscles after forceful contraction (myotonia) and wasting of muscles, as well as other abnormalities.

MYRINGOTOMY. A surgical procedure in which an incision is made in the ear drum to allow fluid or pus to escape from the middle ear.

MYXEDEMA. Severe hypothyroidism, characterized by swelling of the face, hands, and feet, an enlarged tongue, horseness, and physical and mental sluggishness.

N

NAIL BED. The layer of tissue underneath the nail.

NARCOLEPSY. A life-long sleep disorder marked by four symptoms: sudden brief sleep attacks, cataplexy (a sudden loss of muscle tone usually lasting up to 30 minutes), temporary paralysis, and hallucinations. The hallucinations are associated with falling asleep or the transition from sleeping to waking.

NARCOTIC. A drug derived from opium or compounds similar to opium. Such drugs are potent pain relievers and can affect mood and behavior. Long-term use of narcotics can lead to dependence and tolerance. Also known as a narcotic analgesic.

NASAL ENDOSCOPY. A procedure that involves inserting a tiny camera into the nose in order to look at blood vessels and nasal structures.

NASAL SEPTUM. The partition that separates the nostrils.

NASOGASTRIC TUBE. A long, flexible tube inserted through the nasal passages, down the throat, and into the stomach.

NASOPHARYNX. One of the three regions of the pharynx, the nasopharynx is the region behind the nasal cavity.

NEBULIZER. A device that turns liquid forms of medication into a fine spray that can be inhaled.

NECROSIS. Localized tissue death due to disease or injury, such as a lack of oxygen supply to the tissues.

NECROTIZING. Causing the death of a specific area of tissue. Human bites frequently cause necrotizing infections.

NECROTIZING ENTEROCOLITIS. A serious bacterial infection of the intestine that occurs primarily in sick or premature newborn infants. It can cause death of intestinal tissue (necrosis) and may progress to blood poisoning (septicemia).

NEONATAL. Refers to the first 28 days of an infant's life.

NEONATE. A newborn infant, from birth until 28 days of age.

NEONATOLOGIST. A physician (pediatrician) who has special training in the care of newborn infants.

NEOPLASM. An abnormal formation of new tissue. A neoplasm may be malignant or benign.

NEPHROLOGIST. A physician who specializes in treating diseases of the kidney.

NERVE CONDITION VELOCITY (NCV). Technique for studying nerve or muscle disorders, measuring the speed at which nerves transmit signals.

NERVE GROWTH FACTOR. A protein resembling insulin that affects the growth and maintenance of nerve cells

NERVOUS SYSTEM. The system that transmits information, in the form of electrochemical impulses, throughout the body for the purpose of activation, coordination, and control of bodily functions. It is comprised of the brain, spinal cord, and nerves.

NERVOUS TIC. A repetitive, involuntary action, such as the twitching of a muscle or repeated blinking.

NEURODEGENERATIVE DISEASE. A disease in which the nervous system progressively and irreversibly deteriorates.

NEURODERMATITIS. An itchy skin disease (also called lichen simplex chronicus) found in nervous, anxious people.

NEUROFIBROMA. A soft tumor usually located on a nerve.

NEUROFIBROMATOSIS. A progressive genetic condition often including multiple café-au-lait spots, multiple raised nodules on the skin (neurofibromas), developmental delays, slightly larger head size, and freckles in the armpits, groin, and iris. Also known as von Recklinghausen's disease.

NEUROIMAGING. The use of x-ray studies and magnetic resonance imaging (MRI) to detect abnormalities or trace pathways of nerve activity in the central nervous system.

NEUROLEPTIC DRUG. Another name for the older type of antipsychotic medications, such as haloperidol and chlorpromazine, prescribed to treat psychotic conditions.

NEUROLOGICAL. Relating to the brain and central nervous system.

NEUROLOGICAL DISORDERS. Pathological conditions relating to the brain and/or nervous system.

NEUROLOGIST. A doctor who specializes in disorders of the nervous system, including the brain, spinal cord, and nerves.

NEUROLOGY. The study of nerves.

NEUROLYSIS. The destruction of nerve tissue or removal of scar tissue surrounding a nerve.

NEUROMA. Scar tissue that forms around a nerve; a tumor derived from nerve tissue.

NEURONS. Any of the conducting cells of the nervous system that transmit signals.

NEUROPATHY. A disease or abnormality of the peripheral nerves (the nerves outside the brain and spinal cord). Major symptoms include weakness, numbness, paralysis, or pain in the affected area.

NEUROPSYCHOLOGICAL. Referring to the interaction between the nervous system and cognitive function, the influence of one function on the other.

NEUROPSYCHOLOGICAL TESTING. Tests used to evaluate patients who have experienced a traumatic brain injury, brain damage, or organic neurological problems (e.g., dementia). It may also be used to evaluate

the progress of a patient who has undergone treatment or rehabilitation for a neurological injury or illness.

NEUROPSYCHOLOGIST. A clinical psychologist who specializes in assessing psychological status caused by a brain disorder.

NEUROSURGEON. Physician who performs surgery on the nervous system.

NEUROTOXIN. A poison that acts directly on the central nervous system.

NEUROTRANSMITTER. A chemical messenger that transmits an impulse from one nerve cell to the next.

NEUTROPENIA. A condition in which the number of neutrophils, a type of white blood cell (leukocyte) is abnormally low.

NEUTROPHIL. The primary type of white blood cell involved in inflammation. Neutrophils are a type of granulocyte, also known as a polymorphonuclear leukocyte. They increase in response to bacterial infection and remove and kill bacteria by phagocytosis.

NEVUS (PLURAL, NEVI). The medical term for any anomaly of the skin that is present at birth, including moles and birthmarks.

NICOTINE. A colorless, oily chemical found in tobacco that makes people physically dependent on smoking. It is poisonous in large doses.

NICOTINE REPLACEMENT THERAPY. A method of weaning a smoker away from both nicotine and the oral fixation that accompanies a smoking habit by giving the smoker smaller and smaller doses of nicotine in the form of a patch or gum.

NITRATE. A food additive, commonly found in processed meats, that may be a headache trigger for some people.

NITS. The eggs produced by head or pubic lice, usually grayish white in color and visible at the base of hair shafts.

NOCICEPTOR. A nerve cell that is capable of sensing pain and transmitting a pain signal.

NOCTURNAL ENURESIS. Involuntary discharge of urine during the night.

NOCTURNAL LEG CRAMPS. Cramps that may be related to exertion and awaken a person during sleep.

NOCTURNAL MYOCLONUS. A disorder in which the patient is awakened repeatedly during the night by cramps or twitches in the calf muscles. Also sometimes called periodic limb movement disorder.

NONCOMEDOGENIC. A substance that does not contribute to the formation of blackheads or pimples on the skin.

NON-CONDUCTING MATERIALS. Also called insulators, materials through which electric current does not propagate. Examples are ceramics, rubber, wood.

NON-CUSTODIAL PARENT. A parent who does not have legal custody of a child.

NONDISJUNCTION. An event that takes place during cell division in which a chromosome pair does not separate as it should. The result is an abnormal number of chromosomes in the daughter cells produced by that cell division.

NONPHARMACOLOGICAL. Referring to therapy that does not involve drugs.

NONPRODUCTIVE. A cough in which no mucus is coughed up, also called dry cough.

NON-RAPID EYE MOVEMENT (NREM) SLEEP. A type of sleep that differs from rapid eye movement (REM) sleep. The four stages of NREM sleep account for 75–80% of total sleeping time.

NONRHYTHMIC. Having uneven sleep and eating patterns.

NONSTEROIDAL ANTI-INFLAMMATORY DRUGS (NSAIDS). A group of drugs, including aspirin, ibuprofen, and naproxen, that are taken to reduce fever and inflammation and to relieve pain. They work primarily by interfering with the formation of prostaglandins, enzymes implicated in pain and inflammation.

NOREPINEPHRINE. A hormone secreted by certain nerve endings of the sympathetic nervous system, and by the medulla (center) of the adrenal glands. Its primary function is to help maintain a constant blood pressure by stimulating certain blood vessels to constrict when the blood pressure falls below normal.

NORMOCHROMIC. A descriptive term applied to a red blood cell with a normal concentration of hemoglobin.

NORMOCYTIC. A descriptive term applied to a red blood cell of normal size.

NORM-REFERENCED TEST. A test that measures the performance of a student against the performance of a group of other individuals.

NORMS. A fixed or ideal standard; a normative or mean score for a particular age group.

NOSOCOMIAL INFECTION. An infection acquired in a hospital setting.

NUCHAL TRANSLUCENCY. A pocket of fluid at the back of an embryo's neck, visible via ultrasound. When this pocket of fluid is thickened, it may indicate that the infant will be born with a congenital cardiovascular defect.

NUCLEAR FAMILY. The basic family unit, consisting of a father, a mother, and their biological children.

NURSEMAID'S ELBOW. An injury to the ligament (strong band of tissue) that keeps the two bones of the forearm in the correct place.

NURSING UNIT. The floor or section of the hospital where patient rooms are located.

NUTRIENT. Substances in food that supply the body with the elements needed for metabolism. Examples of nutrients are vitamins, minerals, carbohydrates, fats, and proteins.

NUTRITION THERAPY. Nutrition assessment, counseling, and education, usually provided by registered dietitians.

NYSTAGMUS. An involuntary, rhythmic movement of the eyes.

O

OBESITY. An abnormal accumulation of body fat, usually 20% or more over an individual's ideal body weight.

OBSERVATION. Infants and children watch an object, although not actively engaged in it, as in watching a mobile.

OBSESSION. A persistent image, idea, or desire that dominates a person's thoughts or feelings.

OBSESSIVE-COMPULSIVE DISORDER. An anxiety disorder marked by the recurrence of intrusive or disturbing thoughts, impulses, images, or ideas (obsessions) accompanied by repeated attempts to supress these thoughts through the performance of certain irrational and ritualistic behaviors or mental acts (compulsions).

OCCLUSION. The way upper and lower teeth fit together during biting and chewing. Also refers to the blockage of some area or channel of the body.

OCCLUSIONAL. Referring to a type of injury caused by the closing of the teeth on a finger or other body part. Occlusional injuries are also called chomping injuries.

OCCULSION THERAPY. A type of treatment for amblyopia in which the good eye is patched for a period of time, thus forcing the use of the weaker eye.

OCCUPATIONAL THERAPIST. A healthcare provider who specializes in adapting the physical environment to meet a patient's needs. An occupational therapist also assists patients and caregivers with activities of daily living and provide instructions on wheelchair use or other adaptive equipment.

OCULOPHARYNGEAL MUSCULAR DYSTROPHY. A type of muscular dystrophy that affects adults of both sexes, causing weakness in the eye muscles and throat.

ODYNOPHAGIA. Pain in swallowing.

OFF-LABEL USE. Prescribing a drug for a population (e.g., pediatric) or condition for which it was not originally approved by the U.S. FDA. For example, sulfonylurea drugs are not FDA approved for use in children with type 2 diabetes due to a lack of clinical studies in pediatric populations, but a physician may prescribe them in an off-label use of the drug.

OLIGOHYDRAMNIOS. A reduced amount of amniotic fluid, whose causes include non-functioning kidneys and premature rupture of membranes. Without amniotic fluid to breathe, a baby will have underdeveloped and immature lungs.

OMPHALOCELE. A birth defect in which the bowel and sometimes the liver protrudes through an opening in the baby's abdomen near the umbilical cord.

ONCOLOGIST. A physician specializing in the diagnosis and treatment of cancer

OOCYST. A developmental stage of certain parasitic organisms, including those responsible for malaria and toxoplasmosis, in which the zygote of the organism is enclosed in a cyst.

OPEN BITE. A malocclusion in which some teeth do not meet the opposing teeth.

OPHIDIAPHOBIA. Fear of snakes.

OPHTHALMOLOGIST. A physician who specializes in the anatomy and physiology of the eyes and in the diagnosis and treatment of eye diseases and disorders.

OPIUM LATEX. The milky juice or sap of the opium poppy, used to produce morphine.

OPPORTUNISTIC INFECTION. An infection that is normally mild in a healthy individual, but which takes advantage of an ill person's weakened immune system to move into the body, grow, spread, and cause serious illness.

OPPOSITIONAL DEFIANT DISORDER. An emotional and behavioral disorder of children and adolescents characterized by hostile, deliberately argumentative, and defiant behavior towards authority figures that lasts for longer than six months.

OPTIC NERVE. A bundle of nerve fibers that carries visual messages from the retina in the form of electrical signals to the brain.

ORAL LESIONS. A single infected sore in the skin around the mouth or mucous membrane inside of the oral cavity.

ORAL REHYDRATION SOLUTION (ORS). A liquid preparation of electrolytes and glucose developed by the World Health Organization that can decrease fluid loss in persons with diarrhea. Originally developed to be prepared with materials available in the home, commercial preparations have recently come into use.

ORBIT. The eye socket which contains the eyeball, muscles, nerves, and blood vessels that serve the eye.

ORCHIOPEXY. A surgical procedure that places an undescended testicle in the scrotum and/or attaches a testicle to the scrotum.

ORCHITIS. Inflammation of one or both testes, accompanied by swelling, pain, fever, and a sensation of heaviness in the affected area.

OREXIN. Another name for hypocretin, a chemical secreted in the hypothalmus that regulates the sleep/wake cycle. Narcolepsy is sometimes described as an orexin deficiency syndrome.

ORGANELLE. A specialized structure within a cell, which is separated from the rest of the cell by a membrane composed of lipids and proteins, where chemical and metabolic functions take place.

ORGANIC CAUSES. Underlying medical or physical disorders causing failure to thrive.

ORGANISM. A single, independent unit of life, such as a bacterium, a plant, or an animal.

ORGANOGENESIS. The formation of organs during development.

ORGASM. Another word for sexual climax. In the male, orgasm is usually accompanied by ejaculation but may be experienced as distinct from ejaculation.

OROPHARYNX. One of the three regions of the pharynx, the oropharynx is the region behind the mouth.

ORTHOGNATIC SURGERY. Surgery to alter the relationships of the teeth and/or supporting bones, usually in conjunction with orthodontic treatment.

ORTHOPEDIST. A doctor specializing in treatment of the musculoskeletal system.

ORTHOSIS. An external device, such as a splint or a brace, that prevents or assists movement.

OSMOLALITY. The concentration of osmolar particles in the blood (or other solutions) that can help determine if the body is dehydrated.

OSSICLES. The three small bones of the middle ear: the malleus (hammer), the incus (anvil) and the stapes (stirrup). These bones help carry sound from the eardrum to the inner ear.

OSTEOARTHRITIS. A noninflammatory type of arthritis, usually occurring in older people, characterized by degeneration of cartilage, enlargement of the margins of the bones, and changes in the membranes in the joints. Also called degenerative arthritis.

OSTEOCLAST. A large, multinuclear cell involved in the physiological destruction and absorption of bone.

OSTEOGENESIS IMPERFECTA. An inherited disorder of the connective tissues that involves multiple symptoms, including weakened bones that break easily.

OSTEOLOGIST. A doctor who specializes in the skeletal system.

OSTEOMALACIA. A bone disease that occurs in adults due to a prolonged period of vitamin D deficiency. It is characterized by softening of the bone and is sometimes referred to as adult rickets.

OSTEOMYELITIS. An infection of the bone and bone marrow, usually caused by bacteria.

OSTEOPOROSIS. Literally meaning "porous bones," this condition occurs when bones lose an excessive amount of their protein and mineral content, particularly calcium. Over time, bone mass and strength are reduced leading to increased risk of fractures.

OSTOMY. A surgically-created opening in the abdomen for elimination of waste products (urine or stool).

OTALGIA. The medical term for pain in the ear. Impacted cerumen can sometimes cause otalgia.

OTITIS. Inflammation of the ear, which may be marked by pain, fever, abnormalities of hearing, hearing loss, noise in the ears, and dizzy spells.

OTITIS MEDIA. Inflammation or infection of the middle ear space behind the eardrum. It commonly occurs in early childhood and is characterized by ear pain, fever, and hearing problems.

OTOACOUSTIC EMISSION (OAE). Sounds or echoes created by vibrations of hair cells in the cochlea in response to sound; used to screen for hearing impairment in newborns.

OTOLARYNGOLOGIST. A doctor who is trained to treat injuries, defects, diseases, or conditions of the ear,

nose, and throat. Also sometimes known as an otorhinolaryngologist.

OTOSCOPE. A hand-held instrument with a tiny light and a funnel-shaped attachment called an ear speculum, which is used to examine the ear canal and eardrum.

OUTER EAR. Outer visible portion of the ear that collects and directs sound waves toward the tympanic membrane by way of a canal which extends inward through the temporal bone.

OUTPATIENT SURGERY. Also called same-day or ambulatory surgery. The patient arrives for surgery and returns home on the same day. Outpatient surgery can take place in a hospital, surgical center, or outpatient clinic.

OVA. The plural of ovum, it is the female reproductive cell.

OVARY. One of the two almond-shaped glands in the female reproductive system responsible for producing eggs and the sex hormones estrogen and progesterone.

OVERBITE. Protrusion of the upper teeth over the lower teeth.

OVER-THE-COUNTER TREATMENTS. Medications that can be purchased without a prescription.

OVERWEIGHT. Being 25–29% over the recommended healthy body weight for a specific age and height, as established by calculating body mass index.

OVULATE. To release a mature egg for fertilization.

OVULATION. The monthly process by which an ovarian follicle ruptures releasing a mature egg cell.

OXIDATIVE STRESS. A condition where the body is producing an excess of oxygen-free radicals.

OXYGENATE. To supply with oxygen.

OXYGENATED BLOOD. Blood carrying oxygen through the body.

OXYTOCIN. A hormone that stimulates the uterus to contract during child birth and the breasts to release milk.

OZONE. A form of oxygen with three atoms in its molecule (O_3), produced by an electric spark or ultraviolet light passing through air or oxygen. A layer of ozone about 15 mi (24 km) above Earth's surface helps protect living things from the damaging effects of the sun's ultraviolet rays. Ozone is used therapeutically as a disinfectant and oxidative agent.

P

PAGET'S DISEASE. A chronic disorder of unknown cause usually affecting middle aged and elderly people and characterized by enlarged and deformed bones. Changes in the normal mechanism of bone formation occur in Paget's disease and can cause bones to weaken, resulting in bone pain, arthritis, deformities, and fractures. Also known as osteitis deformans.

PAGOPHAGIA. The compulsive eating of ice.

PALATAL LENGTHENING (PALATAL PUSHBACK). A surgical procedure in which tissue from the front part of the mouth is moved back to lengthen it.

PALATE. The roof of the mouth.

PALILALIA. A complex vocal tic in which the child repeats his or her own words, songs, or other utterances.

PALLIATIVE. Referring to a drug or a form of care that relieves pain without providing a cure. Persons in severe pain from terminal cancer are often prescribed narcotics as palliative care.

PALLOR. Extreme paleness in the color of the skin.

PALMAR. Referring to the palm of the hand.

PALMAR GRASP. A young infant's primitive ability to hold an object in the palm by wrapping fingers and thumb around it from one side.

PALPITATIONS. Rapid and forceful heartbeat.

PALSY. Uncontrolable tremors.

PANCARDITIS. Inflammation of the lining of the heart, the sac around the heart, and the muscle of the heart.

PANCREAS. A five-inch-long gland that lies behind the stomach and next to the duodenum. The pancreas releases glucagon, insulin, and some of the enzymes which aid digestion.

PANCREATIC INSUFFICIENCY. Reduction or absence of pancreatic secretions into the digestive system due to scarring and blockage of the pancreatic duct.

PANDAS DISORDERS. A group of childhood disorders associated with such streptococcal infections as scarlet fever and strep throat. The acronym stands for Pediatric Autoimmune Neuropsychiatric Disorders Associated with Streptococci.

PANDEMIC. A disease that occurs throughout a regional group, the population of a country, or the world.

PANHYPOPITUITARISM. Generalized decrease of all of the anterior pituitary hormones.

PANTOPHOBIA. Fear of everything.

PAP TEST. A screening test for precancerous and cancerous cells on the cervix. This simple test is done during a routine pelvic exam and involves scraping cells from the cervix. These cells are then stained and examined under a microscope. Also known as the Papanicolaou test.

PAPULE. A solid, raised bump on the skin.

PARALLEL PLAY. Toddlers play side by side but seldom try to interact with each other, playing separately with similar toy.

PARALYSIS. Loss of the ability to move one or more parts of the body voluntarily due to muscle or nerve damage.

PARAMYXOVIRUS. A genus of viruses that includes the causative agent of mumps.

PARASOMNIA. A type of sleep disorder characterized by abnormal changes in behavior or body functions during sleep, specific stages of sleep, or the transition from sleeping to waking.

PARASYMPATHETIC GANGLION CELL. Type of nerve cell normally found in the wall of the colon.

PARATHYROID GLAND. A pair of glands adjacent to the thyroid gland that primarily regulate blood calcium levels.

PARATHYROID HORMONE. A chemical substance produced by the parathyroid glands. This hormone plays a major role in regulating calcium concentration in the body.

PARENTERAL NUTRITION. Liquid nutrition usually provided intravenously.

PARKINSONISM. A set of symptoms originally associated with Parkinson disease that can occur as side effects of neuroleptic medications. The symptoms include trembling of the fingers or hands, a shuffling gait, and tight or rigid muscles.

PARKINSON'S DISEASE. A slowly progressive disease that destroys nerve cells in the basal ganglia and thus causes loss of dopamine, a chemical that aids in transmission of nerve signals (neurotransmitter). Parkinson's is characterized by shaking in resting muscles, a stooping posture, slurred speech, muscular stiffness, and weakness.

PAROTITIS. Inflammation and swelling of one or both of the parotid salivary glands.

PAROXYSMAL HYPERCYANOIC ATTACKS. Sudden episodes of cyanosis resulting from the circulation of deoxygenated blood to the body.

PARVOVIRUS B19. A virus that commonly infects humans; about 50% of all adults have been infected sometime during childhood or adolescence. Parvovirus B19 infects only humans. An infection in pregnancy can cause the unborn baby to have severe anemia and the woman may have a miscarriage.

PASSIVE IMMUNITY. The body reception of proteins that act as antibodies instead of making the antibodies itself. Immunoglobulins may produce this immunity. All babies have antibodies from their mothers, which give them short-term protection.

PASSIVE MOVEMENT. Movement that occurs under the power of an outside source such as a clinician. There is no voluntary muscular contraction by the individual who is being passively moved.

PASTEURELLOSIS. A bacterial wound infection caused by *Pasteurella multocida*. Pasteurellosis is characterized by inflammation around the wound site and may be accompanied by bacteria in the bloodstream and infection in tissues and organs.

PASTEURIZATION. A process during which milk is heated and maintained at a particular temperature for the purpose of killing, or retarding the development of, pathogenic bacteria.

PATCH TEST. A skin test in which different antigens (substances that cause an allergic reaction) are introduced into a patient's skin via a needle prick or scratch and then observed for evidence of an allergic reaction to one or more of them. Also known as a scratch test.

PATELLA. The kneecap.

PATENT DUCTUS ARTERIOSUS. A congenital defect in which the temporary blood vessel connecting the left pulmonary artery to the aorta in the fetus does not close after birth.

PATENT FORAMEN OVALE (PFO). A congenital heart defect characterized by an open flap that remains between the two upper chambers of the heart (the left and right atria). This opening can allow a blood clot from one part of the body to travel through the flap and up to the brain, causing a stroke.

PATHOGEN. Any disease-producing microorganism.

PATHOGENIC BACTERIA. Bacteria that produce illness.

PATHOLOGIC. Characterized by disease or by the structural and functional changes due to disease.

PATIENT EDUCATION. Instruction and information that helps patients prepare for a procedure, learn about a disease, or manage their health. Patient education may include one-on-one instruction from a health care provi-

der, educational sessions in a group setting, or self-guided learning videos or modules. Informative and instructional handouts are usually provided to explain specific medications, tests, or procedures.

PATIENT RIGHTS AND RESPONSIBILITIES. Every hospital has an established list of patient rights and responsibilities, established by the American Hospital Association. They are usually posted throughout the hospital.

PAVOR NOCTURNUS. Another name for sleep terror disorder, or night terrors.

PEAK FLOW MEASUREMENT. Measurement of the maximum rate of airflow attained during a forced vital capacity determination.

PECTUS CARINATUM. An abnormality of the chest in which the sternum (breastbone) is pushed outward. It is sometimes called "pigeon breast."

PECTUS EXCAVATUM. An abnormality of the chest in which the sternum (breastbone) sinks inward; sometimes called "funnel chest."

PEDIATRIC ALLERGIST. A board certified physician specializing in the diagnosis and treatment of allergic conditions in children.

PEDIATRIC DENTISTRY. The dental specialty concerned with the dental treatment of children and adolescents.

PEDIATRIC INTENSIVIST. A physician who completed a three-year residency in pediatrics after medical school and an additional subspecialty fellowship training in intensive care.

PEDICULICIDE. Any substance that kills lice.

PEDICULOSIS. A lice infestation.

PEER ACCEPTANCE. The degree to which a child or adolescent is socially accepted by peers, usually of about the same age; the level of peer popularity.

PEER INFLUENCE. Peer approval or disapproval of the child's behavior or performance.

PEER PRESSURE. Social pressure exerted by a group or individual in a group on someone to adopt a particular type of behavior, dress, or attitude in order to be an accepted member of a group or clique.

PELVIC INFLAMMATORY DISEASE (PID). Any infection of the lower female reproductive tract (vagina and cervix) that spreads to the upper female reproductive tract (uterus, fallopian tubes and ovaries). Symptoms include severe abdominal pain, high fever, and vaginal discharge. PID is the most common and most serious consequence of infection with sexually transmitted diseases in women and is a leading cause of female fertility problems.

PENICILLAMINE (CUPRIMINE, DEPEN). A drug used to treat medical problems (such as excess copper in the body and rheumatoid arthritis) and to prevent kidney stones. It is also sometimes prescribed to remove excess lead from the body.

PERCUSSION. An assessment method in which the surface of the body is struck with the fingertips to obtain sounds that can be heard or vibrations that can be felt. It can determine the position, size, and consistency of an internal organ. It is performed over the chest to determine the presence of normal air content in the lungs, and over the abdomen to evaluate air in the loops of the intestine.

PERCUTANEOUS UMBILICAL BLOOD SAMPLING (PUBS). A technique used to obtain pure fetal blood from the umbilical cord while the fetus is in utero and also called cordocentesis.

PERICARDITIS. Inflammation of the pericardium, the sac that surrounds the heart and the roots of the great blood vessels.

PERICARDIUM. The thin, sac-like membrane that surrounds the heart and the roots of the great vessels. It has two layers: the inner, serous (or visceral) pericardium and the outer, fibrous (or parietal) pericardium.

PERINATAL. Referring to the period of time surrounding an infant's birth, from the last two months of pregnancy through the first 28 days of life.

PERINATAL MORTALITY. The number of late fetal deaths, 28 weeks or more gestation, and neonatal deaths that occur in the first seven days.

PERINEUM. The area between the opening of the vagina and the anus in a woman, or the area between the scrotum and the anus in a man.

PERIODIC LIMB MOVEMENT DISORDER. A disorder characterized by involuntary flexion of leg muscles, causing twitching and leg extension or kicking during sleep.

PERIODONTAL LIGAMENT. Also called the periodontal membrane, this tough fibrous tissue holds the teeth in place in the gums.

PERIODONTITIS. Inflammation of the periodontium, the tissues that support and anchor the teeth. Without treatment it can destroy the structures supporting the teeth, including bone.

PERIPHERAL NERVOUS SYSTEM (PNS). The part of the nervous system that is outside the brain and spinal cord. Sensory, motor, and autonomic nerves are included. PNS

nerves link the central nervous system with sensory organs, muscles, blood vessels, and glands.

PERISTALSIS. Slow, rhythmic contractions of the muscles in a tubular organ, such as the intestines, that move the contents along.

PERITONEUM. The transparent membrane lining the abdominal and pelvic cavities (parietal peritoneum) and the membrane forming the outer layer of the stomach and interstines (visceral peritoneum). Between the visceral and parietal peritoneums is a potential space called the peritoneal cavity.

PERITONITIS. Inflammation of the peritoneum. It is most often due to bacterial infection, but can also be caused by a chemical irritant (such as spillage of acid from the stomach or bile from the gall bladder).

PERMEABLE. A condition in which fluid or certain other substances are allowed to pass through.

PERMETHRIN. A medication used to rid the scalp of head lice. Permethrin works by paralyzing the lice, so that they cannot feed within the 24 hours after hatching required for survival.

PERSONAL EXPERIENCE SCREENING QUESTIONNAIRE (PESQ). A questionnaire for alcoholism.

PERSONALITY. The organized pattern of behaviors and attitudes that makes a human being distinctive. Personality is formed by the ongoing interaction of temperament, character, and environment.

PERSPECTIVE. The way an artist shows depth or distance in a drawing or painting, usually by drawing figures and buildings larger in the front of the picture and smaller in the back.

PERTUSSIS. Whooping cough, a highly contagious disease of the respiratory system, usually affecting children, that is caused by the bacterium *Bordetella pertussis* and is characterized in its advanced stage by spasms of coughing interspersed with deep, noisy inspirations.

PERVASIVE DEVELOPMENTAL DISORDER. A category of childhood disorder that includes Asperger syndrome and Rett's disorder. The PDDs are sometimes referred to collectively as autistic spectrum disorders.

PETECHIA. Plural, petechiae. A tiny purple or red spot on the skin resulting from a hemorrhage under the skin's surface.

PETROLEUM JELLY OR OINTMENT. Petrolatum, a gelatinous substance obtained from oil that is used as a protective dressing.

PEYRONIE'S DISEASE. A disease of unknown origin which causes a hardening of the corpora cavernosa, the erectile tissue of the penis. The penis may become misshapen and/or curved as a result and erections are painful.

PFEIFFER SYNDROME. This condition includes craniosynostosis, shallow eye sockets, underdevelopment of the midface, short thumbs and big toes, and possible webbing of hands and feet.

PH. A measurement of the acidity or alkalinity of a solution. Based on a scale of 14, a pH of 7.0 is neutral. A pH below 7.0 is an acid; the lower the number, the stronger the acid. A pH above 7.0 is a base; the higher the number, the stronger the base. Blood pH is slightly alkaline (basic) with a normal range of 7.36–7.44.

PHAGOCYTOSIS. A process by which certain cells envelope and digest debris and microorganisms to remove them from the blood.

PHALANX. (plural, plananges) Any of the digital bones of the hand or foot. Humans have three phalanges to each finger and toe with the exception of the thumb and big toe which have only two each.

PHALILALIA. Involuntary echoing by an individual of the last word, phrase, sentence, or sound he/she vocalized.

PHANTOM PAIN. Pain, tingling, itching, or numbness in the place where the amputated part used to be.

PHARMACOLOGICAL. Referring to therapy that relies on drugs.

PHARYNX. The throat, a tubular structure that lies between the mouth and the esophagus.

PHASE CONTRAST MICROSCOPE. A light microscope in which light is focused on the sample at an angle to produce a clearer image.

PHENYLALANINE. An essential amino acid that must be obtained from food since the human body cannot manufacture it. It is necessary for normal growth and development and for normal protein metabolism.

PHENYLKETONURIA (PKU). A rare, inherited, metabolic disorder in which the enzyme necessary to break down and use phenylalanine, an amino acid necessary for normal growth and development, is lacking. As a result, phenylalanine builds up in the body causing mental retardation and other neurological problems.

PHENYTOIN. An anti-convulsant medication used to treat seizure disorders. Sold under the brand name Dilantin.

PHEOCHROMOCYTOMA. A tumor that originates from the adrenal gland's chromaffin cells, causing over-

production of catecholamines, powerful hormones that induce high blood pressure and other symptoms.

PHIMOSIS. A tightening of the foreskin that may close the opening of the penis.

PHLEBOTOMIST. A person who draws blood from a vein.

PHLEGM. Thick mucus produced in the air passages.

PHOBIA. An intense and irrational fear of a specific object, activity, or situation that leads to avoidance.

PHONEMES. The basic units of sound in a language.

PHONICS. A system to teach reading by teaching the speech sounds associated with single letters, letter combinations, and syllables.

PHONOLOGICAL AWARENESS. The ability to hear and manipulate the sounds that make up words.

PHONOLOGY. The science of speech sounds and sound patterns.

PHOTOCOAGULATION. A type of cancer treatment in which cancer cells are destroyed by an intense beam of laser light.

PHOTOPHOBIA. An extreme sensitivity to light.

PHOTOSENSITIZATION. Development of oversensitivity to sunlight.

PHOTOTHERAPY. Another name for light therapy in mainstream medical practice.

PHYSIATRIST. A physician who specializes in physical medicine and rehabilitation.

PHYSICAL THERAPIST. A healthcare provider who teaches patients how to perform therapeutic exercises to maintain maximum mobility and range of motion.

PHYSIOLOGIC. Refers to physiology, particularly normal, healthy, physical functioning.

PHYTOESTROGENS. Compounds found in plants that can mimic the effects of estrogen in the body.

PICA. A desire that sometimes arises in pregnancy to eat nonfood substances, such as dirt or clay.

PIMPLE. A small, red swelling of the skin.

PINCER GRIP. The ability to hold objects between thumb and index finger, which typically develops in infants between 12 and 15 months of age.

PIPERONYL BUTOXIDE. A liquid organic compound that enhances the activity of insecticides.

PITOCIN. A synthetic hormone that produces uterine contractions.

PITUITARY GLAND. The most important of the endocrine glands (glands that release hormones directly into the bloodstream), the pituitary is located at the base of the brain. Sometimes referred to as the "master gland," it regulates and controls the activities of other endocrine glands and many body processes including growth and reproductive function. Also called the hypophysis.

PLACENTA. The organ that provides oxygen and nutrition from the mother to the unborn baby during pregnancy. The placenta is attached to the wall of the uterus and leads to the unborn baby via the umbilical cord.

PLACENTA PREVIA. A condition in which the placenta totally or partially covers the cervix, preventing vaginal delivery.

PLACENTAL ABRUPTION. An abnormal separation of the placenta from the uterus before the birth of the baby, with subsequent heavy uterine bleeding. Normally, the baby is born first and then the placenta is delivered within a half hour.

PLACENTAL INFARCTION. An area of dead tissue in the placenta that is due to an obstruction of circulation in the area.

PLAGIOCEPHALY. A form of craniosynostosis that involves fusion of the right or left side of coronal suture.

PLAGUE. A serious, potentially life-threatening infectious disease caused by the bacterium *Yersinia pestis*. The disease is usually transmitted to humans by the bites of infected rodent fleas. There are three major types: bubonic, pneumonic, and septicemic.

PLANTAR. Relating to the sole of the foot.

PLAQUE. A deposit, usually of fatty material, on the inside wall of a blood vessel. Also refers to a small, round demyelinated area that develops in the brain and spinal cord of an individual with multiple sclerosis.

PLASMA. A watery fluid containing proteins, salts, and other substances that carries red blood cells, white blood cells, and platelets throughout the body. Plasma makes up 50% of human blood.

PLATELET. A cell-like particle in the blood that plays an important role in blood clotting. Platelets are activated when an injury causes a blood vessel to break. They change shape from round to spiny, "sticking" to the broken vessel wall and to each other to begin the clotting process. In addition to physically plugging breaks in blood vessel walls, platelets also release chemicals that promote clotting.

PLAY THERAPY OR THERAPEUTIC PLAY. A type of psychotherapy for young children involving the use of

toys and games to build a therapeutic relationship and encourage the child's self-expression.

PLAY-BASED ASSESSMENT. A form of developmental assessment that involves observation of how a child plays alone, with peers, or with parents or other familiar caregivers, in free play or in special games.

PLEURITIS. Inflammation of the pleura, the membrane surrounding the lungs. Also called pleurisy.

PNEUMATIC OTOSCOPE. An otoscope that can also produce a small puff of air that vibrates the eardrum.

PNEUMOCYSTIS CARINII. A parasite transitional between a fungus and protozoan, frequently occurring as aggregate forms existing within rounded cystlike structures. It is the causative agent of pneumocystosis.

PNEUMONIA. An infection in which the lungs become inflamed. It can be caused by nearly any class of organism known to cause human infections, including bacteria, viruses, fungi, and parasites.

PNEUMOTHORAX. A collection of air or gas in the chest or pleural cavity that causes part or all of a lung to collapse.

POLIO. Poliomyelitis, an acute viral disease marked by inflammation of nerve cells of the brain stem and spinal cord and can cause paralysis.

POLLEN. A fine, powdery substance released by plants and trees; an allergen.

POLYCARBONATE. A very strong type of plastic often used in safety glasses, sport glasses, and children's eyeglasses. Polycarbonate lenses have approximately 50 times the impact resistance of glass lenses.

POLYHYDRAMNIOS. A condition in which there is too much fluid around the fetus in the amniotic sac.

POLYSOMNOGRAPHY. An overnight series tests designed to evaluate a patient's basic physiological processes during sleep. Polysomnography generally includes monitoring of the patient's airflow through the nose and mouth, blood pressure, electrocardiographic activity, blood oxygen level, brain wave pattern, eye movement, and the movement of respiratory muscles and limbs

POLYUNSATURATED FAT. A non-animal oil or fatty acid rich in unsaturated chemical bonds. This type of fat is not associated with the formation of cholesterol in the blood.

POLYURETHANE. A type of synthetic plastic.

PORPHYRIN. An organic compound found in living things that founds the foundation structure for hemoglobin, chlorophyll, and other respiratory pigments. In humans, porphyrins combine with iron to form hemes.

PORTFOLIO. A student-controlled collection of student work products that indicates progress over time.

POSITRON EMISSION TOMOGRAPHY (PET). A computerized diagnostic technique that uses radioactive substances to examine structures of the body. When used to assess the brain, it produces a three-dimensional image that shows anatomy and function, including such information as blood flow, oxygen consumption, glucose metabolism, and concentrations of various molecules in brain tissue.

POSTAXIAL. Situated behind or away from the axis or midline of the body.

POSTEXPOSURE PROPHYLAXIS. Any treatment given after exposure to a disease to try to prevent the disease from occurring. In the case of rabies, postexposure prophylaxis involves a series of vaccines given to an individual who has been bitten by an unknown animal or one that is potentially infected with the rabies virus.

POSTPARTUM. The six-week period following childbirth.

POST-TRAUMATIC STRESS DISORDER (PTSD). A disorder that occurs among survivors of extremely stressful or traumatic events, such as a natural disaster, an airplane crash, rape, or military combat. Symptoms include anxiety, insomnia, flashbacks, and nightmares. Patients with PTSD are unnecessarily vigilant; they may experience survivor guilt, and they sometimes cannot concentrate or experience joy.

POSTURAL. Pertaining to the position of the head, neck, trunk and lower limbs in relation to the ground and the vertical.

POSTURAL DRAINAGE. The use of positioning to drain secretions from the bronchial tubes and lungs into the trachea or windpipe where they can either be coughed up or suctioned out.

PRANAYAMA. The yoga practice of breathing correctly and deeply.

PREAXIAL. Situated in front of the axis or midline of the body.

PREDIABETES. A precursor condition to type 2 diabetes, sometimes called impaired glucose tolerance or impaired fasting glucose. Prediabetes is clinically defined as individuals who have elevated blood glucose levels that are not diagnostic of type 2 diabetes but are above normal (for the fasting plasma glucose test, this measurement would be 100 to 125 mg/dL [5.6 to 6.9 mmol/L]).

PREDNISONE. A corticosteroid medication often used to treat inflammation.

PREECLAMPSIA. A condition that develops after the twentieth week of pregnancy and results in high blood pressure, fluid retention that doesn't go away, and large amounts of protein in the urine. Without treatment, it can progress to a dangerous condition called eclampsia, in which a woman goes into convulsions.

PREGNANCY CATEGORY. A system of classifying drugs according to their established risks for use during pregnancy. Category A: Controlled human studies have demonstrated no fetal risk. Category B: Animal studies indicate no fetal risk, but no human studies, or adverse effects in animals, but not in well-controlled human studies. Category C: No adequate human or animal studies, or adverse fetal effects in animal studies, but no available human data. Category D: Evidence of fetal risk, but benefits outweigh risks. Category X: Evidence of fetal risk. Risks outweigh any benefits.

PREMATURE LABOR. Labor beginning before 36 weeks of pregnancy.

PREMAXILLA. The front central section of the upper gum, containing the four upper front teeth.

PREMOLAR. Bicuspid; the two cupped teeth between the first molars and the cuspids.

PREMUTATION. A change in a gene that precedes a mutation; this change does not alter the function of the gene.

PRENATAL DIAGNOSIS. The determination of whether a fetus possesses a disease or disorder while it is still in the womb.

PRENATAL TESTING. Testing for a disease, such as a genetic condition, in an unborn baby.

PREPUCE. A fold of skin, such as the foreskin of the penis or the skin that surrounds the clitoris.

PRESCHOOL. An early childhood program in which children combine learning with play in a program run by professionally trained adults.

PRESSURE ULCER. Also known as a decubitus ulcer or bedsore, a pressure ulcer is an open wound that forms whenever prolonged pressure is applied to skin covering bony prominences of the body. Patients who are bedridden are at risk of developing pressure ulcers.

PRIAPISM. A painful, abnormally prolonged penile erection.

PRIMARY CAREGIVER. A person who is responsible for the primary care and upbringing of a child.

PRIMARY IMMUNODEFICIENCY DISEASE. A group of approximately 70 conditions that affect the normal functioning of the immune system.

PRIMARY SLEEP DISORDER. A sleep disorder that cannot be attributed to a medical condition, another mental disorder, or prescription medications or other substances.

PROBIOTICS. Bacteria that are beneficial to a person's health, either through protecting the body against pathogenic bacteria or assisting in recovery from an illness.

PROBLEM ORIENTED SCREENING INSTRUMENT FOR TEENAGERS (POSIT). A questionnaire used specifically for teenagers to assess alcohol and drug use.

PRODROME. Early symptoms that warn of the beginning of disease. For example, the herpes prodrome consists of pain, burning, tingling, or itching at a site before blisters are visible while the migraine prodrome consists of visual disturbances.

PROGESTERONE. The hormone produced by the ovary after ovulation that prepares the uterine lining for a fertilized egg.

PROGRESSIVE. Advancing, going forward, going from bad to worse, increasing in scope or severity.

PROGRESSIVE SUPRANUCLEAR PALSY. A rare disease that gradually destroys nerve cells in the parts of the brain that control eye movements, breathing, and muscle coordination. The loss of nerve cells causes palsy, or paralysis, that slowly gets worse as the disease progresses. The palsy affects ability to move the eyes, relax the muscles, and control balance. Also called Steele-Richardson-Olszewski syndrome.

PROJECTIVE PERSONALITY TEST. A personality test in which the participant interprets ambiguous images, objects, stories.

PROJECTIVE TEST. A type of psychological test that assesses a person's thinking patterns, observational ability, feelings, and attitudes on the basis of responses to ambiguous test materials. Projective tests are often used to evaluate patients with personality disorders.

PROLACTIN. A hormone that prepares the breasts during pregnancy for milk production after childbirth.

PROMISCUOUS. Having many indiscriminate or casual sexual relationships.

PRONE. Lying on the stomach with the face downward.

PROPHYLACTIC. Preventing the spread or occurrence of disease or infection.

PROPHYLAXIS. Protection against or prevention of a disease. Antibiotic prophylaxis is the use of antibiotics to prevent a possible infection.

PROSOCIAL BEHAVIORS. Social behavior characterized by positive, cooperative, and reciprocal social exchanges.

PROSTAGLANDINS. A group of hormone-like molecules that exert local effects on a variety of processes including fluid balance, blood flow, and gastrointestinal function. They may be responsible for the production of some types of pain and inflammation.

PROSTHETIC. Referring to an artificial part of the body.

PROSTHETIST. A health care professional who is skilled in making and fitting artificial limbs and other prostheses.

PROTEIN. An important building blocks of the body, a protein is a large, complex organic molecule composed of amino acids. It is involved in the formation of body structures and in controlling the basic functions of the human body.

PROTEIN-LOSING ENTEROPATHY. Excessive loss of plasma and proteins in the gastrointestinal tract.

PROTEINURIA. Having abnormally large quantities of protein in the urine.

PROTOCOL. A plan for carrying out a scientific study or a patient's course of treatment.

PROTON PUMP. A structure in the body that produces and pumps acid into the stomach.

PROTOPORPHYRIN. A kind of porphyrin that links with iron to form the heme of hemoglobin.

PROTOPORPHYRIN IX. A protein the measurement of which is useful for the assessment of iron status. Hemoglobin consists of a complex of a protein plus heme. Heme consists of iron plus protoporphyrin IX. Normally, during the course of red blood cell formation, protoporphyrin IX acquires iron, to generate heme, and the heme becomes incorporated into hemoglobin. However, in iron deficiency, protophoryrin IX builds up.

PROXIMAL MUSCLES. The muscles closest to the center of the body.

PROXIMODISTAL DEVELOPMENT. Motor development which occurs in the first two years of life: head, trunk, and arms before hands and fingers.

PRURITUS. The symptom of itching or an uncontrollable sensation leading to the urge to scratch.

PSEUDOMONAS. A bacterium which can cause ulcers in contact lens wearers.

PSORIASIS. A chronic, noncontagious skin disease that is marked by dry, scaly, and silvery patches of skin that appear in a variety of sizes and locations on the body.

PSYCHODRAMA. A specific form of role play that focuses on acting out "scripts" of unresolved issues within the family, or helping family members adopt new approaches and understanding of one another.

PSYCHOGENIC DISORDERS. A variety of unusual, involuntary movements that occur in children with psychiatric disorders or in response to anxiety, stress, depression, anger, or grief. Psychogenic movements are thought to represent the physical expression of an intolerable mental conflict.

PSYCHOLOGICAL. Pertaining to the mind, its mental processes, and its emotional makeup.

PSYCHOLOGICAL EVALUATION. Examination of a patient by a psychologist through interviews, observation of behavior, and psychological testing with the goal of determining personality adjustment, identifying problems, and helping to diagnose and plan treatment for a mental disorder.

PSYCHOMETRICS. The development, administration, and interpretation of tests to measure mental or psychological abilities. Psychometric tests convert an individual's psychological traits and attributes into a numerical estimation or evaluation.

PSYCHOMOTOR AGITATION. Disturbed physical and mental processes (e.g., fidgeting, wringing of hands, racing thoughts); a symptom of major depressive disorder.

PSYCHOMOTOR RETARDATION. Slowed mental and physical processes characteristic of a bipolar depressive episode.

PSYCHOPATHOLOGY. The study of mental disorders or illnesses, such as schizophrenia, personality disorder, or major depressive disorder.

PSYCHOSES. Mental illness that interferes with an individual's ability to manage life's challenges and everyday activities. The impairment of cognitive ability that distorts reality.

PSYCHOTHERAPY. Psychological counseling that seeks to determine the underlying causes of a patient's depression. The form of this counseling may be cognitive/behavioral, interpersonal, or psychodynamic.

PSYCHOTROPIC DRUG. Any medication that has an effect on the mind, brain, behavior, perceptions, or emotions. Psychotropic medications are used to treat mental

illnesses because they affect a patient's moods and perceptions.

PUBERTY. The point in development when the ability to reproduce begins. The gonads begin to function and secondary sexual characteristics begin to appear.

PULMONARY. Referring to the lungs and respiratory system.

PULMONARY ARTERY. An artery that carries blood from the heart to the lungs.

PULMONARY EDEMA. An accumulation of fluid in the tissue of the lungs.

PULMONARY EMBOLISM. Blockage of an artery in the lungs by foreign matter such as fat, tumor tissue, or a clot originating from a vein. A pulmonary embolism can be a very serious, and in some cases fatal, condition.

PULMONARY HYPERTENSION. A disorder in which the pressure in the blood vessels of the lungs is abnormally high.

PULMONARY HYPOPLASIA. Incomplete or defective development of the lungs.

PULP. The soft, innermost layer of a tooth that contains its blood vessels and nerves.

PULPITIS. Inflammation of the pulp of a tooth that involves the blood vessels and nerves.

PUNCTURE. An injury caused by a sharp, narrow object deeply penetrating the skin.

PUNISHMENT. The application of a negative stimulus to reduce or eliminate a behavior. The two types typically used with children are verbal reprimands and punishment involving physical pain, as in corporal punishment.

PURGING. The use of vomiting, diuretics, or laxatives to clear the stomach and intestines after a binge.

PURIFIED CHICKEN EMBRYO CELL VACCINE (PCEC). A rabies vaccine in which the virus is grown in cultures of chicken embryo cells, inactivated, and purified for IM injection.

PURPURA. A group of disorders characterized by purplish or reddish brown areas of discoloration visible through the skin. These areas of discoloration are caused by bleeding from broken capillaries.

PUS. A thick, yellowish or greenish fluid composed of the remains of dead white blood cells, pathogens, and decomposed cellular debris. It is most often associated with bacterial infection.

PYELONEPHRITIS. An inflammation of the kidney and upper urinary tract, usually caused by a bacterial infection. In its most serious form, complications can include high blood pressure (hypertension) and renal failure.

PYOGENIC. Capable of generating pus. *Streptococcus*, *Staphocococcus*, and bowel bacteria are the primary pyogenic organisms.

PYRETHRIN, PYRETHROID. A naturally occurring insecticide extracted from chrysanthemum flowers. It paralyzes lice so that they cannot feed.

PYREXIA. A medical term meaning fever.

PYRIN. A protein that regulates the body' s inflammatory response to stress or trauma. The MEFV gene involved in FMF produces an unstable form of pyrin that fails to adequately control the inflammatory response.

PYROGEN. A chemical circulating in the blood that causes a rise in body temperature.

PYROMANIA. An impulse control disorder characterized by fire setting.

Q

QUADRIPLEGIA. Paralysis of all four limbs and the trunk below the level of an associated injury to the spinal cord. Also called tetraplegia.

QUANTIFIABLE. A result or measurement that can be expressed as a number. The results of quantifiable psychological tests can be translated into numerical values, or scores.

R

RABIES IMMUNE GLOBULIN (RIG OR HRIG). A human serum preparation containing high levels of antibodies against the rabies virus; used for post-exposure prophylaxis.

RABIES VIRUS ADSORBED (RVA). A rabies vaccine in which the virus is grown in cultures of lung cells from rhesus monkeys, inactivated, and adsorbed to aluminum phosphate.

RACHITIC ROSARY. Beadlike bumps present at the junction of the ribs with their cartilages. It is often seen in children with rickets.

RADIATION THERAPY. A cancer treatment that uses high-energy rays or particles to kill or weaken cancer cells. Radiation may be delivered externally or internally via surgically implanted pellets. Also called radiotherapy.

RADIOFREQUENCY ABLATION (RFA). A procedure in which radiofrequency waves are used to destroy blood vessels and tissues.

RADIOGRAPH. The actual picture or film produced by an x-ray study.

RADIOGRAPHY. Examination of any part of the body through the use of x rays. The process produces an image of shadows and contrasts on film.

RADIOISOTOPE. One of two or more atoms with the same number of protons but a different number of neutrons with a nuclear composition. In nuclear scanning, radioactive isotopes are used as a diagnostic agent.

RADIOLOGIST. A medical doctor specially trained in radiology, the branch of medicine concerned with radioactive substances and their use for the diagnosis and treatment of disease.

RADIOPAQUE. Not penetrable by x-rays. A radiopaque object will look white or light when the x-ray film is developed. Most objects that children swallow can be detected by an x-ray study because they are radiopaque.

RADIOPAQUE DYES, RADIOCONTRAST MEDIA. Injected substances that are used to outline tissues and organs in some x-ray and other radiation procedures.

RADIUS. The bone of the forearm which joins the wrist on the same side as the thumb.

RAGAMUFFINS. A term used in nineteenth-century London to describe neglected or abandoned children who lived on the streets.

RAGGED-RED FIBERS. A microscopic accumulation of diseased mitochondria.

RANGE OF MOTION (ROM). The range of motion of a joint from full extension to full flexion (bending) measured in degrees like a circle.

RAPID EYE MOVEMENT (REM) LATENCY. The amount of time it takes for the first onset of REM sleep after a person falls asleep.

RAPID EYE MOVEMENT (REM) SLEEP. A phase of sleep during which the person's eyes move rapidly beneath the lids. It accounts for 20–25% of sleep time. Dreaming occurs during REM sleep.

RASH. A spotted, pink or red skin eruption that may be accompanied by itching and is caused by disease, contact with an allergen, food ingestion, or drug reaction.

REACTIVE HYPOGLYCEMIA. A rare condition in which blood sugars drop below normal levels approximately four hours after eating.

REACTIVITY. The level or intensity of a person's physical or emotional excitability.

READINESS TEST. A test that measures the extent of a child's acquired skills for successfully undertaking a new learning activity such as kindergarten.

RECEPTIVE APHASIA. A developmental disorder in which a child has difficulty comprehending spoken and written language.

RECEPTIVE LANGUAGE. The comprehension of language.

RECESSIVE. Refers to an inherited trait that is outwardly obvious only when two copies of the gene for that trait are present. An individual displaying a recessive trait must have inherited one copy of the defective gene from each parent.

RECESSIVE DISORDER. Disorder that requires two copies of the predisposing gene one from each parent for the child to have the disease.

RECESSIVE GENE. A type of gene that is not expressed as a trait unless inherited by both parents.

RECOMMENDED DIETARY ALLOWANCE (RDA). The Recommended Dietary Allowances (RDAs) are quantities of nutrients in the diet that are required to maintain good health in people. RDAs are established by the Food and Nutrition Board of the National Academy of Sciences, and may be revised every few years. A separate RDA value exists for each nutrient. The RDA values refer to the amount of nutrient expected to maintain good health in people. The actual amounts of each nutrient required to maintain good health in specific individuals differ from person to person.

RECURRENT. Tendency to repeat.

RED BLOOD CELL INDICES. Measurements that describe the size and hemoglobin content of red blood cells. The indices are used to help in the differential diagnosis of anemia. Also called red cell absolute values or erythrocyte indices.

RED BLOOD CELLS. Cells that carry hemoglobin (the molecule that transports oxygen) and help remove wastes from tissues throughout the body.

RED CELL DISTRIBUTION WIDTH (RDW). A measure of the variation in size of red blood cells.

REDUCIBLE HERNIA. A hernia that can be gently pushed back into place or that disappears when the person lies down.

REDUCTION. The restoration of a body part to its original position after displacement, such as the reduction of a fractured bone by bringing ends or fragments back into original alignment. The use of local or general anesthesia usually accompanies a fracture reduction.

REFERENTIAL LANGUAGE DEVELOPMENT. A style of language development in which a child first speaks single words and then joins words together into two- and three-word sentences.

REFERRED PAIN. Pain that is experienced in one part of the body but originates in another organ or area. The pain is referred because the nerves that supply the damaged organ enter the spine in the same segment as the nerves that supply the area where the pain is felt.

REFLEX. An involuntary response to a particular stimulus.

REFLUX. The backward flow of a body fluid or secretion. Indigestion is sometimes caused by the reflux of stomach acid into the esophagus.

REGIONAL ANESTHESIA. Blocking of specific nerve pathways through the injection of an anesthetic agent into a specific area of the body.

REGISTERED NURSES. Specially trained nurses who provide care during the patient's hospital stay. Registered nurses provide health care, administer medications, monitor the patient's condition, and educate the patient.

REGURGITATION. The flow of material back up the esophagus and into the throat or lungs. Also refers to the backward flow of blood through a partly closed heart valve.

REHYDRATION. The restoration of water or fluid to a body that has become dehydrated.

REIKI. A form of energy therapy that originated in Japan. Reiki practitioners hold their hands on or slightly above specific points on the patient's body in order to convey universal life energy to that area for healing.

RELINQUISHMENT. Giving up parental rights to a child, so someone else can adopt the child.

REMINERALIZATION. Recalcification; the process by which minerals from saliva and food are added to the surface of the enamel or to the dentin.

REMISSION. A disappearance of a disease and its symptoms. Complete remission means that all disease is gone. Partial remission means that the disease is significantly improved, but residual traces of the disease are still present. A remission may be due to treatment or may be spontaneous.

RENAL ARTERY STENOSIS. A disorder in which the arteries that supply blood to the kidneys are narrowed or constricted.

RENDERING. An artist's term for shading or creating texture or shape with markings, usually made with pencil, charcoal, ink, or paint.

REPETITIVE STRESS INJURY. An injury resulting from a repeated movement such as typing or throwing a ball.

REPRESENTATIVE SAMPLE. A random sample of people that adequately represents the test-taking population in age, gender, race, and socioeconomic standing.

RESPIRATORY DISTRESS SYNDROME (RDS). Also known as hyaline membrane disease, this is a condition of premature infants in which the lungs are imperfectly expanded due to a lack of a substance (surfactant) on the lungs that reduces tension.

RESPIRATORY FAILURE. Inability to rid the body of CO_2 or establish an adequate blood oxygen level.

RESPIRATORY SYSTEM. The organs that are involved in breathing: the nose, the throat, the larynx, the trachea, the bronchi, and the lungs. Also called the respiratory tract.

RESTLESS LEGS SYNDROME (RLS). A disorder in which the patient experiences crawling, aching, or other disagreeable sensations in the calves that can be relieved by movement. RLS is a frequent cause of difficulty falling asleep at night.

RESUSCITATION. Reviving a person whose cardiovascular system had slowed or stopped.

RETAINER. An orthodontic appliance that is worn to stabilize teeth in a new position.

RETENTION TREATMENT STAGE. The passive treatment period following orthodontic treatment, when retainers may be used to stabilize the teeth.

RETICULOCYTE. An early, immature form of a red blood cell. Over time, the reticulocyte develops to become a mature, oxygen-carrying red blood cell.

RETINA. The inner, light-sensitive layer of the eye containing rods and cones. The retina transforms the image it receives into electrical signals that are sent to the brain via the optic nerve.

RETINAL HEMORRHAGE. Bleeding of the retina, a key structure in vision located at the back of the eye.

RETINOIC ACID. Vitamin A_1 acid which is used topically to treat acne.

RETINOPATHY. Any disorder of the retina.

RETINOPATHY OF PREMATURITY. A condition in which the blood vessels in a premature infant's eyes do not develop normally. It can, in some cases, result in blindness.

RETRACTIONS. Tugging-in between the ribs when breathing in.

RETROCAVAL URETER. A ureter that is located behind the vena cava blood vessel.

RETROVIRUS. A family of RNA viruses containing a reverse transcriptase enzyme that allows the viruses' genetic information to become part of the genetic information of the host cell upon replication. Human immunodeficiency virus (HIV) is a retrovirus.

REYE'S SYNDROME. A serious, life-threatening illness in children, usually developing after a bout of flu or chickenpox, and often associated with the use of aspirin. Symptoms include uncontrollable vomiting, often with lethargy, memory loss, disorientation, or delirium. Swelling of the brain may cause seizures, coma, and in severe cases, death.

RH BLOOD INCOMPATIBILITY. Incompatibility between the blood of a mother and her baby due the absence of the Rh antigen in the red blood cells of one and its presence in the red blood cells of the other.

RH FACTOR. An antigen present in the red blood cells of 85% of humans. A person with Rh factor is Rh positive (Rh+); a person without it is Rh negative (Rh-). The Rh factor was first identified in the blood of a rhesus monkey and is also known as the rhesus factor.

RHABDOVIRUS. A family of viruses named for their rod- or bullet-like shapes. The rabies virus is a rhabdovirus.

RHEMATOLOGIST. A physician who specializes in the treatment of disorders of the connective tissue structures, such as the joints and related structures.

RHEUMATIC FEVER. An illness that arises as a complication of an untreated or inadequately treated streptococcal infection of the throat. It ususally occurs among school-aged children and cause serious damage to the heart valves.

RHEUMATOLOGIST. A doctor who specializes in the diagnosis and treatment of disorders affecting the joints and connective tissues of the body.

RHINITIS. Inflammation and swelling of the mucous membranes that line the nasal passages.

RHINOPLASTY. Plastic surgery of the nose to repair it or change its shape.

RHINOVIRUS. A group of small RNA viruses that infects the upper respiratory system and causes the common cold.

RICKETS. A condition caused by the dietary deficiency of vitamin D, calcium, and usually phosphorus, seen primarily in infancy and childhood, and characterized by abnormal bone formation.

RIGIDITY. A constant resistance to passive motion.

RODS. Photoreceptors, located in the retina of the eye, that are highly sensitive to low levels of light.

ROLFING. A holistic system of bodywork that uses deep manipulation of the body's soft tissue to realign and rebalance the body's myofacial (connective) structure. It is used to improve posture, relieve chronic pain, and reduce stress.

ROOT CANAL TREATMENT. The process of removing diseased or damaged pulp from a tooth, then filling and sealing the pulp chamber and root canals.

RORSCHACH TEST. A well-known psychological test in which subjects are asked to describe a series of black or colored inkblots. The inkblots allow the patient to project his or her interpretations, which can be used to diagnose particular disorders. Also known as the Rorschach Psychodiagnostic Test.

RUBBERS. A slang name for condoms.

RUBELLA. A mild, highly contagious childhood illness caused by a virus; it is also called German measles. Rubella causes severe birth defects (including heart defects, cataracts, deafness, and mental retardation) if a pregnant woman contracts it during the first three months of pregnancy.

RUSSELL'S SIGN. A scraped or raw area on the patient's knuckles, caused by self-induced vomiting.

S

SACROCOCCYGEAL TERATOMA (SCT). A tumor occurring at the base of the tailbone in a fetus.

SADDLE NOSE. A sunken nasal bridge.

SAGITTAL SUTURE. The suture between the two parietal bones in the top of the skull.

SALICYLATES. A group of drugs that includes aspirin and related compounds. Salicylates are used to relieve pain, reduce inflammation, and lower fever.

SALICYLIC ACID. An agent prescribed to treat a variety of skin disorders, such as acne, dandruff, psoriasis, seborrheic dermatitis, calluses, corns, and warts.

SARCOIDOSIS. A chronic disease that causes the formation of granulomas, masses resembling small tumors composed of clumps of immune cells, in any organ or tissue. Common sites include the lungs, spleen, liver, mucous membranes, skin, and lymph nodes.

SARCOMA. A type of cancer that originates from connective tissue such as bone or muscle.

SCABIES. A contagious parasitic skin disease caused by a tiny mite and characterized by intense itching.

SCAPHOCEPHALY. An abnormally long and narrow skull.

SCHEMAS. Fundamental core beliefs or assumptions that are part of the perceptual filter people use to view the world. Cognitive-behavioral therapy seeks to change maladaptive schemas.

SCHIZOPHRENIA. A severe mental illness in which a person has difficulty distinguishing what is real from what is not real. It is often characterized by hallucinations, delusions, and withdrawal from people and social activities.

SCHOOL PHOBIA. Childhood anxiety about leaving home to attend school.

SCISSORING. Involuntary crossing of the legs.

SCLERA. The tough, fibrous, white outer protective covering of the eyeball.

SCLEROTHERAPY. Injection of an irritating chemical into a blood vessel so that it forms a scar to repair itself.

SCOLIOMETER. A tool for measuring trunk asymmetry that includes a bubble level and angle measure.

SCOLIOSIS. An abnormal, side-to-side curvature of the spine.

SCREENING. A process through which carriers of a trait may be identified within a population.

SCROTUM. The external pouch containing the male reproductive glands (testes) and part of the spermatic cord.

SCURVY. A nutritional disorder caused by vitamin C deficiency that is characterized by tiredness, muscle weakness, joint and muscle aches, a rash on the legs, bleeding gums, and skin bruising.

SEALANT. A thin plastic substance that is painted over teeth as an anti-cavity measure to seal out food particles and acids produced by bacteria.

SEBACEOUS. Related to the glands of the skin that produce an oily substance called sebum.

SEBACEOUS FOLLICLE. A structure found within the skin where a sebaceous gland opens into a hair follicle.

SEBUM. An oily skin moisturizer produced by sebaceous glands.

SECONDHAND SMOKE. A mixture of the smoke given off by the burning end of a cigarette, pipe, or cigar and the smoke exhaled from the lungs of smokers.

SECRETION. A substance, such as saliva or mucus, that is produced and given off by a cell or a gland.

SECURE ATTACHMENT. Type of attachment that usually develops when the primary caregiver is sensitive to the infant's behavior and is emotionally and physically available to the child.

SEDATIVE. A medication that has a calming effect and may be used to treat nervousness or restlessness. Sometimes used as a synonym for hypnotic.

SEIZURE. A sudden attack, spasm, or convulsion.

SEIZURE THRESHOLD. The amount of stimulation required to induce a seizure.

SELECTIVE SEROTONIN REUPTAKE INHIBITORS (SSRIS). A class of antidepressants that work by blocking the reabsorption of serotonin in the brain, thus raising the levels of serotonin. SSRIs include fluoxetine (Prozac), sertraline (Zoloft), and paroxetine (Paxil).

SELF-ESTEEM. A sense of competence, achievement, and self-respect. Maslow felt that the most stable source of self-esteem is genuine accomplishment rather than public acclaim or praise.

SELF-IDENTITY. The awareness that an individual has of being unique.

SEMEN. The thick, whitish liquid released from the penis on ejaculation. It contains sperm and other secretions.

SEMI-STRUCTURED INTERVIEW. A psychiatric instrument characterized by open-ended questions for discussion rather than brief questions requiring yes or no answers.

SENSORIMOTOR. Relating to the combination of sensory and motor coordination.

SENSORINEURAL HEARING LOSS. Hearing loss caused by damage to the nerves or parts of the inner ear governing the sense of hearing. Sound is conducted normally through the external and middle ear.

SENSORY NERVES. Sensory or afferent nerves carry impulses of sensation from the periphery or outward parts of the body to the brain and spinal cord.

SEPARATION ANXIETY. Childhood fear of leaving parents for any reason.

SEPSIS. A severe systemic infection in which bacteria have entered the bloodstream or body tissues.

SEPTAL. Relating to the septum, the thin muscle wall dividing the right and left sides of the heart. Holes in the septum are called septal defects.

SEPTICEMIA. A systemic infection due to the presence of bacteria and their toxins in the bloodstream. Septicemia is sometimes called blood poisoning.

SEPTUM. A wall or partition. Often refers to the muscular wall dividing the left and right heart chambers or the partition in the nose that separates the two nostrils. Also refers to an abnormal fold of tissue down that center of the uterus that can cause infertility.

SEQUELA. Plural, sequalae. An abnormal condition resulting from a previous disease or disorder.

SEQUENTIAL BILINGUALISM. Acquiring first one language and then a second language before the age of three.

SEQUENTIAL MOTOR SKILL. Ability to coordinate different motor skills in sequence, such as running followed by a jump.

SERIAL CASTING. A series of casts designed to gradually move a limb into a more functional position.

SEROPOSITIVE. Showing a positive reaction to a test on blood serum for a disease; exhibiting seroconversion.

SEROSITIS. Inflammation of a serosal membrane (any membrane that lines a body cavity that does not open to the outside of the body). Polyserositis refers to the inflammation of two or more serosal membranes.

SEROTONIN. A widely distributed neurotransmitter that is found in blood platelets, the lining of the digestive tract, and the brain, and that works in combination with norepinephrine. It causes very powerful contractions of smooth muscle and is associated with mood, attention, emotions, and sleep. Low levels of serotonin are associated with depression.

SEROUS. Pertaining to or resembling serum.

SERUM. The fluid part of the blood that remains after blood cells, platelets, and fibrogen have been removed. Also called blood serum.

SEVERANCE OF PARENTAL RIGHTS. The end of parental rights; the involuntarily removal of parental rights of a parent that has abandoned a child; has without just cause failed to support a child; has neglected or abused a child or has stood by and allowed others to neglect or abuse a child; or who because of extended incarceration in prison, is unavailable to parent or nurture the child. Once the parental rights of both parents of a child are removed the child will become available for adoption by another family.

SEXUAL ABUSE. Forced sexual contact through the use of threats or other fear tactics, or instances in which an individual is physically unable to decline sexual activity.

SEXUAL ASSAULT NURSE EXAMINER. A registered nurse who is trained to collect and document evidence from a sexual assault victim, evaluate and treat for sexually transmitted diseases and pregnancy, and refer victims to follow-up medical care and counseling.

SEXUAL ORIENTATION. The direction of somebody's sexual desire, toward people of the opposite sex (heterosexual or straight) or of the same sex (homosexual or gay), or of both sexes (bisexual).

SHAFT. The portion of the hair that extends from the follicle and goes beyond the surface of the epidermis.

SHAKEN BABY SYNDROME. Injuries caused by someone vigorously shaking an infant, usually for five to twenty seconds, which causes brain damage.

SHELTERED ENGLISH. Structured English immersion; English instruction for ELLs that focuses on content and skills rather than the language itself; uses simplified language, visual aids, physical activity, and the physical environment to teach academic subjects.

SHINGLES. An disease caused by an infection with the Herpes zoster virus, the same virus that causes chickenpox. Symptoms of shingles include pain and blisters along one nerve, usually on the face, chest, stomach, or back.

SHOCK. A medical emergency in which the organs and tissues of the body are not receiving an adequate flow of blood. This deprives the organs and tissues of oxygen and allows the build-up of waste products. Shock can be caused by certain diseases, serious injury, or blood loss.

SHUNT. A passageway (or an artificially created passageway) that diverts blood flow from one main route to another. Also refers to a small tube placed in a ventricle of the brain to direct cerebrospinal fluid away from a blockage into another part of the body.

SHYNESS. The feeling of insecurity when among people, talking with people, or asking somebody a favor.

SIBLING RIVALRY. Competition among brothers and sisters in a nuclear family. It is considered to be an important influence in shaping the personalities of children who grow up in middle-class Western societies but less relevant in traditional African and Asian cultures.

SICKLE CELL ANEMIA. An inherited disorder in which red blood cells contain an abnormal form of hemoglobin, a protein that carries oxygen. The abnormal form of hemoglobin causes the red cells to become sickle-shaped. The misshapen cells may clog blood vessels, preventing oxygen from reaching tissues and leading to pain, blood clots and other problems.

SICKLE CELL TRAIT. Condition that occurs in people who have one of two possible genes responsible for the abnormal hemoglobin of sickle cell anemia. People with

this trait may suffer milder symptoms of sickle cell anemia or may have no symptoms. Some scientists believe that the trait actually provides an advantage in tropical environments because the slightly altered shape of the blood cells cause a person to be more resistant to malaria.

SIDESTREAM SMOKE. The smoke that is emitted from the burning end of a cigarette or cigar, or that comes from the end of a pipe. Along with exhaled smoke, it is a constituent of second-hand smoke.

SIGMOID COLON. The final portion of the large intestine that empties into the rectum.

SIGMOIDOSCOPY. A procedure in which a thin, flexible, lighted instrument, called a sigmoidoscope, is used to visually examine the lower part of the large intestine. Colonoscopy examines the entire large intestine using the same techniques.

SILENT REFLUX. An acid reflux problem that does not cause vomiting but can cause chronic, recurrent respiratory symptoms much like asthma.

SIMULTANEOUS BILINGUALISM. Acquiring two languages simultaneously before the age of three.

SINUS. A tubular channel or cavity connecting one body part with another or with the outside. Often refers to one of the air-filled cavities surrounding the eyes and nose that are lined with mucus-producing membranes. They cleanse the nose, add resonance to the voice, and partially determine the structure of the face.

SKIN GRAFTING. A surgical procedure by which skin or a skin substitute is placed over a burn or non-healing wound to permanently replace damaged or missing skin or to provide a temporary wound covering.

SKIN HEMATOMA. Blood from a broken blood vessel that has accumulated under the skin.

SLEEP APNEA. A sleep disorder characterized by periods of breathing cessation lasting for 10 seconds or more.

SLEEP DISORDER. Any condition that interferes with sleep. Sleep disorders are characterized by disturbance in the amount of sleep, in the quality or timing of sleep, or in the behaviors or physiological conditions associated with sleep.

SLEEP LATENCY. The amount of time that it takes to fall asleep. Sleep latency is measured in minutes and is important in diagnosing depression.

SLEEP PARALYSIS. An abnormal episode of sleep in which the patient cannot move for a few minutes, usually occurring on falling asleep or waking up. Often found in patients with narcolepsy.

SLOW-WAVE SLEEP (SWS). Stage of deepest sleep characterized by absence of eye movements, decreased body temperature, and involuntary body movements. Night terrors and sleepwalking occur during this stage of sleep.

SOCIAL ANXIETY DISORDER. Persistent avoidance and/or discomfort in social situations that significantly interferes with functioning.

SOCIAL PHOBIA. An anxiety disorder characterized by a strong and persistent fear of social or performance situations in which the individual might feel embarassment or humiliation.

SOCIAL PROMOTION. Passing a child on to the next grade regardless of readiness in order for the child to remain with his or her age peers.

SOCIAL SKILLS. The knowledge of and ability to use a variety of social behaviors that are appropriate to interact positively with other people.

SOCIAL WITHDRAWAL. Avoidance of social contacts.

SOCIAL WORKER. Health care professional available to help patients and families manage the changes that may occur as a result of the patient's hospitalization. Socials workers provide referrals to community resources and can help the family make arrangements for care in the home as necessary after the patient is discharged from the hospital.

SOCIALIZATION. The process by which new members of a social group are integrated in the group.

SODIUM. An element; sodium is the most common electrolyte found in animal blood serum.

SODOMY. Anal intercourse.

SOMATIC CELLS. All the cells of the body with the exception of the egg and sperm cells.

SOMNAMBULISM. Another term for sleepwalking.

SORE. A wound, lesion, or ulcer on the skin.

SPACE MAINTAINER. An orthodontic appliance that is worn to prevent adjacent teeth from moving into the space left by an unerupted or prematurely lost tooth.

SPASTIC. Refers to a condition in which the muscles are rigid, posture may be abnormal, and fine motor control is impaired.

SPASTICITY. Increased mucle tone, or stiffness, which leads to uncontrolled, awkward movements.

SPATIAL SKILLS. The ability to locate objects in a three-dimensional world using sight or touch.

SPECTROPHOTOMETRY. A testing method that measures the amount of ultraviolet light absorbed by specific substances such as bilirubin pigment. A spectrophotometer can accurately measure how much bilirubin is in a blood sample and the result can be compared to known normal values.

SPEECH PATHOLOGIST. An individual certified by the American Speech-Language-Hearing Association (ASHA) to treat speech disorders.

SPERMATIC CORD. The tissue that suspends the testis inside the scrotum.

SPERMICIDE. A substance that kills sperm. Also called a spermatocide.

SPHINCTER. A circular band of muscle that surrounds and encloses an opening to the body or to one of its hollow organs. These muscles can open or close the opening by relaxing or contracting.

SPHYGMOMANOMETER. An instrument used to measure blood pressure.

SPINA BIFIDA. A birth defect (a congenital malformation) in which part of the vertebrae fail to develop completely so that a portion of the spinal cord, which is normally protected within the vertebral column, is exposed. People with spina bifida can suffer from bladder and bowel incontinence, cognitive (learning) problems, and limited mobility.

SPINAL CANAL. The opening that runs through the center of the spinal column. The spinal cord passes through the spinal canal. Also called the vertebral canal.

SPINAL CORD. The elongated nerve bundles that lie in the spinal canal and from which the spinal nerves emerge.

SPINAL CORD INJURY. Injury to the spinal cord, via blunt or penetrating trauma.

SPIROCHETE. A type of bacterium with a long, slender, coiled shape. Syphilis and Lyme disease are caused by spirochetes.

SPIROMETRY. A test using an instrument called a spirometer that measures how much and how fast the air is moving in and out of a patient's lungs. Spirometry can help a physician diagnose a range of respiratory diseases, monitor the progress of a disease, or assess a patient's response to treatment.

SPIRULINA. A genus of blue-green algae that is sometimes added to food to increase its nutrient value.

SPLENECTOMY. Surgical removal of the spleen.

SPLINT. A thin piece of rigid or flexible material that is used to restrain, support, or immobilize a part of the body while healing takes place.

SPONDYLOSIS. A condition in which one or more to the vertebral joints in the spine become stiff and/or fixed in one position.

SPORE. A dormant form assumed by some bacteria, such as anthrax, that enable the bacterium to survive high temperatures, dryness, and lack of nourishment for long periods of time. Under proper conditions, the spore may revert to the actively multiplying form of the bacteria. Also refers to the small, thick-walled reproductive structure of a fungus.

SPUTUM. The substance that is coughed up from the lungs and spit out through the mouth. It is usually a mixture of saliva and mucus, but may contain blood or pus in patients with lung abscess or other diseases of the lungs.

STANDARD DEVIATION. A measure of the distribution of scores around the average (mean). In a normal distribution, two standard deviations above and below the mean includes about 95% of all samples.

STANDARDIZATION. The process of determining established norms and procedures for a test to act as a standard reference point for future test results.

STANDARDIZED TEST. A test that follows a regimented structure, and each individual's scores may be compared with those of groups of people. In the case of the Cognistat, test taker's scores can be compared to groups of young adults, middle-aged adults, the geriatric, and people who have undergone neurosurgery.

STANFORD-BINET INTELLIGENCE SCALES. A device designed to measure somebody's intelligence, obtained through a series of aptitude tests concentrating on different aspects of intellectual functioning. An IQ score of 100 represents "average" intelligence.

STAPHYLOCOCCAL INFECTION. Infection with one of several species of *Staphylococcus* bacteria. Staphylococcal infections can affect any part of the body and are characterized by the formation of abscesses. Also known popularly as a staph infection.

STAPHYLOCOCCUS. Any of several species of spherical bacteria that occur in groups of four or in irregular clusters. They can infect various parts of the body, especially the skin and mucous membranes.

STATIC ENCEPHALOPATHY. A disease or disorder of the brain that does not get better or worse.

STEATORRHEA. An excessive amount of fat in the feces due to poor fat absorption in the gastrointestinal tract.

STEM CELL. An undifferentiated cell that retains the ability to develop into any one of a variety of cell types.

STENOSIS. A condition in which an opening or passageway in the body is narrowed or constricted.

STENT. A slender hollow catheter or rod placed within a vessel or duct to provide support or to keep it open.

STEPFAMILY. A family formed by the marriage or long-term cohabitation of two individuals, where one or both have at least one child from a previous relationship living part-time or full-time in the household. The individual who is not the biological parent of the child or children is referred to as the stepparent.

STEREOGNOSIS. The ability to recognize objects by sense of touch.

STEREOTACTIC TECHNIQUE. A technique used by neurosurgeons to pinpoint locations within the brain. It employs computer imaging to guide the surgeon to the exact location for the surgical procedure.

STEREOTYPED. Having a persistent, repetitive, and senseless quality. Tics are stereotyped movements or sounds.

STEROID. A class of drugs resembling normal body substances that often help control inflammation in the body tissues.

STEVENS-JOHNSON SYNDROME. A severe inflammatory skin eruption that occurs as a result of an allergic reaction or respiratory infection.

STIMULUS. Anything capable of eliciting a response in an organism or a part of that organism.

STOMATITIS. Inflammation of the mucous lining of any of the structures of the mouth, including the cheeks, gums, tongue, lips, and roof or floor of the mouth.

STOOL. The solid waste that is left after food is digested. Stool forms in the intestines and passes out of the body through the anus.

STRABISMUS. A disorder in which the eyes do not point in the same direction. Also called squint.

STRANGULATED HERNIA. A hernia that is so tightly incarcerated outside the abdominal wall that the intestine is blocked and the blood supply to that part of the intestine is cut off.

STRANGULATED OBSTRUCTION. An obstruction in which a loop of the intestine has its blood supply cut off.

STRAWBERRY TONGUE. A sign of scarlet fever in which the tongue appears to have a red coating with large raised bumps.

STREET DRUG. A substance purchased from a drug dealer. It may be a legal substance, sold illicitly (without a prescription, and not for medical use), or it may be a substance which is illegal to possess.

STREP THROAT. An infection of the throat caused by *Streptococcus* bacteria. Symptoms include sore throat, chills, fever, and swollen lymph nodes in the neck.

STREPTOCOCCUS. Plural, streptococci. Any of several species of spherical bacteria that form pairs or chains. They cause a wide variety of infections including scarlet fever, tonsillitis, and pneumonia.

STREPTOCOCCUS PYOGENES. A common bacterium that causes strep throat and can also cause tonsillitis.

STREPTOMYCIN. An antibiotic used to treat tuberculosis.

STRESS. A physical and psychological response that results from being exposed to a demand or pressure.

STRESSOR. A stimulus, or event, that provokes a stress response in an organism. Stressors can be categorized as acute or chronic, and as external or internal to the organism.

STRICTURE. An abnormal narrowing or tightening of a body tube or passage.

STRIDOR. A term used to describe noisy breathing in general and to refer specifically to a high-pitched crowing sound associated with croup, respiratory infection, and airway obstruction.

STROKE. Interruption of blood flow to a part of the brain with consequent brain damage. A stroke may be caused by a blood clot or by hemorrhage due to a burst blood vessel. Also known as a cerebrovascular accident.

STROMAL. Pertaining to the type of tissue that is associated with the support of an organ.

STRUCTURED ENGLISH IMMERSION. Sheltered English; English-only instruction for ELLs that uses simplified language, visual aids, physical activity, and the physical environment to teach academic subjects.

STUDENTS AGAINST DRUNK DRIVING (SADD). An organization that offers a "Contract for Life" that asks teens to discuss substance use with parents, to call home for a ride if safe transportation is needed, and to wear a seat belt. Parents in turn promise to arrange for that safe transportation home "regardless of the time or circumstances," without discussion of the incident until both teens and parents are calm.

STUPOR. A trance-like state that causes a person to appear numb to their environment.

STY. An external hordeolum caused by an infection of an oil gland on the eyelid.

SUBARACHNOID. Referring to the space underneath the arachnoid membrane, the middle of the three membranes that sheath the spinal cord and brain.

SUBARACHNOID HEMORRHAGE. A collection of blood in the subarachnoid space, the space between the arachnoid and pia mater membranes that surround the brain. This space is normally filled with cerebrospinal fluid. A subarachnoid hemorrhage can lead to stroke, seizures, permanent brain damage, and other complications.

SUBCUTANEOUS. Referring to the area beneath the skin.

SUBDURAL HEMATOMA. A localized accumulation of blood, sometimes mixed with spinal fluid, in the space between the middle (arachnoid) and outer (dura mater) membranes covering the brain. It is caused by an injury to the head that tears blood vessels.

SUBPERIOSTEAL APPOSITION. The process by which bones are made thicker from the outside.

SUBSTANCE ABUSE. Maladaptive pattern of drug or alcohol use that may lead to social, occupational, psychological, or physical problems.

SUBUNGUAL HEMATOMA. Accumulation of blood under a nail.

SUCCIMER. A chelating agent that is used to remove excess lead from the body. Sold under the trade name Chemet.

SUDDEN INFANT DEATH SYNDROME (SIDS). The general term given to ''crib deaths'' of unknown causes.

SUGARS. Those carbohydrates having the general composition of one part carbon, two parts hydrogen, and one part oxygen.

SULFONYLUREA DRUG. A medication for type 2 diabetes that causes the pancreas to produce more insulin, and may trigger hypoglycemia in some people.

SUNSCREEN. A product that blocks the damaging rays of the sun. Good sunscreens contain either para-aminobenzoic acid (PABA) or benzophenone, or both. Sunscreen protection factors range from two to 45.

SUNSTROKE. Heatstroke caused by direct exposure to the sun in which body temperature increases to dangerously high levels.

SUPERIOR MESENTERIC ARTERY SYNDROME. A condition in which a person vomits after meals due to blockage of the blood supply to the intestine.

SUPINE. Lying on the back with the face upward.

SUPRACHIASMATIC NUCLEI (SCN). SCN is that part of the brain that functions as a person's ''biological clock'' to regulate many body rhythms. The SCN is located on top of the main junction of nerve fibers that connects to the eyes.

SUPRAGLOTTITIS. Another term for epiglottitis.

SUPRAVALVULAR AORTIC STENOSIS (SVAS). A narrowing of the aorta.

SURFACTANT. A protective film secreted by the alveoli in the lungs that reduces the surface tension of lung fluids, allowing gas exchange and helping maintain the elasticity of lung tissue. Surfactant is normally produced in the fetal lungs in the last months of pregnancy, which helps the air sacs to open up at the time of birth so that the newborn infant can breathe freely. Premature infants may lack surfactant and are more susceptible to respiratory problems without it.

SUTURE. A ''seam'' that joins two surfaces together, such as is found between the bones of the skull. Also refers to stitching together the torn or cut edges of tissue.

SWADDLING. To wrap the infant securely in clothing or blankets; to provide comfort and control.

SWIMMER'S ITCH. An allergic skin inflammation caused by a sensitivity to flatworms that die under the skin, resulting in an itchy rash.

SYMMETRIC. Occurring on both sides of the body, in a mirror-image fashion.

SYNCOPE. A loss of consciousness over a short period of time, caused by a temporary lack of oxygen in the brain; a faint.

SYNDROME. A group of signs and symptoms that collectively characterize a disease or disorder.

SYNOVIAL JOINT. A fully moveable joint in which a synovial cavity is present between two articulating bones. Also called a diarthrosis.

SYNOVIAL MEMBRANE. The membrane that lines the inside of the articular capsule of a joint, and produces a lubricating fluid called synovial fluid.

SYNOVITIS. Inflammation of the synovial membrane, the membrane that lines the inside of the articular capsule of a joint.

SYPHILIS. This disease occurs in two forms. One is a sexually transmitted disease caused by A systemic infection caused by the spirochete *Treponema pallidum*. It is most commonly transmitted by sexual contact.

SYRINGOMYELIA. Excessive fluid in the spinal cord.

SYRINX. A tubular fluid-filled cavity within the spine.

SYSTEMIC. Relating to an entire body system or the body in general.

SYSTEMIC ABSORPTION. Any substance topical, inhaled, or ingested that is absorbed into the bloodstream and distributed throughout the body.

SYSTEMIC CIRCULATION. Refers to the general blood circulation of the body, not including the lungs.

SYSTOLIC BLOOD PRESSURE. Blood pressure when the heart contracts (beats).

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T LYMPHOCYTES. Also called T cells, a type of white blood cell that is produced in the bone marrow and matured in the thymus gland. These specialized blood cells recognize invading organisms (helper T lymphocytes) and destroy them (killer T lymphocytes).

TACHYPNEA. Rapid breathing.

TAKAYASU ARTERITIS. A disease in which the aorta and its major branches become inflamed. It is often accompanied by high blood pressure, an abnormal pulse, and visual symptoms.

TARTAR. A hardened yellow or brown mineral deposit from unremoved plaque. Also called calculus.

TASK. A goal directed activity used in assessment.

TAY-SACHS DISEASE. An inherited disease caused by a missing enzyme that is prevalent among the Ashkenazi Jewish population of the United States. Infants with the disease are unable to process a certain type of fat which accumulates in nerve and brain cells, causing mental and physical retardation, and, finally, death.

TELANGIECTASIA. Abnormal dilation of capillary blood vessels leading to the formation of telangiectases or angiomas.

TELECOMMUTING. A form of employment in which the employee works at home on a computer linked to the company's central office.

TEMPERAMENT. A person's natural disposition or inborn combination of mental and emotional traits.

TEMPORAL BONES. The compound bones that form the left and right sides of the skull and contain various cavities associated with the ear.

TEMPOROMANDIBULAR JOINT (TMJ). One of a pair of joints that attaches the mandible of the jaw to the temporal bone of the skull. It is a combination of a hinge and a gliding joint.

TEMPOROMANDIBULAR JOINT DISORDER. Inflammation, irritation, and pain of the jaw caused by improper opening and closing of the temporomandibular joint. Other symptoms include clicking of the jaw and a limited range of motion. Also called temporomandibular joint syndrome.

TENDINITIS. Inflammation of a tendon (a tough band of tissue that connects muscle to bone) that is often the result of overuse over a long period of time.

TENDON. A tough cord of dense white fibrous connective tissue that connects a muscle with some other part, especially a bone, and transmits the force which the muscle exerts.

TENOSYNOVITIS. Inflammation of a tendon and its enveloping sheath, usually resulting from overuse injury.

TENOTOMY. A surgical procedure that cuts the tendon of a contracted muscle to allow lengthening.

TERATOGEN. Any drug, chemical, maternal disease, or exposure that can cause physical or functional defects in an exposed embryo or fetus.

TESTICULAR TORSION. A condition involving the twisting of the spermatic cord inside the testicle that shuts off its blood supply and can seriously damage the testicle.

TESTOSTERONE. Male hormone produced by the testes and (in small amounts) in the ovaries. Testosterone is responsible for some masculine secondary sex characteristics such as growth of body hair and deepening voice. It also is sometimes given as part of hormone replacement therapy to women whose ovaries have been removed.

TETANUS. A potentially fatal infection caused by a toxin produced by the bacterium *Clostridium tetani*. The bacteria usually enter the body through a wound and the toxin they produce affects the central nervous system causing painful and often violent muscular contractions. Commonly called lockjaw.

TETRACYCLINE. A broad-spectrum antibiotic.

THALAMUS. A pair of oval masses of gray matter within the brain that relay sensory impulses from the spinal cord to the cerebrum.

THERMOGRAPHY. Use of a heat-sensitive device for measuring blood flow.

THIMEROSAL. A mercury-containing preservative used in some vaccines.

THRESHOLD. The minimum level of stimulation necessary to produce a response.

THROMBOCYTE. Another name for platelet.

THROMBOCYTOPENIA. A persistent decrease in the number of blood platelets usually associated with hemorrhaging.

THROMBOCYTOSIS. An abnormally high platelet count. It occurs in polycythemia vera and other disorders in which the bone marrow produces too many platelets.

THROMBOLYSIS. The process of dissolving a blood clot.

THROMBOLYTICS. Drugs that dissolve blood clots. Thrombolytics are used to treat embolisms.

THROMBOSIS. The formation of a blood clot in a vein or artery that may obstruct local blood flow or may dislodge, travel downstream, and obstruct blood flow at a remote location. The clot or thrombus may lead to infarction, or death of tissue, due to a blocked blood supply.

THROMBUS. A blood clot that forms within a blood vessel or the heart.

THRUSH. An infection of the mouth, caused by the yeast *Candida albicans* and characterized by a whitish growth and ulcers.

THYMIC APLASIA. A lack of T lymphocytes, due to failure of the thymus to develop, resulting in very reduced immunity.

THYMUS GLAND. An endocrine gland located in the upper chest just below the neck that functions as part of the lymphatic system. It coordinates the development of the immune system.

THYROIDECTOMY. Surgical removal of all or part of the thyroid gland.

THYROID-STIMULATING HORMONE (TSH). A hormone produce by the pituitary gland that stimulates the thyroid gland to produce the hormones that regulate metabolism. Also called thyrotropin.

THYROXINE (T_4). The thyroid hormone that regulates many essential body processes.

TIC. A brief and intermittent involuntary movement or sound.

TIME-OUT. A discipline strategy that entails briefly isolating a disruptive child in order to interrupt and avoid reinforcement of negative behavior.

TINNITUS. A noise, ranging from faint ringing or thumping to roaring, that originates in the ear not in the environment.

TITER. The highest dilution of a material (e.g., serum or other body fluid) that produces a reaction in an immunologic test system. Also refers to the extent to which an antibody can be diluted before it will no longer react with a specific antigen. Also spelled titre.

TOCOLYTIC DRUG. A compound given to women to stop the progression of labor.

TOLERANCE. A condition in which an addict needs higher doses of a substance to achieve the same effect previously achieved with a lower dose.

TONGUE THRUSTING. A physiological behavior that causes the tongue to flatten and thrust forward during swallowing and speaking.

TONIC-CLONIC SEIZURE. This is the most common type of seizure among all age groups and is categorized into several phases beginning with vague symptoms hours or days before an attack. These seizures are sometimes called grand mal seizures.

TONSILLECTOMY. A surgical procedure to remove the tonsils. A tonsillectomy is performed if the patient has recurrent sore throats or throat infections, or if the tonsils have become so swollen that the patient has trouble breathing or swallowing.

TONSILS. Common name for the palatine tonsils, which are lymph masses in the back of the mouth, on either side of the tongue. Tonsils act like filters to trap bacteria and viruses.

TOOTH ERUPTION. The emergence of a tooth through the gum.

TOPICAL. Not ingested; applied to the outside of the body, for example to the skin, eye, or mouth.

TOP-LEVEL REACHING. The ability of an infant to grasp an object that is within reach, looking only at the object and not at their hands. Typically develops between four and five months of age.

TOTAL PLEXUS PALSY. Erb/Klumpke palsy; a condition resulting from injury involving all of the brachial plexus nerves and affecting the entire upper extremity of the body.

TOURETTE SYNDROME. A neurological disorder characterized by multiple involuntary movements and uncontrollable vocalizations called tics that come and go over years, usually beginning in childhood and becoming chronic. Sometimes the tics include inappropriate or obscene language (coprolalia).

TOURNIQUET. Any device that is used to compress a blood vessel to stop bleeding or as part of collecting a blood sample. Phlebotomists usually use an elastic band as a tourniquet.

TOXICOLOGY. The branch of medical pharmacology dealing with the detection, effects, and antidotes of poisons.

TOXIN. A poisonous substance usually produced by a microorganism or plant.

TOXOID. A preparation made from inactivated exotoxin, used in immunization.

TOXOPLASMOSIS. A parasitic infection caused by the intracellular protozoan *Toxoplasmosis gondii*. Humans are most commonly infected by swallowing the oocyte form of the parasite in soil (or kitty litter) contaminated by feces from an infected cat; or by swallowing the cyst form of the parasite in raw or undercooked meat.

TRACE ELEMENT. An element that is required in only minute quantities for the maintenance of good health. Trace elements are also called micronutrients.

TRACHEA. The windpipe. A tube composed of cartilage and membrane that extends from below the voice box into the chest where it splits into two branches, the bronchi, that lead to each lung.

TRACHEOESOPHAGEAL FISTULA. An abnormal connection between the trachea and esophagus, frequently associated with the esophagus ending in a blind pouch.

TRACHEOSTOMY. A procedure in which a small opening is made in the neck and into the trachea or windpipe. A breathing tube is then placed through this opening.

TRACHEOTOMY. An surgical procedure in which the surgeon cuts directly through the patient's neck into the windpipe below a blockage in order to keep the airway open.

TRACHOMA. A type of chlamydia that causes blindness.

TRACTION. The process of placing a bone, limb, or group of muscles under tension by applying weights and pulleys. The goal is to realign or immobilize the part or to relieve pressure on that particular area to promote healing and restore function.

TRAIT. A distinguishing feature of an individual.

TRANQUILIZER. A medication that has a calming effect and is used to treat anxiety and mental tension.

TRANS-FATTY ACID. A type of fat created by hydrogenating polyunsaturated oils. This changes the double bond on the carbon atom from a cis configuration to a trans configuration, making the fatty acid saturated, and a greater health concern. For example, stick margarines are known to contain more trans-fatty acids than liquid oils.

TRANSGENDER. Any person who feels their assigned gender does not completely or adequately reflect their internal gender, such as a biological male who perceives himself to be female.

TRANSILLUMINATION. A technique of checking for tooth decay by shining a light behind the patient's teeth. Decayed areas show up as spots or shadows.

TRANSITIONAL BILINGUAL EDUCATION (TBE). Bilingual education that includes ESL and academic classes conducted in a child's primary language.

TRANSLOCATION. The transfer of one part of a chromosome to another chromosome during cell division. A balanced translocation occurs when pieces from two different chromosomes exchange places without loss or gain of any chromosome material. An unbalanced translocation involves the unequal loss or gain of genetic information between two chromosomes.

TRANSPLACENTAL. Passing through or occurring across the placenta.

TRANS-RACIAL ADOPTIONS. Adoption in which a family of one race adopts a child of another race.

TRANSSEXUALISM. A term used to describe a male or female that feels a strong identification with the opposite sex and experiences considerable distress because of their actual sex. Also called gender identity disorder.

TRAUMA. Serious physical injury. Also refers to a disastrous or life-threatening event that can cause severe emotional distress, including dissociative symptoms and disorders.

TRAUMATIC SHOCK. A condition of depressed body functions as a reaction to injury with loss of body fluids or lack of oxygen. Signs of traumatic shock include weak and rapid pulse, shallow and rapid breathing, and pale, cool, clammy skin.

TRAVELER'S DIARRHEA. An illness due to infection from a bacteria or parasite that occurs in persons traveling to areas where there is a high frequency of the illness. The disease is usually spread by contaminated food or water.

TREMOR. Involuntary shakiness or trembling.

TREPONEME. A term used to refer to any member of the genus *Treponema*, which is an anaerobic bacteria consisting of cells, 3–8 micrometers in length, with acute, regular, or irregular spirals and no obvious protoplasmic structure.

TRETINOIN. A drug, used in the treatment of acne, that works by increasing the turnover (death and replacement) of skin cells.

TRIANGLING. A process in which two family members lower the tension level between them by drawing in a third member.

TRICYCLIC ANTIDEPRESSANT. A class of antidepressants, named for their three-ring structure, that increase the levels of serotonin and other brain chemicals. They are used to treat depression and anxiety disorders, but have more side effects than the newer class of antidepressants called selective serotonin reuptake inhibitors (SSRIs).

TRIGGER. Any situation (people, places, times, events, etc.) that causes one to experience a negative emotional reaction, which is often accompanied by a display of symptoms or problematic behavior.

TRIGLYCERIDE. A substance formed in the body from fat in the diet. Triglycerides are the main fatty materials in the blood. Bound to protein, they make up high- and low-density lipoproteins (HDLs and LDLs). Triglyceride levels are important in the diagnosis and treatment of many diseases including high blood pressure, diabetes, and heart disease.

TRIGONOCEPHALY. An abnormal development of the skull characterized by a triangular shaped forehead.

TRIIODOTHYRONINE (T_3). A thyroid hormone similar to thyroxine but more powerful. Preparations of triiodothyronine are used in treating hypothyroidism.

TRIMESTER. The one of three periods of about 13 weeks each into which a pregnancy is divided.

TRINUCLEOTIDE REPEAT EXPANSION. A sequence of three nucleotides that is repeated too many times in a section of a gene.

TRISKAIDEKAPHOBIA. Fear of the number 13.

TRISOMY. An abnormal condition where three copies of one chromosome are present in the cells of an individual's body instead of two, the normal number.

TRUNK. That part of the body that does not include the head, arms, and legs. Also called the torso.

TRYPTOPHAN. An essential amino acid that has to consumed in the diet because it cannot be manufactured by the body. Tryptophan is converted by the body to niacin, one of the B vitamins, and serotonin, a neurotransmitter.

TUBERCULOSIS. Tuberculosis (TB) is a potentially fatal contagious disease that can affect almost any part of the body, but is mainly an infection of the lungs. It is caused by a bacterial microorganism, the tubercle bacillus or *Mycobacterium tuberculosis*. Symptoms include fever, weight loss, and coughing up blood.

TUBEROUS SCLEROSIS. A genetic condition that affects many organ systems including the brain, skin, heart, eyes, and lungs. Benign (non-cancerous) growths or tumors called hamartomas form in various parts of the body, disrupting their normal function.

TUBULE. Tissues and cells associated with the structures that connect the renal pelvis to the glomeruli.

TUMOR. A growth of tissue resulting from the uncontrolled proliferation of cells.

TUMOR-SUPPRESSOR GENE. A gene involved in controlling normal cell growth and preventing cancer.

TURNER SYNDROME. A chromosome abnormality characterized by short stature and ovarian failure caused by an absent X chromosome. It occurs only in females.

TWELVE-STEP PROGRAMS. Several programs to assist in breaking addictions, offering either support to addicted people or to friends and loved ones of addicted people. These programs are spiritual but not religious and are based on the twelve steps that are the basis of Alcoholics Anonymous (AA). Programs include AA, Narcotics Anonymous (NA), Al-Anon, Adult Children of Alcoholics (ACOA), Alateen, and Co-Dependence Anonymous (CODA).

25-HYDROXY-VITAMIN D. The form of vitamin D that is measured in order to assess vitamin D deficiency.

TWIN-TWIN TRANSFUSION SYNDROME (TTTS). A condition in identical monochorionic twins in which there is a connection between the two circulatory systems so that the donor twin pumps the blood to the recipient twin without a return of blood to the donor.

TWO-WAY BILINGUAL EDUCATION. Dual language programs in which English and a second language are both used in classes consisting of ELLs and native-English speakers.

TYMPANIC MEMBRANE. The eardrum, a thin disc of tissue that separates the outer ear from the middle ear. It can rupture if pressure in the ear is not equalized during airplane ascents and descents.

TYMPANOMETRY. A test where air pressure in the ear canal is varied to test the condition and movement of the ear drum. This test is useful in detecting disorders of the middle ear.

TYMPANOSTOMY TUBE. An ear tube. A tympanostomy tube is small tube made of metal or plastic that is inserted during myringotomy to ventilate the middle ear.

TYPE. A category used to define personality, usually based on a theory of some kind. Inhibited and uninhibited are examples of personality types.

TYPHOID FEVER. A severe infection caused by a bacterium, *Salmonella typhi*. People with this disease have a lingering fever and feel depressed and exhausted. Diarrhea and rose-colored spots on the chest and abdomen are other symptoms. The disease is spread through poor sanitation.

TYROSINASE. An enzyme in a pigment cell which helps change tyrosine to dopa during the process of making melanin.

TYROSINE. An amino acid synthesized by the body from the essential amino acid phenylalanine. It is used by the body to make melanin and several hormones, including epinephrine and thyroxin.

U

ULCER. A site of damage to the skin or mucous membrane that is characterized by the formation of pus, death of tissue, and is frequently accompanied by an inflammatory reaction.

ULCERATED. Characterized by the formation of an ulcer.

ULCERATIVE COLITIS. A form of inflammatory bowel disease characterized by inflammation of the mucous lining of the colon, ulcerated areas of tissue, and bloody diarrhea.

ULNA. One of the two bones of the forearm. Two pivot joints join it to the radius, one near the elbow, one near the wrist.

ULTRASONOGRAPHY. A medical test in which sound waves are directed against internal structures in the body. As sound waves bounce off the internal structure, they create an image on a video screen. Ultrasonography is often used to diagnose fetal abnormalities, gallstones, heart defects, and tumors. Also called ultrasound imaging.

ULTRAVIOLET (UV) RADIATION. A portion of the light spectrum with a wavelength just below that of visible light. UV radiation is damaging to DNA and can destroy microorganisms. It may be responsible for sunburns, skins cancers, and cataracts in humans. Two bands of the UV spectrum, UVA and UVB, are used to treat psoriasis and other skin diseases.

UMBILICAL. Refers to the opening in the abdominal wall where the blood vessels from the placenta enter.

UMBILICAL CORD. The blood vessels that allow the developing baby to receive nutrition and oxygen from its mother; the blood vessels also eliminate the baby's waste products. One end of the umbilical cord is attached to the placenta and the other end is attached to the baby's belly button (umbilicus).

UMBILICAL CORD PROLAPSE. A birth situation in which the umbilical cord, the structure that connects the placenta to the umbilicus of the fetus to deliver oxygen and nutrients, falls out of the uterus and becomes compressed, thus preventing the delivery of oxygen.

UNCIRCUMCISED. Not having had the foreskin of the penis removed.

UNDERLYING CONDITION. Disorder or disease that causes the appearance of another medical disorder or condition.

UNDESCENDED TESTICLE. A testicle that is still in the groin and has not made its way into the scrotum.

UNIFOCAL. Only one tumor present in one eye.

UNILATERAL. Refers to one side of the body or only one organ in a pair.

UNILATERAL CLEFT. A cleft that occurs on only the right or left side of the lip.

UNILATERAL NEGLECT. Also called one-sided neglect. A side effect of stroke in which the stroke survivor ignores or forgets the weaker side of the body caused by the stroke.

UNIPARENTAL DISOMY. Chromosome abnormality in which both chromosomes in a pair are inherited from the same parent.

URETER. The tube that carries urine from the kidney to the bladder; each kidney has one ureter.

URETEROVESICAL JUNCTION. The point where the ureter joins the bladder.

URETEROVESICAL VALVE. A sphincter (an opening controlled by a circular muscle), located where the ureter enters the bladder, that keeps urine from flowing backward toward the kidney.

URETHRA. A passageway from the bladder to the outside of the body for the discharge of urine. In the female this tube lies between the vagina and clitoris; in the male the urethra travels through the penis and opens at the tip. In males, seminal fluid and sperm also pass through the urethra.

URETHRAL MEATUS. The opening of the urethra on the body surface through which urine is discharged.

URETHRITIS. Inflammation of the urethra, the tube through which the urine moves from the bladder to the outside of the body.

UROGENITAL. Refers to both the urinary system and the sexual organs, which form together in the developing embryo.

UROLOGIST. A physician who specializes in the anatomy, physiology, diseases, and care of the urinary tract (in men and women) and male reproductive tract.

URTICARIA. An itchy rash usually associated with an allergic reaction. Also known as hives.

URUSHIOL. The oil from poison ivy, oak, and sumac that causes severe itching, blistering, and rash.

UTEROPLACENTAL INSUFFICIENCY. Designates the lack of blood flow from the uterus to the placenta, resulting in decreased nourishment and oxygen to the fetus.

UTERUS. The female reproductive organ that contains and nourishes a fetus from implantation until birth. Also called the womb.

UVEITIS. Inflammation of all or part the uvea. The uvea is a continuous layer of tissue that consists of the iris, the ciliary body, and the choroid. The uvea lies between the retina and sclera.

V

VACCINATION. Another word for immunization.

VACCINE. A substance prepared from a weakened or killed microorganism which, when injected, helps the body to form antibodies that will prevent infection by the natural microorganism.

VACCINE ADVERSE EVENT REPORTING SYSTEM (VAERS). A federal government program for reporting adverse reactions to the administration of a vaccine.

VACCINE INJURY COMPENSATION PROGRAM (VICP). A program through which victims of vaccine-induced injury or death can be awarded financial compensation.

VACCINE INJURY TABLE. The guidelines by which claims to the VICP are evaluated; includes the vaccines, injuries or other conditions, and the allowable time periods for coverage by the VICP.

VARICELLA ZOSTER. The virus that causes chickenpox (varicella).

VARICELLA-ZOSTER IMMUNE GLOBULIN. A substance that can reduce the severity of chickenpox symptoms.

VARIVAX. The brand name for varicella virus vaccine live, an immunizing agent used to prevent infection by the *Herpes (varicella) zoster* virus. The vaccine works by causing the body to produce its own protection (antibodies) against the virus.

VASCULAR. Pertaining to blood vessels.

VASCULAR MALFORMATION. Abnormally formed blood or lymph vessels.

VASCULOPATHY. Any disease or disorder that affects the blood vessels.

VECTOR. A carrier organism (such as a fly or mosquito) which serves to deliver a virus (or other agent of infection) to a host. Also refers to a retrovirus that had been modified and is used to introduce specific genes into the genome of an organism.

VEGAN. A vegetarian who does not eat eggs or dairy products.

VEIN. A blood vessel that returns blood to the heart from the body. All the veins from the body converge into two major veins that lead to the right atrium of the heart. These veins are the superior vena cava and the inferior vena cava. The pulmonary vein carries the blood from the right ventricle of the heart into the lungs.

VELAMENTOUS INSERTION OF THE UMBILICAL CORD. The attachment of the umbilical cord close to the membranes (bag of water) or in the membranes.

VENOARTERIAL (V-A) BYPASS. The type of extracorporeal membrane oxygenation that provides both heart and lung support, using two tubes (one in the jugular vein and one in the carotid artery).

VENOVENOUS (V-V) BYPASS. The type of extracorporeal membrane oxygenation that provides lung support only, using a tube inserted into the jugular vein.

VENTILATOR. A mechanical device that can take over the work of breathing for a patient whose lungs are injured or are starting to heal. Sometimes called a respirator.

VENTRICLES. The lower pumping chambers of the heart. The ventricles push blood to the lungs and the rest of the body.

VENTRICLES OF THE BRAIN. The spaces within the brain where cerebrospinal fluid is made.

VENTRICULAR FIBRILLATION. An arrhythmia characterized by a very rapid, uncoordinated, ineffective series of contractions throughout the lower chambers of the heart. Unless stopped, these chaotic impulses are fatal.

VENTRICULAR SEPTAL DEFECT. An opening between the right and left ventricles of the heart.

VENULES. The smallest veins.

VERMILION BORDER. The line between the lip and the skin.

VERTEBRAE. Singular, vertebra. The individual bones of the spinal column that are stacked on top of each other. There is a hole in the center of each bone, through which the spinal cord passes.

VERTEX. The top of the head or highest point of the skull.

VERTIGO. A feeling of dizziness together with a sensation of movement and a feeling of rotating in space.

VESICLE. A bump on the skin filled with fluid.

VESTIBULAR SYSTEM. The brain and parts of the inner ear that work together to detect movement and position.

VIBRATION. The treatment that is applied to help break up lung secretions. Vibration can be either mechanical or manual. It is performed as the patient breathes deeply. When done manually, the person performing the vibration places his or her hands against the patient's chest and creates vibrations by quickly contracting and relaxing arm and shoulder muscles while the patient exhales. The procedure is repeated several times each day for about five exhalations.

VIBROACOUSTIC STIMULATION. In the biophysical profile, use of an artificial larynx to produce a loud noise to "awaken" the fetus.

VIDEO GAME RELATED SEIZURES (VGRS). Seizures thought to be brought on by the flashing lights and complex graphics of a video game.

VILLI. Tiny, finger-like projections that enable the small intestine to absorb nutrients from food.

VIRAL MENINGITIS. Meningitis caused by a virus. Also called aseptic meningitis.

VIRILIZING SYNDROMES. Abnormalities in female hormone production that produce male characteristics.

VIRUS. A small infectious agent consisting of a core of genetic material (DNA or RNA) surrounded by a shell of protein. A virus needs a living cell to reproduce.

VISCOSITY. Thickness of a liquid.

VISUAL ACUITY. Sharpness or clearness of vision.

VISUAL PERCEPTION (VP). The ability to perceive or understand what is being seen; the integration of an image with an idea of what it represents.

VISUOSENSORY. Pertaining to the perception of visual stimuli.

VITAMIN STATUS. The state of vitamin sufficiency or deficiency of any person. For example, a test may reveal that a patient's folate status is sufficient, borderline, or severely inadequate.

VITAMINS. Small compounds required for metabolism that must be supplied by diet, microorganisms in the gut (vitamin K) or sunlight (UV light converts pre-vitamin D to vitamin D).

VITREOUS. The transparent gel that fills the back part of the eye.

VITREOUS SEEDING. Small pieces of tumor have broken off and are floating around the vitreous.

VOCATIONAL. Relating to an occupation, career, or job.

VOID. To empty the bladder.

VOIDING CYSTOGRAM. A radiographic image of the mechanics of urination.

VOLUNTARY MUSCLES. Muscles that can be moved by conscious thought.

VOLVULUS. A twisting of the intestine that causes an obstruction.

VON WILLEBRAND FACTOR (VWF). A protein found in the blood that is involved in the process of blood clotting.

VULVA. The external genital organs of a woman, including the outer and inner lips, clitoris, and opening of the vagina.

W

WARFARIN. An anticoagulant drug given to treat existing blood clots or to control the formation of new blood clots. Sold in the United States under the brand name Coumadin.

WATER-SOLUBLE VITAMINS. Vitamins that are not stored in the body and are easily excreted. They must, therefore, be consumed regularly as foods or supplements to maintain health.

WEBBING. A tissue or membrane that connects two digits at their base or for the greater part of their length.

WECHSLER INTELLIGENCE SCALES. A test that measures verbal and non-verbal intelligence.

WEPMAN'S AUDITORY DISCRIMINATION TEST (WADT). A commonly used test for evaluating auditory discrimination skills.

WHEAL. A smooth, slightly elevated area on the body surface that is redder or paler than the surrounding skin.

WHITE BLOOD CELLS. A group of several cell types that occur in the bloodstream and are essential for a properly functioning immune system; they fight infection.

WHITE MATTER. A substance, composed primarily of myelin fibers, found in the brain and nervous system that protects nerves and allows messages to be sent to and from the brain and various parts of the body. Also called white substance.

WHOLE BLOOD. Blood which contains red blood cells, white blood cells, and platelets in plasma.

WHOOPING COUGH. An infectious disease of the respiratory tract caused by a bacterium, *Bordetella pertussis*. Also known as pertussis.

WILSON'S DISEASE. A rare inherited disease in which excessive amounts of copper accumulate in the liver or brain. It is fatal unless the patient complies with lifelong treatment with penicillamine and zinc oxidase. Wilson's disease is also known as inherited copper toxicosis.

WISDOM TEETH. The third molars at that back of the mouth.

WITHDRAWAL SYMPTOMS. A group of physical and/or mental symptoms that may occur when a person suddenly stops using a drug or other substance upon which he or she has become dependent.

WORLD HEALTH ORGANIZATION (WHO). An international organization within the United Nations system that is concerned with world health and welfare.

X

X CHROMOSOME. One of the two sex chromosomes (the other is Y) that determine a person's gender. Normal males have both an X and a Y chromosome, and normal females have two X chromosomes.

XENOPHOBIA. Fear of strangers or foreigners.

XEROSIS. The medical term for dry skin. Many children diagnosed with atopic dermatitis have a history of xerosis even as newborns.

X-LINKED. A gene carried on the X chromosome, one of the two sex chromosomes.

X RAYS. High-energy radiation used in high doses, either to diagnose or treat disease.

XYY SYNDROME. A chromosome disorder that affects males.

Y

YELLOW FEVER. An infectious disease caused by a virus. The disease, which is spread by mosquitoes, is most common in Central and South America and Central Africa. Symptoms include high fever, jaundice (yellow eyes and skin) and dark-colored vomit, a sign of internal bleeding. Yellow fever can be fatal.

YOGI (FEMALE, YOGINI). A trained yoga expert.

Z

ZOONOSIS. Any disease of animals that can be transmitted to humans. Rabies is an example of a zoonosis.

ZOOPHOBIA. Fear of animals.

ZYGOTE. The result of the sperm successfully fertilizing the ovum. The zygote is a single cell that contains the genetic material of both the mother and the father.

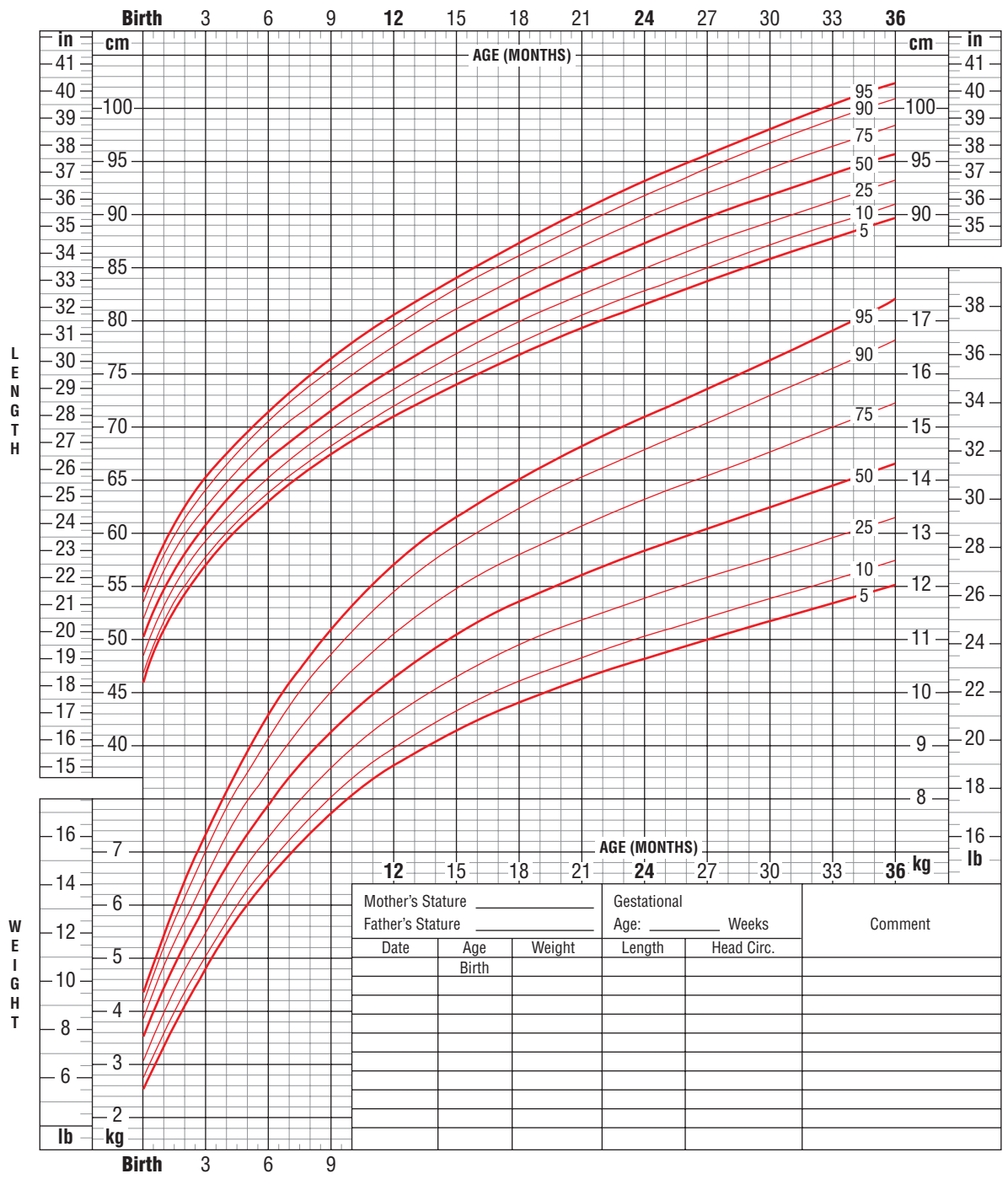
GROWTH CHARTS APPENDIX

The following growth charts were obtained from the United States Centers for Disease Control. These charts, updated in 2000, include norms for head circumference, weight, and length for boys and girls from birth through age three. Separate charts display percentiles for height, weight, and body mass index (BMI) for boys and girls through age 20.

Birth to 36 months: Boys
Length-for-age and Weight-for-age percentiles

NAME _____

RECORD # _____



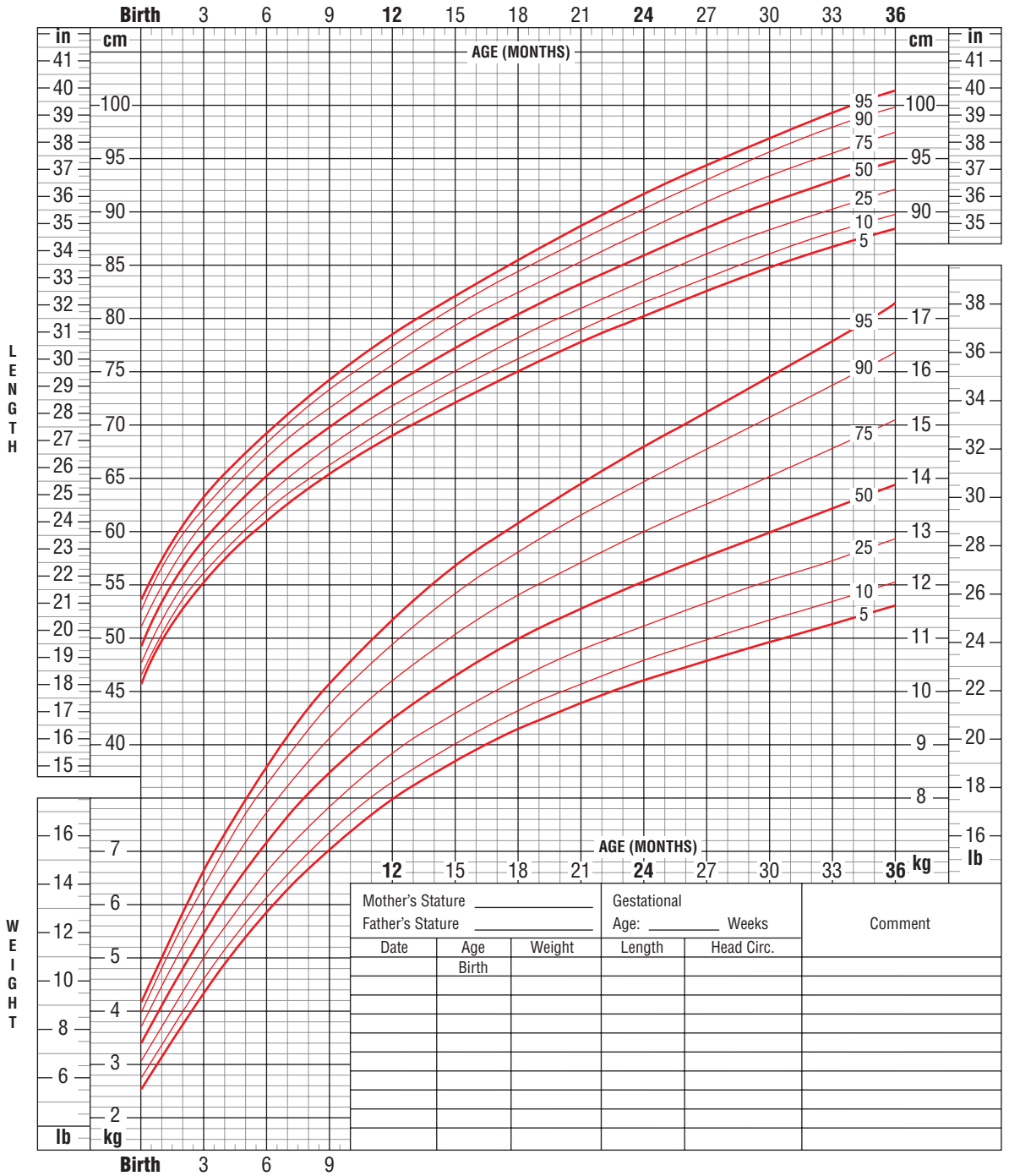
Published May 30, 2000 (modified 4/20/01).

SOURCE: Developed by the National Center for Health Statistics in collaboration with the National Center for Chronic Disease Prevention and Health Promotion (2000). http://www.cdc.gov/growthcharts

Birth to 36 months: Girls
Length-for-age and Weight-for-age percentiles

NAME _____

RECORD # _____



Mother's Stature _____
Father's Stature _____

Gestational Age: _____ Weeks

Date	Age	Weight	Length	Head Circ.	Comment
	Birth				

Published May 30, 2000 (modified 4/20/01).

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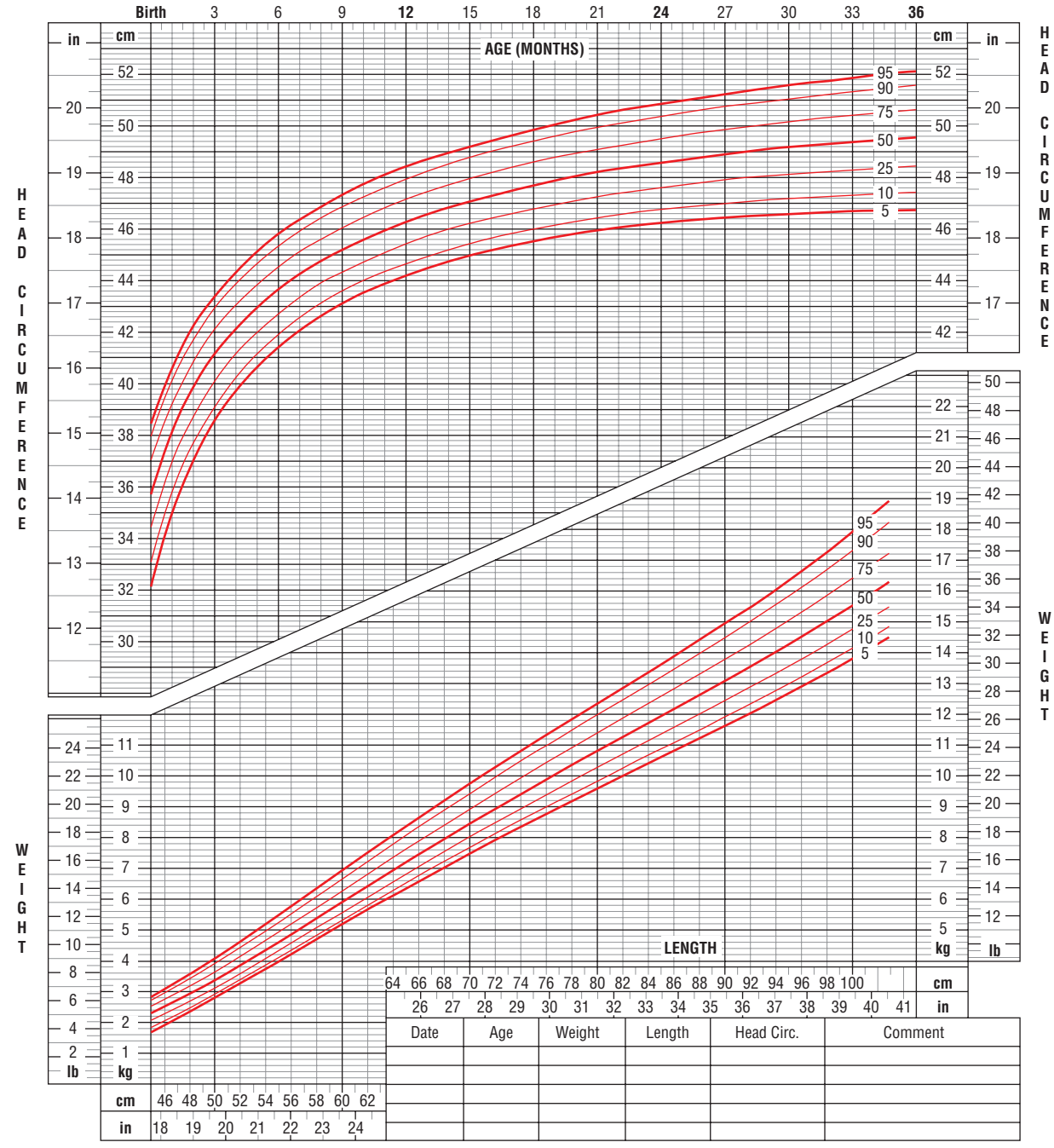
Birth to 36 months: Boys
Head circumference-for-age and
Weight-for-length percentiles

NAME _____

RECORD # _____



Published May 30, 2000 (modified 10/16/00).

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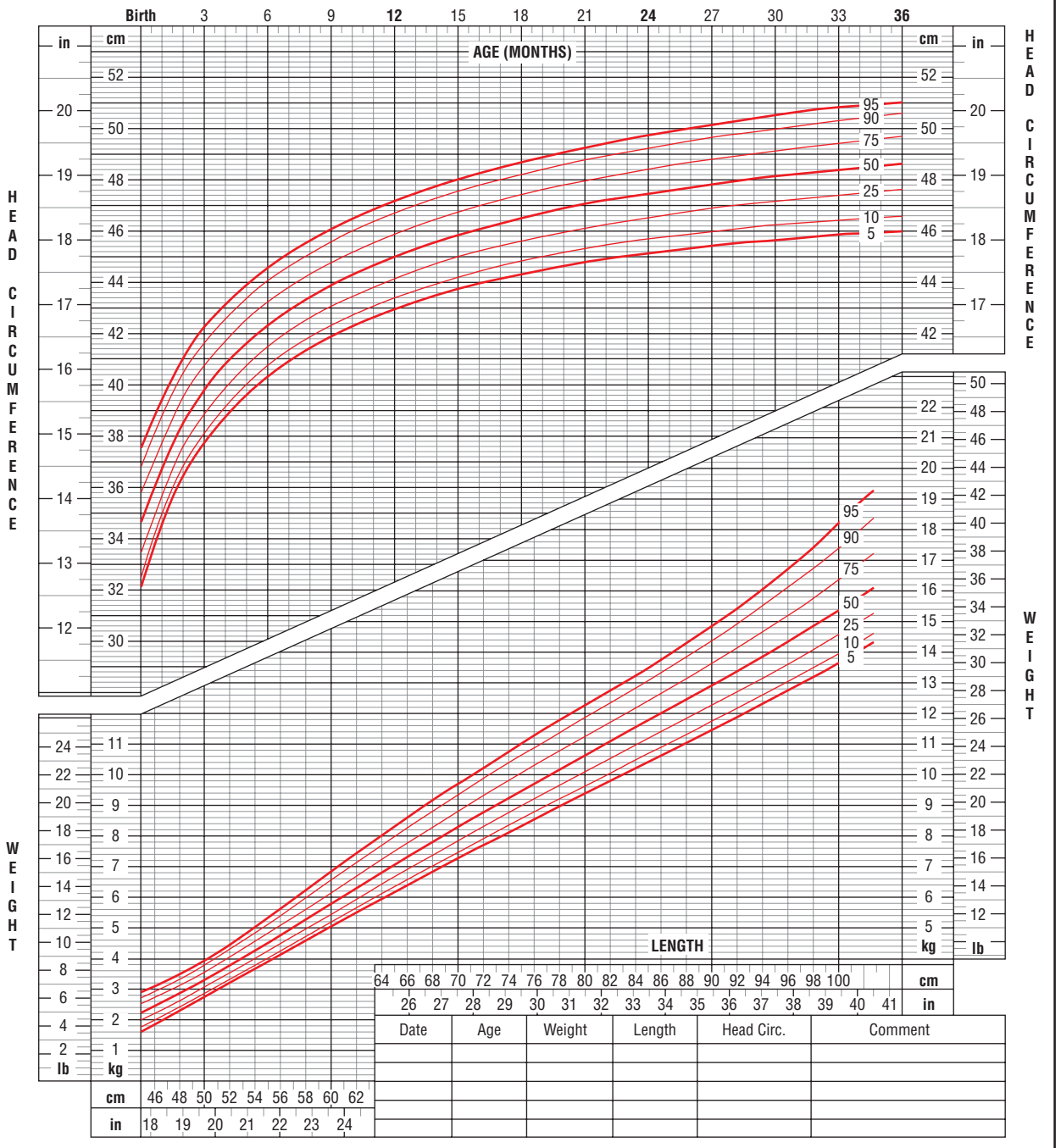
Birth to 36 months: Girls
Head circumference-for-age and
Weight-for-length percentiles

NAME _____

RECORD # _____



Published May 30, 2000 (modified 10/16/00).

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2 to 20 years: Boys
Stature-for-age and Weight-for-age percentiles

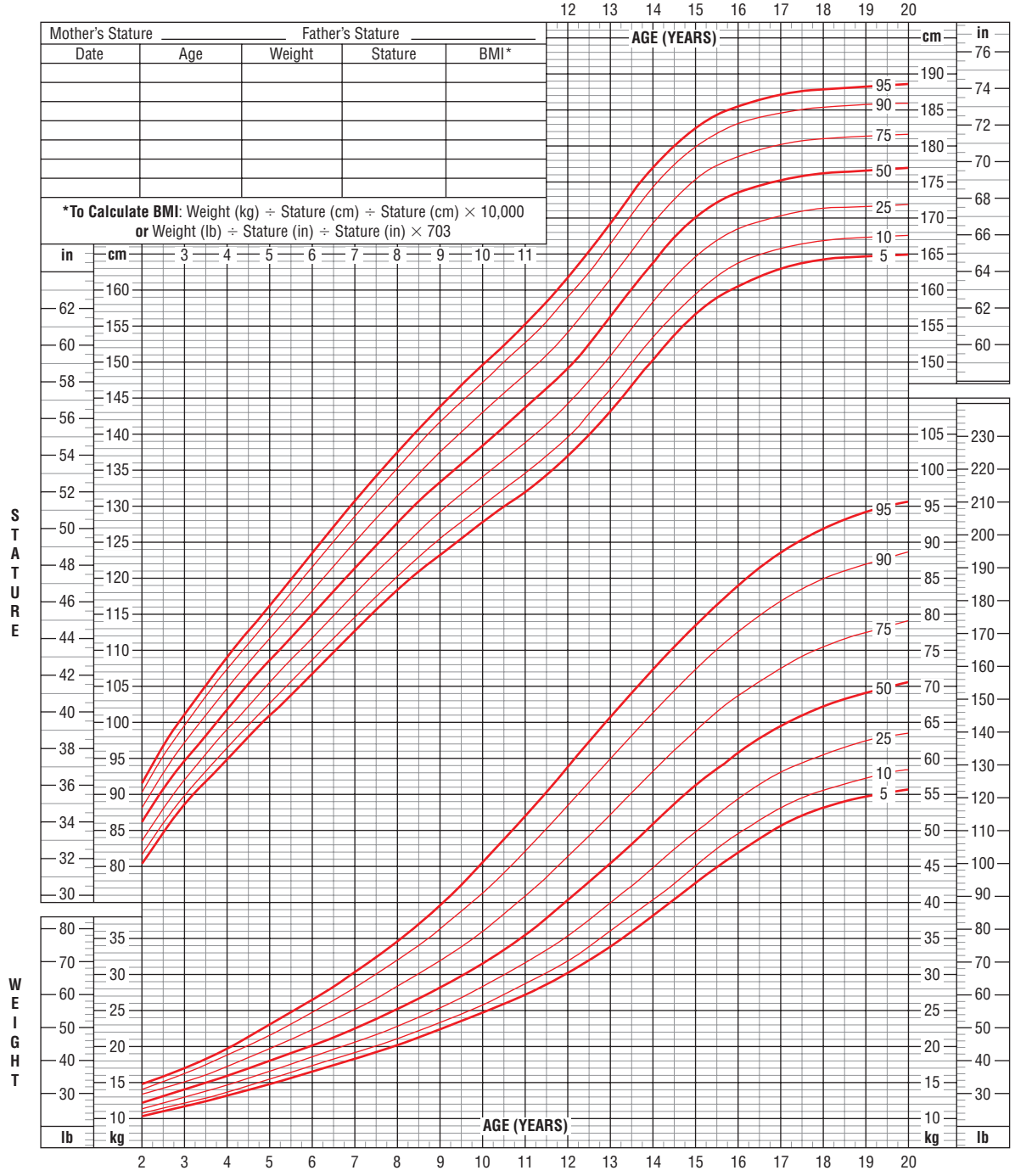
NAME _____

RECORD # _____

Mother's Stature _____ Father's Stature _____

Date	Age	Weight	Stature	BMI*

***To Calculate BMI**: Weight (kg) ÷ Stature (cm) ÷ Stature (cm) × 10,000
or Weight (lb) ÷ Stature (in) ÷ Stature (in) × 703



Published May 30, 2000 (modified 11/21/00).

SOURCE: Developed by the National Center for Health Statistics in collaboration with the National Center for Chronic Disease Prevention and Health Promotion (2000). http://www.cdc.gov/growthcharts

2 to 20 years: Girls
Stature-for-age and Weight-for-age percentiles

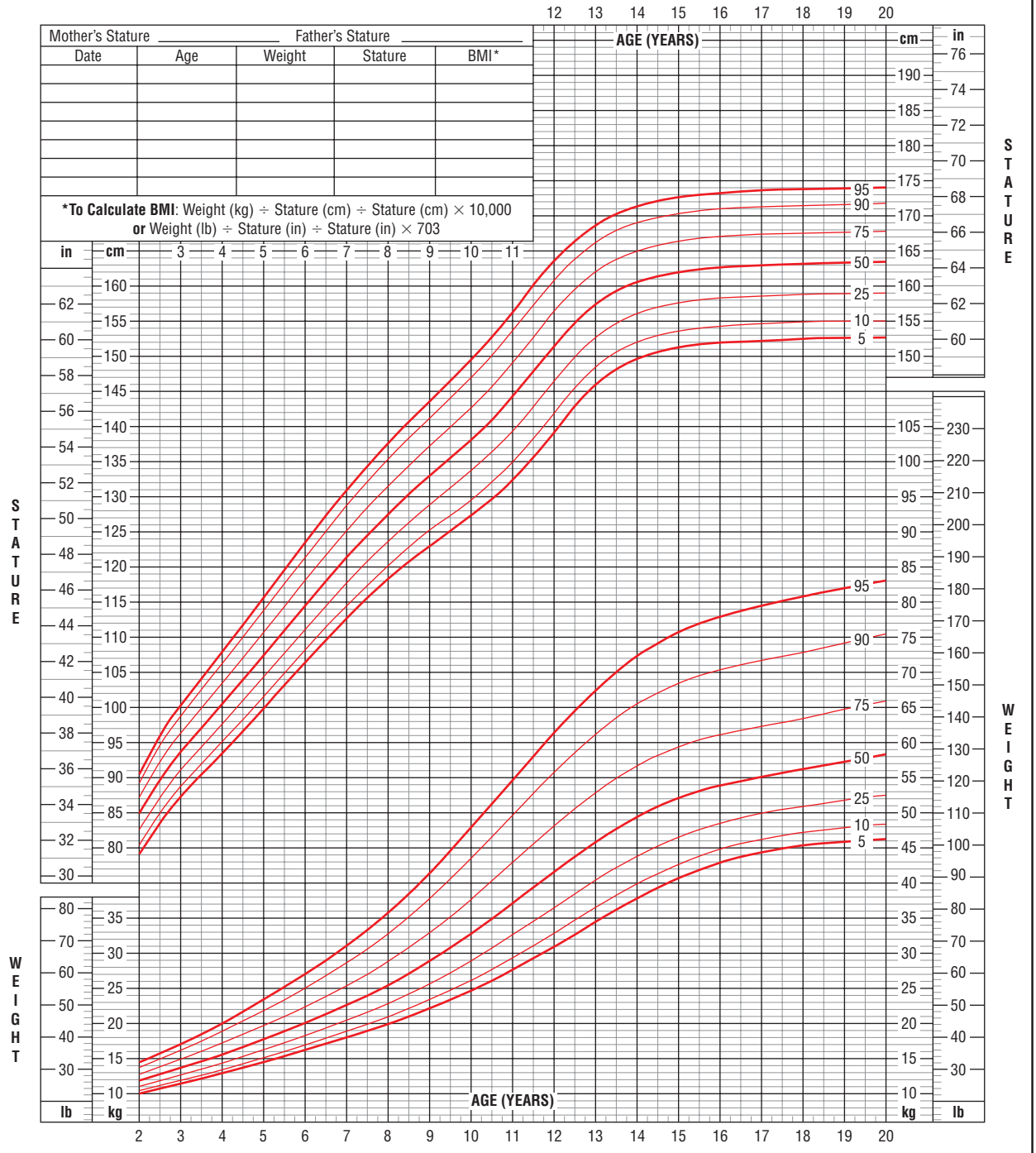
NAME _____

RECORD # _____

Mother's Stature _____ Father's Stature _____

Date	Age	Weight	Stature	BMI*

***To Calculate BMI**: Weight (kg) ÷ Stature (cm) ÷ Stature (cm) × 10,000
or Weight (lb) ÷ Stature (in) ÷ Stature (in) × 703



Published May 30, 2000 (modified 11/21/00).

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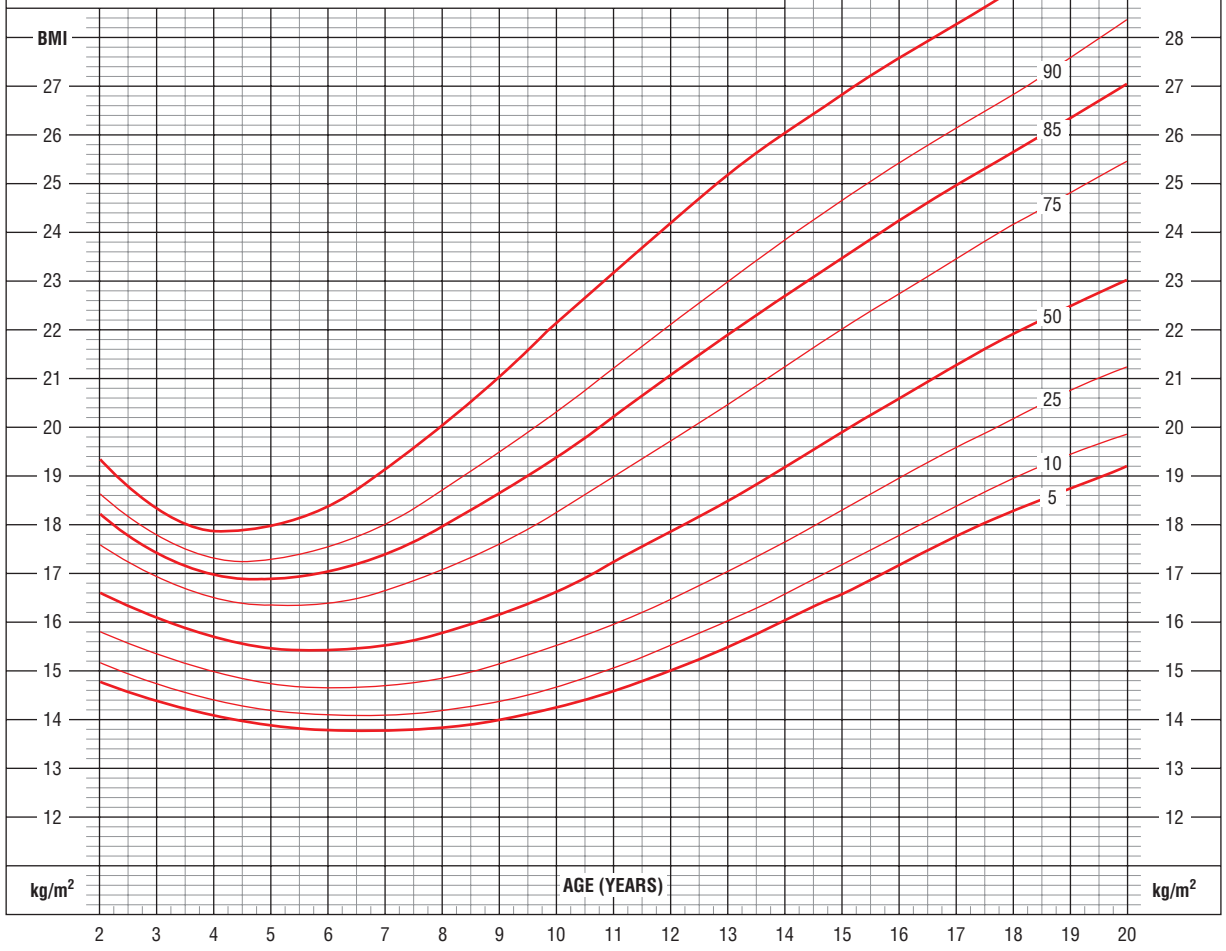
2 to 20 years: Boys
Body mass index-for-age percentiles

NAME _____

RECORD # _____

Date	Age	Weight	Stature	BMI*	Comments

***To Calculate BMI**: Weight (kg) ÷ Stature (cm) ÷ Stature (cm) × 10,000
or Weight (lb) ÷ Stature (in) ÷ Stature (in) × 703



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2 to 20 years: Girls
Body mass index-for-age percentiles

NAME _____

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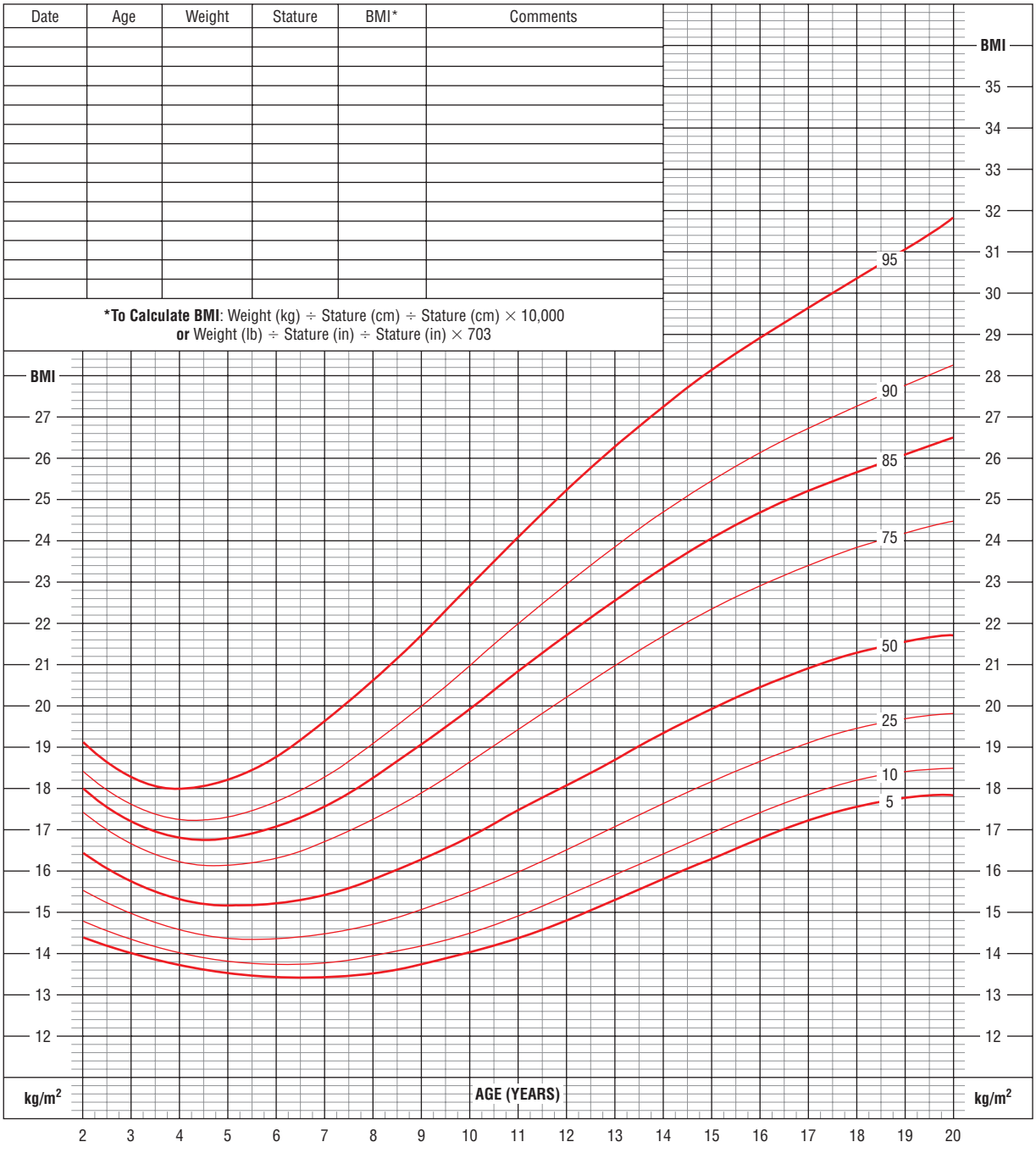

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APPENDIX OF COMMON CHILDHOOD MEDICATIONS

This list of childhood medications was created from the drugs listed in the main body of the text. Short descriptions and any relevant cautionary information are included.

ABACAVIR.....Antiretroviral drug (for treatment of AIDS virus) in patients over the age of three months.

ACCUTANE.....*See* isotretinoin

ACETAMINOPHEN.....Non-narcotic analgesic and fever reducer without anti-inflammatory effects. *Use care in selecting the dosage form and measuring doses. Note that the infant drops have a higher concentration of acetaminophen than the children's formula, and should only be given in drops, never by the teaspoonful.*

ACIPHEX.....*See* rabeprazole

ACYCLOVIR.....An antiviral agent used against herpes infections. It has been used in infants, and even in newborns. Acyclovir has been reported on favorably in the treatment of chickenpox.

ADAPALENE.....A prescription drug for treatment of acne, adapalene is applied to the skin and works by keeping pores clear.

ADRENALIN.....*See* epinephrine

ADRIAMYCIN.....*See* doxorubicin

ADRUCIL.....*See* 5-fluorouracil

AFRIN.....*See* oxymetazoline

AGENERASE.....*See* amprenavir

ALEVE.....*See* naproxen

ALLEGRA.....*See* fexofenadine

ALUMINUM AND MAGNESIUM HYDROXIDES.....An antacid combination used for a number of stomach disorders.

AMANTIDINE.....Although best known for treatment of Parkinson's disease, amantidine provides protection against influenza virus in children as young as one year.

AMINOPHYLLINE.....A bronchodilator used in treatment of asthma.

AMITRIPTYLINE.....A tricyclic antidepressant. *Tricyclic antidepressants are not recommended for children under the age of 12 years. Parents should discuss the safety of all antidepressants before using them in children or adolescents.*

AMLEXANOX.....The only drug approved for use in canker sores. It may work by reducing inflammation.

AMOXICILLIN.....Amoxicillin is a semi-synthetic penicillin similar to ampicillin. The semi-synthetics are effective against more bacteria than are the natural penicillins, and amoxicillin causes less diarrhea in children than does ampicillin.

AMOXICILLIN-CLAVULANATE.....A broad-spectrum antibiotic combination that is effective against bacteria which would be resistant to penicillin alone.

AMOXIL.....*See* amoxicillin

AMPHOTERICIN B.....An antifungal drug, given by vein, which may be used to

treat sinus infections caused by fungi as well as other fungal infections. This drug may cause severe discomfort in patients.

AMPICILLIN ..... A type of penicillin which is effective against more types of bacteria than the original drug. May cause diarrhea in children.

AMPRENAVIR ..... Antiretroviral drug (for treatment of AIDS virus) in patients over the age of four years.

ANADROL ..... *See* oxymetholone

ANAPROX ..... *See* naproxen

ANTHRALIN ..... An ingredient in creams for treatment of psoriasis or some types of hair loss.

APHTHASOL ..... *See* amlexanox

ASPIRIN ..... Non-narcotic pain reliever and fever reducer with anti-inflammatory effects. *Aspirin should not be given to children except on a physician's order.*

ASPIRIN-FREE ANACIN ... *See* acetaminophen

ASTALIN ..... *See* azalastin

AUGMENTIN ..... *See* amoxicillin-clavulanate

AVENTYL ..... *See* nortriptyline

AXID ..... *See* nizatidine

AZACTAM ..... *See* aztreonam

AZELAIC ACID ..... A prescription drug for treatment of acne. It kills bacteria and keeps pores clear.

AZELEX ..... *See* azelaic acid

AZITHROMYCIHN ..... An antibiotic similar to erythromycin, but with better absorption into the lungs, and greater effectiveness against some types of bacteria.

AZTREONAM ..... An antibiotic which may be useful in treating infections caused by bacteria resistant to other drugs.

BABYLAX ..... *See* glycerin

BAL ..... Acronym for British Anti-Lewisite—*See* dimercaprol

BAYER SELECT MAXIMUM STRENGTH HEADACHE PAIN RELIEF FORMULA ..... *See* acetaminophen

BENADRYL ..... *See* diphenhydramine

BENZAMYCIN ..... Benzoyl peroxide and erythromycin combination used in the treatment of acne.

BENZOYL PEROXIDE ..... Skin drying agent, used for treatment of acne. May be mixed with other compounds such as antibiotics.

BEXTRA ..... *See* valdecoxib

BIAXIN ..... *See* clarithromycin

BISACODYL ..... An irritant cathartic (laxative) not recommended for children under six years old. Available as tablets and suppositories.

BLENOXANE ..... *See* bleomycin

BLEOMYCIN ..... An anti-cancer drug of the antibiotic type, bleomycin has been used to treat germ-cell cancers (ovaries or testis) and some other cancers in children.

BLEPH ..... *See* sulfacetamide

BLISTEX ..... *See* phenol

BONINE ..... *See* meclizine

BOTOX ..... *See* botulinum toxin

BOTULINUM TOXIN ..... Although most widely used for reduction of wrinkles, botulinum toxin may be injected as a means of reducing excessive perspiration. Its most important use in children is reduction of spasticity in children with cerebral palsy.

BRITISH ANTI-LEWISITE .. *See* dimercaprol

BROMPHENIRAMINE .... An antihistamine with only moderately sedative effects. The product is available in liquid form, and one of the branded products has a high

level of taste acceptability among children.

BUPROPION..... An antidepressant which is unrelated to other types of antidepressant drugs. Bupropion has been used to treat attention deficit disorder and bipolar disorder. It is also used to help stop smoking. Although approved only for use in adults, the drug has been used to treat children. *The United States Food and Drug Administration has issued a warning that antidepressants may increase the risk of suicide when used to treat children and adolescents. The FDA warns that the risk of suicide is increased in children with bipolar disorder.*

CALAMINE LOTION ..... A preparation of zinc oxide, lime water, glycerin, and water that may relieve some insect stings and irritants like poison ivy. Preparations that contain phenol or antihistamines may be more effective than the original formula.

CALCIUM CARBONATE .. An antacid for treatment of stomach disorders. It may also be used as a diet supplement to provide extra calcium.

CALCIUM CARBONATE AND MAGNESIUM HYDROXIDE ..... An antacid combination used for a number of stomach disorders.

CALCIUM DISODIUM EDETATE ..... A chelating agent used as an antidote to lead poisoning.

CALCIUM DISODIUM VERSENATE ..... *See* calcium disodium edetate

CARBAMAZEPINE ..... An anti-epileptic drug that has been used for a wide range of neurologic conditions. It is approved for use in children above the age of six years, but has been used in younger children as well.

CASCARA SAGRADA .... An irritant cathartic (laxative) not recommended for children under six years old.

CECLOR..... *See* cefaclor

CEFACLOR..... An antibiotic given by mouth. The drug is available in a pediatric suspension that is very well accepted by children.

CEFOBID ..... *See* cefotaxime

CEFOTAXIME ..... A broad-spectrum antibiotic given by injection into a vein.

CEFTRIAXONE ..... A broad-spectrum antibiotic given by injection into a vein. Ceftriaxone is considered to be the best treatment for late Lyme disease.

CELEBREX..... *See* celecoxib

CELECOXIB ..... A COX-2 inhibitor; a type of non-steroidal anti-inflammatory drug that is often used to treat arthritis. No studies have been done on children.

CELEXA..... *See* citalopram

CELONTIN..... *See* methosuximide

CETIRIZINE ..... An antihistamine with very little sedating effect. It is used to treat hay fever and other allergies.

CHLORAMPHENICOL.... A broad-spectrum antibiotic, unrelated to other antibiotics. Because this drug can cause very serious adverse reactions, it should be reserved for cases which cannot be treated with any other antibacterial agent.

CHLOROMYCETIN ..... *See* chloramphenicol

CHLORPHENIRAMINE ... An antihistamine used in the treatment of allergies that carries mild risk of drowsiness.

CHLOR-TRIMETRON..... *See* chlorpheniramine

CHOLESTYRAMINE ..... Cholesterol reducer for children over the age of two years.

CHRONULAC..... *See* lactulose

CIMETIDINE ..... A gastric acid reducer used to treat gastroesophageal reflux and diseases in which the stomach produces too much

acid. An over-the-counter form is available.

CITALOPRAM ..... An antidepressant of the SSRI class (serotonin specific reuptake inhibitor). These drugs are used to treat depression, and may be prescribed for bipolar disorder. *Warning: Antidepressants may increase depression or even lead to suicide when used in children and adolescents. Careful observation is essential.*

CITRUCEL ..... *See* methylcellulose

CLARITHROMYCIN ..... Clarithromycin is an antibiotic similar to erythromycin, but with better absorption into the lungs, and greater effectiveness against some types of bacteria.

CLARITIN ..... *See* loratidine

CLEMASTINE ..... An antihistamine used to treat allergies that carries a mild risk of drowsiness.

CLEOCIN-T ..... *See* clindamycin. This form is for application to the skin, commonly used for treatment of acne.

CLINDAMYCIN ..... Antibiotic, used for anaerobic infections and for treatment of acne.

CLONAZEPAM ..... A benzodiazepine used as a tranquilizer and anti-seizure medication. This is a controlled drug under law.

CLORAZEPATE ..... A benzodiazepine used as a tranquilizer, for treatment of alcoholism, and as an anti-seizure medication. This is a controlled drug under law.

CLOTRIMAZOLE ..... An antifungal drug that may be used as an oral lozenge, skin cream, lotion, or vaginal cream, depending on the location of the fungal infection.

CLOZAPINE ..... Clozapine is an anti-psychotic drug which has been used in treatment of bipolar disorder. Although it is approved only for adults, it has been studied in children as young as nine years. For safe use, weekly blood counts are essential.

CLOZARIL ..... *See* clozapine

COLACE ..... *See* docusate

COLCHICINE ..... A medicine for gout that has found uses in several other conditions, such as familial mediterranean fever. Colchicine has been used to treat children as young as three years.

CORTIZONE ..... *See* hydrocotisone

COUMADIN ..... *See* warfarin

CRIXAVAN ..... *See* indinavir

CROMOLYN ..... A mast-cell stabilizing drug used for prevention of asthmatic and allergic attacks.

CUPRIMINE ..... *See* penicillamine

CYCLIZINE ..... An over-the-counter antihistamine used in treatment of motion sickness in children over the age of six years.

CYCLOPHOSPHAMIDE ... An alkylating agent used in treatment of cancer. It may be used for some types of childhood leukemias.

CYTOVENE ..... *See* ganciclovir

CYTOXAN ..... *See* cyclophosphamide

DARVON ..... *See* propoxyphene

DDAVP ..... *See* desmopressin acetate

DECADRON ..... *See* dexamethasone

DECADURABOLIN ..... *See* nandrolone

DELTASONE ..... *See* prednisone

DEMEROL ..... *See* meperidine

DEPAKENE ..... *See* valproic acid

DEPAKOTE ..... *See* valproic acid

DEPEN ..... *See* penicillamine

DESFERAL ..... *See* desferoxamine

DESFEROXAMINE ..... A drug which binds iron, it is used to reduce the blood iron

levels in thalassemia.. It is given only by injection.

DESIPRAMINE..... A tricyclic antidepressant, used to treat depression, but which may also be useful in treatment of bulimia. *Warning: Antidepressants may increase depression or even lead to suicide when used in children and adolescents. Careful observation is essential.*

DESMOPRESSIN..... A synthetic compound similar to antidiuretic hormone (ADH). It may be used to treat bedwetting. Because desmopressin increases the count of blood platelets, it has been used to control some types of coagulation disorders.

DEXAMETHASONE..... An anti-inflammatory steroid that is widely used. It may be given by mouth, by vein, applied to the skin, or included in eye drops or ear drops.

DEXTROMETHORPHAN . A cough suppressant that is the active ingredient in most over-the-counter cough remedies. It is usually indicated in product names as DM.

DIAZEPAM..... A benzodiazepine used as a tranquilizer, muscle relaxant, and anti-seizure medication. This is a controlled drug under law.

DIBASIC SODIUM PHOSPHATE..... A saline cathartic (laxative), it acts by drawing water into the intestine, and produces a watery stool. Children may be more sensitive than adults to toxic effects. Do not use in children under the age of six years.

DIDANOSINE..... Antiretroviral drug (for treatment of AIDS virus) in patients over the age of two weeks, although the drug has been used in newborn infants as well.

DIFFERIN..... *See* adapalene

DIGOXIN..... Derived from digitalis, this drug increases the strength of heart contractions.

DILANTIN..... *See* phenytoin

DILAUDID..... *See* hydromorphone

DIMENHYDRINATE..... An over-the-counter antihistamine used in treatment of motion sickness in children over the age of two years.

DIMERCAPROL..... A chelating agent which is used as an antidote for arsenic, gold, mercury and lead poisoning.

DIMETANE..... *See* brompheniramine

DIPHENHYDRAMINE..... An antihistamine that is very effective but commonly causes drowsiness.

DOCUSATE..... A stool softener to relieve constipation (not a laxative). It may take as long as three days to show any benefit. Not recommended for children under six years old.

DOXORUBICIN..... An anti-cancer drug of the antibiotic type, doxorubicin has been used to treat Ewing's sarcoma and some types of lymphoma in children.

DTP VACCINE..... A vaccine that protects against diphtheria, tetanus, and pertussis. It has largely been replaced by diphtheria and tetanus toxoids and acellular pertussis (DTaP) vaccine, which has fewer unwanted side effects.

DRAMAMINE..... *See* dimenhydrinate

DRITHOCREME..... *See* anthralin

DRYSOL..... *See* aluminum chloride

DTAP VACCINE..... A vaccine that protects against diphtheria, tetanus, and pertussis. It is safer than the old DTP vaccine.

DULCOLAX..... *See* bisadocyl

DUPHALAC..... *See* lactulose

DURAGESIC..... *See* fentanyl

DUVOID ..... *See* bethanechol

EFAVIRENZ ..... Antiretroviral drug (for treatment of AIDS virus) in patients over the age of three years.

EFFEXOR ..... *See* venlafaxine

ELAVIL ..... *See* amitriptyline

ELIDEL ..... *See* pimecrolimus

EPINEPHRINE ..... A hormone that is used as a stimulant and vasoconstrictor in the treatment of anaphylaxis. It may also be used to stop bleeding during surgery.

EPIVIR ..... *See* lamivudine

EPSOM SALTS ..... *See* magnesium sulfate

ERTAPENEM ..... A prescription antibiotic used to treat penicillin-resistant bacteria. Has not been well studied in children.

ERYTHROMYCIN ..... An antibiotic which may be taken by mouth or injected to treat infections, or applied to the skin to treat acne. Erythromycin is widely used for patients who are allergic to penicillin.

ESCITALOPRAM ..... An antidepressant of the SSRI class (serotonin specific reuptake inhibitor) These drugs are used to treat depression, and may be prescribed for bipolar disorder. *Warning: May increase depression or even lead to suicide when used in pediatrics. Careful observation is essential.*

ESKALITH ..... *See* lithium carbonate

ESOMEPRAZOLE ..... A prescription gastric acid reducer of the proton-pump inhibitor class. It has been given to children as young as three years of age.

ETHOSUXIMIDE ..... A drug used in treatment of epilepsy, usually for absence seizures.

ETHOTOIN ..... A drug in the same family as phenytoin, used to control epilepsy.

ETOPOSIDE ..... An anti-cancer drug of the topoisomerase class. It may be used to treat some types of leukemia and other cancers in children.

FAMCICLOVIR ..... An anti-viral drug which has been useful in treatment of herpes infections, including those associated with chickenpox. At present, it is not recommended that this drug be used in patients under the age of 18 years.

FAMOTIDINE ..... A gastric acid reducer of the H-2 receptor blocker class used to treat gastroesophageal reflux and diseases in which there is an excess of stomach acid. An OTC form is available

FAMVIR ..... *See* famciclovir

FELBAMATE ..... An anti-epileptic drug approved for use in children over the age of 14 years, but which has been used in younger children as well. Should be reserved for cases not responsive to other drugs.

FELBATOL ..... *See* felbamate

FENTANYL ..... A narcotic pain reliever, fentanyl is often given to children to relax them before surgery. This is a controlled drug under federal law.

FEO-SOL ..... *See* ferrous sulfate

FERROUS SULFATE ..... The preferred form of iron for treatment of iron deficiencies. It comes in tablet and liquid form.

FEXOFENADINE ..... A non-sedating antihistamine used in the treatment of allergies. It has not been tested in children younger than six years.

FINEVIN ..... *See* azelaic acid

5-FLUOROURACIL ..... An antimetabolite used in the treatment of cancer. The injection has limited use in common childhood cancers,

but a skin cream has been used for some types of skin cancers.

5-FU ..... *See* 5-fluorouracil

FLUMADINE ..... *See* rimantidine

FLUOXETINE ..... An antidepressant of the SSRI class (serotonin specific reuptake inhibitor). These drugs are used to treat depression, and may be prescribed for bipolar disorder. Fluoxetine has also been used to treat bulimia. *Warning: May increase depression or even lead to suicide when used in children. Careful observation is essential.*

FLUVAX ..... *See* influenza virus vaccine

FOSCARNET ..... An antiviral drug used for treatment of cytomegalovirus and severe herpes virus infections. It is appropriate for adolescents, but not recommended for children.

FORTOVASE ..... *See* saquinavir

FOSCAVIR ..... *See* foscarnet

FULVICIN ..... *See* griseofulvin

FUNGIZONE ..... *See* amphotericin B

GAMIMUNE ..... *See* immune serum globulin

GAMMAGARD ..... *See* immune serum globulin

GANCICLOVIR ..... Antiviral agent for cytomegalovirus infections of the eye. Although not recommended for patients below the age of 12 years, the drug is has been used in children as young as three months.

GEODON ..... *See* ziprasidone

GLUCAGON ..... A hormone manufactured by the pancreas that triggers the release of blood glucose by the liver, often in diabetics. It is only administered by injection.

GLUCAGON EMERGENCY KIT ..... See glucagon

GLYCERIN ..... A hyperosmotic laxative that is used in either rectal solution or suppository form. Glycerin is used in children as young as newborns. The chemical has many uses in drug formulation since it helps products retain moisture (in lotions and creams), while in oral liquids it acts as a thickening agent and provides a slightly sweet taste.

GLYCOLIC ACID ..... A chemical peel used to treat acne and for other cosmetic purposes.

GRANISETRON ..... An anti-emetic drug used to control nausea and vomiting associated with cancer therapy. The drug may be given by mouth or by vein, and is used in children over the age of two years.

GRISACTIN ..... *See* griseofulvin

GRISEOFULVIN ..... An antifungal agent used to treat infections of the hair and nails.

GRIS-PEG ..... *See* griseofulvin

GUAIFENESIN ..... Guaifenesin is commonly included in over-the-counter cough remedies as an expectorant, a drug which breaks up mucus. Although the drug has been used for a long time, there is little proof that it works.

GYNE-LOTRIMIN ..... *See* clotrimazole

HALDOL ..... *See* haloperidol

HALOPERIDOL ..... A prescription drug commonly used for behavioral disorders such as excessive rage. It has also been useful in the treatment of Tourette's syndrome.

HEXADROL ..... *See* dexamethasone

HIVID ..... *See* zalcitabine

HUMULIN ..... *See* insulin

HYDROCORTISONE ..... An anti-inflammatory steroid that may be taken by mouth, or in creams and lotions to be

applied to skin and mucous membranes. Hydrocortisone creams may be used on infants and young children. Low concentrations of hydrocortisone are available in over-the-counter preparations.

HYDROMORPHONE ... Hydromorphone is a narcotic pain reliever and cough suppressant for use in children over the age of six years. It is a controlled drug under federal law.

HYDROXYCHLOROQUINE ... An antialarial compound that has been used to treat some autommune conditions including lupus and juvenile dermatomyositis.

HYTONE ... *See* hydrocortisone

IBUPROFEN ... Non-narcotic pain-reliever and fever reducer with anti-inflammatory effects.

IMIPRAMINE ... A tricyclic antidepressant, used primarily to treat depression, but which may also be used in treatment of bulimia. Imipramine may also be useful in control of bedwetting. *Warning: May increase depression or even lead to suicide when used in children and adolescents. Careful observation is essential.*

IMITREX ... *See* sumatriptan

IMMUNE SERUM GLOBULIN ... This is a preparation of the portion of the blood that contains antibodies. It helps the immune system prevent or fight off infections.

IMODIUM ... *See* loperamide

INDINAVIR ... Antiretroviral drug for treatment of AIDS virus. According to the manufacturer, safety in children has not been established, but the drug has been listed in standard pediatric references.

INDOCIN ... *See* indomethicin

INDOMETHACIN ... A non-steroidal anti-inflammatory drug (NSAID) available by prescription only, usually used to treat arthritis and other inflammatory conditions. Not recommended for children under the age of 14 except in special circumstances. Indomethacin has special application in some infants born with heart problems.

INFLUENZA VIRUS VACCINE ... A vaccine used to prevent infection with the influenza virus. It may be given to infants as young as six months of age.

INSULIN ... A hormone which is essential for glucose (sugar) utilization. There are many types of modified insulins designed to give either more rapid effect or longer duration of action. Insulin must be given by injection.

INTAL ... *See* cromolyn

INVANZ ... *See* ertapenem

INVIRASE ... *See* saquinavir

ISONIAZID ... Anti-tuberculosis drug that may be used to treat tuberculosis and to protect people in close contact with tuberculosis patients. Infants and young children can tolerate higher doses of this drug than do older children and adults.

ISOTRETINOIN ... An anti-acne drug.

IVERMECTIN ... A prescription treatment for parasites which is effective against lice, although it is not FDA-approved for this purpose.

KALETRA ... *See* lopinavir/ritonavir fixed combination

KENALOG ... *See* triamcinolone

KETOCONAZOLE ... An antifungal agent that may be used as a cream for skin fungus, or in tablet form for systemic infections.

KETOPROFEN . . . . . A non-steroidal anti-inflammatory drug (NSAID), available by prescription only. Not given to children under the age of 16 unless directed by a physician.

KETOROLAC TROMETHAMINE . . . . . A non-teroidal anti-inflammatory drug (NSAID) given by injection. This is an effective pain reliever for short-erm use only. Its main advantage over narcotics for short-erm use is that it does not cause sedation. This drug is not normally used for treatment of children.

KLONOPIN . . . . . *See* clonazepam

KONSIL . . . . . *See* psyllium

KWELL . . . . . *See* lindane

KYTRIL . . . . . *See* granisetron

LACTULOSE . . . . . A hyperosmotic laxative that is also used for other conditions, including liver and brain problems. It has been used to treat children, but is not recommended for frequent use.

LAMICTAL . . . . . *See* lamotrigene

LAMIVUDINE . . . . . An antiretroviral drug, used to treat the AIDS virus, in children above the age of three months. Because viral resistance develops when lamivudine is used alone, the drug should be administered in combination with other antiretroviral drugs.

LAMOTRIGENE . . . . . A anti-epileptic drug primarily for use in adults, but which has been used in children with severe epilepsy as young as two years of age. The drug has also been used to treat bipolar disorder.

LAMICTAL . . . . . *See* lamotrigene

LANOXIN . . . . . *See* digoxin

LANSOPRAZOLE . . . . . A prescription gastric acid reducer of the proton-pump inhibitor class, used to treat gastroesophageal reflux, and other disorders of excess stomach acid. A pediatric suspension is available, and the flavor had been reported to be popular with children.

LEVO-DROMORAN . . . . . *See* levorphanol

LEVORPHANOL . . . . . A narcotic pain reliever for severe pain. Its use in children has not been well established. It is a controlled drug under federal law.

LEXAPRO . . . . . *See* escitalopram

LIDOCAINE . . . . . A local anesthetic that may be injected, or applied to skin or mucous membranes. It relieves pain from wounds and sores, including canker sores.

LINDANE . . . . . A prescription drug for removal of lice. Lindane may cause nerve damage even when used as directed. It should never be used on premature infants.

LINEZOLID . . . . . A prescription antibiotic used to treat penicillin-resistant bacteria. Has not been well studied in children. Because it may cause blood problems, regular blood tests are needed while taking this drug.

LITHIUM CARBONATE . . . A drug used in treatment of some psychiatric conditions including bipolar mood disorder and anorexia.

LONITEN . . . . . *See* minoxidil (Loniten designates tablets for high blood pressure only.)

LOPERAMIDE . . . . . An antidiarrheal drug that is available without prescription. It has been given to children as young as two years, but should not be used without first consulting a physician.

LOPINAVIR/RITONAVIR FIXED COMBINATION . A combination of antiretroviral drugs (anti-IDS drugs) that may be used in children as young as six months.

LORATIDINE ..... A non-sedating antihistamine used in the treatment of asthma.

LOTRIMIN ..... *See* clotrimazole

MAALOX ..... *See* aluminum and magnesium hydroxides

MAGNESIUM CITRATE (CITRATE OF MAGNESIA) ... A saline cathartic (laxative), it acts by drawing water into the intestine, and produces a watery stool. Children may be more sensitive than adults to toxic effects. The oral liquid should be chilled before use. Do not use in children under the age of two years.

MAGNESIUM HYDROXIDE ..... A saline cathartic (laxative), it acts by drawing water into the intestine, and produces a watery stool. Children may be more sensitive than adults to toxic effects. Do not use in children under the age of two years. May be used as an antacid.

MAGNESIUM SULFATE ... A saline cathartic (laxative), it acts by drawing water into the intestine, and produces a watery stool. Children may be more sensitive than adults to toxic effects. Do not use in children under the age of two years. May be used as a soak for bruises and sprains. Flavored forms available.

MAREZINE ..... *See* cyclizine

MARIJUANA ..... Abuse drug, the active component, tetrahydrocannabinol, or THC, may be used as an antinauseant to relieve some of the effects of cancer chemotherapy. Under United States law, marijuana has no legitimate medical use.

MECLIZINE ..... An over-the-counter antihistamine used in treatment of motion sickness in children over the age of 12 years.

MECLOFENAMATE SODIUM ..... A non-teroidal anti-inflammatory drug (NSAID), available by prescription only. Safety and efficacy in children under 14 years of age has not been established.

MEDROL ..... *See* methylprednisolone

MEFENAMIC ACID ..... A non-steroidal anti-inflammatory drug (NSAID), available by prescription only. Safety and efficacy in children under 14 years of age has not been established.

MENABOL ..... *See* stanzolol

MEPERIDINE ..... A synthetic narcotic analgesic. This is a controlled drug under federal law.

MEPHENYTOIN ..... A drug in the same family as phenytoin, used to control epilepsy.

MESANTOIN ..... *See* mephenytoin

METADATE ..... *See* methylphenidate

METAMUCIL ..... *See* psyllium

METHSUXIMIDE ..... A drug used in treatment of epilepsy, usually for absence seizures.

METHYLCELLULOSE ..... A bulk-forming laxative. Should be given in divided doses, with at least eight ounces of fluid. Not recommended for children under six years old.

METHYLPHENIDATE ..... A stimulant which is used in treatment of attention deficit/hyperactivity disorder in children. This is a controlled drug under federal law.

METHYLPREDNISOLONE An anti-inflammatory steroid which may be administered by mouth or injection. The drug may be given to infants and children for many diseases.

METOCLOPRAMIDE ..... A prokinetic agent, metoclopramide accelerates emptying of the stomach into the intestine. It may be used for stomach disorders or in conjunction with anticancer drugs.

MICANOL ..... *See* anthralin

MICROSULFON ..... *See* sulfadiazine

MILK OF MAGNESIA ..... *See* magnesium hydroxide

MILONTIN..... *See* phensuximide

MINERAL OIL ..... An emollient cathartic (laxative) it acts by lubricating the intestine. Not recommended for children under six years old. Routine use may reduce absorption of essential vitamins.

MINOXIDIL..... An antihypertensive drug which has been widely used as a lotion to treat androgenetic hair loss.

MIRTAZAPINE..... An antidepressant, chemically different from most other antidepressants. *Warning: May increase depression or even lead to suicide when used in children and adolescents. Careful observation is essential.*

MODAFINIL ..... A central nervous system stimulant which is chemically unrelated to other drugs such as caffeine or methylphenidate. Although officially not approved for children, the drug has been used to treat childhood narcolepsy.

MOTRIN..... *See* ibuprofen

MYCELEX ..... *See* clotrimazole

MYLANTA ..... *See* calcium carbonate and magnesium hydroxide

MYSOLINE..... *See* primidone

NANDROLONE ..... An anabolic steroid, used for treatment of some diseases that cause breakdown of the body, kidney disease, and anemia. May be subject to abuse. This is a controlled drug under federal law.

NAPHAZOLINE..... A vasoconstrictor, most often used as eye drops to treat eye redness associated with colds.

NAPHCON ..... *See* naphazoline

NAPROSYN..... *See* naproxen

NAPROXEN..... A non-steroidal anti-inflammatory drug (NSAID), available over-the-counter and by prescription. Safety and efficacy in children under two years of age has not been established.

NEFAZODONE..... An antidepressant which is unrelated to other types of antidepressant drugs. *Warning: May increase depression or even lead to suicide when used in children and adolescents. Careful observation is essential.*

NELFINAVIR ..... Nelfinavir is an anti-retroviral drugs (anti-AIDS). The manufacturer does not recommend use of this drug for children younger than two years, but it has been studied with some success in children as young as newborns.

NEO-SYNEPHRINE..... *See* phenylephrine

NEXIUM..... *See* esomeprazole

NILSTSTAT..... *See* nystatin

NITROSTAT..... *See* nitroglycerin

NIZATIDINE ..... A gastric acid reducer of the H-2 receptor blocker class. An OTC form is available.

NIZORAL..... *See* ketoconazole

NOLVADEX..... *See* tamoxifen

NORPRAMINE ..... *See* desipramine

NORTRIPTYLINE ..... A tricyclic antidepressant. *Warning: May increase depression or even lead to suicide when used in children and adolescents. Careful observation is essential.*

NORVIR..... *See* ritonavir

NUMORPHAN ..... *See* oxymorphone

NYSTATIN ..... An antifungal drug which may be used in several forms: swallowed, used as a mouth rinse, applied to the skin, or used as a vaginal suppository,

depending on the location of the fungal infection.

OLANZAPINE ..... Olanzapine is an anti-psychotic drugs which has been used in treatment of bipolar disorder. Although it is approved only for adults, there have been reports of its use in children as young as eight years. The frequency of adverse effects is higher in children than in adults.

ONDANSETRON ..... An anti-emetic drug used to control of nausea and vomiting that are associated with cancer therapy. The drug may be given by mouth or by vein, and is used in children over the age of four years.

ORAP ..... *See* pimozide

ORUDIS ..... *See* ketoprofen

ORUVAIL ..... *See* ketoprofen

OSELTAMIVIR ..... Antiviral drug used to treat influenza infections in patients over the age of 13 years.

OTRIVIN ..... *See* xylometazoline

OXANDRIN ..... *See* oxandrolone

OXANDROLONE ..... An anabolic steroid used for treatment of some diseases that cause breakdown of the body, kidney disease, and anemia. May be subject to abuse.

OXCARBAZEPINE ..... An anti-epileptic drug similar to carbamazepine. Approved for use in children over 16 years, but has been used in younger children as well.

OXYCODONE ..... Oxycodone is a narcotic pain reliever which is approved for children above the age of six years. It is a controlled drug under federal law.

OXYCONTIN ..... *See* oxycodone

OXYMETAZOLINE ..... A vasoconstrictor which may be used as a nasal spray to relieve stuffy nose, or as an eye drop to treat eye redness associated with colds.

OXYMETHOLONE ..... An anabolic steroid used for treatment of some diseases that cause breakdown of the body, kidney disease, and anemia. May be subject to abuse.

OXYMORPHONE ..... A narcotic pain reliever for children over the age of two years. It is administered by injection or rectal suppository. It is a controlled drug under federal law.

PAMELOR ..... *See* nortriptyline

PANADOL ..... *See* acetaminophen

PANTOPRAZOLE ..... A prescription gastric acid reducer of the proton-pump inhibitor class. It is used to treat gastroesophageal reflux and other disorders in which the stomach produces too much acid.

PARNATE ..... *See* tranylcypromine

PAROXETINE ..... An antidepressant of the SSRI class (serotonin specific reuptake inhibitor). These drugs are used to treat depression, and may be prescribed for bipolar disorder. *Warning: May increase depression or even lead to suicide when used in children and adolescents. Careful observation is essential.*

PAXIL ..... *See* paroxetine

PEGANONE ..... *See* ethotoin

PENICILLAMINE ..... A drug used as a treatment for rheumatoid arthritis, Wilson's disease (excess copper in the blood), and as an antidote for heavy metal poisoning (iron, mercury, lead, and arsenic).

PENICILLIN ..... An antibiotic; the parent compound of a large class of antibiotics that may be used in treatment or prevention of bacterial infections.

PEPCID ..... *See* famotidine

PERTOFRANE ..... *See* desipramine

PHENACEMIDE ..... An anti-epileptic drug no longer in common use.

PHENOL ..... A barbiturate used in treatment of epilepsy.

PHENSUXIMIDE ..... A drug used in treatment of epilepsy, usually for absence seizures.

PHENYLEPHRINE ..... A nasal decongestant.

PHENYTOIN ..... An anti-seizure (anti-epileptic) agent used to control many different types of seizures. It is available in liquid and chewable tablets for children.

PHOSPO-SODA ..... *See* dibasic sodium phosphate

PIMECROLIMUS ..... Medication originally used to prevent rejection in organ transplants. An ointment of this compound is used to treat moderate to severe atopic dermatitis in adults or children two years of age and older.

PIMOZIDE ..... A prescription drug for treatment of Tourrette's syndrome in children over the age of 12 years.

PLAQUENIL ..... *See* hydroxychloroquine

PODOFILOX ..... An agent used for wart removal, it should be used only by a physician, since it is too strong for self application. It has been used in children as young as two years old.

PODOPHYLLUM ..... An agent used for wart removal, it should be used only by a physician, since it is too strong for self application.

PONSTEL ..... *See* mefenamic acid

PREDNISONE ..... An anti-inflammatory steroid. It is most often given by mouth and has many applications.

PREVACID ..... *See* lansoprazole

PRIMIDONE ..... A drug used for several different types of epilepsy. In some children it may cause agitation.

PROPOXYPHENE ..... A synthetic narcotic analgesic for mild pain.

PROTONIX ..... *See* pantoprazole

PROTOPIC ..... *See* tacrolimus

PROVIGIL ..... *See* modafinil

PROZAC ..... *See* fluoxetine

PSEUDOEPHEDRINE ..... A nasal decongestant, may be inhaled or taken by mouth.

PSYLLIUM ..... A bulk-forming laxative. Should be given in divided doses, with at least eight ounces of fluid. Not recommended for children under six years old.

QUESTRAN ..... *See* cholestyramine

QUETIAPINE ..... An antipsychotic agent approved only for use in adults, but has been used in children both for psychosis and bipolar disorder.

RABEPRAZOLE ..... A prescription gastric acid reducer of the proton-pump inhibitor class, used in the treatment of gastroesophageal reflux and other disorders in which the stomach produces too much acid.

RANITIDINE ..... A gastric acid reducer of the H-2 receptor blocker class. An over-the-counter form is available. A pediatric suspension is available, but in one study, children did not like the flavor.

REBETOL ..... *See* ribavirin

REGLAN ..... *See* metoclopramide

RELENZA ..... *See* zanamivir

RETROVIR ..... *See* zidovudine

RIBAVIRIN ..... Antiviral drug used to treat respiratory viruses in infants and young children.

RIMANTIDINE ..... Antiviral drugs used to treat influenza virus infections in children one year old or older.

RISPERDAL ..... *See* risperidone

RISPERIDONE ..... An antipsychotic drug which has been used in treatment of bipolar disorder. Although it is

approved only for adults, it has been studied in children as young as nine years.

RITALIN ..... *See* methylphenidate

RITONAVIR ..... An antiretroviral (anti-AIDS) drug for use in children over the age of two years.

ROCEFIN ..... *See* ceftriaxone

ROGAIN ..... *See* minoxodil (Rogain is a solution, applied to the scalp, for hair loss only. Minoxidil in tablet form is used for treatment of high blood pressure.)

SALICYLIC ACID ..... Used as a skin peel, for removal or warts and treatment of acne.

SAQUINAVIR ..... An anti-retroviral (anti-AIDS) drug. The manufacturer does not recommend use of this drug for children younger than 16 years, but it has been studied with some success in children as young as two years.

SEROQUEL ..... *See* quetiapine

SERTRALINE ..... An antidepressant of the SSRI class (serotonin specific reuptake inhibitor). *Warning: May increase depression or even lead to suicide when used in children and adolescents. Careful observation is essential.*

SERZONE ..... *See* nefazodone

SOLU-MEDROL ..... *See* methylprednisolone

SPIRONOLACTONE ..... Potassium-sparing diuretic used in heart disease; also has anti-androgenic effects which may be used to treat acne.

STANZOLOL ..... An anabolic steroid used for treatment of some diseases that cause breakdown of the body, kidney disease, and anemia. May be subject to abuse.

STAVUDINE ..... An anti-retroviral (anti-AIDS) drug that may be given to infants as young as newborns.

STERAPRED ..... *See* prednisone

STROMECTOL ..... *See* ivermectin

SUDAFED ..... *See* pseudoephedrine

SULAMYD ..... *See* sulfacetamide

SULFACETAMIDE ..... A sulfa drug with antibacterial effects. It may be applied to the skin to treat acne, or used in eye drops to treat pink eye (conjunctivitis) infections.

SULFADIAZINE ..... A sulfa drug that may be used to treat toxoplasmosis in infants.

SUMATRIPTAN ..... A prescription drug used for treatment of migraine headaches.

SUMYCIN ..... *See* teracycline

SURFAK ..... *See* docusate

SURMONTIL ..... *See* trimipramine

SUSTIVA ..... *See* efavirenz

SYMMETREL ..... *See* amantidine

TACROLIMUS ..... A medication originally used to prevent rejection in organ transplants. An ointment of this compound is used to treat moderate to severe atopic dermatitis in adults or children two years of age and older.

TAGAMET ..... *See* cimetidine

TAMIFLU ..... *See* oseltamivir

TAMOXIFEN ..... An anti-estrogen drug normally used for treatment of breast cancer in adult women. It has been reported on favorably for treatment of gynecomastic (enlarged breasts in the male) in adolescent boys.

TAVIST ..... *See* clemastine

TCA ..... *See* trichloracetic acid

TEGRETOL ..... *See* carbamazepine

TETRACYCLINE ..... An antibiotic which is effective against many types of bacteria and other pathogens. It is not recommended for young children because it can cause

discoloration of the teeth. A solution or paste made of tetracycline may relieve the discomfort of a canker sore. It may be used for treatment of acne in adolescents.

TETRACYN.....*See* tetracycline

TOFRANIL.....*See* imipramine

TOLECTIN.....*See* tolmetin sodium

TOLMETIN SODIUM.....A non-steroidal anti-inflammatory drug (NSAID), available by prescription only. Safety and efficacy in children under two years of age has not been established.

TORADOL.....*See* ketorolac tromethamine

TRANXENE.....*See* clorazepate

TRANYLCYPROMINE.....An antidepressant of the mono-amine oxidase inhibitor class (MAOI), has been used to treat bipolar disorder as well as depression.. Special diet restrictions are essential for safe use. *Warning: May increase depression or even lead to suicide when used in children and adolescents. Careful observation is essential.*

TRETINOIN.....A prescription drug used for treatment of acne.

TRIAMCINOLONE.....A steroid cream which is used to treat various skin conditions, including canker sores.

TRICHLORACETIC ACID.....An agent used for wart removal, it should be used only by a physician, since it is too strong for self application.

TRILEPTAL.....*See* oxcarbazepine

TRIMIPRAMINE.....A tricyclic antidepressant, not widely used. *Warning: May increase depression or even lead to suicide when used in children and adolescents. Careful observation is essential.*

TRIMOX.....*See* amoxicillin

TUMS.....*See* calcium carbonate

TYLENOL.....*See* acetaminophen

VALACYCLOVIR.....An antiviral drug used to treat all types of herpes virus infections, it may be used in adolescents, but has not been studied in young children.

VALDECOXIB.....A COX-2 inhibitor; a type of non-steroidal anti-inflammatory drug that has a reduced risk of stomach ulcers. *Drugs in this class have been linked to an increased frequency of heart attacks and strokes. Valdecoxib has been associated with a higher frequency of severe skin reactions than other drugs in the same class.*

VALIUM.....*See* diazepam

VALPROATE SODIUM.....*See* valproic acid

VALPROIC ACID.....Anti-epileptic drug for use in a variety of seizure types. It has been used for mood disorders, bipolar disorder, and other purposes as well. Approved for use in children over the age of 10 years, but has been used in much younger children as well.

VALTREX.....*See* valacyclovir

VENLAFAXINE.....An antidepressant which is unrelated to other types of antidepressant drugs. Warning: Antidepressants may increase depression or even lead to suicide when used in children and adolescents. Careful observation is essential.

VEPESID.....*See* etoposide

VIDARABINE.....Antiviral drug used to treat severe herpes infections in newborns, but its primary value is in the form of an eye ointment to treat herpes infections of the eye.

VIDEX.....*See* didanosine

VIR-A.....*See* vidarabi

VIRACEPT ..... *See* nelfinavir

VIRAZOL ..... *See* ribavirin

VISINE ..... *See* oxymetazoline

WARFARIN ..... A "blood thinner," a drug that helps keep blood from clotting. It is given to children who have had heart surgery until the heart has fully healed.

WELLBUTRIN ..... *See* bupropion

WINSTROL ..... *See* stanzolol

XYLOCAINE ..... *See* lidocaine

XYLOMETAZOLINE ..... A vasoconstrictor to treat nasal congestion which may be used as a nasal spray or nose drops.

ZALCITABINE ..... Zalcitabine is an anti-retroviral (anti-AIDS) drug. The manufacturer does not recommend use in patients under the age of 13 years, but it may be prescribed in younger children.

ZANAMIVIR ..... Antiviral drug used to treat influenza infections caused by viruses types A and B in adults and children over the age of seven.

ZARONTIN ..... *See* ethosuximide

ZERIT ..... *See* stavudine

ZIAGEN ..... *See* abacavir

ZIDOVUDINE ..... Antiretroviral agents (anti-AIDS drug) which may be appropriate for infants and children, best used in combination with other drugs.

ZIPRASIDONE ..... An antipsychotic drug. It has been approved only for use in adults, but has been used in children for treatment of schizophenia and bipolar disorder.

ZITHROMAX ..... *See* azithromycin

ZOFRAN ..... *See* ondansetron

ZOLOFT ..... *See* sertraline

ZONEGRAN ..... *See* zonisamide

ZONISAMIDE ..... A drug used to treat epilepsy, particularly partial seizures. Safety in children under 16 has not been established, but this drug has a history of use in pediatrics.

ZOVIRAX ..... *See* acyclovir

ZYPREXA ..... *See* olanzapine

ZYRTEC ..... *See* cetirizine

ZYVOX ..... *See* linezolid

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